Southern Regional Meeting Abstracts

Cardiovascular 1
Concurrent Session
11:00 AM
Thursday, February 26, 2015

1

NEUROFIBROMIN REGULATES MONOCYTE/MACROPHAGE FUNCTION
Stansfield B1, Ingram D2. Georgia Regents University, Augusta, GA and
Indiana University, Indianapolis, IN.

Purpose of Study: Neurofibromin results from mutations in the NF1 gene and functions as a negative regulator of Ras activity. Loss of neurofibromin, as observed in persons with neurofibromatosis type 1 (NF1), sensitizes cells to growth factor stimulation and abberant Ras activation. Some NF1 patients develop arterial stenosis in adolescence and early adulthood. We recently showed that loss of a single NF1 gene copy in myeloid cells is sufficient to induce arterial stenosis after arterial injury and enhance the mobilization of pro-inflammatory Ly6Chi CCR2+ monocytes. Therefore, we tested the hypothesis that activation of the MCP-1/CCR2 signaling cascade mediates the recruitment of neurofibromin-deficient myeloid cells to induce arterial stenosis.

Methods Used: We utilized a carotid artery ligation model to induce neointima formation. The left common carotid artery was ligated proximal to the bifurcation and mice were allowed to recover for 28 days. Control and ligated arteries were analyzed for neointima formation.

Nf1+/- mice with genetic deletion of MCP-1 or CCR2 were subjected to arterial injury separately, chimeric Nf1+/- were generated with specific deletion of CCR2 in bone marrow cells or vascular wall cells and carotid artery ligation was performed. Nf1+/- and WT macrophages and smooth muscle cells were isolated from the long bones and aorta, respectively. Functional assays were performed on Nf1+/- and WT macrophages and SMC. Finally, a competitive inhibitor of CCR2 signaling was administered daily following arterial injury and arteries were analyzed for neointima formation.

Summary of Results: Nf1+/- mice develop a robust neointima after arterial injury. Genetic deletion of either MCP-1 or CCR2 completely abrogates neointima formation in Nf1+/- mice. Loss of CCR2 in bone marrow cells of Nf1+/- mice abolishes neointima formation in Nf1+/- conditioned mice, while CCR2 deficient vascular wall cells partially reduces Nf1+/- arterial stenosis. Nf1+/- macrophages exhibit a masked sensitivity to MCP-1 and MCP-1 is secreted in the growth media of Nf1+/- macrophages. Finally, daily administration of a CCR2 antagonist after arterial injury significantly reduced Nf1+/- neointima formation.

Conclusions: Nf1+/- monocytes/macrophages exhibit increased sensitivity to MCP-1 and are recruited to sites of vascular injury via the MCP-1/CCR2 signaling axis.

2

IMPACT OF CATION DYSHOMEOSTASIS ON CORRECTED QT INTERVAL AND ARRHYTHMOCENICITY

Purpose of Study: The electrical activity of the heart depends on transmembrane ionic gradients and electrolyte abnormalities may facilitate arrhythmias. Hypokalemia is the most common electrolyte abnormality encountered in clinical practice. It can cause electrocardiographic (ECG) changes, prominent amongst which are its effects on action potential duration and prolongation of corrected QT interval (QTc). Hypomagnesemia has been known to also prolong QTc. Herein, we addressed serum K+ and Mg2+ concentrations in patients having prolonged QTc on standard ECG.

Methods Used: A retrospective chart analysis of 1400 patients who presented to Regional One Center Memphis between July 1, 2013 and June 30, 2014 having an ECG with prolonged QTc (>440 msec). Patients who were on medications that could prolong QTc were excluded. Serum K+ and Mg2+ concentrations obtained at the time of ECG were recorded for evaluation.

Summary of Results: A direct correlation was noted between K+ concentration and QTc. 18% (n=176) of patients with QT prolongation had K+ <3.5 meq/L whereas 52% (n=512) with QT prolongation had K+ >4.0 meq/L. 34% (n=336) with K+ between 3.6 and 4.0 meq/L had prolonged QTc interval. Similarly 8% (n=80) of patients with prolonged QTc had hypomagnesemia when normal Mg2+ was considered to be above 1.5 meq/L as compared to 60% (n=596) when normal Mg2+ was considered above 2.0 meq/L. 52% (n=516) of patients with QTc prolongation had Mg2+ values between 1.5 and 2.0 meq/L. Very few patients with higher K+ and Mg2+ values had QTc prolongation (8 and 3% respectively).

Conclusions: Both hypokalemia and hypomagnesemia can prolong QTc and thereby raise arrhythmogenic potential. This includes a broader definition of reduced K+ concentrations of 3.6-4.0 meq/L and Mg2+ concentrations of 1.5-2.0 meq/L. Hence, to minimize the risk of arrhythmias K+ and Mg2+ concentrations should be kept above 4.0 and 2.0 meq/L respectively, which we refer to as the 4/2 rule.

3

GLUCOCORTICOID LEVELS IN RESISTANT HYPERTENSIVE PATIENTS WITH AND WITHOUT ALDOSTERONE EXCESS
Ghazi L, Dudenbostel T, Calhoun D, Oparil S. University of Alabama Birmingham, Birmingham, AL.

Purpose of Study: Resistant hypertension (RHTN) is a prevalent and growing clinical problem. Aldosterone excess is common in patients with RHTN. We aim in this study was to evaluate cortisol, cortisone levels and the urinary cortisol to urinary cortisol ratio in resistant hypertension patients with and without aldosterone excess.

Methods Used: We retrospectively analyzed 77 patients seen at the referral Clinic at the University of Birmingham Alabama who were evaluated for RHTN. Tests included blood pressure measurement, physical exam, complete metabolic profile, plasma aldosterone, plasma renin activity, and 24 hour metabolic profile, plasma aldosterone, plasma renin activity, and 24 hour urinary aldosterone (UAldo, ug/24 hr), sodium (U-Na+, mEq/24 hr), potassium (U-K+, mEq/24 hr), urinary cortisol (U-C, ug/24 hr), and urinary cortisone (U-Cn, ug/24 hr) levels.

Summary of Results: In this study, 30 patients had aldosterone excess and 47 had no biochemical evidence of aldosteronism. Patients with aldosterone excess were significantly younger (51.3±11.5 vs 58.6±14.4 yrs, p=0.018) and had more males (63.33 vs 34.05 %, p=0.012). There was no difference for race, BMI, or duration of hypertension. The biochemical evaluation revealed that for patients with aldosterone excess; UAldo (23±12.6 vs. 6.96±3.17, p=0.001), U-C (17.94±14.8 vs 11.52±7.65, p=0.037), U-Cn (88.4±4.74 vs 58.5±29.0, p=0.0038), U-Na+ (223.7±121.4 vs 164.2±76.5, p=0.022), and U-K+ (85.45±3 vs 49.1±24.2, p=0.0033) values were higher than in patients without aldosteronism. The U-Cn to U-C ratio was not significantly different in patients with aldosterone excess and without aldosteronism. However, there was a wide range of U-Cn to U-C ratios in patients with moderately abnormal cortisol and cortisone levels.
MEASURES OF OBESITY AND OXIDATIVE STRESS: THE BOGALUSA HEART STUDY
Barshop RP1, Fernandez-Alonso C1,2, Chen W1, Siniruvain SR1, Berenson GS2.
1Tulane University, New Orleans, LA; 2Tulane University, New Orleans, LA and Tulane University, New Orleans, LA.

Purpose of Study: Oxidative stress is considered to be associated with obesity, and more specifically the accumulation of adipose tissue. F2-isoprostanes are synthesized through the peroxidation of fatty acids, a process which also creates reactive oxygen species. As such, F2-isoprostanes serve as useful biomarkers of oxidative stress originating from adipose tissue and lipid metabolism. There is currently a number of obesity indices employed within scientific literature, each with its own utility. However, the relationship between measures of obesity and oxidative stress has not been fully examined.

Methods Used: Urinary F2-isoprostane levels were collected from 898 adults, mean age of 43 years (29-50 years); 42.2% male 68.2% white, as part of the Bogalusa Heart Study. Sex and race specific independent associations were tested through multivariable-adjusted linear regression analyses. Because the different obesity indices have different scales, standardized Z-scores were used in the regression analyses.

Summary of Results: When using standardized Body Mass Index (BMI) as the measure of obesity, it was found to be independently associated with isoprostane (β=32.6, p<0.0001) after controlling for sex, race, gamma-glutamyl transferase, and triglycerides. Standardized waist-height ratio has a similar association with oxidative stress; an independent association was found (β=33.1, p=0.0001) after adjusting for the same covariates. Similar associations were found using triceps and subscapular skinfolds. However, the Z-standardized measurement of Body Shape Index (ABSI) was not found to be associated with isoprostane through linear regression.

Conclusions: These findings indicate that the relationship between obesity and oxidative stress is dependent on the metric used for measuring obesity. It is of note that ABSI, which uses waist circumference and is considered to be more associated with mortality events than other obesity metrics, is not associated with oxidative stress. This calls for a need to understand better the effect of body fatness and obesity related oxidative damage to the risk of morbidity and mortality.

Adult Clinical Case Symposium

Case Report: A 53-year-old male was hospitalised for presenting with chief complaint of sudden onset “worse headache of my life” one hour after first dose of leuprolide injection. This was associated with light sensitivity, nausea and vomiting. On physical examination, pupils were equal, round, and reactive to light. Extra-ocular motion, peripheral vision, and cranial nerves II-XII were intact without any sensory or motor deficits. Laboratory analysis showed thyroid stimulating hormone (TSH) of 0.13 μIU/ml (0.35-5.5 μIU/ml), random cortisol 2.5 μg/dl (4.5-22.7 μg/dl), adrenocorticotropic hormone 6.2 μg/ml (7.2-63.3 μg/ml), Prolactin 2.7 ng/ml (4.0-15.2 ng/ml), with normal levels of follicle stimulating hormone, luteinizing hormone, and growth hormone. Magnetic resonance imaging showed pituitary hemorrhage within an adenoma measuring 19 x 16 x 12 mm concerning for pituitary apoplexy. Patient was given intravenous hydrocortisone with improvement of his symptoms over 24 hours. He was discharged on po hydrocortisone 20 mg in the morning and 10 mg at night and continued to stay asymptomatic after 1 month follow up.

Pituitary apoplexy is a rare but serious life-threatening condition. Patients usually present with sudden onset of severe headache followed by rapidly worsening visual field defects. Although the pathophysiology is ill defined at this time, several factors associated with this phenomenon have been suggested: pituitary vasculature abnormalities, size of the adenoma, and elevated intrasellar pressure. Pituitary apoplexy after gonadotropin releasing hormone (GnRH) agonist administration is very rare. This may be because most incidental pituitary adenomas are microadenomas rather than macroadenomas which would be less likely to develop into a symptomatic pituitary apoplexy even if hemorrhagic necrosis did occur. However, given the seriousness of this condition and frequency of pituitary adenomas in the general population of 10-20%, physicians should be cautious and pay close attention if patients present with signs and symptoms associated with pituitary apoplexy. Screening patients with pituitary adenoma prior to treatment would likely not be cost effective given its rare occurrence.

DIABETIC CHRONIC EOSINOPHILIC PNEUMONIA - A DIAGNOSTIC CONUNDRUM
Ali RA1, Baldeo C1, Stem borrskis L1, Cury J1, Siddiqui A2. 1 UF Health, Jacksonville, Jacksonville, FL and 2 UF Health, Jacksonville, FL.

Case Report: A 63 year old African American female with Hypertension, Diabetes Mellitus and end stage renal disease was admitted for evaluation of recurrent pneumonia after being treated for a prior episode one month earlier. She had bilateral rales and an eosinophil predominant leukocytosis. Chest x-ray (CXR) showed venous congestion and patchy opacity at the left base.
She was started on Vancomycin/Zosyn. CT angiogram (CTA) chest was suggestive of early interstitial lung disease, and mediastinal and hilar adenopathy. Infectious work-up was negative. Despite nine days of antibiotics, the eosinophilia worsened and respiratory symptoms progressed. Prednisone was started with favorable clinical and serologic responses. Bronchoscopy revealed whitish secretions and lavage (BAL) was unremarkable. Bronchial biopsy revealed interstitial fibrosis. During the steroid taper, eosinophilia and respiratory symptoms recurred. Repeat CT revealed a large pericardial effusion, bilateral pleural effusions and worsening interstitial lung opacities. Echocardiogram confirmed tamponade and urgent pericardial window was performed. Thoracentesis revealed an eosinophilic effusion.

She was readmitted 2 months later for possible unresolved pneumonia. She had bilateral crackles. CXR suggested pulmonary edema. Eosinophilia was again noted, peaking at 46%. Repeat bronchoscopy was unchanged. BAL and paratracheal lymph node and bronchial biopsies revealed eosinophilic pneumonia. Bone marrow biopsy showed myeloid hyperplasia and normal flow cytometry and cytogenetics.

She was started on prednisone 40mg daily on a prolonged taper with dramatic improvement in eosinophilia and symptoms.

Idiopathic chronic eosinophilic pneumonia (ICEP) is a rare disorder associated with intense infiltration of the lungs with eosinophils. Diagnosis is based on subacute respiratory symptoms, alveolar and/or peripheral blood eosinophilia and peripheral pulmonary infiltrates, after eliminating other causes.

Treatment of ICEP hinges mainly on steroids with a prolonged taper. Response is impressive, however premature discontinuation can precipitate relapses, as demonstrated in our case. Long term sequelae include asthma and peribronchial fibrosis with more than 50% of patients requiring long term corticosteroid therapy.

RA MAY NOT CONFER ADDITION RISK OF OSTEOPOROSIS IN MALE RA PATIENT AS COMPARED TO FEMALE

Bhutia J, Majithia V. UMMC, Jackson MS, MS.

Purpose of Study: Osteoporosis (OP) is a common disease and is increasingly being recognized in males. A number of underlying diseases including rheumatoid arthritis (RA) have been associated with it and this relationship has been well proven in females but unclear in males with RA. This study is done to evaluate association between RA and OP in male population.

Methods Used: 585 men who underwent DEXA scan performed at UMC from 2005-2012 were included in the analysis of retrospective cohort with documented RA. PubMed literature search was performed using keywords: Male, OP and RA yielding 775 articles (limiting data to English and publication year > 1960), 663 articles were excluded with abstract review, 112 underwent full text review with 7 having relevant data and were included in analysis.

Summary of Results: as below

Conclusions: This analysis suggests that relationship of RA and OP in male patients remains confusing. Although no firm conclusions can be drawn, the results of cohort analysis and review of literature suggest that RA diagnosis and disease activity may not add any increase in the risk of developing OP above a low BMI, older age and steroid use. There was only one study out of 8 where RA itself seemed to add additional risk of developing OP while other studies suggested that this was not the case or possibly was only contributory in older age

FIGURE 1.
This is the first reported case of isolated diplopia secondary to FK506 and CyA in recommended therapeutic doses for treatment of Idiopathic MGN. Previously reported cases have occurred in the setting of much higher levels of CNIs, in patients with solid organ or bone marrow transplants. This case highlights the variable and unpredictable nature of calcineurin inhibitor neurotoxicity even at levels not considered to be toxic. Early detection and discontinuation of therapy led to resolution of the symptoms with no sequel in this case.

11
HEMOPHAGOCYTIC LYMPHOHISTIOCYTOSIS: PEDIATRIC DISEASE IN AN ADULT
Provo J1, Kiefer AC1, Vasquez R2, Warrier R2. 1Tulane University School of Medicine, New Orleans, LA and 2Ochsner Medical Center, New Orleans, LA.

Case Report: Hemophagocytic Lymphohistiocytosis (HLH) is a rare hyper-inflammatory state more common in children. We report a 21 year old Caucasian female in the Adult ICU with a fulminate illness following a Streptococcus A infection. The course of her illness proves that a high index of suspicion for HLH needs to be considered even in adults with multi-system and multi-organ failure.

Case: A 21 year old Caucasian female with morbid obesity presented to the emergency department with a fever of 40.1°C, sore throat, vomiting, rigors, cough, arthralgia, myalgia, rash, decreased oral intake, and lethargy. Her primary physician had diagnosed her to have strep throat with a positive culture 4 days earlier. She had fever, sore throat, and rash and started on Amoxicillin. She had no history of autoimmune disorders, sick contacts, pets, other medications or drug allergies. Physical Exam was remarkable for erythema and diffuse exudate of the tonsils bilaterally. Bimanual exam did not reveal the presence of a retained foreign body. Tender, violaceous macules were noted on the dorsum of the hands, feet, and chest wall. Labs demonstrated thrombocytopenia and leukocytosis. She quickly decompensated, developing respiratory distress requiring ventilator support and vasopressors. Her course was further complicated by kidney injury requiring dialysis, and DIC requiring blood products. Pediatric hematologists came to the rescue and were noted on the dorsum of the hands, feet, and chest wall. Labs demonstrated thrombocytopenia and leukocytosis.

Discussion: Hospitalists admit patients with stroke on a daily basis and more than 25% have an associated aphasia. Traditionally, aphasia is categorized as one of the following: Broca’s, Wernicke’s, global, transcortical or amnestic. The area of the brain affected typically correlates quite closely with the type of aphasia seen. Wernicke’s aphasia, also called fluent or receptive aphasia, occurs when the temporoparietal cortex is damaged in the dominant hemisphere. In right handed patients, the left hemisphere is dominant 95% of the time (60% of left handed). Our patient’s large left sided defect correlates with destruction of Wernicke’s area, however, patients with Wernicke’s aphasia usually have difficulty with both spoken and written word comprehension. It is important to note that some patients with schizophrenia or psychosis can have speech that resembles Wernicke’s aphasia, however, their speech often involves fixation on a theme and their neologisms are more contextually appropriate. Rarely, as in our patient, one can still understand written language. Recent studies suggest that anterior temporal regions sustain semantic processing of visually represented language when posterior language areas are injured. This challenges the conventional neurobiological model of language, and new models are taking its place. It is important for physicians to be aware of both typical and atypical presentations of Wernicke’s aphasia. Attempts to communicate with written, rather than strictly verbal cues, can be rewarding in certain instances.

12
AN UNUSUAL TIME WHEN WERNICKE’S MAKES SENSE
Lemley RJ, Jenkins M, Smalligan RD. Texas Tech University HSC, Amarillo, TX.

Case Report: A 58yo right handed man with a history of bipolar disorder with psychosis and multiple CVAs presented with abnormal speech. Other PMH, FH, P/S were unremarkable. On physical exam he had a nonfocal detailed neurologic exam but his speech was abnormal. He spoke fluently with normal articulation and form, yet the speech contained no meaning. He repeated words and phrases in an attempt to convey his ideas. He had poor auditory processing and could not answer verbal questions. However, the patient understood written queries and provided short, appropriate verbal responses. The first few words of his response were meaningful but then he shifted into senseless speech. He did not have any recurrent theme to his answers. Head CT showed old left temporoparietal as well as multifocal right sided strokes.

Discussion: Hospitalists admit patients with stroke on a daily basis and more than 25% have an associated aphasia. Traditionally, aphasia is categorized as one of the following: Broca’s, Wernicke’s, global, transcortical or amnestic. The area of the brain affected typically correlates quite closely with the type of aphasia seen. Wernicke’s aphasia, also called fluent or receptive aphasia,
14 IMMUNE SYSTEM GONE WILD
Fidone EJ, Mirkes C. Baylor Scott and White Health care, Temple, TX.
Case Report: Hemophagocytic lymphohistiocytosis (HLH) is a rare hematologic disorder, with an estimated incidence of 1.2 cases per million per year, characterized by an exaggerated immune response leading to marked proliferation of reactive lymphohistiocytes, excessive release of inflammatory cytokines and eventual cytokine-induced, multi-organ failure. HLH presents a diagnostic challenge to physicians due to its ability to mimic more common hematologic, infectious and rheumatologic etiologies. However, prompt diagnosis of HLH is crucial, as this disorder progresses quickly and is invariably fatal without treatment.

A 28-year-old African-American male presents to the ED with a 3-day history of spiking fevers, and severe abdominal pain. He reports having a three month history of abdominal pain, night sweats, and a 100-pound weight loss. Vital signs on presentation were normal except for a heart rate of 126 and a blood pressure of 104/66. Physical exam revealed a diaphoretic male with scleral icterus, axillary and inguinal lymphadenopathy, abdominal tenderness, and hepatosplenomegaly. Laboratory evaluation demonstrated pancytopenia, a ferritin level of 11,884 ng/mL and a fibrinogen level of <60 mg/dL. Over the next few days, the patient continued to decline clinically, requiring multiple units of blood and cryoprecipitate. An axillary lymph node core biopsy and a bone marrow aspirate and biopsy demonstrated hemophagocytic lymphohistiocytosis. After ruling out infecting etiologies, the patient was initiated on high-dose corticosteroids. His clinical picture improved dramatically in the following days.

This case emphasizes the importance of early detection when confronted with HLH. The average time to diagnosis for HLH can range from 2 weeks to 3 months. However, patients suffering from HLH do not have the luxury of time. This disease is almost uniformly fatal within 2 months if left untreated, yet, dramatic clinical response can occur once treatment is initiated. Thus, prompt consideration of HLH is paramount. This patient illustrates that a dramatically elevated ferritin strongly suggests an autoimmune hemolytic process. When coupled with hypofibrinogenemia, pancytopenia, generalized lymphadenopathy and hepatosplenomegaly, one must first consider a consumptive, rather than an infiltrative, process.

15 ISOLATED CNS HISTOPLASMOSIS IN IMMUNOCOMPETENT HOST: MIMICKING BRAIN METASTASIS
Mohamed A1, Edriss H1, Fenire M2, Ali E1, Mazek H1, Nugent K1, 1Texas Tech university health sciences center; Lubbock, TX and 2East Tennessee state University, East Tennessee, TN.
Purpose of Study: Histoplasmosis is a disease caused by the dimorphic fungus Histoplasma capsulatum. Most patients with histoplasmosis have no symptoms; however when symptomatic, it usually manifests with acute or chronic lung disease. Progressive disseminated disease is rare and almost always occurs in immunosuppressed hosts.
Methods Used: Case analysis and literature review.
Summary of Results: We report a 56-year-old man from New Mexico with a 6 month history of weight loss, night sweats and personality changes. He also reported new onset headache and progressive right-sided weakness. His past medical history included a three month history of abdominal pain, night sweats, and a 100-pound weight loss. Laboratory values included an eosinophilia of 1510 (normal 0-390/UL), elevated ferritin level of 11,884 ng/mL and a fibrinogen level of <60 mg/dL. Over the next few days, the patient continued to decline clinically, requiring multiple units of blood and cryoprecipitate. An axillary lymph node core biopsy and a bone marrow aspirate and biopsy demonstrated hemophagocytic lymphohistiocytosis. After ruling out infecting etiologies, the patient was initiated on high-dose corticosteroids. His clinical picture improved dramatically in the following days.

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16 A CASE OF HYponatRemia POTENTIATED BY NSAIDS
Pournourmoeza M1, Pournourmoeza M2, Patel B1, Peiris A2, Patel P1, 1ETSU Quillian College of Medicine, Johnson City, TN and 2ETSU Quillian College of Medicine, Johnson City, TN.
Case Report: Introduction: Non-steroid anti-inflammatory agents (NSAIDs) as a sole source of hyponatremia is uncommon. They do so by inhibiting pros-thaglandin synthesis leading to a potentiation of vasopressin, water reabsorption, enhanced fluid retention and ultimately hyponatremia. We report an unusual case of hyponatremia due to enhancement of desmopressin effect by NSAIDS.
Case Report: A 47 year old male admitted to the hospital for 2 weeks of back pain, headache, nausea, acral edema. Past medical history included resected pituitary macroadenoma, hypocortisolism, hypogonadism, Diabetes insipidus. He was on desmopressin, Hydrocortisone and Testosterone Injections. Vitals were within normal limits. Physical exam demonstrated 10 lb weight gain in two weeks, mild facial and extremity swelling. Laboratory analysis showed sodium 117 mmol/L, plasma osmolality 253 mOsm/Kg, sodium excretion 63 meq/L, urine osmolality 406 mOsm/Kg. Patient began taking Ibuprofen 600 mg every 4 hours two weeks ago for back pain. After excluding other causes of hyponatremia, enhanced desmopressin effect by Ibuprofen was the likely cause of hyponatremia. Ibuprofen was discontinued as desmopressin regimen was reduced to twice daily with free water restriction. Sodium increased gradually to 137 on outpatient follow up. Subsequently the desmopressin was changed to prevent TID dose and patient has done well.

Discussion: Hyponatremia usually develops during the first two weeks after drug initiation. NSAIDs as a sole source of hyponatremia is uncommon and are more commonly observed in the setting of altered renal function. NSAIDs like ibuprofen cause a reduction in renal prostaglandin synthesis which normally antagonize antidiuretic hormone (ADH). Such inhibition potentiates the effects of ADH which ultimately leads to decreased water excretion. This case represents a unique cause of NSAID-induced hyponatremia in the setting of Diabetes insipidus treated with desmopressin. Mortality rates tend to increase as the serum Na falls from 134 to 120 meq/L. Therefore, clinicians should be aware of patients with altered fluid status, since common over the counter medication such as NSAIDs impact fluid balance and pose a great risk for induction of hyponatremia.

Pediatric Clinical Case Symposium
1:00 PM Thursday, February 26, 2015
17 A LUNG MASS IN A TEENAGE MALE WITH ASTHMA
Mirani G1, Valerie E2, Smith RK3, Greer DL4, Craver R1, Begue R5, 1 Tulane University Health Sciences Center, New Orleans, LA; 2 Children’s Hospital, New Orleans, LA; 3 Children’s Hospital, New Orleans, LA; 4 Louisiana Health Sciences Center, New Orleans, LA and 5 Children’s Hospital, New Orleans, LA.
Case Report: A 17-year-old male with a history of asthma and allergic rhinitis presented with cough, chest pain, and shortness of breath for several weeks. An outpatient chest x-ray revealed a right middle lobe lung mass. The patient underwent an open thoracotomy with a wedge resection of the 4 x 2 cm lesion, extending from right middle lobe into the hilum. The pathology of the resected mass was reported as necrotizing granulomatous inflammation with fungal hyphae, marked eosinophilia, and bronchiectasis. The fungal culture eventually grew Bipolaris species (Figure 1). The susceptibility testing showed following MICs: AMB 0.25, ITRA 0.25, and VORI 0.25 (µg/mL, University of Texas Health Science Center at San Antonio). The patient tolerated a three-month course of itraconazole with resolution of symptoms. Significant laboratory values included an eosinophilia of 1510 (normal 0-390/UL), elevated
IgE level of 4920 (normal 0-158 IU/mL), and Bipolaris australiensis IgE of 54.80 kU/L (normal < 0.35, ARUP Laboratories). Subsequent skin scratch tests to various mold spores including Bipolaris showed marked reactivity. This clinical picture would fit the diagnosis of allergic bronchopulmonary mycosis with Pulmonary Phaeohyphomycosis due to Bipolaris spp.

**FIGURE 1.**

PERSISTENT FEVERS IN AN 18-YEAR-OLD RENAL TRANSPLANT RECIPIENT

Mirani G1, McNaughton J2, Yosypiv IV1, Schmieg JJ2, Robinson J1. 1Tulane University Health Sciences Center, New Orleans, LA and 2Tulane University Health Sciences Center, New Orleans, LA.

**Case Report:** An 18-year-old male, with a history of kidney transplant in 2006, presented with fevers up to 104°F (normal) in the 2-3 days prior to hospital admission in January 2014. The patient also reported a mild cough and headaches. The cough was non-productive. His headaches were non-focal and non-persistent and were present upon waking. His sister at home had a recent upper respiratory infection. There was no vomiting, diarrhea, dysuria, urinary frequency or urgency, or hematuria. The patient did not report intake of unusual foods or recent travel. His immuno-suppressive medications included cyclosporine, mycophenolate mofetil, and prednisone. CT scan of the chest revealed multiple lymph nodes including a large right-sided axillary lymph node. Excisional axillary lymph node biopsy performed on the 7th day of hospitalization, revealed necrotizing granulomatous lymphadenitis with features most consistent with cat scratch disease. Warthin-Starkey stains highlighted rare possible bacilli (Figure 1). Bartonella DNA PCR (qualitative) from plasma was positive. Bartonella Henslae IgM was negative, and IgG was 1:640 (negative < 1:320 titer). The patient was treated with azithromycin 250 mg orally daily for 6-8 weeks. He became afebrile within 24 hours of starting antibiotic. Repeat CT scan a month later showed improved lymphadenopathy.

**FIGURE 1.**

UCUTANEOUS LYMPHOMA MIMICKING BENIGN SKIN RASH

Sharma A, Radulescu VC. University of Kentucky, Lexington, KY.

**Case Report:** A previously healthy 10-year-old female presented with low-grade intermittent fever and multiple areas of red tender rash since last 3 months. On examination, she had multiple erythematous, well-circumscribed indurated plaques and nodules, 3 to 5 cm in diameter present on both arms, right leg, right flank and back.

CBC was normal except for a low WBC count (1900/uL). Systemic markers of inflammation (ESR, CRP, procalcitonin) were within normal limits. Serum electrolytes were normal but liver enzymes were mildly elevated (AST 187U/L, ALT 172U/L, ALP 155U/L). Extensive infectious disease and immunological work up remained negative. Imaging studies of chest, abdomen and pelvis did not show any abnormality or lymph node enlargement. Bone marrow microscopy was normal as well.

Skin biopsy of the lesions showed an epidermal perivascular infiltrate consisting of atypical lymphocytes and histiocytes surrounding individual fat cells in a necklace-like pattern along with fat necrosis and karyorrhexis. Immunohistochemical staining led to the diagnosis of subcutaneous paninculitiss-like T-cell lymphoma (SPTCL). She was treated with cyclophosphamide, doxorubicin, vincristine and prednisone (CHOP) with good results.

Discussion: SPTCL is a T-cell lymphoma localized primarily to the subcutaneous adipose tissue without lymph node involvement. In SPTCL, T cells have a CD4-CD8+CD56- TCR-β phenotype. Cutaneous lymphoma with CD4-CD8-CD56+ TCR γδ phenotype, previously included under SPTCL, is now classified as cutaneous γδ T-cell lymphoma as it has an aggressive course with worse prognosis. The incidence of SPTCL is <1% of the non-Hodgkin lymphomas. It is exceedingly rare in children and has a female predominance. Patients usually have leukopenia with mildly elevated liver enzymes. Biopsy histology as described above is pathognomonic of SPTCL. It has a good prognosis with complete resolution of the lymphoid infiltrate either spontaneously or after chemotherapy.

**FIGURE 1.**

**ACUTE KIDNEY INJURY AND HYPERTENSIVE EMERGENCY IN A PATIENT WITH CLASS V LUPUS NEPHRITIS**

Cantu MS1, Kidd L2, Singh D1, Yosypiv IV1. 1Tulane School of Medicine, New Orleans, LA and 2Tulane School of Medicine, New Orleans, LA.

**Case Report:** A 13 year old African American female with a past medical history of class V lupus nephritis (LN) with normal kidney function well-controlled with mycophenolate, hydroxychloroquine and enalapril, presented with recent facial abcess, progressive body swelling and right-sided flank pain. Physical examination showed elevated blood pressure of 171/117 mmHg, periorbital and lower extremity edema. Laboratory studies showed elevated
Case Report:
GRU, Augusta, GA.

SELF-RESOLVING ORTNER’S SYNDROME IN A 22-NEONATE WITH BILATERAL PNEUMOTHORACES
Connor EE 1, Craver R 1,2, McGoey R 1.

NEONATAL PERICARDIAL CONGLOMERATE (CPL) IS NOT A CONTAMINANT
Connor EE 1, Craver R 1,2, McGoey R 1.

NEONATE WITH BILATERAL PNEUMOTHORACES
Connor EE 1, Craver R 1,2, McGoey R 1.
Louisiana State University Health Sciences Center, New Orleans, LA and 2Children's Hospital of New Orleans, New Orleans, LA.

Case Report: A female neonate delivered via caesarian section at 26 weeks gestation experienced immediate respiratory distress after birth requiring transfer to the neonatal intensive care unit and intubation. Imaging showed bilateral pneumothoraces, and chest tubes were placed. Despite these interventions and aggressive supportive care, the infant expired within 24 hours of birth. An autopsy was authorized by the family and later performed by an attending and resident pathologist. Immunohistochemical stains were subsequently selected and performed.

The autopsy was unremarkable with the exception of the lung findings. The lungs were hyperinflated and had a combined weight of 34gm [expected 18-69gm]. The pleural surfaces were finely nodular with visible faint white streaks. Sectioning revealed a firm tan-pink parenchyma with a diffuse, subtle cystic pattern that extended to the pleural surface. Microscopic analysis of all five lung lobes showed dilated cystic spaces in three contiguous sites: subpleural, interlobar, and periairial. In most spaces, the lining was disrupted or fragmented. Immunohistochemical staining showed these lining cells to be positive for CD31 and D2-40. These findings are consistent with the diagnosis of primary pulmonary lymphangiectasia.

Congenital pulmonary lymphangiectasia (CPL) is a rare disorder and is often not diagnosed until autopsy. Literature reports over the past roughly 150 years cite less than 100 total cases. Thought of as a uniformly fatal disorder, CPL is thought to account for 0.5% to 1% of all stillborn and neonatal deaths. The cause is unknown but postulated to be due to an inherent developmental abnormality of the lymphatic system leading to dilated lymphatic spaces and lymphatic dysfunction. Primary CPL is limited to the lungs and does not affect in utero development, but presentation immediately after birth includes severe respiratory distress, pneumothoraces and pleural effusions. Greater awareness of CPL is needed and further reports in the literature would undoubtedly enhance our understanding of the condition and focus efforts towards the development of new therapeutic models.

SELF-RESOLVING ORTNER’S SYNDROME IN A TERM NEONATE
Pierce E, Bhatia J, Chan A. GRU, Augusta, GA.

Case Report: Ortner’s syndrome, defined as left recurrent laryngeal nerve dysfunction resulting from cardiovascular pathology, is a rarely reported phenomenon in neonates. This case report describes a term infant who presented with hoarse cry and stridor at 33 hours of life accompanied by desaturations and respiratory distress. Work up included an echocardiogram which demonstrated the presence of a moderately large ductus arteriosus aneurysm; MRI confirmed that the mass was exerting an effect on the left recurrent laryngeal nerve. Bedside nasolaryngoscopy confirmed hypomobility of the left vocal cord. The infant was observed for several days since feedings were tolerated and desaturation episodes improved. After seven days of observation and serial echocardiograms, the aneurysm showed signs of regression without surgical intervention. This case emphasizes the importance of considering alternative pathologies for otherwise narrow differential diagnosis of hoarse cry in the neonatal population. Furthermore, unlike some previously reported cases, no surgical intervention was necessary confirming the current recommendations for several days of observation prior to pursuing surgical repair.

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A CASE OF GRANULICATELL A ADIACENS INFECTIVE ENDOCARDITIS IN A SIX YEAR OLD
Neemuchwala F 1, Struk M 2, Burns J 1, Whittingham E 1. 1Florida State University, Pensacola, FL and 2Highland Regional Medical Center, Sebring, FL.

Case Report: Timely diagnosis of infective endocarditis (IE) in children can be a difficult clinical challenge. Herein we report a case of IE caused by an uncommon organism.

A six year old female was admitted with a history of persistent fevers and fatigue of one month duration. She had a past medical history of congenital aortic stenosis, ventricular septal defect, status-post Norwood and Damus-Kaye-Stansel procedure as an infant and 6 months prior to admission had a bovine conduit placed for pulmonic stenosis.

Four weeks prior to admission she was seen at the primary care clinic for fever 103.2°F diagnosed as a viral syndrome. Three days later she had a dental procedure and although prophylaxis for endocarditis was prescribed, the patient did not take the medication. Five days later she was evaluated in the ED for persistent fever for 8 days where a blood culture obtained grew Granulicatella adiacens (GA). This was felt to be a contaminant and no antibiotics were started. Patient had repeat blood culture in ED 5 days later which again grew GA but was not treated because at that time child was clinically well. Finally, an additional 2 weeks passed and child was admitted due to recurrent fever.

On admission, child had fever 101.8, and there was a harsh systolic murmur which was documented previously. Labs revealed elevated ESR and CRP with anemia and thrombocytopenia. The patient was started on Ceftriaxone and Vancomycin. Echocardiogram showed an echogenic density in the conduit. Blood cultures obtained on admission grew GA which was Vancomycin sensitive but Penicillin and Cephalexin resistant. Hence decision was made to discontinue Ceftriaxone and complete therapy with intravenous Vancomycin.

GA is a genus of nutritionally variant streptococci. It is often difficult to isolate in clinical laboratories and is reported to be resistant to standard treatments for streptococci. Previous reports of combined treatment with Vancomycin and Meropenem have been documented but our patient had successful treatment with single agent therapy. In conclusion, GA should not be considered a contaminant as it can cause endocarditis in pediatric patients and prophylaxis for endocarditis must be witnessed in high risk patients.

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ACUTE GENERALIZED EXANTEMATOUS PUSTULOSIS: A RARE PEDIATRIC CASE DUE TO CLINDAMYCIN
Edgar-Zarate C 1, Kaliki V 1, Shalin S 2. 1University of Arkansas Medical Sciences, Little Rock, AR and 2University of Arkansas for Medical Sciences, Little Rock, AR.

Case Report: Acute generalized exanthematous pustulosis (AGEP) is an acute reaction characterized by the sudden eruption of hundreds of sterile, non-follicular pinhead sized pustules predominantly in the main folds of the skin on a background of erythema. The incidence of AGEP is estimated to be around 1 to 5 cases per million per year with a slight female predominance. Previous studies have shown that the majority of cases are caused by adverse drug reactions and infections. AGEP has been reported only in a handful of pediatric cases, with drugs such as beta-lactams and cephalosporin as the suspected causative agent. Clindamycin has been suspected to be the etiology of AGEP only in adults. We here report the first case of AGEP associated with clindamycin exposure in a child.

A previously healthy 9-year-old boy presents after taking 1 dose of clindamycin prescribed for cellulitis to his left lower leg. The following day, he developed diffuse erythema and fever. The rash progressed the next day to a pustular appearance. Once admitted to the hospital he developed hypotension, and was treated for toxic shock syndrome. A skin biopsy was performed and...
showed subcorneal pustules, intradermal neutrophils and adjacent epidermal edema which is consistent with AGEP. His hospitalization was complicated by anemia and hypoalbuminemia, for which he did receive an albumin infusion. He made a full recovery with spontaneous resolution of the rash in about 9 days after cessation of the clindamycin.

The diagnostic criteria of AGEP are an acute purpuric eruption, fever, leukocytosis, subcorneal or intra-dermal pustules on skin biopsy, and spontaneous resolution in less than 15 days. Our patient met all of these criteria. No treatment is necessary with AGEP as it was evidenced in the presented case. While systemic manifestations are rare in AGEP, abnormal liver function tests, renal insufficiency, respiratory distress, and agranulocytosis have been reported in the past. The hypotension, anemia and hypoalbuminemia in this case were secondary to AGEP, although this remains unclear. We believe AGEP is a rare but potentially severe adverse reaction to clindamycin and prompt discontinuation should be considered if highly suspected.

26 PORCELAIN GALLBLADDER IN A CHILD WITH NEPHROTIC SYNDROME PRESENTING WITH SPONTANEOUS BACTERIAL PERITONITIS

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Purpose of Study: To Report the First Case of Porcelain Gallbladder in A Patient with Nephrotic Syndrome Presenting with Spontaneous Bacterial Peritonitis.

Methods Used: PUBMED Literature search, single case report.

Summary of Results: Porcelain gallbladder, or gallbladder wall calcification, is a rare condition, the etiology of which is not well understood. It has been hypothesized that it results from inflammation or irritation of the gallbladder from causes such as disordered calcium metabolism, chronic cholecystitis, cholelithiasis, abdominal trauma, or surgery. The clinical significance of porcelain gallbladder has recently been debated secondary to recent literature revealing a lesser but still significant association with gallbladder cancer than was previously recognized in older studies. This is significant because gallbladder cancer carries a poor prognosis. We report the first case of a child presenting with porcelain gallbladder without a prior episode of cholecystitis, cholelithiasis, abdominal surgery, trauma, or a known disorder of calcium metabolism and demonstrate relevant imaging findings.

Conclusions: The pathogenesis of porcelain gallbladder is not yet fully understood, but the role of gallbladder wall inflammation appears to be a common factor most often thought to be from direct gallstone irritation. This pediatric case of nephrotic syndrome with spontaneous bacterial peritonitis demonstrates our hypothesis that, perhaps, any process leading to inflammation in the peritoneal cavity or generalized edema may be a risk factor for gallbladder wall inflammation. Furthermore, the recurrence of edema, such as from hypoalbuminemia associated with nephrotic flares, may be the catalyst for gallbladder wall calcification, even without overt symptoms of gallbladder disease.

27 PRIMARY HIV MANIFESTING AS ACUTE PANCREATITIS IN AN ADOLESCENT: SYMPTOM OF A LARGER PROBLEM?

Lamb G, Dietz S, Graham R. University of Texas Southwestern, Dallas, TX.

Case Report: AF is a 16 year old African American male who presented to the hospital in August for acute abdominal pain in the setting of 3 months abdominal pain, anorexia, fever and weight loss. Symptoms began in May, but were considered insignificant and treated with analgesics and antipyretics until June 12 when the patient’s mother noted that his pain and fatigue became significant enough to keep him bed-ridden, he continued to have intermittent fevers and began to lose weight. Between June 23 and July 2 he saw his PCP twice and each time was found to have mildly elevated liver enzymes and lipase trended from 821 to 1079. Symptoms improved until 8/1 when he presented to the ED with worsening abdominal pain. At this lipase was 980, abdominal US showed no abnormalities, he denied alcohol use and a lipid profile was normal. On 8/9 he had fever of 104.5 and developed emesis and watery diarrhea. He was unable to tolerate PO and urine output decreased. He developed fatigue and dizziness and presented to the ED on 8/13. Labs showed ALT: 521, AST: 257, T.Bili: 1.3, D.Bili 0.6, GGT: 92 and lipase: 490. CBC was unremarkable. He reported a 13kg weight loss over the past month. CT showed inflammation of the pancreas, but no necrosis and no tumors. Pt denied illicit drug use or sexual activity, but HIV screen was positive. Quantitative PCR returned with >10,000,000 copies. The patient’s symptoms improved and HAART was deferred to outpatient treatment in order to prevent recurrence of pancreatitis.

Pancreatitis in the pediatric population is most commonly caused by trauma, medications, biliary tract disease and infections. Acute pancreatitis due to primary infection with HIV-1 has been rarely reported (7 adults, 1 adolescent). Although it has been well described as a complication of HIV secondary to medications and opportunistic infections, case reports now suggest that acute pancreatitis can be due to direct invasion of the pancreas by HIV. Dallas County has the highest rate of new HIV diagnoses in the state of Texas. The highest increase in new diagnoses is in persons aged 15-24. This extremely rare presentation has focused attention on the rapid growth of HIV + adolescents in the region and the need for a high level of suspicion in adolescent patients presenting to the ED.

Case Reports in Cardiovascular Medicine

2:00 PM Thursday, February 26, 2015

28 ACUTE ABDOMINAL PAIN DUE TO SPONTANEOUS RENAL ARTERY THROMBUS

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Background: Acute abdominal pain is commonly encountered in the emergency department (ED); spontaneous renal artery thrombosis is a very rare cause of acute abdominal pain.

Case report: A 56-year-old male with no past medical history presented to the ED with sudden onset sharp and constant abdominal pain, starting one hour prior without associated symptoms or trauma. His vital signs were within normal limits and physical exam revealed right lower quadrant abdominal tenderness. Blood work showed elevated lactate dehydrogenase (LDH), and normal complete blood count, renal function, liver function, urinalysis, lipase, amylase, and toxicology.Computed tomography angiogram of the abdomen showed thrombus in the right renal artery branch. He was treated with intravenous heparin, invasive angiography confirmed occlusion of the right renal artery. Attempts were done to recanalize the vessel, but were unsuccessful. Due to the small amount of kidney in jeopardy, the procedure was terminated.

Transthoracic and transesophageal echocardiography were negative for right-to-left intracardiac shunts or evidence of intracardiac thrombus. His aorta was not suggestive of an atheroembolic source. Hypercoagulability workup was negative. Patient was switched to rivaroxaban and discharged home in stable condition. 30-day event monitor showed sinus rhythm with no atrial or ventricular arrhythmias.

Discussion: Symptoms of acute renal infarction (ARI) due to renal artery occlusion are nonspecific, such as abdominal pain, flank pain, nausea or vomiting. It is usually underdiagnosed, so a high suspicion of this diagnosis is always warranted in high risk patients (1). The source of renal artery thrombosis is usually a thromboembolic event (2-4) or trauma (5). Spontaneous thrombosis of the renal artery can also be attributed to idiopathic dissection of the renal artery (6), underlying hypercoagulable state (7), and other conditions that have been associated with renal artery thrombosis and ARI such as nephrotic syndrome (8). However, renal artery thrombosis without any obvious underlying cause in an otherwise healthy patient is extremely rare (9-11). Revascularization of the occluded artery should be considered if possible, although was unsuccessful in our case (12, 13). 30

SHOULD WE REVISIT ANTICOAGULATION GUIDELINES DURING THYROID STORM?

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Case Report: 49 year old female with history of alcohol and illicit substance abuse evaluated due to palpitations, shortness of breath and lower extremity edema. On physical exam, heart rate irregular 174 bpm, BP 126/98 mmHg, RR 36 per minute, and T 96.9 F. Was anxious and diaphoretic with slight elevation in her jugular venous pressure and bibasilar rales. Point of maximal impulse was hyperkinetic and apically displaced. 2+ bilateral pitting edema noted as well as horizontal nystagmus and fine resting tremor. Echocardiogram showed atrial fibrillation with 184bpm. Initial blood workup remarkable for BNP 1277 pg/ml, TSH 0.02 mIU/L, Free T4 above 7.0 ng/dl. Thyrotopin receptor antibodies elevated at 18.91 IU/L confirming diagnosis of Grave's disease. CXR with mild pulmonary edema, dilatation of the cardiac silhouette and bilateral pleural effusions. Initial echocardiogram with mildly dilated left ventricle and severe reduction in left ventricular systolic function of 30%. Ventricular response remained difficult to control for which high doses of propranolol, metoprolol and digoxin were unsuccessfully attempted. CHADS2 score of 1 and aspirin started. Methimazole, potassium iodide and hydrocortisone Initiated which controlled her symptoms and decreased free T4 level. As her ventricular response remained poorly controlled, transesophageal echocardiography confirmed dilatation with global hypokinesis and severely reduced ejection fraction, also 7 mm thrombus identified with significant spontaneous echo contrast within the left atrial appendage. Heparin and Warfarin were immediately initiated. She responded to standard heart failure therapy with Lisinopril, Toprol XL and Furosemide. On six month follow-up, she was euthyroid with normal sinus rhythm. Repeat transesophageal echocardiogram showed normal left ventricular cavity size and ejection fraction 60% with complete dissolution of her left atrial appendage clot. In Thyroid Storm, heart failure occurs in approximatley 6% of cases and less than 1% develop dilated cardiomyopathy with Left Ventricular Systolic Dysfunction.

AN UNUSUAL CASE OF TRIGEMINAL NEURALGIA AND THE HEART

Dhakal P, Carhart R, Villareal D. SUNY Upstate Medical University; Syracuse, NY.

Case Report: A 33-year-old Caucasian male presented to his primary care doctor with pain involving left side his face for three months, precipitated by chewing and talking. This was associated with 15 pounds weight loss. Trigeminal neuralgia was diagnosed and a subsequent CT of the neck revealed no pathology in the affected site but showed bilateral upper lobe pulmonary infiltrates. A follow up CT chest revealed a left pleural effusion along with the incidental finding of a large lesion occupying nearly entire left lower lobe. He underwent liver biopsy, which revealed melanoma.

OBSTRUCTING RIGHT ATRIAL TUMOR

Gordon S1, Pekulovic A, Hess M2 1Virginia Commonwealth University, Richmond, VA and 2Virginia Commonwealth University, Richmond, VA.

Case Report: Mr. D. is a 54 year old male with past medical history of rheumatoid arthritis, who presented with fatigue and shortness of breath for 2-3 months. He had a trans-thoracic echocardiogram, which demonstrated a large right atrial mass, filling the entire atrium and prolapsing into the right ventricle through the tricuspid orifice. He was found to have liver lesions and underwent liver biopsy, which revealed melanoma.

His symptoms of cardiac dysfunction increased and the patient showed hemodynamically significant signs and symptoms of venous hypertension. Coronary angiograms demonstrated a large right atrial tumor being supplied from an RCA branch. He underwent surgical intervention with excision of the right atrial mass, radical resection the right atrium and reconstruction of the right atrium with pericardial patch. Surgery revealed a large mass growing in the trabeculated area, not involving the tricuspid valve, but attached to the myocardium close to the coronary artery. The tumor was growing down the inferior vena cava, but no attachments to the hepatic tumors. Pathology confirmed melanoma.

Post-operative course was uneventful. He was agreeable to starting treatment with CTLA4 Therapy and is currently being treated with ipilimumab. He

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had improvement in his liver lesions after one cycle. Cycle 3 was delayed due to rash and he re-started ipilimumab after a three week delay. He is doing well.

We present the case of a 25 year-old man who was admitted to our institution due to a one-year history of progressive fatigue with moderate exertion. He had a past medical history of an upper back "tumor" which was excised at another institution due to severe abdominal pain and nausea and vomiting. He was hypertensive and developed respiratory distress. He was later intubated and placed on broad spectrum antibiotics and intravenous fluid. His laboratory showed increased lipase level of 1,682 units/L. CT abdomen revealed acute pancreatitis with peripancreatic fluid without masses or pseudocyst. Severe pancreatitis was diagnosed. His clinical status was complicated by septic shock, acute respiratory distress syndrome, acute kidney injury and disseminated intravascular coagulopathy with platelet count of 48,000/µL, INR of 1.26, d-dimer of 16,552 ng/mL, and fibrinogen level of 447mg/dL.

On day 4 of admission, the patient became restless and twelve leads electrocardiography showed ST elevation in V3-V6. Cardiology was consulted for emergent coronary angiography. There was a total occlusion at the mid left anterior descending artery (LAD). Percutaneous coronary intervention of LAD with aspiration thrombectomy and three bare metal stents placement were performed successfully. Patient was transferred to the medical intensive care unit for ongoing critical care management with the continuation of ticagrelor and aspirin. There was no significant major bleeding. He improved slowly from his severe condition and was finally discharged from hospital after 44 days of hospital stay. He presented to 4-week follow up visit with significant recovery.

Etiology of PE was an underlying malignancy resulting in a hypercoagulable state.

Acute massive PE is defined as more than 30% obstruction of the pulmonary arterial bed. TTE reveals RV dysfunction due to dilatation or a hypercoaguable state. Etiology of PE was an underlying malignancy resulting in a hypercoagulable state. Acute massive PE is defined as more than 30% obstruction of the pulmonary arterial bed. TTE reveals RV dysfunction due to dilatation or a hypercoaguable state. Etiology of PE was an underlying malignancy resulting in a hypercoagulable state.

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Acute massive PE is defined as more than 30% obstruction of the pulmonary arterial bed. TTE reveals RV dysfunction due to dilatation or a hypercoaguable state. Etiology of PE was an underlying malignancy resulting in a hypercoagulable state.
BLOCKADE OF GLI1/2 DIMINISH HUMAN RHABDOMYOSARCOMA GROWTH IN XENOGRAFT MODEL

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Purpose of Study: Rhabdomyosarcoma (RMS) typically arises from skeletal muscle. Currently RMS in patients with recurrent and metastatic disease have no successful treatment. The molecular pathogenesis of RMS varies based on cancer sub-types. Among them a small percentage of embryonal RMS are driven by the sonic hedgehog (Shh) signaling pathway. However, inhibitors of this signaling pathway particularly those which inhibit smoothened have not found to be highly effective in animal testing.

Methods Used: In this study we have investigated the effect of Gant-61, a Gli-1 & 2 inhibitor on established RMS cell lines RD and RH30 both in vivo and in vitro.

Summary of Results: We found that Shh pathway effectors GLI1 and/or GLI2 are over-expressed in the majority of RMS cells and that GANT-61, a specific GLI1/2 inhibitor dampens the proliferation of both embryonal RMS and alveolar RMS cells-derived xenograft tumors thereby blocking their growth. As compared to vehicle-treated control, about 50% tumor growth inhibition occurs in mice receiving GANT-61 treatment. The proliferation inhibition was associated with slowing of cell cycle progression which was mediated by the reduced expression of cyclins D1/2/3 & E and the concomitant induction of p21. GANT-61 not only reduced expression of GLI1/2 in these RMS but also significantly diminished AKT/mTOR signaling. The chemotherapeutic action of GANT-61 was significantly augmented when combined with temsirolimus. Finally, reduced expression of proteins driving epithelial mesenchymal transition (EMT) characterized the residual tumors.

Conclusions: Targeting Gliomas-associated oncogene transcription factors represent a novel target in both alveolar and embryonal rhabdomyosarcoma and their blockade diminish the growth of these tumors in xenograft model. Gant-61 represents a novel molecule with possible indications to be tested in combination with other known chemotherapeutic agents to improve cure and long term survival.
males to be diagnosed with ADHD was more than twice that of females (OR 2.4, 95% CI 2.1-2.8).

Conclusions: In the past decade, the research evaluating ADHD across gender and race is limited. Our review indicates that AA and female children are significantly less likely to be diagnosed with ADHD. These disparities may be the consequence of rating scale bias, difference in symptom presentation, or a cultural difference in the approach to mental illness. An understanding of which etiological and cultural mechanisms account for the observed racial and gender differences would allow for more appropriate diagnosis and care for these children.

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IS MICROARRAY CGH A USEFUL DIAGNOSTIC TEST IN AUTISM SPECTRUM DISORDERS?

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Purpose of Study: Autism spectrum disorder (ASD) has a heterogeneous clinical presentation that involves problems with social understanding, language acquisition, and repetitive behaviors. ASD is a multifactorial disease caused by a combination of genetic and epigenetic factors and environmental triggers. Chromosomal abnormalities, rearrangements, deletions, and duplications are associated with ASD; however information on phenotypic presentations of genomic variants is limited. The purpose of this study is to identify phenotypes associated with chromosomal alterations in children with ASD.

Methods Used: ArrayCGH, Fragile X, and chromosome analysis were performed on 28 children between the ages of 2-18 seen at Tulane University Hospital for ASD or autistic features. Metabolic studies were requested (amino acids, ammonia, lactate, uric acid and urine organic acids), and information was collected on dysmorphic features, speech delay, learning problems, developmental delay, behavior problems, and aggressive behavior.

Summary of Results: Of the 28 children in the study 18 had an additional speech delay, 20 had a developmental delay, 20 had behavior problems, 9 had aggressive behavior, 8 had seizures or epilepsy, and 4 had decreased muscle tone. Dysmorphic features included 8 children with a high forehead and 9 children with macrocephaly. Eight children had chromosome alterations; two had a 16p13.11 microdeletion associated with autistic features including developmental, speech, and learning delays. However, the two patients did not share other phenotypic features. Other chromosome alterations in the cohort included deletion of 21q22.3 and duplications at 1q25, 9q34.3, 12q23, 15q13, and 6p22.

Conclusions: Our cohort had a high number of patients with chromosomal abnormalities, 28%, compared to the 10% reported in the literature. This may be due to selection bias as the patients in our cohort were referred for genetic consultation by Tu. The psychiatry department possibly for suspected genetic abnormalities. The small sample size in the study limited our ability to create subgroups with common chromosome alterations. Future work will involve increasing the sample size to allow classification of subgroups based on phenotypic similarities.

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LYMPHOPROLIFERATIVE DISORDER IN A CHILD WITH CROHN’S DISEASE AND THIOPURINE USE

Messer A, Raulji C, Fletcher M, Keith B, Velez M. LSU School of Medicine - Health Sciences Center, New Orleans, LA.

Case Report: Thiopurines are widely used in the treatment of patients with inflammatory bowel disease (IBD). Concern for associated lymphoproliferative (LPD) disorders must be addressed when deciding appropriate treatment. Multiple large studies have shown a strong association of increased risk of LPD in IBD patients receiving thiopurine for treatment. However, this association is not frequently described in the pediatric population and pediatirc data are limited.

An 11-year-old African-American female with one-year history of Crohn's Disease (CD) presented with fever, respiratory distress and seizure activity. Prior to admission, she had URI symptoms for one month and intermittent fevers for two weeks. Her CD treatment consisted of 6-mercaptopurine; however, it was discontinued ten days earlier due to neutropenia.

On admission, CBC showed pancytopenia with WBC 1.66 x 10^3 with ANC 460, hemoglobin 7.7 g/dL, and platelets 36,000. Ferritin level was elevated at 1823ng/ml. EBV PCR was positive in plasma (24,700 copies), bone marrow (19,900 copies) and CSF (400 copies). A head CT and MRI showed multifocal, subcortical white matter lesions, although neurological exam remained normal. Bone marrow aspirate and biopsy revealed hypocellular marrow with focal hemophagocytosis. Upon further evaluation CT scans showed multiple pulmonary nodules, an 8x5cm anterior mediastinal mass, and enlarged adenoids and tonsils. Biopsy of the anterior mediastinal mass was consistent with LPD.

The patient's treatment included high dose methylprednisolone, which was later changed to dexamethasone for better CNS penetration, and rituximab. End of treatment demonstrated normal CBC and ferritin, negative EBV PCR with improvement of brain lesions.

This patient is among the youngest documented to have EBV-associated LPD due to thiopurine use for CD. Her new-onset seizures and CNS lesions made her case at first puzzling. The development of LPD in children with CD can occur as a complication of the treatment, due to the immunosuppression associated with thiopurine.

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CRITICAL CONGENITAL HEART DEFECTS AMONG INFANTS BORN IN ARKANSAS HOSPITALS: IMPLICATIONS FOR NEWBORN SCREENING

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Purpose of Study: The U.S. Department of Health and Human Services recommends the addition of pulse oximetry (PO) screening for critical congenital heart defects (CCHD) to the newborn screening panel. We determined the number of infants born in Arkansas with CCHDs and identified the number of those infants recognized beyond 3 days of life who could potentially benefit from earlier PO screening.

Methods Used: The prevalence of CCHDs among Arkansas’ livebirth population was computed for birth years 2000-2010 using data obtained from the Arkansas Reproductive Health Monitoring System. Timing of initial diagnosis of CCHD was evaluated for 7 CCHD phenotypes among term infants. Infant mortality rates were computed and stratified by gestational age and timing of initial diagnosis. Delays in initial diagnosis, defined as diagnosis made after day 3 of life, were compared across birth hospital neonatal level of care.

Summary of Results: There were 559 livebirths in Arkansas having at least one CCHD diagnosis. The birth prevalence of CCHD was 13 per 10,000 livebirths. Approximately 20% of term neonates with CCHD experienced a delay in initial diagnosis. Mortality among infants with CCHD was 24.9%. Infants with CCHD born prematurely had a higher infant mortality (41.6%) than term infants (21.5%). A disproportionate number of late diagnoses occurred in neonatal care level 1 hospitals.

Conclusions: These results characterize the CCHD prevalence in Arkansas prior to implementation of universal PO screening. Universal screening may improve outcomes for CCHD term infants, where 1 in 5 experience delays in initial diagnosis. Many infants experiencing delays in diagnosis are born at level 1 neonatal facilities; thus supporting the need for universal screening, particularly in the smaller, birthing hospitals found in most rural parts of Arkansas. These results establish a pre-screening baseline that may be used to assess the effectiveness of PO screening in reducing infant mortality and severe morbidity.

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STILL SEARCHING: UNDERLYING GENETIC CAUSE OF CUTIS LAXA AND MULTIPLE ANEURYSMS

Preston GJ1, Scott K1, Settown C2, Marble M2, Baskind H1, Yosypiv IV1, Anderson H1, Morava E1, 1Tulane University, New Orleans, LA and 2LSU, New Orleans, LA.
Purpose of Study: Vascular aneurysms associated with Cutis Laxa can cause life-threatening complications in young patients. Several genes including SLC2A10, CBS, ACTA2, FBN1, FBN2, COL3A1, TGFB1, TGFB2, SMAD3, GLUT1, LTBP4 and MYH11 have been implicated. We evaluated a patient with CL, cardiovascular aneurysms and severe hypertension with suspected connective tissue disorder. DNA analysis revealed a heterozygous mutation in MYH11, a gene coding for myosin heavy chain, a contractile protein in smooth muscle.

Methods Used: Mutations of MYH11 are associated with thoracic aortic aneurysm, aortic dissection, and abnormal elastic formation. The patient’s mutation (c.3651+5→+11del7insM) results in the deletion of seven nucleotides and the insertion of a single G nucleotide, creating five base pairs into intron 27. To evaluate pathogenicity we performed RT-PCR for abnormal MYH11 splice variants, cDNA sequencing for cryptic splice sites, long range PCR for intron retention and additional sequencing for ABCC6 and GGCX. Patient expired at the age of 5 and underwent autopsy. We also performed histology for elastin structure and Golgi secretion studies.

Summary of Results: Autopsy confirmed severe atherosclerotic vascular aneurysms of the distal abdominal aorta, bilateral renal arteries, and bilateral internal carotids. Pathology identified fragmented elastin fibers in skin biopsy, and extreme atherosclerosis of vascular aneurysms, characterized by ossification and bone marrow formation in the calcified plaques. Molecular analysis of MYH11 cDNA revealed only normally spliced, mature mRNA transcripts. Sequencing of ABCC6 and GGCX revealed no sequence anomalies in the protein-coding region. Histology in fibroblast revealed delayed elastic lamina and Golgi secretion.

Conclusions: Based on functional data the MYH11 mutation is non-pathogenic. Histology confirmed the diagnosis of Cutis Laxa. The underlying genetic cause is still unknown. This patient represents an ideal candidate for next-generation sequencing analysis, and whole-exome sequencing is our next goal. The case highlights the inherent limitations in single-gene sequencing approach and the ability of next-generation techniques to provide crucial leads in perplexing cases, tapping previously hidden genomic data toward new genetic diagnoses.

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MISSING THE MARK? INTESTINAL PARASITE PREVALENCE IN INFORMAL SETTLEMENTS IN LIMA, PERU

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Purpose of Study: The WHO suggests large scale preventive chemotherapies for soil-transmitted helminthiasis (STH) for communities in which prevalence is estimated to be greater than 20%. WHO lists Peru as a country potentially needing STH prevention. Prevalence varies widely regionally and remains understudied in the newest urban informal settlements in Lima.

Objective: (a) Determine the STH prevalence in the new informal settlements in Lima, Peru
(b) Estimate seasonal (summer/winter) variation in the prevalence of STH in 3 informal settlements in Lima, Peru.

Methods Used: Children aged 1 to 10 were recruited from 3 new informal settlements in Lima Peru. After informed consent, participants were interviewed, weighed and measured. Stool samples were examined using macroscopic examination, direct microscopic examination, technique of spontaneous sedimentation in tube, and Baermann technique. We attempted to obtain 2 stool samples from each participant during 2 seasons (summer/winter).

Summary of Results: The prevalence of STH and pathogenic protozoa did not differ between summer and winter (p>0.05). Only 3% (summer) to 9% (winter) of study participants had STH. The prevalence of pathogenic protozoa was 25% during the summer and 17% during the winter. Approximately 1% (summer) and 5% (winter) of participants had both pathogenic protozoa and STH.

Conclusions: Even using the broadest definition of STH, the prevalence in this study is considerably lower than the WHO recommended threshold (20%) for large scale preventive chemotherapy for STH. Although pathogenic protozoa prevalence was higher than STH prevalence, recommended empirical treatment for STH by groups active in these newest invasion settlements is not supported by the data and could contribute to unnecessary medication exposures and poor resource allocation.

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A CURIOUS CASE OF CUTIS LAXA WITH HOLOPROSENCEPHALY: COULD A SEEMINGLY INNOCUOUS INTRON DELETION BE THE CULPRIT?

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Purpose of Study: We report a new syndrome of cutis laxa, alobar holoprosencephaly, and cerebellar agenesis observed in a baby girl born at term. Microarray CGH analysis revealed a small deletion in intron 3 of Neuregulin 3 gene (NRG3). The deletion was confirmed to be present in the patient but not the mother. Mutations in NRG3 gene have been associated with cognitive impairment. Additionally, transgenic mice overexpressing NRG3 in the skin
exhibited a cutis laxa phenotype. While there is evidence supporting the pathogenicity of the intronic deletion, the mechanism is unknown. The purpose of the study is to establish the clinical significance of the intronic deletion, with the end goal of providing informed genetic consultation for the patient.

**Methods Used:** Using immunohistochemistry, we conducted skin pathohistological studies to confirm the clinically observed cutis laxa. To investigate whether the deleted intronic DNA has a role in modulating NRG3 expression, we examined the known transcription factor binding sites within the deleted intron and designed in vitro experiments with luciferase reporter assay. We also directly reprogrammed the patient's skin fibroblasts to neuroblasts and investigated expression levels of endogenous NRG3.

**Summary of Results:** Abnormal wrinkling and altered elastin and beta-integrin was found in the patient's skin. Remarkably, the findings resemble the histology of the adult skin. Our preliminary in vitro data suggested that 3 regions within the deleted intron exhibited moderate repressive property and 1 region, potent repressive property. Preliminary expression study in the reprogrammed neuroblasts revealed abnormal NRG3 expression.

**Conclusions:** Our pathohistological study confirmed skin developmental anomaly in our patient and suggested premature skin aging. Our in vitro data demonstrated that the deleted intronic DNA may have a role in regulating NRG3 expression. Interestingly, MYC may have a repressor function in NRG3 expression. Additional studies will help elucidate the mechanisms which led to the remarkable anomaly in our patient, and even more importantly, offer closure to the patient's mother.

**Adult Clinical Case**

**Joint Plenary Poster Session and Reception**

5:00 PM Thursday, February 26, 2015

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**PROMETHAZINE TOXICITY MISDIAGNOSED AS MENINGOENCEPHALITIS**

Ali E, Mazeik H, Mohamed A, Baser K, Mulkey Z. Texas Tech University Health Science Center/Lubbock, Lubbock, TX.

**Introduction:** Promethazine is a commonly used medication in practice, but toxicity is rare with only a few cases of intoxication reported in the literature. We report a misdiagnosed case initially treated as meningencephalitis due to overlap of findings.

**Case description:** A 62-year-old man was transferred from outside facility with confusion, fever, and jerking movements. A lumbar puncture had been done, and antibiotics had been started. Upon arrival he was agitated and had dysarthria and visual hallucinations. His blood pressure was 89/72 mmHg, pulse 98 beats/minute, respiratory rate 22/minute, and temperature 101°F. The examination demonstrated disorientation, dilated pupils, dysmetria, truncal ataxia, positive Babinski's sign, and nystagmus. Lab work revealed an elevated creatine kinase and leukocytosis which were all normal. Drug screen was negative; as was a hepatitis C virus, CMV, and had normal glucose, protein, and cell counts. Given the rapid resolution of his symptoms and exclusion of infection we concluded that the diagnosis was promethazine overdose.

**Discussion:** Promethazine intoxication may result in wide range of symptoms, including CNS depression, agitation, or delirium, and anticholinergic findings, such as dilated pupils, dry mouth, fever, seizures, and coma. Treatment is supportive.

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**CRYOglobulinemia as an Idiosyncratic Drug Reaction of Moringa oleifera**


**Case Report:** A 43 year old Caucasian male with treatment-naive Hepatitis C presented for evaluation of a rash of four days duration. Two days prior to onset he started an herbal medication, Moringa oleifera, for headache. The rash was a palpable purpura with scant pus-filled blisters that densely populated the ankles. The trunk, face, and genitals were spared. There was no mucosal involvement. He denied constitutional symptoms, arthralgias and tick bites. Previous drug screen was positive for oxycodone.

He had a mild peripheral eosinophilia and elevated inflammatory markers. Biochemistry, urinalysis, coagulation profile and prothrombin screen were all unremarkable. Infectious work-up for HIV, CMV, syphilis and Lyme disease were negative. ANA, ANCA's and Rheumatoid factor (RF) titers were negative, and C3/C4 were normal. Hepatitis C antibody was positive but viral load was undetectable. Cryoglobulins were elevated at 8%, with immunofixation electrophoresis revealing a type III cryoglobulinemia. Skin biopsy showed leukocytoclastic vasculitis.

He was treated with clindamycin for the cellulitis. No specific treatment was instituted for the cryoglobulinemia and the rash resolved independently.

Cryoglobulinemia refers to the presence in the serum of single or mixed immunoglobulins, which precipitate at temperatures below 37°C and re-dissolve on re-warming, with types II and III representing mixed cryoglobulinemia (MC).

The triad of MC is purpura, weakness and arthralgia. Orthostatic purpura ranges from petechiae to severe cutaneous lesions which are attributed to the deposition of circulating immune complexes on vessel walls. Leukocytoclastic vasculitis is the histopathological hallmark of MC. Hepatitis C virus (HCV) exerts a chronic stimulus on the immune system kindling the production of monoclonal IgM RF which is implicated in the development of cryoprecipitate immune complexes, along with complement.

Considering the undetectable viral load, negative RF titers and normal complement levels, it is unlikely that HCV was responsible for the precipitation of MC in this case. As the introduction of Moringa oleifera coincided with disease onset, it postulates that this may be an idiosyncratic drug reaction and that alternate immune stimuli may influence the structure of circulating immune-complexes responsible for MC.
anemia and thrombocytopenia. There are association between solid tumors and TTP/HUS. Prostate cancer and TTP/HUS. The diagnosis of thrombotic thrombocytopenic purpura and hemolytic uremic syndrome (TTP/HUS) is made clinically and requires only thrombocytopenia and microangiopathic hemolytic anemia without other clinically apparent etiology. Idiopathic TTP/HUS is a medical emergency that is almost always fatal if appropriate treatment is not initiated promptly. Treatment should involve plasma pheresis and may add steroid and immunotherapy.

The ESР was elevated at (60-80 mm/hr), ANA , Dasna, SSA/SSB were strongly positive and RF, CCP were strongly positive. A 3 mm punch biopsy was done on her elbow and knee which showed lymphocytes and neutrophils cutting the blood vessel in mid and upper dermis; no frank leukocytoclastic vasculitis was seen. The patient was started on oral prednisone 40 mg daily with tapering to 10 mg daily with almost complete resolution of her skin lesions. Because of persistent synovitis, she was switched from methotrexate to azathioprine.

Summary of Results: as below

Conclusions: Rheumatoid arthritis may affect skin and its common manifestations are rheumatoid nodules, Bywater lesions, pyoderma gangrenosum, the true incidence of adult onset Still disease and vasculitis is unknown. Both disease manifestations include cryoglobulinemia, as well as rheumatoid neutrophilic dermatitis. Rheumatoid neutrophilic dermatitis is a prototype of sweet syndrome (neutrophilic vascular reaction). Histology shows dense dermal neutrophilic infiltrate with or without vasculitis, there may be dermal papillae with microabscess; the infiltrate may extend to the penile apex. Therapy consist of topical or systemic glucocorticoids, sulfamethoxazole and trimethoprim (100 to 200 mg/day), and hydroxychloroquine. In resistance case steroid sparing agents azathioprine , mycophenolate or cyclophosphamide may be tried.

A CASE OF RHEUMATOID NEUTROPHILIC DERMATOSIS

Bhawal J. UMMC, Jackson MS, MS.

Purpose of Study: Rheumatoid Neutrophilic dermatitis (RND) is cutaneous manifestation of RA, It can se in patients who are both positive and negative for RF and common in middle age females, it usually Present as vesiculopustular, plaques and nodules.

Methods Used: 37 y/o AAF with PMH of RA diagnosed in 2007 severe arthralgia and synovitis. In 2010 she developed a vesicular rash on her right; hydrocortisone and Benadryl were prescribed which did not help. Later it worsened and she developed sever itching with a burning sensation which disseminated to her b/l arms and legs. On physical examination, firm nodules 3 to 5 mm in size were observed over her both elbows; these lesions were erythematous papules and plaques, some in an annular configuration, with central umbilation over extensor arms and knees. In addition she had many of hyperpigmented lesions over her entire body. MCPS showed synovitis and she was unable to make fist. The nails and mouth were clear of lesions.

The esr was elevated at 60-80 mm/hr, ANA, DPNA, SSA/SSB were strongly positive and RF, CCP were strongly positive. A 3 mm punch biopsy was done on her elbow and knee which showed lymphocytes and neutrophils cutting the blood vessel in mid and upper dermis; no frank leukocytoclastic vasculitis was seen. The patient was started on oral prednisone 40 mg daily with tapering to 10 mg daily with almost complete resolution of her skin lesions. Because of persistent synovitis, she was switched from methotrexate to azathioprine.

Summary of Results: as below

Conclusions: Rheumatoid arthritis may affect skin and its common manifestations are rheumatoid nodules, Bywater lesions, pyoderma gangrenosum, the true incidence of adult onset Still disease and vasculitis is unknown. Both disease manifestations include cryoglobulinemia, as well as rheumatoid neutrophilic dermatitis. Rheumatoid neutrophilic dermatitis is a prototype of sweet syndrome (neutrophilic vascular reaction). Histology shows dense dermal neutrophilic infiltrate with or without vasculitis, there may be dermal papillae with microabscess; the infiltrate may extend to the penile apex. Therapy consist of topical or systemic glucocorticoids, sulfamethoxazole and trimethoprim (100 to 200 mg/day), and hydroxychloroquine. In resistance case steroid sparing agents azathioprine, mycophenolate or cyclophosphamide may be tried.
Among causes of pneumonia, CEP must be included in the differential diagnosis, particularly when there is a history of autoimmune disease. We present a case of CEP diagnosed in the setting of undifferentiated connective tissue disease (UCTD).

A 71-year-old male with a history of UCTD and common variable immunodeficiency on monthly intravenous-immunglobulin (IVIG) infusions, presented to the emergency department with episodic fever and dyspnea over the previous three months. Of note, patient missed past month’s IVIG treatment due to travel to California. He was hospitalized at an outside medical center the previous week, where he was treated with IV antibiotics for community-acquired pneumonia (CAP) and discharged with mild symptomatic improvement. In the following days, his symptoms急性 worsened. On exam, he had labored breathing, ronchi in the right middle lobe of lung, and a 2+ pitting edema in legs bilaterally. Vancomycin and piperacillin-tazobactam were started prophylactically for broad spectrum antibacterial coverage. Spiral CT ruled out pulmonary embolism. Bronchoscopy revealed an elevated eosinophil count (28%). Patient had recent negative work up for Churg-Strauss. Drug-induced and fungal causes were excluded with appropriate tests. Despite the lack of peripheral eosinophilia, CEP was diagnosed given duration of illness. Labs for serum IgG to Strongyloides, ELISA to Giardia antigen, and stool parasites came back negative. Following initiation of steroid treatment, patient showed drastic improvement in presenting symptoms.

While the exact pathophysiologic mechanism is not definitively established, it has been shown the production of proinflammatory cytokines in auto-immune disease can induce infiltration of eosinophils into various tissues. Absence of peripheral eosinophilia is unusual in CEP; however it can be present in up to 10-20% of cases. Steroid treatment should not begin for CEP until parasitic infection is unequivocally excluded to avoid potentially life-threatening steroid induced hyperparasitic infection.

GLOBAL STRESS-INDUCED CARDIOMYOPATHY WITH LEFT AND RIGHT VENTRICULAR DYSFUNCTION IN THE SETTING OF SEVERE CHRONIC OBSTRUCTIVE PULMONARY DISEASE (COPD) EXACERBATION

Hritani A, Dakkak M, Baxi K, Nakshabendi R, Paterick T. University of Florida-Jacksonville, Jacksonville, FL.

Introduction: Stress-induced Cardiomyopathy (SIC) is defined as a reversible, stress-induced left ventricular (LV) regional hypokinesis, or akinesis, in the absence of coronary artery disease (CAD). An interesting and rarely reported phenomenon is global SIC with right ventricular (RV) involvement. Here, we present a case of global LV and RV SIC in the setting of acute hypercapnic respiratory failure from COPD exacerbation.

Case description: A 70 year old male with a PMH of COPD on home oxygen presented after he was found in tripped position with empty nebulizer. Patient was intubated and brought to emergency department, where he was found to be hypotensive with bilateral diffuse inspiratory and expiratory wheezes on exam. Initial Electrocardiogram revealed 2 mm ST-elevation in V3-V4, which subsequently resolved. Troponin I was elevated. The patient received intravenous fluids, norepinephrine, and antibiotics. Transhoracic echocardiogram revealed severe global LV and RV dysfunction. The left ventricular ejection fraction (LVEF) was 15% and there was an akinetic RV.

He was treated for COPD exacerbation, as well as type II non-Segment elevation myocardial infarction with heparin. The patient was weaned off pressers and extubated on day 3. Cardiac catheterization revealed non-obstructive CAD. Repeat echo on day 5 revealed LVEF of 65% and normal RV function. The patient was discharged with the diagnosis of global SIC in the setting of COPD exacerbation.

Discussion: This case represents global biventricular SIC secondary to COPD exacerbation. The pathophysiology of SIC is thought to be due to coronary vasospasm secondary to catecholamine excess, or coronary microvascular dysfunction. RV involvement has been reported in patients with lower LVEF. Since SIC is regarded as a myocardial stunning-like phenomenon, patients with biventricular involvement appear to have had a more severe inciting event. Connection of COPD exacerbation and SIC may be due to exaggerated sympathetic activation, and the use of sympathomimetic bronchodilators. Additionally, the increased adrenergic activity from severe hypoxia, hypercapnea, use of beta agonists, and acidosis all increase the risk of SIC.

MULTIPLE BILATERAL KIDNEY INFARCTS AS INITIAL PRESENTATION OF PAROXYSMAL ATRIAL FIBRILLATION

Hritani A, Baidoun F, Poenaria A. University of Florida-Jacksonville, Jacksonville, FL.

Introduction: Acute renal infarction is an uncommon cause of abdominal pain and is under-diagnosed as a cause of kidney dysfunction. Early diagnosis is important, since renal function can be restored with revascularization of an occluded vessel. Bilateral renal infarctions secondary to atrial fibrillation is a rare thromboembolic complication and only one reported case in the literature was found.

Case description: A 71 year old caucasian female with a past medical history of hypertension and peripheral arterial disease presented to the hospital complaining of right-sided flank pain and hematuria.

On presentation her blood pressure was 165/77 mmHg and her cardiac exam revealed a regular rate and rhythm; she had bilateral costovertebral angle tenderness. The rest of her exam was not significant. Her urinalysis showed microscopic hematuria and a serum creatinine of 1.26 mg/dl up from her baseline of 0.72 mg/dl. Her initial Electrocardiogram (EKG) showed unchanged left bundle branch block bundle block with normal sinus rhythm. A CT angiogram of the chest and abdomen showed no aortic pathology and bilateral kidney infarcts.

On the second day of hospitalization her telemetry monitor showed Atrial fibrillation confirmed by a stat EKG. A diagnosis of paroxysmal Atrial fibrillation was made and the patient was started on anticoagulation in addition to beta blockers for heart rate control.

Discussion: The renal infarction is a rare disease, difficult to diagnose because the symptoms are often misleading. Mechanisms of renal infarction are multiple and varied. The embolic pathogenesis involves the occlusion of renal artery or branch vessels by cholesterol or blood clots. Imaging is the key to diagnose renal artery occlusion. The CT scan has a good sensitivity about 85% mainly with reconstruction thickness of 5 mm. Treatment guidelines for renal infarction are yet to be established but anticoagulation is being employed so far.

Conclusion: The renal circulation is considered the final resting place for up to 2.3% of systemic arterial embolizations making bilateral renal infarctions a rare thromboembolic complication. But because of the necessity for prompt treatment interventions it should always be on the differential diagnosis list for patients presenting with flank pain and hematuria.

PRIMARY EWING’S SARCOMA/PRIMITIVE NEUROECTODERMAL TUMOR OF THE PENIS: AN UNUSUAL PRESENTATION

Hsu J, Hamilton R. University of Mississippi, Jackson, MS.

Case Report: 22 year old white male with history of anxiety and seizure disorder presented with a one week history of left inguinal and testicular pain. He was treated for epididymitis after a negative scrotal ultrasound. He presented one month later with fatigue, low-grade fevers, malaise, and shortness of breath without weight loss or night sweats.

CT chest/abdomen/pelvis showed a right-sided pleural effusion with multiple bilateral nodules and a cm heterogeneously enhancing inguinal canal mass indistinguishable from the penis with normal appearing testicles.

A needle core biopsy was obtained which showed a small round blue cell tumor that was CD3, CD30, LCA, Cam 5.2, Desmin, S-100, CD20, NSE, CD56, WT-1, Myogenin, and Cytokeratin AE1/AE3 negative. Immunostains were CD99 and synaptophysin positive. EWSR1-FLI1 fusion was detected by reverse transcriptase-polymerase chain reaction which confirmed the diagnosis of Ewing’s Sarcoma.

Patient was treated with vincristine 1.4 mg/m2 on Day 1, doxorubicin 25 mg/m2 on days 1 and 2, cyclophosphamide 1200 mg/m2 on day 1, and mesna 500mg IV q6h x 48 hours in 21 day cycles. Patient completed the first
HEMOBILIA: A RARE CAUSE OF MELENA

Joglekar KP, Loveless CA. Georgia Regents University, Augusta, GA.

Case Report: A 63-year-old African American male with past medical history of hypertension, ESRD and esophageal adenocarcinoma with liver metastasis presented with 2 episodes of large bloody bowel movements. He had undergone a CT guided percutaneous liver biopsy of the metastatic liver lesions 4 weeks prior to presentation without any complications.

His physical exam revealed right upper quadrant tenderness. There were no signs of portal venous hypertension. Digital rectal exam revealed melena in the rectal vault. His hemoglobin (Hb) on presentation was 11g/dL, which was normal. Laboratory testing revealed leukocytosis and an erythrocyte sedimentation rate of 65 mm/hr. Three blood cultures were negative for aerobic and anaerobic bacteria. Fungal stains were negative as was a serum HIV test. Three sputum samples were negative for AFB. The clavicular mass was then biopsied and showed similar results to the paraspinal mass. The patient was dx’d with skeletal TB and was started on rifampin, isoniazid, pyrazinamide and ethambutol and his symptoms gradually improved.

Discussion: Although global TB incidence has decreased steadily by 2% per year for the past decade, it remains a significant public health problem, particularly in underdeveloped countries. Extrapulmonary TB can affect many tissues but 30% of the time it will be a skeletal site. This case is noteworthy for its presentation and patient population.

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WHEN APPARENT METASTASES ARE NOT DISTURBING

Lemley RJ, Singh M, Smalligan RD. Texas Tech University HSC, Amarillo, TX.

Case Report: A 35yo previously healthy African American male presented with a 2-month hx of progressive back pain. Two weeks prior he had noticed tingling in his right leg that progressed to weakness in the leg. He had also noticed a nontender right clavicular mass for one week. He denied fever, sweats, chills, night sweats and cough though he had noticed loss of appetite and an 11 pound weight loss. The day of admission he suddenly could not walk due to profound weakness in both legs. PMH, FH, P/S unremarkable except for hx of incarceration and no recent drug use. Physical exam nontender mass palpable on the medial aspect of R clavicle, clear lungs, heart regular, abd normal and neuro exam revealed increased patellar DTRs and bilateral positive Babinski signs. CT of the spine showed a 3x3cm mass anterior to T8 with bony destruction and spinal canal invasion. CT chest showed osteolytic lesions in the R clavicle, multiple ribs, hilar adenopathy, and some small lung nodules. Hospital course: Emergent surgery for excision of the spinal mass showed caseous necrosis without malignancy; AFB and fungal stains were negative as was a serum HIV test. Three sputum samples were negative for AFB. The clavicular mass was then biopsied and showed similar results to the paraspinal mass. The patient was dx’d with skeletal TB and was started on rifampin, isoniazid, pyrazinamide and ethambutol and his symptoms gradually improved.

Discussion: Although global TB incidence has decreased steadily by 2% per year for the past decade, it remains a significant public health problem, particularly in underdeveloped countries. Extrapulmonary TB can affect many tissues but 30% of the time it will be a skeletal site. This case is noteworthy for its presentation and patient population.

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SPLENOMAS MASCARADER OF METASTATIC DISEASE

Mazek H, Verma R, Mohamed A, Ali E, Hosford S, Cobos E. Texas Tech University Health Science Center, Lubbock, TX.

Purpose of Study: Splenosis is a heterogeneous autotransplantation of splenic tissue usually following splenectomy and traumatic rupture of spleen. They
The Artery of Percheron (AOP) is a small perforating artery.

Conclusions: Splenules are incidental findings of little clinical significance in most patients. However, they must be properly identified in various situations, such as: differentiating them from metastatic lesions, torsion, infarction, endometriosis, mesenchymal tumors and peritoneal mesothelioma. These differential diagnoses can be differentiated using clinical history and radiological appearance. On noncontract CT, the attenuation of splenules is comparable to that of the spleen. However, small splenules (<1 cm) may be hypodense and if calcified as in our patient can be mistaken for metastatic disease. Fused SPECT/CT imaging with Tc-99m labeled denatured red blood cells facilitates the definitive diagnosis of a splenule and may also help in identifying splenules that have relocated within the abdominal cavity.

Methods Used: Case analysis and literature review.

Summary of Results: A 69-year-old man underwent coronary angiography for evaluation of chest pain. During the procedure, he received 2 mg of midazolam and 1 mg of hydromorphone. The patient did not wake up at the end of the procedure and remained minimally responsive. The patient withdrew from painful stimulation but had no spontaneous movements. One dose of naloxone 0.4 mg intravenous did not change his clinical examination. Immediate neurology consult was done, his pupils (left-3 mm, right-2 mm) were minimally reactive to light and that he had left medial rectus palsy. His reflexes were normal and there were no other focal deficits. His immediate CT scan of brain showed no acute hemorrhage or infarction. MRI of the brain confirmed bilateral medial thalamic and left midbrain infarctions consistent with an artery of Percheron infarction. The patient didn’t receive TPA because of an arterial puncture at a non-compressible site within 7 days. Although stroke is uncommon, we should all monitor patients closely throughout the procedure.

One of exclusion criteria for use of TPA is arterial puncture at a non-compressible site within 7 days. Although stroke is uncommon, we should all monitor patients closely throughout the procedure.

Case: A 36-year-old female with metastatic, triple positive, invasive ductal carcinoma of the right breast was admitted to our Hospital in June 2014. She was originally diagnosed 18 months back with lymph node and osseous metastasis and four cycles of weekly gemcitabine, paclitaxel, and trastuzumab, along with monthly denosumab treatment, resulting in a complete response. Patient continued on maintenance tamoxifen, trastuzumab and denosumab after confirming a normal left ventricular ejection fraction (LVEF). In March 2014 she was found with generalized lymphadenopathy indicative of progressive disease. Patient refused cytotoxic therapy and pertuzumab was added to her trastuzumab regimen. In June 2014 she developed severe dyspnea and an echocardiogram confirmed the presence of a large pericardial effusion with a LVEF of 40%. Patient underwent pericardiectomy with marked clinical improvement. Pericardial fluid was negative for malignant cells.

Discussion: Pericardial effusion is a consequence of inflammation of the pericardium due to neoplastic agents such as doxorubicin, cyclophosphamide, and trastuzumab. Pertuzumab is a monoclonal antibody that selectively binds to the human epidermal growth factor receptor-2 protein (HER-2) inhibiting its dimerization and subsequent activation. Pertuzumab has been approved by the FDA in combination with trastuzumab (a monoclonal antibody that also inhibits HER-2 activation), and docetaxel as first-line treatment for metastatic HER-2-expressing breast cancer. Since HER-2 is involved in the growth and survival of adult cardiomyocytes treatment with trastuzumab may result in a reversible decrease in LVEF in up to 20% of patients. Our patient’s new-onset pericardial effusion developed only after pertuzumab was added to trastuzumab. Thus, our case report illustrates the importance of close follow-up and monitoring of patients receiving dual anti-HER-2 therapy for early detection of this life threatening complication.

My Patient Can’t Talk After the Cath

Maze H1, Edlaiw M2, Mohamed A1, Ali E1, Nugent K1, Meyerrose G1

Texas Tech University Health Science Center, Lubbock, TX

Purpose of Study: Coronary angiography is generally considered safe procedure. However, it can cause complications. One uncommon complication is a stroke. Periprocedural strokes often occur during or immediately after the procedure when the femoral artery sheath is still in place, but the diagnosis can be delayed up to 36 hours in some cases. Periprocedural stroke occurs in 0.03% to 0.3% of diagnostic procedures and 0.3-0.4% of percutaneous coronary interventions.

Methods Used: Case analysis and literature review.

Summary of Results: A 69-year-old man underwent coronary angiography for evaluation of chest pain. During the procedure, he received 2 mg of midazolam and 1 mg of hydromorphone. The patient did not wake up at the end of the procedure and remained minimally responsive. The patient withdrew from painful stimulation but had no spontaneous movements. One dose of naloxone 0.4 mg intravenous did not change his clinical examination. Immediate neurology consult was done, his pupils (left-3 mm, right-2 mm) were minimally reactive to light and that he had left medial rectus palsy. His reflexes were normal and there were no other focal deficits. His immediate CT scan of brain showed no acute hemorrhage or infarction. MRI of the brain confirmed bilateral medial thalamic and left midbrain infarctions consistent with an artery of Percheron infarction. The patient didn’t receive TPA because of an arterial puncture at a non-compressible site within the last 7 days. The patient had slow improvement in his cognitive function.

Conclusions: The Artery of Percheron (AOP) is a small perforating artery supplying paramedian thalamus and mid-brain. AOP infarction is rare and typically has a triad of altered mental status, vertical gaze palsy, and memory impairment. In the case of intraprocedural stroke, the major advantages of performing a cerebral angiogram is that the diagnosis of embolic stroke can be confirmed and treatment can be given immediately. The occurrence of postprocedural stroke requires urgent cerebral imaging (well before 4.5 hours have elapsed) to confirm an ischemic cause and to plan subsequent treatment.

Newer Drugs, Better Treatment Options Come With a Price

Maze H1, Verma R, Mohamed A, Ali E, Figueroa J

Texas Tech University Health Science Center, Lubbock, TX

Case Report: Introduction:

Pericardial effusion is an excessive accumulation of fluid in the pericardial space that results as a complication of malignancy and/or antinecancer therapy. We report a case of a patient who developed a non-malignant peri-cardial effusion after receiving treatment with the combination of pertuzumab with trastuzumab.

Case: A 36-year-old female with metastatic, triple positive, invasive ductal carcinoma of the right breast was admitted to our Hospital in June 2014. She was originally diagnosed 18 months back with lymph node and osseous metastasis and four cycles of weekly gemcitabine, paclitaxel, and trastuzumab, along with monthly denosumab treatment, resulting in a complete response. Patient continued on maintenance tamoxifen, trastuzumab and denosumab after confirming a normal left ventricular ejection fraction (LVEF). In March 2014 she was found with generalized lymphadenopathy indicative of progressive disease. Patient refused cytotoxic therapy and pertuzumab was added to her trastuzumab regimen. In June 2014 she developed severe dyspnea and an echocardiogram confirmed the presence of a large pericardial effusion with a LVEF of 40%. Patient underwent pericardiectomy with marked clinical improvement. Pericardial fluid was negative for malignant cells.

Discussion: Pericardial effusion is a consequence of inflammation of the pericardium due to neoplastic agents such as doxorubicin, cyclophosphamide, and trastuzumab. Pertuzumab is a monoclonal antibody that selectively binds to the human epidermal growth factor receptor-2 protein (HER-2) inhibiting its dimerization and subsequent activation. Pertuzumab has been approved by the FDA in combination with trastuzumab (a monoclonal antibody that also inhibits HER-2 activation), and docetaxel as first-line treatment for metastatic HER-2-expressing breast cancer. Since HER-2 is involved in the growth and survival of adult cardiomyocytes treatment with trastuzumab may result in a reversible decrease in LVEF in up to 20% of patients. Our patient’s new-onset pericardial effusion developed only after pertuzumab was added to trastuzumab. Thus, our case report illustrates the importance of close follow-up and monitoring of patients receiving dual anti-HER-2 therapy for early detection of this life threatening complication.

Approaching Patient with Chronic Total Occlusion of Coronary Artery Using a 5 French Catheter Sheath Radially

Maze H1, Sherif K1, Ali E1, Mohamed A1, Fenere M2, Suarez J1

Texas Tech University Health Science Center, Lubbock, TX

Case Report: Introduction:

Chronic total occlusion (CTO) intervention of coronary artery is both complex and time consuming, but has been associated with better long term outcomes. Limited techniques are available for CTO’s of coronary arteries including LASER atherectomy, rotational atherectomy devices and re-entry catheters. Most of these approaches utilize femoral approach. We present a case of CTO approached with LASER endovascular intervention (EVI) by radial artery approach using a 5 French sheath.

Case: A 57-year-old man presented with retrosternal chest pain for two days. Patient has history of coronary angiography two years before this presentation that showed diffuse right coronary disease at that time. Examination was normal at time of presentation. EKG showed no acute change. Cardiac enzymes...
within normal limit. Patient had myocardial perfusion stress test that revealed inferior wall ischemia. Decision was made to perform coronary angiography. Access in right radial artery was achieved by modified Seldinger technique and a 5-French vascular sheath was placed. Coronary artery angiography showed right coronary artery (RCA) CTO. 5-French JR4 guide catheter successfully engaged RCA. We were unable to advance balloon for angioplasty and at that time decision to perform Eximer Laser Coronary Angioplasty was made. Laser angioplasty was performed across the CTO into the distal RCA; a marked improvement of flow was evident thereafter and demonstrated significantly excellent results with TIMI III flow in RCA.

Discussion: To best of our knowledge this is the first case report describing significantly excellent results with TIMI III flow in RCA. To our knowledge this is the first case report describing significantly excellent results with TIMI III flow in RCA.

Methods Used: To best of our knowledge this is the first case report describing significantly excellent results with TIMI III flow in RCA. The patient was hemodynamically stable. She was given heparin, aspirin, and clopidogrel. The CTO was crossed with a 4.0 mm coronary guidewire. The guidewire was exchanged for a 6 French long guidewire. This approach shows that smaller sheath sizes can be utilized despite the procedural complexities that come with LASER EVI in coronary arterial system. There are two rationales to use 5 Fr system guiding catheter. First, is to reduce amount of contrast used during the procedure. This is an important strategy in patients with chronic kidney disease to reduce risk of acute kidney injury in those patients. Second reason is the favorable impact of using 5 Fr guiding catheter on vascular complication.

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DYSPHAGIA IN PATIENT WITH BULLOUS PEMPHIGOID
Mohamed A, Mazek H, Panikkath D, Rakvit A, Klonjji S. Texas Tech University Health Science Center, Lubbock, TX.
Purpose of Study: Bullous pemphigoid is an acquired autoimmune skin disease that predominantly affects elderly. It is characterized by the presence of blisters resulting from defect of the adhesion of the basal epidermal layer with the basement membrane. It affects any part of the skin. Mucus membranes are rarely affected. Esophageal involvement has only been reported in a few cases.

Methods Used: case analysis and literature review.

Summary of Results: 56 Years old farmer male with medical history of Bullous Pemphigoid diagnosed three years ago presented with progressive dysphagia to solid food and odynophagia for one year. The bullous lesions involved his upper and lower extremities and were worsen with friction from work in his farm. He also had mouth ulcers, but denies any genital or anorectal lesions. No weight loss. His medications were Prednisone, Colchex, and Methotrexate. Skin exam revealed multiple widespread, symmetrically distributed Bullous lesions involving his upper and lower extremities. Complete blood count, and complete metabolic panel were unremarkable. Esophagogram showed esophageal stricture. Upper endoscopy showed esophageal stricture extending from 25-40 cm from the incisors. Ballon dilation was performed. Pathology showed evidence of acute and chronic inflammation. Patient was started on Proton Pump inhibitor and reported improvement of symptoms.

Conclusions: Bullous pemphigoid is an autoimmune disease associated with circulating antibodies against hemidesmosomes that leads to disruption of the adhesive material which leads to intraepithelial bullous formation. It is predominantly affects the skin. In contrast to Pemphigus vulgaris, mucosal involvement is rare and extra oral involvement is exceptional. Our case is interesting as the disease presents with severe esophageal involvement with subsequent dysphagia. Esophagoeal involvement has been only reported in few cases. Our patient has severe esophageal disease with very long strictures. This case highlights the importance of considering esophageal disease in those patients with suggestive symptoms. Early diagnosis could reduce the risk of esophageal strictures development.

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IS IT REALLY A MALIGNANCY? A CASE OF CHRONIC PANCREATITIS WITH VITAMIN D DEFICIENCY MASQUADING AS MALIGNANCY
Mohamed A, Verma R, Mazek H, Tijiani L, Rakvit A. Texas Tech University Health Science Center, Lubbock, TX.

Purpose of Study: Clinicians should know the causes of incidental bone lesions. The most common differential for patients with bone lesions is malignancy. Other possible causes include vitamin D deficiency.

Methods Used: case analysis and literature review.

Summary of Results: 49-year-old African-American inmate with no significant medical history presented to our hospital with the diagnosis of possible metastatic cancer after bone scan showed osteoblastic lesions in the humeri and ribs. The patient had significant weight loss, night sweats, generalized weakness, and bone pain for one year. He also had rib fractures secondary to simple falls. He reports a history of heavy alcohol abuse before he was incarcerated in prison in 1995. On physical exam he looked cachectic and anemic. He had chest wall tenderness with palpation over the ribs. He had no abdominal organomegaly. Rectal exam showed a normal prostate with no nodules. His admission labs were remarkable for calcium 5.9 mg/dl and vitamin D level < 5 ng/ml. His HIV serology was negative. Repeated bone scan shows pathologic radiotracer uptake in the ribs and pubic bones consistent with healing fractures. Malignancy work up, including CT scans of the chest, abdomen and pelvis, was unremarkable. Tumor markers including PSA were negative. The pancreas was atrophic in the abdominal CT scan. On further questioning, patient gave history of chronic abdominal pain and diarehal episodes. His PTH was 1590 pg/ml; a parathyroid sestamibi scan was negative. It was concluded that his clinical presentation was secondary to severe pancreateitis with malabsorption and severe vitamin D deficiency leading to brown tumors of the bone with pathological fractures. The patient then was started on pancreatic enzymes replacement, vitamin D, and calcium supplementation.

Conclusions: We are reporting a rare presentation of chronic pancreatitis. Clinical manifestation, such bone pain, fractures, weight loss, and bone lesions in bone scan, could suggest malignancy. Clinician should be alert to the late presentations of chronic pancreatitis causing vitamin D deficiency with subsequent secondary hyperparathyroidism leading to brown tumors. These patients need simple lab tests, such as vitamin D levels, and, if low, then PTH levels.

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DYSPNEA IN A COPD PATIENT - NOT ALWAYS AN EXACERBATION
Panikkath D, Panikkath R, Gadwala S, Edris H, Nugent K. Texas Tech University Health Sciences, Lubbock, TX.

Case Report: Inhaled B2 agonists is the first line therapy in COPD exacerbation. Repetitive or continuous administration of these agents reverse airflow obstruction. Below we describe a patient who presented with acute exacerbation of COPD and had worsening dyspnea despite appropriate treatment with beta agonists. A 53-year-old woman with COPD presented in respiratory distress of 4 days duration. She had taken multiple albuterol inhalers and nebulization treatments at home with no relief. She was febrile, dyspneic, tachycardic, and wheezy. Initial lab work was unremarkable except for significant lactate of 7.2 mmol/L. She was given multiple albuterol treatments with fluid boluses. Her dyspnea however did not improve even though her wheezing resolved and she did not need increased oxygen requirement. Lactate levels rechecked 4 hours later was 8.5 mmol/L. ABG drawn showed mixed respiratory alkalosis and metabolic acidosis. This was inconsistent with the possible explanation of worsening of COPD as the cause of dyspnea in which case she would have shown primarily respiratory acidosis. Beta agonists were then suspected as the possible cause of elevated lactate levels. We believe that increase in lactate level was due to inhaled β-adrenergic agonists. There are several case reports that suggest high lactate concentrations can develop during the inhaled β agonist treatment. Several mechanisms might explain the lactacidemia, stimulation of β-adrenergic receptors causes increase in adenylate cyclase activity, increased lipolysis; increased free fatty acids, which inhibit conversion of pyruvate to lactate. As a result, lactate increases in lactic acid; and increased glycogenolysis and gluconeogenesis. It is unclear as to why only few patients treated with β-agonists develop lactic acidosis. Lactic acidosis due to its metabolic consequences can increase the sensation of dyspnea and compensatory hyperventilation. This could be easily misinterpreted as a sign of treatment failure and lead to inappropriate intensification of treatment. Any discrepancy between exacerbation of dyspnea and improvement of bronchospasm when using beta agonists should alert us to possible lactic acidosis. Serial blood gas analyses should detect evolving lactic acidosis.
RUPTURED SINUS OF VALSALVA ANEURYSM: AN UNUSUAL CAUSE OF ACUTE HEART FAILURE

Perez R, Calderon R. University of Puerto Rico, San Juan.

Case Report: 45 y/o male with no past medical history who developed epigastric discomfort and progressive shortness of breath. At ER, patient was tachypneic and had no evidence of JVD or cyanosis. Auscultation revealed clear lung fields and a new non-radiating IV/VI holosystolic murmur throughout the left sternal border. A low pitch I/IV diastolic murmur and S3 gallop were also audible. Bedside echocardiogram showed an unspecified left to right shunt.

Transesophageal echocardiogram revealed a large ruptured right sinus of valsalva aneurysm (7.4 cm) with a significant left to right shunt from the aortic root into the right ventricle. Left heart volume overload and a preserved left ventricular ejection fraction were present. Moderate aortic valve regurgitation noted. Mild tubular aortic root dilation (3.9cm) and a small atrial septal defect (ASD) with bi-directional flow were also identified. Patient immediately underwent Bentall procedure and patch closure of the ASD with no complications at 6 months.

Sinus of valsalva aneurysms are very uncommon, with an incidence of 0.1 to 3.5% of all congenital heart defects. Such aneurysms account for only 0.14% of all open-heart surgical procedures. It is frequently associated with other degenerative conditions such as aortic valve regurgitation and ascending aortic aneurysm, which must be ruled out before time of surgery. Rupture is the major cause of death and should be clinically suspected in all patients with subacute onset of atypical chest pain and shortness of breath in association with a new heart murmur.

EXPECTED AND UNEXPECTED ELECTROLYTE DISTURBANCES WITH BATH SALT INGESTION

Pourmorteza M1, Al Shathir M2, Roy T2, Byrd R2. 1East Tennessee State University, Johnson City, TN and 2East Tennessee State University, Johnson City, TN.

Case Report: A 61-year-old male with history of drug abuse was admitted to the intensive care unit with mental status changes. His family reported that the patient had ingested "bath salts". His blood pressure was 210/112mmHg, heart rate 107 bpm and respiratory rate 24 bpm. He was confused and combative. Laboratory analysis showed: potassium 2.3 meq/L, corrected calcium 16.9 mg/dl, magnesium 1.4 mg/dl, creatinine 2.0 mg/dl, blood urea nitrogen 10 mg/dl, and creatinine phosphokinase 189 U/L. His serum bicarbonate was 34 mEq/L, pH 7.56 and partial pressure of carbon dioxide 37 mmHg. His blood alcohol, salicylate, acetaminophen, ammonia and liver function test were normal. Urine drug screen was positive for benzodiazepine. Parathyroid hormone, vitamin D and Lithium levels were normal. Intravenous infusion of lorazepam, labetolol, and normal saline were initiated. The hypercalcemia and alkalemia resolved slowly over three days. An analysis of the substance he ingested could not be accomplished.

A 54 year old female presented with lower back pain, bilateral lower extremity weakness, and bladder incontinence. Patient denied any recent trauma, illness, or lumbar injections. Vitals were unremarkable. Physical exam was significant for motor strength 2/5 left lower extremity, 1/5 right lower extremity, and absent bilateral patella tendon reflexes. Labs were unremarkable. Magnetic resonance imaging (Fig. 1) of lumbar spine revealed L4-L5 posterior extradural cystic lesion causing severe thecal sac stenosis and compression of the cauda equine. Neurosurgery performed a hemilaminotomy, foraminotomy, and drainage of epidural fluid collection, which was visually purulent, but gram stain and culture were negative for organisms. After surgery there was marked improvement in strength and sensation of lower extremities, with complete resolution of bladder incontinence. Patient was discharged to a rehabilitation facility where she continued physical therapy.

Benign lumbar epidural cyst is an uncommon cause of acute cauda equina syndrome. Prompt diagnosis and neurosurgical intervention may reduce long term sequela associated with this condition.
Synthetic cathinones are sold under the name “bath salts”. Electrolyte disturbances due to the use of these agents have been documented. Hypokalemia was reported in 10 out of 236 patients in one study. Hyponatremia is also rarely reported. Hypercalcemia associated with synthetic cathinone use has not been demonstrated. Bupropion is the only cathinone derivative that has a medical indication. Despite widespread clinical use of this medication, hypercalcemia has not been observed. Causes of hypercalcemia were investigated and ruled out in our patient. It is doubtful that our patient’s hypercalcemia was due to direct cellular toxicity. Given the extensive experience with bupropion, it seems unlikely that his hypercalcemia was due to a metabolic effect of the synthetic cathinone. His biochemical profile is consistent with calcium-alkali syndrome. We suspect that the bath salt our patient ingested had been adulterated with a calcium-containing antacid and resulted in his metabolic and electrolyte disturbances.

INTRODUCTION: Encephalopathy is an uncommon but serious side-effect of prolonged metronidazole use. It is important for clinicians to be aware of metronidazole-induced encephalopathy and recognize its clinical findings.

CASE DESCRIPTION: A 55-year-old male was admitted to the hospital for vertigo and weakness. He had a recent history of a liver abscess for which he was taking oral metronidazole. He complained of dizziness described as the room spinning that worsened with change in position, which had become progressively worse. He also had nausea, vomiting, and headache. He denied fever, neck pain, and tinnitus. Vital signs were within normal limits, and physical exam was remarkable only for globally mild strength reduction. He had no nystagmus or nuchal rigidity. Laboratory data was non-contributory. During his hospital stay his dizziness worsened. His weakness increased to the point that he could not walk or grip objects. He began to have slurred speech, dysphagia, and trismus. He developed dysmetria with abnormal heel-to-shin and finger-to-nose tests, Babinski sign, and ataxia. Reflexes were absent in a stocking and glove distribution. Cranial nerves remained intact. Computed tomography of the head without contrast was unremarkable. Lumbar puncture revealed mildly elevated protein. Magnetic resonance imaging (MRI) of the brain with contrast revealed bilateral abnormal signals in the dentate nuclei and punctate foci in the posterior pons including diffusion abnormality. This pattern is consistent with documented metronidazole encephalopathy. The metronidazole was immediately discontinued. His symptoms began to dissipate over the next few days. On reconciliation of his drug regimen, he had been given metronidazole 500 mg three times daily for 33 days when his symptoms abruptly started. The metronidazole was discontinued after 42 days of use.

CONCLUSION: Metronidazole-induced encephalopathy is an uncommon but serious side-effect of prolonged use of metronidazole. Clinicians that encounter a patient with new-onset encephalopathy (particularly with cerebellar signs and symptoms) as well as a current metronidazole use should consider obtaining an MRI of the brain, followed by discontinuation of metronidazole if deemed contributory.

Case Report:

A 55-year-old male was admitted to the hospital with a three-day history of nausea, vomiting, and non-bloody diarrhea and developed chest pain and shortness of breath while in triage. Evaluation demonstrated hyperthyroidism, as well as the presence of hematuria, proteinuria, and elevated troponin levels. Cardiac catheterization established a diagnosis of takotsubo cardiomyopathy. The patient was treated with clopidogrel and aspirin with complete resolution of her cardiomyopathy over several weeks. This case illustrates the possible association of takotsubo cardiomyopathy with subclinical hyperthyroidism.

METRONIDAZOLE-INDUCED ENCEPHALOPATHY
Randall G. Kansas City University of Medicine and Biosciences, Kansas City, MO.

Case Report:

INTRODUCTION: Encephalopathy is an uncommon but serious side-effect of prolonged metronidazole use. It is important for clinicians to be aware of metronidazole-induced encephalopathy and recognize its clinical findings.

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SUCCESSFUL TREATMENT OF AN ELDERLY PATIENT WITH PSYCHOSIS VIA VIDEO TELECONFERENCE
Tan SY, Wells J. LSUHSC New Orleans, New Orleans, LA.

Case Report: Patients in medically underserved areas experience significant barriers to mental health care. In the management of an acutely psychotic patient, the quality and frequency of interaction with the mental health provider is of the utmost importance. By leveraging advanced telecommunication technology, in concert with a patient’s primary care provider, a remotely located psychiatrist may effectively provide care for patients in psychiatric emergencies.
We present the case of an elderly patient who presented to a rural primary care center with complaint of depression and PTSD as a consequence of spousal abuse. At the time of initial presentation, the patient was tearful, malnourished, and complained of anxiety and insomnia. She had a remote history of psychiatric hospitalization.

We were consulted by the primary care team to provide psychiatric care via video teleconferencing, from our academic center in a nearby metropolitan area. We performed initial assessment, medication management, and counseling via teleconferencing. The patient was thereby initially stabilized. During routine telephone follow-up to assess medication efficacy and adherence, the patient's family reported waning adherence to the medication regimen, and overt psychotic symptoms including auditory and visual hallucinations, anorexia, and extreme paranoia.

Through video teleconferencing with the primary care team and patient, we were able to effectively assess this patient’s need for hospitalization, while simultaneously managing her psychotic symptoms. Weekly telemedicine follow-up visits were scheduled to monitor symptoms. Frequent weight checks and care management team phone calls were made to ensure compliance and safety.

The patient was ultimately stabilized with aripiprazole, mirtazapine, and chlorzoxazone. Her paranoia and hallucinations resolved, and patient was subsequently able to attend various social functions and resume care for herself and other family members.

High fidelity video teleconferencing, while still a relatively novel technology, is a very effective modality for the delivery of psychiatric care to medically underserved areas. Current research suggests that this promising technology decreases hospitalization, reduces costs, and improves patient adherence and satisfaction.

Cephalospin Side Chain Idiosyncrasies: Ceftriaxone Induced Agranulocytosis

Uy N,1, Mushar D1,2. Baylor College of Medicine, Houston, TX and 1Michael E. DeBakey Veterans Affairs Medical Center, Houston, TX.

**Case Report:** Drug-induced agranulocytosis is an adverse event characterized by a sharp drop of absolute neutrophil count (ANC) to <500/mm3 due to immunologic or cytotoxic mechanisms. Although associated with a large variety of drugs, these adverse effects remain rare, occurring in 2.4-15.4 cases per million courses of therapy. We describe a patient with ceftriaxone-induced agranulocytosis whose ANC fully recovered after treatment was switched to cefepime.

A 49 year old male with polysubstance abuse was admitted with altered mental status after taking multiple Viconin pills for severe lower back pain. Imaging showed osteomyelitis (L4-L5), phlegmon (L3-S1), and epidural abscess (L4). Four blood cultures revealed Streptococcus intermedius. A 6-week course of ceftriaxone, 2 gm intravenously (IV) every 12 hours, was prescribed. On day 25 of his therapy, a routine complete blood count documented an ANC of 480 per mm3, down from 4435 per mm3 12 days prior. The following day he developed a fever of 103.3F with tachycardia to the 130s; antibiotic therapy was changed to IV vancomycin 2 gm every 12 hours and cefepime 2gm every 8 hours. Neutropenia reached its lowest point on day 28, with an ANC of 0. On day 32, one week after ceftriaxone discontinuation and while receiving cefepime, his ANC recovered to 2024 per mm3. MRI of the spine revealed markedly improved L4-L5 osteomyelitis. The patient was discharged with oral clindamycin 450 mg 3 times daily for 2 weeks to complete 6 weeks of therapy.

This patient developed neutropenia and fever during a prolonged ceftriaxone course and improved shortly after cessation. There is limited literature on ceftriaxone-induced agranulocytosis, especially within the last decade; most reports describe a time course of development and recovery. A unique feature of our patient is his neutrophil recovery during treatment with cefepime; to our knowledge, this is the only such reported case. Cefepime and ceftriaxone have remarkably similar structures with an identical core and R1 side chain; thus, his neutropenia was likely associated with ceftriaxone’s unique R2 side chain. Overall, neutropenia during treatment with a beta lactam antibiotic is uncommon but remains an important consideration in any patient whose ANC falls during an extended course.

PSoriasis and Coronary Artery Disease: A Hispanic Population Study

Alcifer P1,2, Nieves I1,2, Sulia C1, Figueroa L1, Bangs HL1,2, Crespo M1, Escobales N1, 1University of Puerto Rico, Medical Sciences Campus, San Juan and 2Cardiovascular Center of Puerto Rico and the Caribbean, San Juan.

**Purpose of Study:** Psoriasis is an immune mediated disorder characterized by chronic elevated levels of systemic inflammation. Recent studies have found that this chronic inflammatory state may contribute to the development of coronary artery disease (C.A.D.). There is a suggestion that psoriasis itself increases the risk of myocardial infarction (M.I.) and cerebrovascular disease (C.V.D.) compared to the general population. Psoriasis is relatively common among Hispanics, who are known to have a less aggressive C.A.D. compared to Caucasians in the United States. We intend to find the incidence of C.A.D. in Puerto Ricans with psoriasis.

**Methods Used:** Retrospective study analyzing the coronary angiography reports of 46 patients with moderate-to-severe psoriasis. Findings were recorded and statistically analyzed using the student's T test.

**Summary of Results:** The male to female ratio found was 7:3 and the median age was 56 years. Of the 46 patients, 27 had C.A.D. by angiographic criteria (59%). Twelve patients were referred for bypass graft surgery, 8 underwent coronary stent placement and 7 were treated medically. None showed electrocardiographic changes suggesting a M.I. or had history or physical findings suggesting C.V.D.

**Conclusions:** Whether psoriasis can be considered a risk factor for developing C.A.D. or strokes (S.) is still on debate. At least 50% of our patients showed angiographic C.A.D., but no clinical M.I. or C.V.D. This is particularly important because the most frequent cause of death in the Island is C.V.D., followed by M.I. The HDL-C and LDL levels were normal. Half of patients had Diabetes Mellitus Type 2 (D.M.), when in Puerto Rico is 16% of the total population. We believe inflammation is an important factor.

Psoriasis and Coronary Artery Disease: A Hispanic Population Study

Altieri PI 1,2, Nieves J 1,2, Sulia C 1, Figueroa L 1, Banchs HL 1,2, Crespo M 1, Escobales N1, 1University of Puerto Rico, Medical Sciences Campus, San Juan and 2Cardiovascular Center of Puerto Rico and the Caribbean, San Juan.

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RELATIONSHIP BETWEEN THE ECG PATTERN OF LEFT VENTRICULAR HYPERTROPHY AND PROLONGED QT INTERVAL


**Purpose of Study:** Many patients with electrocardiographic evidence of LVH will have concurrent prolongation of corrected QT interval (QTc) of the electrocardiogram. Myocardial hypertrophy may lead to electrical remodeling and delayed repolarization with prolonged QTc. The objective of this study is to determine whether one ECG pattern of left ventricular hypertrophy (LVH) is more susceptible than another to QTc prolongation. ECG-LVH pattern was defined as limb lead, concentric (RI+SIII>25 mm and RaVL>13 mm) or chest lead, eccentric (SV1+RV5>35 mm). We sought to determine if one ECG pattern of LVH would be more likely associated with prolonged QTc than the other and hence may have an increased propensity for supra- and ventricular arrhythmogenicity.

**Methods Used:** A retrospective analysis of 989 patients who presented to Regional One Medical Center, Memphis between July 1, 2013 and June 30, 2014. ECG pattern of LVH was recognized as concentric, eccentric or combined. An independent sample t-test was performed to compare the average length of QTc with each pattern. Patients taking QT prolonging medications were excluded from the study. Statistical analysis was performed using IBM SPSS v20.0.

**Summary of Results:** Of the 869 patients with prolonged QTc, 496 had LVH with a mean QTc of 477±1 msec; 214 (43%) had concentric LVH with a
mean QTc of 474±1 msec while 234 (47%) patients had eccentric LVH with a mean QTc of 482±2 msec. 48 (10%) patients met both criteria with a mean QTc of 469±2 msec. A t-test showed that the difference in QTc interval between the two groups were statistically significant (p<0.05).

Conclusions: The length of the QTc interval was statistically different in favor of eccentric LVH being more prolonged. Thus, there is an association between electrocardiographic pattern of LVH and length of QTc interval. Further studies will be needed to address ECG and echocardiographic patterns of LVH and their relation to QTc interval and propensity for cardiac arrhythmias.

RISK OF ATRIAL ARRHYTHMIAS IN PATIENTS WITH PROLONGED QTc INTERVAL AND LEFT ATRIAL ENLARGEMENT


Purpose of Study: The association between prolongation of the electrocardiographic QTc interval and ventricular arrhythmias is well established. A growing body of work suggests a link between prolonged QTc interval and atrial arrhythmias. Left atrial enlargement is considered an additional risk factor implicated in the occurrence of atrial arrhythmias. This study examined the association between prolonged QTc interval, ECG-diagnosed left atrial enlargement (LAE) and atrial arrhythmias.

Methods Used: Data was collected from 12-lead standard ECGs obtained from an inpatient and ambulatory pool of adult patients followed at an urban medical center over a one-year period from July 2013 to June 2014. A prolonged QTc interval was defined as greater than 440 ms. LAE was defined as P wave duration in any lead greater than 0.11 s or notched P wave with interpeak duration greater than 0.04 s.

Summary of Results: In patients with prolonged QTc interval, chi-square analysis showed a statistically significant negative association between the presence of left atrial enlargement and incidence of atrial arrhythmias (p<0.01). Among patients with prolonged QTc, those with no evidence of LAE on ECG were 8.3 times more likely to develop atrial arrhythmias than patients with evidence of LAE on ECG (p<0.05).

Conclusions: Both prolonged QTc interval and electrocardiographic evidence of left atrial enlargement are implicated as risk factors associated with atrial arrhythmias. In this study, patients with both prolonged QTc and LAE were significantly less likely to demonstrate atrial arrhythmias than patients with prolonged QTc interval alone.

CONTROLLING ATRIAL FIBRILLATION BY ATRIAL FLUTTER ABLATION IN THE PRESENCE OF LA ENLARGEMENT

Ibrahim S, Al-Saffar F. University of Florida-Jacksonville, Jacksonville, FL.

Purpose of Study: Typical isthmus dependent atrial flutter (AFL) ablation targeting the isthmus between the inferior vena cava and the tricuspid annulus is an established therapy for the treatment of AFL. AFL is often complicated by new coexistent atrial fibrillation or new incident arrhythmias with reported incidence of up to 50%. Our study investigated the benefit of AFL ablation on atrial fibrillation (AF) burden after isthmus dependent AFL radiofrequency ablation with Left Atrium (LA) dilation taken into account.

Methods Used: Records were reviewed for the last 10 years on adult patients who had isthmus dependent AFL ablation and implanted cardiac device. The general characteristics and comorbidities of the patient population were summarized. LA diameter was assessed at the time of ablation and at 12 months. Daily AF burden before AFL ablation was extracted through device interrogation. AF delta (daily frequency before minus frequency after ablation was the primary outcome). The correlation between these predictors and the outcome was tested using t-test and generalized linear models.

Summary of Results: 80 patients were included. Mean AF per day before ablation was 23.15 and after ablation was 4.74 (p value 0.204). 71.42% of patients had LA diameter (> 4 cm). Patients with normal LA diameter had stable AF delta (mean 0.01, standard deviation 0.156), while patients with dilated LA had a great increase in AF delta (mean +30.19, standard deviation 73.82) (p = 0.425). It was surprising to see that hemoglobin levels lower than 12 gm/dl had a beneficial effect in reducing AF recurrence with odds ratio (0.18, 95% CI 0.036 - 0.89, p value 0.03).

Conclusions: AFL ablation showed a trend toward reducing AF burden in this sample of patients. This effect; however, did not achieve statistical significance. Most of those patients had concomitant LA dilation, which may have reduced the efficacy of AFL ablation as dilated LA can increase the odds and the severity of AF. The protective effects of lower hemoglobin levels seen in our study can be attributed to the fact that patient with more comorbidities (like COPD, obesity hyperventilation, or heart failure) have higher probability of being hypoxic and develop secondary polycythemia.

AN UNUSUAL CASE OF COINCIDING ANOMALOUS CORONARY ARTERY AND PAPILLARY FIBROELASTOMA

Ibrahim S, Al-Saffar F. University of Florida - Jacksonville, Jacksonville, FL.

Case Report: Introduction: The primary cardiac tumors are extremely rare tumors that arise from the normal tissues of the heart. More than 90% of primary cardiac tumors that arise from the normal tissues of the heart are benign and most commonly are papillary fibroelastomas. A growing body of work suggests a link between prolongation of the electrocardiographic QTc interval and left atrial enlargement (LAE) and atrial arrhythmias. Among patients with prolonged QTc, those with no evidence of LAE on ECG were 8.3 times more likely to develop atrial arrhythmias than patients with evidence of LAE on ECG (p<0.05).

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An Unusual Case of Coinciding Anomalous Coronary Artery and Papillary Fibroelastoma

Ibrahim S, Al-Saffar F. University of Florida-Jacksonville, Jacksonville, FL.

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RELATIONSHIP BETWEEN LEFT VENTRICULAR HYPERTROPHY AND CORRECTED QT INTERVAL OF THE ELECTROCARDIOGRAM


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RELATIONSHIP BETWEEN LEFT VENTRICULAR HYPERTROPHY AND CORRECTED QT INTERVAL OF THE ELECTROCARDIOGRAM

Purpose of Study: Prolongation of the corrected QT interval of the electrocardiogram increases the risk of ventricular arrhythmias. Whether left ventricular hypertrophy raises the potential for QTc prolongation remains uncertain. Herein, we examined electrocardiograms of patients both with and without ECG evidence of left ventricular hypertrophy to determine if its presence was associated with QTc prolongation.

Methods Used: A retrospective chart analysis of 1400 consecutive patients who presented to the Regional One Health Medical Center between July 1, 2013 and June 30, 2014 and received an electrocardiogram. Patients who were taking medications known to prolong the QTc interval were excluded. After exclusion, 988 patients (53±0.41 yrs; 46% male) qualified for review. The corrected QT interval of each electrocardiogram was recorded as well as the presence or absence of left ventricular hypertrophy by standard ECG criteria using voltage in limb and chest leads.

Summary of Results: Of 988 patients, 371 were noted to have left ventricular hypertrophy (LVH) by ECG criteria. The mean corrected QT interval in those patients was significantly higher (489.5±32.0 ms) than those who did not have LVH (469.7±26.2 ms). The 95% confidence interval for patients with or without LVH was 486.2-492.8 ms and 467.6-471.8 ms, respectively.

Conclusions: Left ventricular hypertrophy appears to be an independent risk factor for corrected QT interval prolongation of the electrocardiogram. Patients having LVH should be carefully monitored for additional risk factors that prolong QT interval (e.g., drugs, hypokalemia, hypomagnesemia) to prevent the heightened potential for supra- and ventricular arrhythmias.

THE RELATIONSHIP BETWEEN QTc PROLONGATION AND VENTRICULAR ECTOPIC BEATS

Purpose of Study: Herein, we addressed the relationship between QTc-prolongation and premature ventricular contractions in an urban medical center patient cohort presenting with prolonged QTc on their standard ECG.

Methods Used: 1500 patients who presented to Regional One Medical Center Memphis were evaluated for prolonged QTc (>440 ms). Patients on medications that would prolong QTc were excluded. A total of 989 patients had prolonged QTc. The ECG was evaluated for presence of premature ventricular contractions. Statistical analysis was conducted using IMB SPSS V 20.0.

Summary of Results: A paired two-sample t-test was performed with equal variances. Of the total of 989 patients with prolonged QTc, 61 had left ventricular hypertrophy (L VH) by ECG criteria. The mean corrected QT interval of each electrocardiogram was recorded as well as the presence or absence of left ventricular hypertrophy by standard ECG criteria using voltage in limb and chest leads.

Conclusions: Left ventricular hypertrophy appears to be an independent risk factor for corrected QT interval prolongation of the electrocardiogram. Patients having LVH should be carefully monitored for additional risk factors that prolong QT interval (e.g., drugs, hypokalemia, hypomagnesemia) to prevent the heightened potential for supra- and ventricular arrhythmias.

RACIAL DIVERGENCES IN ASSOCIATIONS OF C-REACTIVE PROTEIN WITH CENTRAL AND PERIPHERAL BLOOD PRESSURES—THE BOGALUSA HEART STUDY
Leeone N, Fernandez-Alonso C, Chen W, Srinivasan SR, Berenson GS. Tulane University, New Orleans, LA; Tulane University, New Orleans, LA and Tulane University, New Orleans, LA.

Purpose of Study: C-reactive protein (CRP), a biomarker for systemic inflammation, is noted to have associations with various cardiovascular disease risk factors, but CRP’s relationship and racial differences with central systolic blood pressure (cSBP), peripheral systolic blood pressure (SBP) and diastolic blood pressure (DBP) are uncertain. This study examines those relationships and racial contrasts in the biracial (black-white) community of Bogalusa, Louisiana.

Methods Used: cSBP (Omron HEM9000Alt), peripheral blood pressures and serum CRP levels were attained on a biracial population of 735 individuals (range of age 29-51 years, 57% female, 70% white) from 2007 to 2010. Linear regression was performed to determine CRP’s relationship with central and peripheral blood pressure levels in white and black subjects.

Summary of Results: In whites log CRP was associated with cSBP (P=0.001, R²=0.097), SBP (P=0.003, R²=0.172) and DBP (P=0.001, R²=0.140) when adjusted for glucose, age, sex, BMI and HDL-C. In blacks log CRP did not have any significant associations with cSBP, SBP or DBP after adjusting for confounders. When comparing cSBP, SBP and DBP to the different CRP risk prediction categories (low, intermediate, high) for cardiovascular disease (CVD), increasing trends were found in whites for cSBP, SBP and DBP but were not present in blacks.

Conclusions: cSBP, peripheral systolic blood pressure and diastolic blood pressure were all associated with CRP in whites, but these associations were not present in blacks. The racial divergences found suggests a variation of central nervous system control of blood pressure detected by inflammation. This observation may have implications for control of blood pressure and should also be taken into account when developing clinical applications for CRP.

A CONGENITAL DILEMMA: LATE PRESENTATION OF EBSTEIN’S ANOMALY
Marcial JM, Lopez JE, Maldonado J. University of Puerto Rico, San Juan and University of Puerto Rico, San Juan.

Case Report: An active 49-year-old male nurse with a history of an untreated heart murmur was talking to a colleague at work when he experienced a sudden episode of nausea, light-headedness and loss of consciousness followed by a rapid recovery. His ECG showed a right bundle branch block, extreme right axis deviation and a 1st degree AV block. Transhoracic echocardiography demonstrated a massive dilation of the right heart chambers, apical displacement of the septal tricuspid valve leaflet and severe tricuspid regurgitation. Agitated saline injection and cardiac MRI excluded shunts and confirmed adequate biventricular function. No arrhythmias were evidenced during 72 hours on cardiac telemetry. In view of the non-specific and isolated nature of the syncope, absence of pre-excitation on the ECG and adequate biventricular function, the patient was deemed to be at a low risk for sudden cardiac death and an electrophysiology study was not performed. Moreover, evidence-based medicine did not warrant the initiation of new medications or surgical intervention. Most patients with this complex congenital condition, Ebstein’s Anomaly, are very symptomatic in adulthood due to recurrent cardiac arrhythmias, congestive heart failure or cyanosis. Our patient, however, was free from disease burden and thus is illustrative of an important and sometimes forgotten rule in medicine: “don’t fix it if it ain’t broke”.

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A 35 year-old woman with history of bronchial asthma presented to our institution with shortness of breath and chest pain. She had started an oral contraceptive 2 months before presentation. She denied recent surgeries, immobilization, smoking, and alcohol/drug abuse. Physical exam showed an obese woman with tachycardia, tachypnea, hypoxemia and borderline hypotension. Initial blood work revealed elevated D-Dimer, troponin and BNP levels. ECG was relevant for sinus tachycardia. Chest CT with intravenous contrast showed bilateral pulmonary artery embolism. Venous duplex scan of the lower extremities was negative for deep vein thrombosis. Transthoracic echocardiography showed flattening of the interventricular septum, right ventricular dilatation, and elevated tricuspid regurgitation doppler velocity consistent with right ventricular pressure overload. A right ventricular thrombus was also noted. Based on the echocardiography findings and a large clot burden we decided to administer systemic thrombolytic therapy. Alteplase 100 mg was given in 2 hours, followed by heparin infusion. Echocardiography at 48 hours post-thrombolysis showed normalization of the right ventricular dimension and tricuspid regurgitation velocity. It also showed interval resolution of the right ventricle thrombus. A pulmonary angiography showed complete resolution of the pulmonary emboli with normal pulmonary artery pressure. Warfarin was started and titrated to goal INR of 2-3. Hypercoagulable state workup ordered before warfarin administration was normal. She was discharged in stable condition and was advised to use non-hormonal contraception. Venous thromboembolism is a serious condition with considerable morbidity and mortality. Thrombolytic therapy for acute pulmonary embolism is recommended for hemodynamically unstable patients. Thrombolysis is also indicated in hemodynamically stable patients with evidence of a large clot burden, severe hypoxemia, right atrial/ventricular thrombus and right ventricular dysfunction. Systemic thrombolysis is generally preferred over catheter directed thrombolysis. Several risk factors exist for DVT/PE and it is important to counsel patients after a thromboembolic event in order to minimize these risks when possible.

**SUCCESSFUL THROMBOLYSIS FOR PULMONARY EMBOLISM WITH ASSOCIATED RIGHT VENTRICULAR THROMBUS**

Banchs-Vilas H, Calderon R, Gutierrez Y. *University of Puerto Rico School of Medicine, San Juan.*

**Case Report:** A 35 year-old woman with history of bronchial asthma presented to our institution with shortness of breath and chest pain. She had started an oral contraceptive 2 months before presentation. She denied recent surgeries, immobilization, smoking, and alcohol/drug abuse. Physical exam showed an obese woman with tachycardia, tachypnea, hypoxemia and borderline hypotension. Initial blood work revealed elevated D-Dimer, troponin and BNP levels. ECG was relevant for sinus tachycardia. Chest CT with intravenous contrast showed bilateral pulmonary artery embolism. Venous duplex scan of the lower extremities was negative for deep vein thrombosis. Transthoracic echocardiography showed flattening of the interventricular septum, right ventricular dilatation, and elevated tricuspid regurgitation doppler velocity consistent with right ventricular pressure overload. A right ventricular thrombus was also noted. Based on the echocardiography findings and a large clot burden we decided to administer systemic thrombolytic therapy. Alteplase 100 mg was given in 2 hours, followed by heparin infusion. Echocardiography at 48 hours post-thrombolysis showed normalization of the right ventricular dimension and tricuspid regurgitation velocity. It also showed interval resolution of the right ventricle thrombus. A pulmonary angiography showed complete resolution of the pulmonary emboli with normal pulmonary artery pressure. Warfarin was started and titrated to goal INR of 2-3. Hypercoagulable state workup ordered before warfarin administration was normal. She was discharged in stable condition and was advised to use non-hormonal contraception. Venous thromboembolism is a serious condition with considerable morbidity and mortality. Thrombolytic therapy for acute pulmonary embolism is recommended for hemodynamically unstable patients. Thrombolysis is also indicated in hemodynamically stable patients with evidence of a large clot burden, severe hypoxemia, right atrial/ventricular thrombus and right ventricular dysfunction. Systemic thrombolysis is generally preferred over catheter directed thrombolysis. Several risk factors exist for DVT/PE and it is important to counsel patients after a thromboembolic event in order to minimize these risks when possible.

**PURULENT PERICARDITIS CAUSING CARDIAC TAMPOONADE**

Banchs-Vilas H, Ortega-Gil J, Claudio H, Perez R, Abreu J. *University of Puerto Rico School of Medicine, San Juan.*

**Case Report:** A 36 year-old man with history of Hepatitis C and intravenous drug abuse was transferred to our institution from a community hospital for further management. Upon arrival to our institution he was tachycardic, tachypneic, with clear lungs and muffled heart sounds. ECG showed sinus tachycardia with low voltages. Transthoracic echocardiography showed a large pericardial effusion with right sided chambers collapse. He had emergent pericardiocentesis and a yellow/purulent material was obtained. Pericardial window was performed and 1,500 mL of purulent fluid were drained. Pericardial fluid analysis showed an inflammatory exudate with 316,500 WBCs/μL with neutrophilic predominance. Pericardial fluid and biopsy were negative for Mycobacterium, and fluid culture grew Streptococcus intermedius. He was treated with intravenous antibiotics and made an uneventful recovery. Purulent pericarditis is a rare condition in the antibiotic era. It is most commonly a complication of a primary infectious process such as pneumonia, with hematogenous or direct spread into the pericardium. Predisposing factors include immunosuppression, thoracic trauma/surgery, thoracic infection, and less commonly endocarditis. Treatment consists of pericardial drainage with antibiotic therapy for 2-4 weeks. Even with prompt drainage and antibiotic therapy this condition is associated with high morbidity and mortality.
In otherwise normal patients, there may be variations in the number, shape and location of the ostia or origins of the coronary arteries. Origin of the right coronary artery from the left sinus of valsalva can lead to sudden cardiac death. This anatomical defect is uncommon occurring in 0.17% of patients.

**COMPLETE ATRIOVENTRICULAR CANAL IN A PATIENT WITH DOWN SYNDROME**
Claudio H, Calderon R. University of Puerto Rico School of Medicine, San Juan.

**Case Report:** 32 y/o male patient with past medical history of Down Syndrome was evaluated in the Emergency Room due to worsening shortness of breath and dyspnea with minimal exertion since 1 month prior to evaluation. Physical examination showed classic features of Down Syndrome including flat nasal bridge, macroglossia, epicanthal folds, and single palmar crease. Cardiovascular findings show pulmonary crackles in both bases and a 3/6 holosystolic murmur heard throughout the precordium. Electrocardiogram showed sinus tachycardia at 110 bpm, right axis deviation with complete right bundle branch block and biatrial enlargement. The patient was admitted to the telemetry unit with a diagnosis of decompensated congestive heart failure and was started on standard medical therapy including diuretics and vasodilators. Echocardiographic evaluation was notable for a complete atrioventricular canal. The patient's condition improved and he was discharged home several days later.

Atrioventricular (AV) canal defects are a group of congenital cardiac defects involving the AV septum and AV valves. Atrioventricular canal defects account for about 4 to 5 percent of congenital heart defects with a reported prevalence of 0.3 to 0.4 per 1000 live births. Complete AV canal defects result in increased pulmonary blood flow because of left-to-right shunting, eventually resulting in pulmonary hypertension and heart failure.

**A MAN WITHOUT A LEFT ATRIUM: REGIONAL CARDIAC TAMPOONADE**
Sparks K, Engel LS. LSU Health Sciences Center, New Orleans, LA.

**Case Report:** A 29 year old man underwent extensive surgery including an emergent thoracotomy for bilateral pneumothorax and cardiac tamponade followed by placement of pleural and pericardial drains, and a craniotomy for treatment of subarachnoid hemorrhage following a motor vehicle accident. Following and exceptional recovery, he became short of breath and tachycardic. A CT-Angiogram of the chest revealed a left lower lobe segmental embolism. He was started on intravenous heparin with bridging to warfarin. His symptoms initially improved; however one week later he had sinus tachycardia (heart rate of 110 bpm at rest and 150 bpm upon standing), dizziness, and a 12mmHg drop in systolic blood pressure. On exam, his jugular venous pressure was found to be 9mmHg, and there was no variation in his systolic blood pressure with respiration, and no pericardial rub or distance of heart sounds. An elevated serum atria natriuretic peptide raised concerns for pericardial effusion. A repeat trans-thoracic echocardiogram was not diagnostic. Anti-coagulation was discontinued due to continued concern for pericardial effusion. Trans-esophageal echocardiogram revealed a large pericardial effusion with loculated fibrin strands posterior to the left heart resulting in diastolic left ventricular collapse and near complete left atria compression with obstruction of inflow to left ventricle. He subsequently received an emergent subxyphoid pericardial window for drainage of the effusion. The patients symptoms improved and a repeat trans-thoracic echo revealed complete resolution of the effusion.
Discussion: Diagnosis of cardiac tamponade is usually clinically made with specific findings such as the classic Beck’s Triad (distant heart sounds, distended neck veins, low blood pressure) in addition to findings such as Pulsus Paradoxus. Any loculated, eccentric effusion or localized hematoma can produce regional cardiac tamponade in which only selected chambers are compressed. Establishing the diagnosis can often become challenging as physical exam findings and echocardiography are not always typical of tamponade physiology. Regional tamponade is often seen after pericardiodytomy (or myocardial infarction) and therefore the clinical suspicion should remain heightened.

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TRANSIENT MID-INFERIOR WALL AKINESIA: A VARIANT FORM OF TAKOTSUBO CARDIOMYOPATHY

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Case Report: Background: Takotsubo cardiomyopathy (TCM) is one of the conditions that mimic acute coronary syndrome (ACS). This condition is usually characterized by transient hypokinesis of the left ventricular apex with compensatory hyperkinesis of the basal segments associated with emotional or physical stress. We are reporting a case of atypical presentation of Takotsubo cardiomyopathy.

Case Report: A 49-year-old Caucasian non-smoker woman presented with severe intermittent left-sided chest pain radiated to left arm for 4 hours. Her past medical history included hypertension and one episode of deep vein thrombosis 8 years ago. There was no significant family history. She denied recent physical or emotional stress. On physical examination, vital signs were normal. Cardiac examination revealed normal heart rate and regular rhythm. There were no murmurs or extra sounds. Lungs were clear to auscultation. Laboratory findings revealed an elevated CK of 336 IU/L, elevated CK-MB of 27 ng/ml, elevated brain natriuretic peptide of 333 pg/ml, normal Cr of 0.9 mg/dl, normal complete blood count, and unremarkable lipid profile. Electrocardiogram indicated ST-segment elevation in leads II, III, and aVF. Diagnosis of ACS was made. She underwent emergent cardiac catheterization, which showed normal coronary arteries. Left ventriculography revealed akinesis of mid portion of inferior wall. Transthoracic echocardiography was performed 2 days after admission, which demonstrated normal cardiac chambers size and ejection fraction with resolution of inferior wall akinesis. The patient was discharged after 3 days of observation.

Conclusion: TCM is found in 1% to 3% of patients with suspicion of ACS. It occurs mostly in postmenopausal women and causes by a rapid elevation of catecholamine level during stress and microvasculature dysfunction. TCM is generally transient and resolved within a few days to several, however, recent studies have reported mortality rate up to 8%. To date, the mainstay of treatment is symptomatic treatment. There is no consensus regarding long-term treatment to prevent recurrences. B-blockers and ACE inhibitors are recommended during ventricular recovery period.

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TAKOTSUBO CARDIOMYOPATHY WITH HEART BLOCK- CASE SERIES

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Case Report: Takotsubo cardiomyopathy (TC) is a reversible cardiomyopathy with a clinical presentation indistinguishable from myocardial ischemia. TC is estimated to represent 1-2% of patients presenting with features suggestive of MI. Chest pain and dyspnea are the typical presenting symptoms. Transient ST segment elevation and a small rise in cardiac biomarkers are common. Regional wall motion abnormality which extends beyond the territory of a single epicardial coronary artery in the absence of obstructive coronary lesions is the characteristic finding. Supportive treatment leads to spontaneous rapid recovery in nearly all patients. The prognosis is excellent, and recurrence occurs in <10% of patients. Complete heart block accompanying TC is a very rare presentation. The wall motion abnormality that accompanies TC recovers completely but the AV block might persist.

68 y/o Women heavy smoker presented with SOB, chest tightness and nausea/vomiting. Physical exam revealed tachycardia, coarse breath sounds and lower extremity edema. Labs showed elevated troponins, ECG was significant for T-wave inversion in V1 and V2. EF 34-39% with hypokineti kinetic ventricular wall. ACS protocol was initiated; next day telemetry illustrated two episodes of high grade AV block and ST depression in anterior leads. She had a cath showing apical ballooning with no coronary stenosis suggestive of TC.

87 y/o Women with PMH of HTN and MV endocarditis presented with sudden onset sub sternal chest pain. In ER, she had an episode of symp tomat ic bradycardia (HR 30) which responded to atropine. Physical exam was unremarkable. Labs revealed elevated troponins. ECG showed Q waves in inf and ant leads. Cath showed apical ballooning. EF cannot be calculated. She remained bradycardic; post cath ECG revealed high grade AV block and a dual chamber PPM was implanted.

These cases demonstrate the possible link between AV block and TC and the management dilemma it poses. Although the LV dysfunction recovers in almost all patients with TC, it is not well known whether AV conduction normalizes with time. It is not clear whether the AV block is the result of TC. Long term follow up is necessary to monitor resolution of AV conduction abnormalities and hence to avoid unnecessary pacing.

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PACEMAKER LEAD PREVENTING COMPLETE DECOMPENSATION IN CARDIAC TAMPOONADE

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Case Report: INTRODUCTION: Cardiac tamponade remains a feared complication of pericardial effusions. The hemodynamic effect of any effusion is related to the acuity of which it occurs as well as the volume of the effusion. Regardless of the scenario, recognizing this condition quickly and acting on it with the help of an experienced cardiologist may result in a remarkable outcome.

CASE: A 53 year old incarcerated man with Marfan’s syndrome status-post recent AVR and TVR with epicardial DDDR pacemaker placement (1 month prior) presented with a three day history of difficulty breathing. He was in moderate respiratory distress with heart rate of 110 bpm, respiratory rate of 20/min, blood pressure 85/57. Physical exam revealed an aortic click, a II/VI holosystolic murmur (atrial and tricuspid areas), JVP 20 mmHg, and bilateral lower extremity edema. Significant labs include: Cr 1.59 mg/dl, Hg 3.2 g/dl, Hct 9.6 %, INR 9.3, BNP 454 pg/ml, and Troponin 0.04 ng/ml. CTA did not demonstrate any acute aortic dissection. Trans thoracic echocardiogram revealed: a very large anterior pericardial effusion; mid-right ventricular free wall anchored to the chest due to epicardial pacemaker lead; and evidence of cardiac tamponade with right ventricular collapse in diastole (except the anchored part). The patient was admitted to the MICU and received FFP and pRBCs prior to undergoing pericardiocentesis; 1650mL of serosanguinous fluid was drained. He tolerated the procedure well and after a few days, was discharged in stable condition.

DISCUSSION: To date, there have been many cases revealing cardiac tamponade as a complication of pacemaker insertion but none that clearly describe a pacemaker as a tool that actually saves the patient from complete decompensation. This case of subacute tamponade was complicated by the fact that not only did this patient have Marfan’s syndrome with a history of aortic root and multiple cardiac valve replacements, he was also severely anemic with an increased risk of bleeding from his supratherapeutic INR. Fortunately, life-saving pericardiocentesis was able to be performed in a controlled setting and his outcome - due to his pacer wire preventing complete obstructive shock - was indeed remarkable.

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ARTERIO-RIGHT ATRIAL COMMUNICATION AFTER TRICUSPID ANNULOPLASTY

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Case Report: A 45-year-old man with dyspnea was admitted for the management of atrial septal defect (ASD). ASD repair together with tricuspid...
annuloplasty (TA) was performed due to moderate tricuspid regurgitation (TR). On one month follow up, patient was asymptomatic however a continuous murmur over left lower sternal area was heard. There was no residual defect on the interatrial septum and minimal TR was observed by transesophageal echocardiography (TTE), however aorto-right atrial fistula immediately above the annuloplasty suture (Fig1A) was found. Transesophageal echocardiography confirmed aorto-right atrial fistula (Fig1B) and patient underwent surgery for fistula repair(Fig2).Discussion: In patients with an ASD, corrective surgery to the tricuspid valve with moderate to severe regurgitation in the presence of high pulmonary pressures may ameliorate postoperative persistent TR. In our case ASD was repaired with a primary suture. TR was repaired with double needle sutures. We believe formation of a fistula between the right atrium and aorta was the result of a nodule that was deeply threaded above anteroseptal commissure of the tricuspid valve into the adjacent aorta, two close anatomic landmarks. To the best of our knowledge, aorto-right atrial communication has not been reported in literature after TA. Conclusion: A continuous flow detected in the right atrium after TA should raise suspicion for an aorto-right atrial fistula.

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SEVERE RIGHT VENTRICULAR DILATATION AFTER ROSS PROCEDURE

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Case Report: A 31-year-old woman with history of aortic valve replacement at 5 years of age due to congenital aortic stenosis and a Ross procedure with a pulmonary homograft and coronary re-implantation due to prosthetic aortic valve stenosis eleven years ago develops ten months of progressive dyspnea on exertion. Transthoracic echocardiography demonstrates severe pulmonary regurgitation which progressed from moderate severity assessed three years ago, as well as severe right ventricular dilatation with newly diagnosed severe tricuspid regurgitation. Invasive measurements confirm normal pulmonary pressures and left ventricular function, however, coronary angiography evidences a chronic and total occlusion of the ostium of the right coronary artery with collateral filling and normal left coronary anatomy. The patient underwent porcine pulmonary valve re-implantation, tricuspid annuloplasty and coronary artery bypass graft to the right coronary artery. After one month of follow up, she reports great improvement in symptoms and quality of life. Severe right ventricular dilatation and dysfunction after Ross procedure, even in the presence of significant pulmonary homograft dysfunction, is uncommon and should prompt the search for alternative etiologies such as pulmonary hypertension or coronary artery disease, at any age.

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RESISTANT HYPERTENSION: A CHALLENGE IN HYPERTENSION MANAGEMENT

Vargas PE, Cox RA, Ortega-Gil J. University of Puerto Rico School of Medicine, San Juan.

Case Report: A 39-year-old woman with arterial hypertension (HTN), type 2 diabetes (DM), obesity and asthma came to our center with chest pain, headaches and nausea. Exam was remarkable for blood pressure (BP) 210/105 mmHg (both arms); pulse 125/min; BMI 30 kg/m2. One week before had been discharged from another hospital after 41-day stay for hypertensive crisis on 8 antihypertensive drugs. After admission to our center had ICU monitoring while on IV antihypertensive therapy. Cardiac markers, ECG, 2-D and Doppler echo and coronary angiography were all normal. Secondary causes of HTN (renal artery disease, pheochromocytoma, primary aldosteronism and renal disease) were excluded. BP control was achieved on 2g Na+ diet, chlorthalidone, irbesartan, metoprolol succinate and spironolactone. Lifestyle changes (LSC) were advised. On follow up BP has remained slightly above goal. Resistant hypertension (RH) refers to BP over goal despite adherence to at least 3 optimally-dosed antihypertensive drugs of different classes, including a diuretic. While prevalence is unknown, it is felt to affect nearly 30% of hypertensives. Has been related to obesity, high Na+ intake, chronic kidney disease (CKD), DM, obstructive sleep apnea and old age. Studies have shown higher risk for cardiovascular (CV) and renal events. Accurate diagnosis requires confirmed compliance with therapy, adequate BP measurement and exclusion of white coat HTN. Search for secondary causes and evidence of target organ damage is essential. Due to multifactorial origin, management requires implementation of LSC, identification and treatment of secondary causes and effective multi-drug regimen. Though therapy is mostly empiric, multi-drug regimens with adequate doses of diuretics (mostly thiazides, unless GFR <30 mL/min when loop diuretic is required), a calcium channel blocker and either an ACE inhibitor or ARB may be effective; use of a mineralocorticoid receptor antagonist may be considered in less responsive patients. Due to a growing number of patients with comorbidities (DM, CKD and obesity) in our daily practice, adequate identification and management of RH is imperative to avert risk of CV and renal events. RH offers an area in need of active research, so that we could more successfully meet this clinical challenge in the future.
CHAGAS CARDIOMYOPATHY IN NEW ORLEANS AND SOUTHEAST U.S.
Hsu R, Chakraborti C, Sander GE. Tulane University School of Medicine, New Orleans, LA.
Case Report: Chagas disease, caused by Trypanosoma cruzi, affects 7-8 million people primarily in Central and South America and over 300,000 people in the United States. This report describes two cases of Chagas disease identified in a single hospital in the southeastern United States, which demonstrate of myocardial involvement suggestive of ischemic infarction but normal coronaries. There are three stages to Chagas disease—acute, intermediate, and chronic. Hallmarks of acute Chagas disease include fever, myalgias, diaphoresis, hepatosplenomegaly, and myocarditis while symptoms of chronic Chagas disease include pathological involvement of heart, esophagus, and colon. Myocardial involvement is identifiable by ECG and cardiac MRI showing inflammation and left ventricular wall functional abnormalities. With a growing Hispanic population in the United States who are immigrants from Central and South America, it is imperative for clinicians to keep Chagas disease as part of the differential diagnosis in patients presenting with heart disease with a history of exposure from endemic areas.

TORSADES DE POINTES PROVOKED BY INGESTION OF TONIC WATER AND GRAPEFRUIT JUICE
Mizeracki AM, Askari R, Kerikos N, Jha S, Khouzam RN. University of Tennessee Health Science Center, Memphis, TN.
Case Report: Purpose: Many drugs and over-the-counter supplements can prolong the QTc interval of the ECG to raise the risk of arrhythmias. Herein, we present a case of Torsades de Pointes in a patient with ischemic cardiomyopathy precipitated by ingestion of tonic water (containing the plant alkaloid quinine) and grapefruit juice, a P450 inhibitor.
Case Report: A 66-year-old male presented with an episode of syncope and applied electrical shock from his wearable external defibrillator (LifeVest®). He recalled feeling dizzy and then waking up on the floor. For leg cramps, he reported drinking tonic water in place of tap water for a week and continued his regular consumption of grapefruit juice for months. Medications included aspirin, prasugrel, carvedilol, lisinopril, atorvastatin and furosemide. Vital signs and physical examination were unremarkable. Wave form analysis from LifeVest showed an episode of Torsades de Pointes which lasted 34 seconds. QTc interval duration prior to initiation of the arrhythmia was 516 ms. A 150-joule shock was followed by restoration of sinus rhythm. EKG in the emergency room showed a QTc of 481 msec with a right bundle branch block that had been found on previous tracings. Laboratory examination was unremarkable with potassium 3.9 mmol/L and magnesium 2.0 mg/dL. After cessation of quinine and grapefruit juice ingestion over the next 48 hours, no further arrhythmias were observed on telemetry and QTc interval trended downward to 460 ms. Follow-up after 3 months revealed no further arrhythmias or recurrence of symptoms.
Conclusions: People at risk for ventricular arrhythmias (e.g., ischemic cardiomyopathy) should have their medication profile, including herbal supplements, scrutinized for any potential agents whose presence or interaction can cause QTc prolongation and raise the risk of arrhythmias.

IMMUNOMODULATION OF ATRIAL FIBRILLATION IN THE SETTING OF POSTOPERATIVE PERICARDITIS
Mizeracki AM, Fan T. University of Tennessee Health Science Center, Memphis, TN.
Case Report: Purpose: The appearance of atrial fibrillation (a-fib) in the days following coronary artery bypass surgery (post-op CABG) is a common occurrence that increases the length of hospital stay and often changes the traditional medical management of such patients. Herein, we present a patient with post-op CABG in whom difficult-to-control a-fib appeared and which ultimately converted to sinus rhythm with the addition of colchicine.

Case Report: A 69-year-old male with diabetes, benign prostatic hyperplasia, gout, hypertension and neuropathy underwent successful CABG. His postoperative course was uneventful. A week after discharge, the patient experienced weakness and shortness of breath. Blood pressure 80/40, heart rate was noted to be bradycardic; troponin I.5, and WBC 19. Echocardiogram showed a large loculated, compressive pericardial effusion and he therefore underwent surgical intervention with creation of a pericardial window and drainage of the effusion. Given possible sepsis, the patient was started on broad-spectrum antibiotics. Days later, he developed atrial fibrillation with rapid ventricular response. The initial rate control strategy included digoxin followed by increasing doses of diltiazem followed by metoprolol eventually achieving relative rate control. The patient was subsequently given colchicine to modify inflammatory cell response; normal sinus rhythm was achieved within 24 hours. At 6 months follow-up the patient remains in sinus rhythm.
Conclusions: Our 69-year-old patient with post-operative pericarditis and atrial fibrillation was receiving multi-drug treatment with digoxin, metoprolol and diltiazem with only relative success. The addition of colchicine to this regimen was associated with the conversion of atrial fibrillation to normal sinus rhythm. A growing body of evidence would suggest that plant alkaloid is efficacious in modulating the inflammatory response that accompanies postoperative pericarditis with pericardial effusion and which, in turn, contributes to the appearance of atrial fibrillation.

A RELIABLE ECG SIGN OF WIDOW MAKER ARTERY DISEASE, THE WELLENS’ SIGN
Panikkath D, Panikkath R, Sultan A, Adiga AG, Klomjit S, Nugent K. Texas Tech University of Health Sciences, Lubbock, TX.
Case Report: Wellens’ sign is a reliable sign of significant proximal left anterior descending artery (LAD) disease, but the awareness of this sign among internists, emergency room physicians, and family practitioners is low. It can be helpful in identifying patients who require cardiac catheterization among those who present with atypical chest pain.
This is a case report and review of literature.
A 54-year-old man with no significant past medical history presented with short lasting episodes of sharp stabbing chest pain. Pain resolved with conservative management. However, his ECG showed biphasic T inversions in leads V1 and V2 (Wellens’ sign). Cardiac angiography was done which showed a 99% lesion in the proximal left anterior descending artery. The other coronary arteries were normal. Percutaneous intervention with a drug eluting stent was done to treat the lesion. The patient did not have any recurrence of pain and was discharged the next day after the procedure and was asymptomatic on follow up. Identification of this sign helped in early conditioning of the procedure and likely prevented the occurrence of a massive anterior wall myocardial infarction in this patient.
Wellens’ sign is a useful sign of significant disease in the proximal LAD. It has a sensitivity of 69% and specificity of 89% for significant (≥70%) stenosis of the LAD with a positive predictive value 86%. It is not uncommon and is reported in 14%-18% of patients with unstable angina. This sign indicates pre infarction state of coronary artery disease, and progression to anterior wall myocardial infarction is likely. Medical management alone is not considered adequate. This sign may not be present at the time of presentation and might appear later after the resolution of chest pain. Patients with this sign can develop anterior wall myocardial infarction in a mean time of 8.5 days after the onset of this sign. Massive anterior wall myocardial infarction places the patients at risk of left ventricular dysfunction, heart failure, arrhythmias, and death. Awareness of this sign among physicians should help in early detection of proximal LAD lesions.

CENTRAL ADRENAL INSUFFICIENCY AND HYPOTHYROIDISM PRESENTING AS UNSTABLE CARDIAC ARRHYTHMIAS
Brown MC, Bolorunduro OB, Garg N. University of Tennessee Health Science Center, Memphis, TN.
Case Report: Purpose: To demonstrate central adrenal insufficiency and hypothyroidism presenting as hemodynamically unstable arrhythmias.

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Case Report: Two cases who developed unstable arrhythmias associated with central adrenal insufficiency and hypothyroidism are described. A 63 y/o male with panhypopituitarism developed atrial fibrillation with rapid ventricular response (RVR) and hypotension following dental abscess incision and drainage. Despite receiving intravenous beta blockers, his hypotension remained refractory to IV hydration. He was initially treated for suspected sepsis with intravenous antibiotics, but his condition did not improve. Persistent hypotension, hyperkalemia and hypomagnesemia were noted. Following scheduled stress dosages of IV steroids, his blood pressure, heart rate, and metabolic panel improved. Oral hydrocortisone was restarted and he was discharged home, but returned 5 days later with mental status changes, vomiting, decreased oral intake, atrial fibrillation with RVR (150-160), hypotension (89/65), and electrolyte derangements. Despite aggressive IV hydration he remained hypotensive. It was not until oral steroids were restarted (after noncompliance) that his blood pressure and heart rate stabilized.

A 23-year-old male with a history of Raynaud’s phenomenon, bipolar disorder, hypogonadism, and diabetes insipidus presented with altered mental status, constipation, and decreased urinary output. His initial HR was 42/min, with a blood pressure of 99/42. ECG revealed junctional bradycardia. Throughout his hospital course, his systolic blood pressure remained in the 70s. He was started on a continuous dopamine infusion. To determine causes of symptomatic bradycardia, his workup included echocardiograph, MRI of the brain, cortisol, testosterone and prolactin levels, and antibodies for CREST syndrome, and electrolytes; all were within normal limits. Thyroid panel, however, revealed central hypothyroidism and was considered responsible for his bradycardia.

Conclusions: Disorders of the adrenal, thyroid and pituitary glands can be associated with significant arrhythmias and therefore should be considered in patients presenting with difficult-to-manage, hemodynamically unstable cardiac arrhythmias.

ST-SEGMENT ELEVATION MYOCARDIAL INFARCTION IN A 23-YEAR-OLD FEMALE FOLLOWING PLASMAPHERESIS

Bolorunudo OB, Valasaraydy P, Khourazm RN. University of Tennessee Health Science Center, Memphis, TN.

Case Report: A 23-year-old African-American female with a history of asthma presented with complaints of severe chest pain that began an hour prior to her arrival at the hospital. Earlier in the day, she had voluntarily donated 1 liter of plasma; the plasmapheresis was uneventful and standard precautions had been taken. Soon afterwards she developed chest pain associated with dyspnea and a witnessed transient loss of consciousness. She was unconscious for less than 1 minute with no seizure-like activity. She gave no history of recreational drug abuse, or use of contraceptive medications. No personal or family history of cardiac disease, clotting disorders or chronic inflammatory disease was elicited and she was not pregnant. Social history was significant for smoking one pack of cigarettes a day for 5 years.

Her EKG showed ST-segment elevation in anterior and inferior leads, without PR segment depression. Troponin I on arrival was 2.6 ng/mL (0-0.05 ng/mL) and peaked at 17.8 ng/mL. Urine drug screen was negative. The constellation of signs and symptoms specific to end-organ damage and a systemic inflammatory response with urinary eosinophilia. Histopathologic confirmation by biopsy is the only definitive diagnosis. There is no specific therapy for this disorder and its main goals should include supportive care for tissue injury and modification of risk factors.

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A CASE OF THE RARE “BLUE TOES SYNDROME”

Brown MC, Khourazm RN. University of Tennessee Health Science Center, Memphis, TN.

Case Report Purpose: Ateroembolism refers to embolization from a primary atherosclerotic plaque of a large-caliber conduit artery to small to medium-sized arteries causing end-organ damage. Blue toe syndrome is one of the most common dermatologic manifestations of cholesterol crystal embolization. In a prospective study of 1579 patients undergoing coronary angioplasty in the US, clinical evidence of cholesterol embolization was found in 1 patient (0.06%). The purpose of this report is to raise awareness and recognition of this rare condition following coronary or peripheral angioplasty and to avoid confusion with more common acute limb ischemia.

Case Report: A 76-year-old male with history of coronary and peripheral vascular disease had undergone a recent percutaneous peripheral angioplasty for severe bilateral iliac disease. Upon follow up, he complained of worsening left lower extremity pain and discoloration of his left toes for the past 5 days. ECG showed left ventricular hypertrophy. He was admitted to rule out recurrent limb ischemia and potential need for another peripheral angioplasty and possible angioplasty. A non-invasive work up, including arterial ultrasound, did not reveal limb vascular stenosis. Instead, the constellation of symptoms suggested cholesterol embolization and the patient was started on a high-intensity statin, along with dual antiplatelet therapy. Six weeks later, he was noted to have complete resolution of left lower extremity pain and major improvement in toe discoloration.

Conclusions: In contrast to acute lower limb ischemia, the clinical presentation of the rare cholesterol embolization syndrome is based on a combination of signs and symptoms specific to end-organ damage and a systemic inflammatory response with urinary eosinophilia. Histopathologic confirmation by biopsy is the only definitive diagnosis. There is no specific therapy for this disorder and its main goals should include supportive care for tissue injury and modification of risk factors.

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A CASE OF LEVO-TRANPOSITION OF THE GREAT ARTERIES

Vargas PE, Claudio H, Camargo E, Lopez JE. University of Puerto Rico School of Medicine, San Juan.

Case Report: A 68-year-old woman with Levo-Transposition of the Great Arteries (LTGA), mesocardia and situs inversus came to our center with dyspnea on exertion and leg edema since one week prior. Physical exam was remarkable for loud systolic and diastolic murmurs, jugular vein distention, and decreased breath sounds. Initial workup included echocardiography and CT chest. Echocardiography showed aortic arch with mirror image branching, atrial septal defect, absent pulmonary valve, and a 3.5 x 2.4 cm hyperechogenic mass in the left ventricle. CT chest showed a right aortic arch, left pulmonary artery, and a right-sided azygos vein. An emergent cardiac catheterization revealed extensive thrombosis of the proximal left anterior descending artery (LAD) and a distal LAD occluded by fresh thrombus. A thrombectomy in the proximal and distal segments of the LAD was performed with improvement of blood flow in the distal segment; however, the proximal segment had significant residual thrombosis. A 3.5 x 24 mm drug-eluting stent was placed in the proximal LAD and she was given eptifibatide for 18 hours.

Conclusions: This case describes the increased thrombotic potential after plasmapheresis, where blood is exposed to a foreign surface, and the heightened risk of thrombosis in cigarette smokers.

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Aortic valve stenosis or insufficiency, aortopathy, or IE are the most common presentations of BAV disease. However, sometimes it presents atypically. BAV with complete heart block was reported by Karbashi-Afshar et al in 2014 and Suzuki et al in 1993. In both cases, the aortic valve was heavily calcified and free of IE. In our case, a less substantial amount of aortic valvular calcification was present mainly limited to the right coronary cusp. We believe that slow extension of the calcific boundary surrounding the aortic annulus to include the conduction system resulted in CHB.

CASE REPORT OF AN ATYPICAL PRESENTATION OF CALCIFIED BICUSPID AORTIC VALVE

Ababneh B, Jain N. LSU Health Sciences Center, New Orleans, LA.

Case Report: Background:Bicuspid aortic valve is the most common congenital heart disease anomaly. BAV is usually an incidental finding by echocardiography and at autopsy. We report in this case an atypical presentation of a patient who has a BAV with complete heart block (CHB).

Case Report: A 37-year-old male with no significant past medical or surgical history who presented to the emergency department complaining of 24-hour history of intermittent atypical chest pain followed next day with dizziness. He denied any other symptoms at the time of evaluation. Upon arrival to the ER, the patient was complaining of dizziness. His vital signs showed marked bradycardia with normal blood pressure. An electrocardiogram (ECG) showed sinus rhythm with third degree AV block with narrow QRS escape complex; no ischemic changes. Transcutaneous pacing was initiated to treat symptomatic bradycardia, resolving his dizziness. An urgent coronary angiogram was done which showed normal coronary arteries and a temporary intravenous pacemaker was also placed. All blood work that was obtained including a complete blood count, serum electrolytes, kidney function test, liver function test, cardiac markers, and thyroid stimulating hormone were within normal limits. Urine drug screen was negative. Chest x-ray was also normal. A transthoracic echocardiography and the findings were significant for calcified bicuspid aortic valve with moderate aortic insufficiency and dilated aortic root measured 4.7 cm. Patient's underlying complete heart block remained unchanged for several days after admission prompting dual chamber pacemaker implantation. He was discharged home in a stable condition.

Discussion: Aortic valve stenosis or insufficiency, aortopathy, or IE are the most common presentations of BAV disease. However, sometimes it presents..
without any chest pain, shortness of breath, leg pains, nausea or vomiting. At triage, his blood pressure was 160/124 with a heart rate of 54. Physical exam revealed bradycardia with normal S1 and S2, non-distended jugular venous pulsation, and equal distal pulses. Lab results revealed a stable chronic anemia, an acute kidney injury with a creatinine of 1.53 mg/dL and a negative troponin. EKG was negative for any ischemic changes but demonstrated LVH. Initial chest X-ray was significant for a moderately enlarged pericardial silhouette and prominence of the ascending arch of the aorta. A transthoracic echo demonstrated a circumferential pericardial effusion without tamponade physiology and a dilated ascending aorta up to 45mm. CT scan showed interval development of an ascending aortic rupture with an associated mural thrombus. Emergent ascending and transverse arch replacement with aortic valve resuspension was successfully performed.

**Discussion:** Management of aortic aneurysms relies on early recognition, aggressive medical management and prophylactic surgery when indicated to avoid dissection and rupture. The classic presentation of aortic rupture includes severe chest pain radiating to the back with hemodynamic compromise. Painless dissections account for 6.4% (63 out of 977 total patients) of the population. High clinical suspicion in conjunction with a high risk past medical history allowed the expedient diagnosis and repair of asymptomatic aortic rupture in our patient who presented with likely unrelated leg weakness.

**Case Report:** A 58-year-old female with hypertrophic obstructive cardiomyopathy (HOCM) was also being treated for acute myelocytic leukemia (AML). Her medical management for HOCM included metoprolol and dil-tiazem. During induction chemotherapy she experienced atrial fibrillation and worsening dyspnea. Several weeks later she was admitted to the hematology service for consolidative chemotherapy, where she experienced an episode of chest pressure that lasted for 30 minutes.

Echocardiography confirmed asymmetric septal hypertrophy and systolic anterior motion of the mitral valve, with a peak instantaneous left ventricular outflow tract (LVOT) gradient of 66 mmHg at rest. Coronary angiography was normal. Invasive hemodynamic assessment showed a resting LVOT gradient that ranged from 50-70 mmHg, which increased to 100-110 mmHg during Valsalva maneuver. Post-provocation the gradient increased to 200-230 mmHg with an aortic tracing showing the classic decreased pulse pressure and "spike and dome" appearance of dynamic obstruction.

Due to ongoing chemotherapy and unstable blood counts, we proceeded with percutaneous alcohol septal ablation rather than surgical myectomy. Timing was coordinated with completion of chemotherapy, prior to nadir of the patient's cell counts. Cell counts the morning of the procedure were a white blood cell count of 1.7 10^9/L, hemoglobin of 7.7 g/dL, platelets of 30 10^9/L, for which she received pre-procedural transfusions of red blood cells and platelets. During the procedure, the left main coronary artery was engaged via a right radial approach. A patient-specific septal perforator was occluded with a balloon, and after echocardiography confirmed appropriate localization with contrast injection, 2.1 mLs of pure ethanol was injected. Follow-up assessment confirmed excellent result. The resting LVOT gradient was 5-10 mmHg and was less than 70 mmHg with provocation. The remainder of her hospital course was uneventful. She subsequently received consolidative chemotherapy without cardiac complications.

This is, to the best of our knowledge, the first report of percutaneous alcohol septal ablation for HOCM in a patient actively undergoing chemotherapy for leukemia.

**Case Report:** AN EMERGING CAUSE OF AORTITIS

Foster MB, Jain N, Atluri P. LSU-New Orleans, New Orleans, LA.

**Case Report:** We report a particularly unusual case of a 66- year male with a past history of hypertension and dyslipidemia who was found to have panuveitis involving the right eye. The patient presented to Ophthalmology clinic with 4 days of decreased vision and increasing pain, redness, and discharge from his right eye and otherwise normal vitals. CT chest revealed an aneurysmal aortic root estimated at 4cm in diameter with a second proximal descending aortic aneurysm that was 5.2cm. Further workup revealed a bicuspid aortic valve and severe aortic stenosis. The left ventricular size and function were normal by transthoracic echocardiography.

His CBC and chemistry panel was normal. His initial inflammatory markers were abnormal with a CRP of 25 mg/dL and an ESR of 85 mm/hr. Quantiferon gold testing was within normal limits. Rheumatoid factor, anti-nuclear antibody, double stranded-DNA, complement levels were within normal limits. The patient had elevated immunoglobulin levels measuring 1700 mg/dL. IgG subclass level was elevated at 204 mg/dL. The patient underwent aortic valve replacement with mechanical prosthesis. Transaortic endovascular graft was also performed at level of descending aortic aneurysm. Pathology revealed lymphoplasmacytic infiltrate within the aorta and testing revealed plasma cells staining positive for IgG4.

Noninfectious aortitis is made up of three primary categories: Systemic vasculitides, isolated aortitis, and chronic periaortitis. The differential diagnosis of inflammatory aortitis caused by rheumatologic disease and vasculitides includes giant cell arteritis, Takayasu arteritis, rheumatoid arteritis, lupus, psoriatic arthritis, ankylosing spondylitis, Behcet’s disease, polychondritis, reactive arthritis, and the more recently recognized IgG4-RSD. This disease is an autoimmune disorder characterized by a diffuse inflammatory reaction rich in IgG4-positive plasma cells associated with sclerosis and obliterative phlebitis. Initially described as a cause of autoimmune pancreatitis, IgG4-RSD has now been recognized as a protein condition capable of insidiously involving multiple organs. Elevated serum globulin, IgG, and IgG4 are commonly seen. The three pathologic hallmarks of this disease are lymphoplasmacytic infiltration, sclerosis, and obliterative phlebitis.

**SUBVALVULAR AORTIC STENOSIS**

Joseph G. Louisiana State University; New Orleans, LA.

**Case Report:** Review of the literature revealed limited evidence and guideline on the management of asymptomatic subvalvular aortic stenosis.

This case is presented to help revisit the literature on the diagnostic approach and management of patient with asymptomatic subvalvular aortic stenosis. A 30 yo male presented to clinic for bacterial skin infections. Physical exam revealed Systolic Ejection murmur grade 2/6 with no radiation. Patient was asymptomatic with no chest pain, syncope, nor heart failure symptoms. Patient denied any history of congenital heart diseases and no family history of sudden cardiac death. Laboratory evaluation was unremarkable. ECG normal sinus rhythm. 2D echocardiogram showed evidence of sub-aortic membrane and subaortic stenosis.

Left Heart catheterization showed LVEDP(left ventricular end diastolic pressure) 18-20 mmHg , LV 133/18 mmHg Ao 127/77 mmHg. A gradient across the subvalvular membrane of 6 mmHg. Decision was made to defer surgical intervention at this time and follow patient clinically with periodic imaging.

Subvalvular Aortic stenosis is the 2nd most common form of AS. Among children with AS, subvalvar AS accounts for 10 to 14 percent of cases. Of these, it is more common in males with about 67 to 75 % of cases. There are a variety of lesions that can occur alone or in combination including: 1. thin membrane (the most common lesion). 2. Thick fibromuscular ridge. 3. Diffuse tunnel-like obstruction. 4. Abnormal mitral valve attachments. 5. Occasionally, accessory endocardial cushion tissue.

Recommendation for Asymptomatic patient are:

Surgical intervention is indicated for: 1. Infant (1st Decade) with LVOT Peak gradient >50 mmHg. 2. Children with gradient >30 mmHg. 3. At least Mild Aortic regurgitation.

Regular Follow up is recommended for: 1. Children with LVOT(left ventricular outflow tract) gradient <30 mmHg and no significant LVH/left ventricular hypertrophy). 2. We performed echocardiography for any change in symptoms or the examination. 3. Unoperated adults with stable gradients < 50 mmHg without LVH.

**ALCOHOL SEPTAL ABLATION FOR HYPERTROPHIC OBSTRUCTIVE CARDIOMYOPATHY IN A PATIENT WITH ACUTE MYELOCYTIC LEUKEMIA**

Owens JT, Hess M, Gertz Z. VCU Medical Center, Richmond, VA.

**Case Report:** SUBV AL VULAR AORTIC STENOSIS

Joseph G. Louisiana State University; New Orleans, LA.

**Case Report:** Review of the literature revealed limited evidence and guideline on the management of asymptomatic subvalvular aortic stenosis. This case is presented to help revisit the literature on the diagnostic approach and management of patient with asymptomatic subvalvular aortic stenosis. A 30 yo male presented to clinic for bacterial skin infections. Physical exam revealed Systolic Ejection murmur grade 2/6 with no radiation. Patient was asymptomatic with no chest pain, syncope, nor heart failure symptoms. Patient denied any history of congenital heart diseases and no family history of sudden cardiac death. Laboratory evaluation was unremarkable. ECG normal sinus rhythm. 2D echocardiogram showed evidence of sub-aortic membrane and subaortic stenosis.

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5:00 PM
Thursday, February 26, 2015
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RARE PRESENTATION OF A PARATHYROID CARCINOMA ASSOCIATED WITH BROWN TUMORS
Babbar A, Bohatch L, House J. University of Florida College of Medicine, Jacksonville, FL.
Case Report: A 36 year old African American man with no medical history was referred to the hospital for abnormal labs. The patient went to his PCP for evaluation for a painless neck mass that had been enlarging for ten months. Associated symptoms were right sided chest pain with inspiration, nausea, vomiting, polyuria, mild constipation, and intermittent arm and leg pain for two months. Vital signs on admission were significant for a BP of 152/108. Physical exam revealed a 4 cm firm, painless, well circumscribed, mobile mass on the left anterior neck, lateral to the thyroid cartilage. Initial labs in the ED revealed a calcium level of 19 and the patient was admitted. EKG revealed normal sinus rhythm and a QTC of 404 ms. Chest X-ray showed a 5.6 cm right upper lobe mass with adjacent rib destruction. CT chest showed diffuse sclerosis of the regional skeleton with multiple lytic bony lesions. CT neck showed a heterogeneous mass with solid and cystic components with peripheral calcifications. PTH level was 61% male. Compared to the pre-pathway group, children on the pathway were more likely to be admitted and to receive more nebulizer treatments while in the ED. More study is needed to determine the effects of this pathway ED metrics.

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while less than 1% of the time parathyroid carcinoma is implicated. The age of onset is 45-59. The exact pathogenesis is unknown. It can occur as part of a hereditary syndrome. Unlike adenomas, parathyroid carcinoma can manifest as a palpable neck mass and hypercalcemic crisis with calcium >14, altered mental status, weakness, azotemia, and cardiac arrhythmia. Our patient had a unique presentation with only mild symptoms of hypercalcemia and a normal EKG with normal QTc, despite a calcium level of 19. Our patient also had multiple brown tumors present. Brown tumors are present in only 4-15% of patients with parathyroid adenomas. They are slow growing and typically present late. There are only a few reported cases of brown tumors secondary to parathyroid carcinoma. Carcinomas grow at a more rapid pace, which leaves inadequate time for brown tumors to form.

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SIGNIFICANTLY INCREASED BASE DEFICIT ON PRESENTATION IS ASSOCIATED WITH SEVERE COURSE OF DIABETIC KETOACIDOSIS IN CHILDREN
Bhatt P1, Singh D1, Akingbola O1, Srivastava S2, Tulane University School Of Medicine, New Orleans, LA; Tulane University School of Public Health and Tropical Medicine, New Orleans, LA and Tulane University School of Public Health and Tropical Medicine, New Orleans, LA.

Purpose of Study: To determine whether significantly increased baseline deficit (BD >20) predicts severity of DKA course in PICU

Methods Used: This is a retrospective review of medical records performed in children ≤18 years old admitted to PICU with DKA from January 2007 through December 2013. Demographic, hemodynamic, laboratory, and treatment data was collected from the time of admission to ED till discharge from the hospital.

Summary of Results: 31 patients with a total of 52 episodes presented with DKA during study period but only 25 patients with 37 episodes (age range 3-17 years; Mean±2;3.05 yrs) were eligible and included in the study. 51% had BD> -20 (Group (Gr) 1; n=19;Mean -24.31 ± 3.99) and 49% had a BD> -20 (Group (Gr) 2; n=18;Mean -13.70±3.82). 26% had new onset DKA in Gr1 compared to 16.66% in Gr2. Gr1 was associated with increased hospital LOS (mean 5.84 days vs 4.22 days; OR 1.77; p<0.001), increased PICU LOS (mean 49.68 hours vs 29.89 hours; OR 1.03; 95%CI 1.03 - 1.18; p<0.003), increased duration of insulin infusion in PICU (mean 29.68 hours vs 14.50 hours; OR 1.13; 95%CI 1.03 - 1.23; p<0.005), higher cumulative i.v. insulin/kg (mean 55.13 units/kg vs 11.00 units/kg; OR 9.23; 95%CI 2.06 - 41.40; p<0.003), higher cumulative i.v. fluids/kg (mean 160.26 ml/kg vs 77.81 ml/kg; OR 1.02; 95%CI 1.00 - 1.05; p=0.003), higher corrected anion gap mean (55.16 vs 27.40 ml/kg; OR 1.04; CI 1.010-1.083; p=0.01) & required longer duration of i.v. insulin infusion & duration of stay in PICU.

Conclusions: Younger children with DKA with lower BMI on presentation are more severely dehydrated & have more severe ketoacidosis as evidenced by increased negative BD & corrected anion gap & lower pH, & require longer duration of i.v. insulin infusion & duration of stay in PICU.

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METFORMIN HAS A POSITIVE THERAPEUTIC EFFECT ON PROSTATE CANCER IN DM2 PATIENTS
Chong RW1, Vasudevan V1,2, Zubir J1, Solomon S1,2. 1University of Tennessee Health Science Center, Memphis, TN; 2Veterans Association Medical Center, Memphis, TN and Veterans Association Medical Center; Memphis, TN.

Purpose of Study: Prostate cancer and type 2 diabetes mellitus are both common diseases found in the elderly male population. The diabetic drug, metformin, has been shown to have anti-neoplastic properties and demonstrated better treatment outcomes when used as adjuvant therapy in breast cancer patients. Some proposed signaling pathways include inhibition of mTOR and activation of tumor suppressor genes p53 and LKB1 via AMPK. The analogous hormonally-sensitive cancer in men is prostate. We investigate survival, risks of recurrences, and levels of prostate specific antigen (PSA) in DM2 patients with prostate cancer on metformin using CPRS in the VA system (≥287 patients).

Methods Used: Prostate cancer patients with type 2 diabetes that remained on metformin were compared to controls not on metformin matched by age, weight, race, and Gleason score cancer staging. The endpoints of our study included final PSA values, number of recurrences, metastases, secondary cancers, and number living for each group. Final hemoglobin A1C (HbA1C) and creatinine levels were obtained and compared in both groups. Inclusion and exclusion criteria were identified. Statistical analysis included unpaired t-test and Chi-squared test.

Summary of Results: There were significantly fewer deaths (23% vs10%), fewer recurrences (15% vs 8%), and fewer metastases (5% vs 0%), and fewer secondary cancers (17% vs 6%) in the metformin group (p<0.004). The final PSA value was lower in the metformin-treated group with a result approaching significance (p=0.067). The primary treatments for prostate cancer (i.e. surgery, radiation, androgen depletion) were found to be comparable in both groups.

Conclusions: Our retrospective study shows that adjuvant use of metformin leads to a significantly improved prognosis in prostate cancer. Not only are PSA levels controlled for several years, but there are fewer cancer recurrences in metformin treated patients. HbA1C levels were similar in both groups, so increased mortality in the control group was not due to diabetes. Overall, these results are promising on their own but should be followed up with a prospective study or clinical trial.
STOPPING THE STORM, WHEN THYROID STORM REQUIRES PLASMAPHERESIS
Coleman-Pierre R, Gneisert A, Caruthers C, Tadin D, Sparks K, Dinh P, Engel LS. LSU Health Sciences Center, New Orleans, LA.

Case Report: A 51 year old woman with Graves’ disease presented with abdominal pain with progressive distention, worsening dyspnea on exertion to shortness of breath at rest and palpitations for 4 weeks. The patient was noncompliant with atenolol and methimazole. On triage, she was in atrial fibrillation with a heart rate (HR) of 160 to 170 beats per minute. TSH was 0.1 (0.35-5.50) with free T4 = 6.01 (0.62-1.43) ng/dL, total T3 5.94 (0.6-1.81) ng/mL and free T3 32.2 (2.4-4.2) pg/mL. Esmolol drip and methimazole were started. On day 2, her HR remained 140 to 150. After receiving iodine drops, the patient developed asystole. ACLS was initiated. After intubation, the patient spontaneously converted to sinus rhythm. Hydrocortisone was started and methimazole was increased. Atrial fibrillation, HR 160 to 170, returned on day 5 and was refractory to labetalol, digoxin, diltiazem, esmolol and procainamide. Otolaryngology was consulted, but the patient was too hemodynamically unstable for thyroidectomy. Given no improvement in HR on maximum medications, plasmapheresis was initiated. After day 1 of plasmapheresis, sinus rhythm returned and HR dropped to 30. Atropine was administered, esmolol was discontinued, and dopamine was started. On day 6, atrial fibrillation, HR 160 to 180, returned refractory to same medications. After 3 days of plasmapheresis, free T4 decreased from 3.98 to 1.58 ng/dL. Heart rate improved. Oral propranolol and sotalol were started but discontinued for bradycardia. Hydrocortisone wean was initiated. The patient discharged to home asymptomatic with free T4 of 1.6 ng/dL.

Discussion: Thyroid storm typically presents with exaggeration of hyperthyroid symptoms. Tachycardia greater than 140 and congestive heart failure are common. Mortality rate of thyroid storm is 75% to 90% untreated and 10% to 30% treated. Conventional therapy reduces the release of thyroid hormone from the thyroid gland with iodine, blocks the formation of thyroid hormone with thioureas and reduces the effects of thyroid hormone with adrenergic blocking agents. Plasmapheresis can reduce thyroxine levels by 25% by the direct removal of free thyroid hormones. Plasmapheresis can be a life saving bridge to definitive therapy.

INFLAMMATORY BIOMARKERS AND CLINICAL OUTCOMES IN PEDIATRIC TYPE 1 DIABETES PATIENTS
Coulon SJ 1, Hempe J 1, Velasco-Gonzalez C 2, Scribben R 1, Stender S 1. LSU Health Sciences Center, New Orleans, LA and ‘LSU Health Sciences Center, New Orleans, LA.

Purpose of Study: This pilot study examines correlations between inflammation in youth with type 1 diabetes (T1D), glycemic control (mean blood glucose (MBG), HbA1c), and social determinants of health (SDH). Inflammation is linked to the development of T1D and progression of diabetic complications. Hyper/hypoglycemia and insulin increase levels of inflammatory cytokines; these and hyperglycemia-induced free radicals can cause oxidative stress. Likewise, sub-optimal SDH may create an inflammatory milieu that exacerbates chronic inflammation. A public health model for studying SDH categorizes these conditions: physical activity/food/social environment. Concentrated disadvantage index (CDI) is a combined variable that is a proxy for these conditions. We hypothesized associations between plasma cytokines and CDI impacting diabetes control.

Methods Used: Children with T1D attending diabetes clinic at Children’s Hospital, New Orleans (suburban academic) were randomly recruited and enrolled (IRB consent) (n = 35; male/female: 20/18; black/white: 19/19; mean age (y): 14 ± 3.4; mean HbA1c (%): 9.8 ± 2.3; mean MBG (mg/dL): 224.3 ± 77.5; mean BMI-z score: 0.51). Inflammatory biomarkers (pro-inflammatory: IFNy, IL1b, IL6, IL8, IL12p70, TNFα, VEGF, CRP; anti-inflammatory: IL1ra, IL4, IL10) were measured in the plasma (immunoassay).

Summary of Results: MBG was significantly inversely correlated with TNFa (r = -0.35, p = 0.02) and IL10 (r = -0.68, p = 0.03) (remained after adjustment for age, gender, race, CDB, or BMI-z). CRP was marginally positively correlated with HbA1c (r = 0.30, p = 0.07) (disappeared after adjustment for CDB or BMI-z).

Conclusions: Some plasma cytokines have a significant indirect relationship with MBG while others have a direct relationship with HbA1c. Increasing sample size in this ongoing study may determine if sub-optimal diabetes control is associated with inflammatory biomarkers and whether or not these relationships are confounded by SDH.

A CASE OF BILATERAL PRIMARY ADRENAL LYMPHOMA
Smith MM, Chiang MC, Spiegel JC, Suer AW, Engel LS, Uwaiso G, de Silva TN, Guirly SG. LSU Health Sciences Center, New Orleans, LA.

Case Report: A 57 year old woman with a history of essential hypertension and primary hypothyroidism presented with an 8 month history of bilateral upper and lower extremity edema. Symptoms included shortness of breath, increased abdominal size, and a door sign of thyroid storm. Treatment includes therapy with a beta blocker to block hyper/hypoglycemia and insulin increase levels of inflammatory cytokines; these and hyperglycemia-induced free radicals can cause oxidative stress. Likewise, sub-optimal SDH may create an inflammatory milieu that exacerbates chronic inflammation. A public health model for studying SDH categorizes these conditions: physical activity/food/social environment. Concentrated disadvantage index (CDI) is a combined variable that is a proxy for these conditions. We hypothesized associations between plasma cytokines and CDI impacting diabetes control.

Discussion: Thyroid storm typically presents with exaggeration of hyperthyroid symptoms. Tachycardia greater than 140 and congestive heart failure are common. Mortality rate of thyroid storm is 75% to 90% untreated and 10% to 30% treated. Conventional therapy reduces the release of thyroid hormone from the thyroid gland with iodine, blocks the formation of thyroid hormone with thioureas and reduces the effects of thyroid hormone with adrenergic blocking agents. Plasmapheresis can reduce thyroxine levels by 25% by the direct removal of free thyroid hormones. Plasmapheresis can be a life saving bridge to definitive therapy.
initiated on day 3 and continued for 4 days. CT obtained on day 6 revealed large cystic rim-enhancing lesion with internal septations in the thyroid gland concerning for an abscess. US guided neck biopsy was performed on day 7. Twenty mL of purulent fluid was aspirated from an abscess in a branchial cleft cyst. Fluid cultures grew streptococcus anginosus, eikenella corrodens, and multiple anaerobic bacteria.

EG’s thyrotoxicosis was precipitated by destruction of thyroid tissue due to an abscess in the left 3rd/4th branchial cleft sinus. Cysts of the third and fourth branchial cleft can cause recurrent abscesses or suppurative thyroiditis. The cleft will often become infected with anaerobes and other oral flora as seen in EG’s case. Only with appropriate treatment of the infection, drainage and antibiotic therapy, thyrotoxicosis resolves. Importantly, this type of presentation resolves independently of anti-thyroid medication because the storm is precipitated by thyroid tissue destruction.

Galactose was administered to reduce the half-life of thyroxine and as a trial to reduce the inflammatory response. The patient was continued on MMI and PTU. A large cystic rim-enhancing lesion with internal septations in the thyroid gland was noted on the CT scan performed on day 3. A course of antibiotics was initiated on day 3 and continued for 4 days. CT obtained on day 6 revealed a large cystic rim-enhancing lesion with internal septations in the thyroid gland concerning for an abscess. US guided neck biopsy was performed on day 7. Twenty mL of purulent fluid was aspirated from an abscess in a branchial cleft cyst. Fluid cultures grew streptococcus anginosus, eikenella corrodens, and multiple anaerobic bacteria.

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**ORAL GALACTOSE SUPPLEMENTATION IN PGM1-CDG: AS SWEET AS IT SOUNDS?**

Gadomski TE, Wong S, Preston GJ, Scott K, Morava E.

**AS WEE SWEET AS IT SOUNDS?**

Gadomski TE, Wong S, Preston GJ, Scott K, Morava E. Tulane University School of Medicine, New Orleans, LA.

**Purpose of Study:** Congenital disorders of glycosylation (CDGs) are diverse genetic syndromes caused by impaired glycoprotein synthesis. Phosphoglucomutase 1 deficiency (PGM1-CDG) is a newly recognized CDG with a multi-system phenotype. Patients present with decreased protein glycosylation, hypoglycemia, abnormal liver function, muscle involvement and endocrine dysfunction.

PGM1 enzyme is essential for glycosylation as well as glucose and galactose metabolism. In PGM1-CDG, the glycans chains on proteins lack galactose. Previous in vitro studies in patient fibroblasts showed restored galactose in the glycans chains after galactose supplementation. We designed an observational study to characterize the clinical and biochemical diversity in PGM1-CDG and to evaluate the outcomes of PGM1-CDG patients receiving dietary galactose supplementation.

**Methods Used:** Participants in our study received oral galactose supplementation over 18 weeks. D-Galactose intake started at 0.5 g/kg per day, increasing to 1.0 g/kg per day after 6 weeks and to 1.5 g/kg per day after 12 weeks. Maximal daily dose of galactose was 50.0 g, an amount that is within the recommended daily intake. Laboratory and biochemical analyses were completed every 6 weeks to assess glycosylation, growth, biochemical and metabolic parameters as well as endocrine and liver function. Dietary and clinical evaluations were performed before and after galactose supplementation.

**Summary of Results:** Noted benefits of galactose supplementation varied with PGM1-CDG phenotype. Clinical changes observed were improved exercise tolerance and coagulation status. Laboratory data demonstrated that liver function and coagulation abnormalities responded quickly to galactose supplementation but endocrine changes required longer treatment periods. Participant creatine kinase and glucose levels remained variable.

**Conclusions:** Our study confirms the beneficial effects of galactose supplementation in patients with PGM1-CDG. Controlling hypoglycemia and muscle dysfunction remain as challenges due to glycogen release anomalies in PGM1-CDG. Patient compliance to supplementation alters the effects of galactose supplementation. Further studies are in progress to improve dosing efficacy and to determine which clinical parameters respond best to galactose supplementation.

**PTU-INDUCED HEPATITIS IN GRAVES’ DISEASE**

Henderson D, Steinberg H. University or Tennessee health Science Center, Memphis, TN.

**Case Report:** Graves’ disease (GD) is one of the most common causes of thyrotoxicosis. Hepatic injury in this setting has been observed and can be due to multiple factors. Liver toxicity due to thionamides, the mainstay of treatment, is estimated to occur in less than 0.5% of patients. It is more common with propylthiouracil (PTU) than with methimazole (MMI). Liver dysfunction sets limitations on therapeutic options in patient with thyrotoxicosis. Evidence has highlighted, amongst others, treatment strategies such as steroids, beta-blockers, potassium iodide, and plasmapheresis in the setting of severe thyrotoxicosis refractory to thionamides or in cases of liver injury. Our case highlights the dilemma of treating thyrotoxicosis with goiter in the setting of PTU induced liver toxicity.

A 26 year old female presented to the emergency room with complaints of weakness, tremors, weight loss, palpitations, hair loss, and bulging eyes. She had dysphagia to solids and dyspnea while supine. She was diagnosed with GD in 2013 and, at that time treated with PTU inpatient and discharged on MMI. She developed a diffuse rash to MMI and was changed to PTU 50 mg every 8 hours. She ran out of PTU and propranolol 6 weeks prior to the current admission. Physical exam noted proptosis and goiter with mild right upper quadrant tenderness. Thyroid was not palpable and liver edge was not felt. She had diffuse hyper trophy of the thyroid that was not palpable and liver edge was not felt. She had diffuse hyper trophy of the thyroid that was not palpable.

**Case Report:** We report a case of severe thyrotoxicosis refractory to thionamides or in cases of liver injury. Our case highlights the dilemma of treating thyrotoxicosis with goiter in the setting of PTU induced liver toxicity.

A 26 year old female presented to the emergency room with complaints of weakness, tremors, weight loss, palpitations, hair loss, and bulging eyes. She had dysphagia to solids and dyspnea while supine. She was diagnosed with GD in 2013 and, at that time treated with PTU inpatient and discharged on MMI. She developed a diffuse rash to MMI and was changed to PTU 50 mg every 8 hours. She ran out of PTU and propranolol 6 weeks prior to the current admission. Physical exam noted proptosis and goiter with mild right upper quadrant tenderness. Thyroid was not palpable and liver edge was not felt. She had diffuse hyper trophy of the thyroid that was not palpable and liver edge was not felt. She had diffuse hyper trophy of the thyroid that was not palpable.

**A CASE OF SEVERE PRIMARY HYPERPARATHYROIDISM IN A SIX YEAR OLD RESULTING IN A FALSE POSITIVE SESTAMIBI SPECTRIGRAPHY IN AREAS OF BROWN TUMOR**

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Case Report: Primary hyperparathyroidism (PHPT) causes hypercalcemia secondary to exaggerated secretion of parathyroid hormone (PTH). It is very rare in children and because of its low prevalence and ambiguity of symptoms often remains undiagnosed until the child experiences end organ damage. Most often PHPT is caused by an adenoma and these lesions are commonly detected by ultrasound or MRI in children. In adults, technetium-99m sestamibi scintigraphy is commonly used to detect adenomas and this imaging modality is becoming more popular in children.

A 6 year old African American male presented with hematuria, leg pain, and headache. Physical exam was normal except for a 0.5 cm fixed nodule on the right side of his neck. Serum calcium was 15 mg/dl (8.8-10.1 mg/dl) and PTH intact 343 pg/ml (9-59 pg/ml). Sestamibi scintigraphy showed an intense area of uptake overlaying the right thyroid gland and increased activity over the right and left clavicles and the left shoulder area. This was concerning for a metastatic lesion or a brown tumor. Thyroid ultrasound and neck/chest MRI ruled out a metastatic lesion making the increased areas of uptake most consistent with a brown tumor.

His pituitary adenoma was removed and he suffered from hungry bone syndrome evidenced by a precipitous drop in his calcium and phosphorus. Knowing that he had a brown tumor on imaging enhanced preparation and closer monitoring of his electrolytes.

Primary hyperparathyroidism is rare in children and clinicians must have a high index of suspicion when patients present with vague symptoms such as fatigue, hematuria, abdominal pain and headache which are unexplained by other pathology. The child in this case was young for this disease process (median age 36 years) and had rather extreme hypercalcemia (peak at 174 mg/dl). His sestamibi scan revealed areas of uptake near the clavicles. False positive findings with this imaging modality are rare and can represent other tissues such as lung, brain, bone, carcinoid tumors, lymphoma, and thymomas. In this case the increased area of uptake represented a brown tumor which suggested more long standing disease. Knowing this helps one to anticipate more drastic post-operative electrolyte changes and intervene appropriately.

A SWEET CASE OF CHOREA
Modica MD, Nesh KJ, Engel LS, Masri N. LSU Health Sciences Center, New Orleans, LA.

Case Report: INTRODUCTION: Chorea is an involuntary, irregular, poorly patterned movement that often times can take on a worm like appearance termed athetosis. This movement disorder is associated with a wide variety of illness ranging from primary hereditary disease such as Huntington’s, to secondary causes including cerebrovascular, para-neoplastic, metabolic, inflammatory and immunologic diseases. Of the metabolic causes, hyperparathyroidism and obesity stand out.

Case: A 71 year old African American woman with past history of diabetes mellitus type II presented to the emergency room with a chief complaint of uncontrollable spasm and odd movements in her left wrist of five days duration. Her blood sugar had been running high over the last several after a recent change in medication to iraglutide and glipizide. Physical exam showed sporadic choreiform movement of the left wrist and left arm with no other significant findings, including no other focal findings on the rest of her neurologic examination. Labs at the time of admit were significant for blood glucose of 1013 mg/dl with normal anion gap and without presence of ketones in the blood or urine. The patient was started on a basal insulin regimen. Her blood glucose decreased to 563 mg/Dl and the choreiform movements dissipated. MRI/MRA of the brain showed no acute findings. Pneumocystis pneumonia. A cortisol stimulation test demonstrated a post stimulation cortisol level of 10 mcg/dl. The patient was placed on hydrocortisone 100 mg every 6 hours and his blood pressure was 204/109 mmHg. Her blood pressure improved with an adjusted medication regimen and she was discharged feeling much better, headache free. Unfortunately, her headache quickly returned the day after discharge. She reported feeling very dizzy and almost fell, and she then returned to the ED. CT without contrast showed no acute intracranial abnormality, CSF studies were within normal limits with negative cultures, Renal U/S was done and was noted be within the normal limits. A 24 hour urine Metanephrine revealed an elevated value of 2327 mg, a 24 hour urine Normetanephrine showed a result of 9988 , and a urine 24hr catecholamine fraction of 555 ug/m2. The patient was seen in clinic and initially prescribed an alpha blocker and later a beta blocker with referrals to Endocrinology and General Surgery. A CT of her Abdomen and Pelvis with contrast confirmed a 4.6 cm portocaval mass most consistent with a extra adrenal phaeochromocytoma. General surgery was able to remove the mass without any complications. Surgical pathology demonstrated a benign phaeochromocytoma, 5cm in greatest dimensions with negative surgical margins for tumor.

Discussion: Patients with phaeochromocytomas may have either persistently high blood pressure or episodic peaks in blood pressure related to catecholamine surges. Secondary causes are hyperparathyroidism which can present with symptomology such as flushing/sweating (Phaeochromocytoma), presence of a renal bruit (renal artery stenosis), or laboratory abnormalities such as hypokalemia (Alddosteronism). Furthermore, patients with resistant hypertension or early or late onset of hypertension should undergo screening for secondary etiologies.

CENTRAL ADRENAL INSUFFICIENCY WITH CONCURRENT PRIMARY ADRENAL INSUFFICIENCY
Smith MM, Varughese AG, Engel LS, Lo-Blais B, Uwaiifo G. LSU Health Sciences Center, New Orleans, LA.

Case Report: A 43-year-old man with AIDS (CD4 count 13 cells/mm3, viral load > 1 million copies) presented with acute onset of lower back weakness, which progressed to whole body generalized weakness. He had anorexia, shortness of breath, and difficulty standing after micturition, followed by post micturition syncope and persistent hypotension. Physical exam demonstrated oral thrush, bibasilar rales, and hyperesthesia of the feet.

CT Brain demonstrated no acute intracranial abnormalities but lumbar puncture was positive for C. neoformans. He had multiple lab abnormalities including Hgb 7.1 g/dl, HCT 20.6%, and Platelets 99 x10^9/L. CMP showed Potassium of 5.9mmol/L, Chloride 114 mmol/L, CO2 19 mmol/L, and BUN/ Creatinine of 37 mmol/L/ 3.29mg/dl. Sputum studies demonstrated pneumocystis pneumonia. He underwent treatment for Cryptococcus meningitis and Pneumocystis pneumonia. A cortisol stimulation test demonstrated a subnormal response with baseline cortisol value of 2 mcg/dl and 60 min post stimulation cortisol level of 10 mcg/dl. The patient was placed on maintenance steroid replacement. The hypothalamic-pituitary axis work up showed repeated low levels of ACTH (<11) consistent with central adrenal insufficiency. A full adrenocortical-hypothalamic screening profile revealed FSH (1.2IU/L) and LH (2.7 IU/L), which were inappropriately low in view of the patient’s very low testosterone level (<10ng/dl), consistent with central hypogonadism. The prolactin level was 8.7 ng/ml, IGF-1 153ng/ml, and GH 6.6ng/ml, which were all normal levels. He had a low aldosterone (<1 ng/dl) and plasma renin activity (0.68ng/ml/hr) level consistent with a coexisting primary adrenal insufficiency. The anti-adrenal antibody screen was negative. The patient’s clinical condition improved with steroid and androgen replacement.
ABSI AS A PREDICTIVE DIAGNOSTIC TOOL

Tucker W, de Silva TN, Usuafa G. LSUHSC-New Orleans, New Orleans, LA.

Purpose of Study: Body mass index (BMI) is a widely used index to predict morbidity and mortality linked to obesity. A limitation of BMI is its failure to account for fat distribution. Visceral adiposity is a more potent indicator of morbidity and mortality than BMI. Waist circumference (WC) provides information regarding degree of visceral adiposity. "A Body shape Index" (ABSI) includes WC, height and weight in describing degree of adiposity and clinical risk. ABSI can be used to determine severity and complications associated with obesity.

The intent of this study was to quantify the correlation of ABSI to comorbidities associated with obesity. We compared the correlations of these variables with ABSI and BMI in predicting clinical surrogates of obesity. Data was obtained from the COSMOS (Clinical Outcomes Study of Morbidly Obese Subjects) program, an underserved population of patients with severe obesity. Data repositories in EPIC and CLIQ were interrogated and collated in an Excel spread sheet.

The study investigated correlations between ABSI and chosen parameters as compared to BMI. This study illustrates the need for further evaluation in larger populations and could influence clinical guidelines regarding the addition of WC to basic vital signs.

Methods Used: 131 patients in the COSMOS program met the inclusion criteria. We used the published equation for ABSI, based on WC and adjusted for height and weight: ABSI = WC / BMI^2 / height^0.5. Data analyses were done using JMPin and Microsoft Excel 2008.

Summary of Results: ABSI had statistically significant correlation with ALT and triglycerides (R^2 = 0.02 and 0.09 respectively with p-values < 0.05) compared to BMI. BMI had an increased correlation with cholesterol (R^2 = 0.053 with a p-value < 0.05) compared to ABSI. In a multivariate correlation analyses ABSI had a stronger correlation with hyperinsulinemia, hypertriglyceridemia, and coronary artery disease (R^2 = 0.52, 0.51, 0.46 respectively with p-values < 0.05) compared to BMI. In this multivariate correlation analyses, BMI had higher correlation with hyperglycemia (R^2 = 0.412) compared to ABSI.

Conclusions: ABSI correlates with single and multivariate variables associated with co-morbidities of obesity. While further studies of the clinical application of ABSI are needed, given the ABSI is a cost efficient tool with low procedural risk, it could be a valuable diagnostic tool in the evaluation of obese patients.

CHARACTERISTICS OFEmerging ADULTS ENROLLED IN DIABETES TRANSITION PROGRAM

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Purpose of Study: Diabetes mellitus (DM) in the pediatric population has significantly increased in the past 50 years. Therefore, there is an increasing number of young adults transferring from pediatric to adult diabetes providers. The Diabetes Transition Program (DTP) at the Medical University of South Carolina (MUSC) works to improve the transition of care from pediatrics to adult services. This program provides a multidisciplinary team approach that supports patients with a goal of increasing the number of annual clinic visits, improving A1C, and decreasing the number of diabetic ketoacidosis (DKA) admissions. This study was designed to characterize the initial cohort of emerging adults that have transitioned into adult care.

Methods Used: A retrospective chart analysis was conducted from June 2013 to May 2014. 53 total patients were examined, 26 were identified as no longer active pediatric patients and thus considered "failure to capture". The remaining 37 patients were divided into two groups: 1) completed the transition into adult care (n=20) and 2) received a referral to adult care and were lost to follow-up (n=17). The patients' demographics, diabetes and other health characteristics were collected.

Summary of Results: The average age of the overall cohort (n=37) was 19.3 ± 0.7 years, 51% female, with Non-Hispanic whites being 84%. The median A1C was 9.2%, and 11% reached their A1C goal of < 7.5%. There were 4 patients with DKA admissions in the past year, 75% of which were in the lost to follow up group; 22% of patients had ADHD and 11% had depression. 70% had an annual urine microalbumin, 60% of patients had annual thyroid tests completed, 54% had an annual lipid panel. Only 15% had 4 or more clinic visits within the past year.

Conclusions: Overall, less than half of patients transitioned into the adult system. The patients that did transition into adult care had lower A1C and fewer DKA admissions. Future initiatives include, hiring a patient navigator in order to increase the capture rate of patients and improving clinical outcomes by decreasing A1C, decreasing DKA admissions, and increasing annual screening.

GASTRIC CARCINOID TUMOR TYPE II IN THE SETTING OF METASTATIC GASTRINOMA-A RARE COEXISTENCE

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Case Report: A 60 yr old WM was evaluated for chronic diarrhea and anemia. He reported nausea, emesis, early satiety, weight loss and episodic facial flushing. Physical examination revealed only epigastric tenderness. Lab results showed mild anemia. An EGD showed a 6 mm nodule in the gastric body and biopsy confirmed gastric carcinoid. A CT abdomen showed multiple metastatic liver lesions. Endoscopic ultrasound of UGI showed a hyperechoic mass at the pancreaticoduodenal area suspicious of a Gastrinoma. Serum Gastrin level was 47000pg/mL. A 24 hr urine HBAA level was 10.1. An Octreotide (Oct) scan was positive for Oct activity within the pancreatoduodenal mass and liver lesions suggesting a neuroendocrine tumor (NET). Liver biopsies were positive for Chromogranin A (CgA), Synaptophysin (SNP) and Gastrin (G) suggesting metastatic Gastrinoma. He was treated with high dose PPI and Lanreotide.

Diabetes mellitus (DM) in the pediatric population has significantly increased in the past 50 years. Therefore, there is an increasing number of young adults transferring from pediatric to adult diabetes providers. The Diabetes Transition Program (DTP) at the Medical University of South Carolina (MUSC) works to improve the transition of care from pediatrics to adult services. This program provides a multidisciplinary team approach that supports patients with a goal of increasing the number of annual clinic visits, improving A1C, and decreasing the number of diabetic ketoacidosis (DKA) admissions. This study was designed to characterize the initial cohort of emerging adults that have transitioned into adult care.

Methods Used: A retrospective chart analysis was conducted from June 2013 to May 2014. 53 total patients were examined, 26 were identified as no
Case Report: 64 yr old AAM who presented to the emergency room with a history of a fall and right arm pain for 5 days. The patient has no significant history of recent illness. ROS revealed a 20 lb weight loss and new onset diplopia over the last six months. Physical exam was normal.

X-rays showed multiple lesions in the right humerus with a fracture. A skeletal survey revealed widely distributed lytic lesions including the rib cage and femurs. CBC demonstrated: HCT 30.8, Hgb 10.3, WBC 5.3x10^3 3* platelet count, 175,000/mm^3*, serum calcium 9.4, Bun 13, CR 1.9. Serum electrolysis confirmed the presence of a monoclonal gammopathy with a spike of 3.8mg/dl. Bone marrow biopsy was consistent with Multiple Myeloma.

The patient underwent medullary fixation of the right humerus and prophylactic fixation for impending fracture of the left, and was d/ced home. He was later admitted with fever of 102 F, chills, rigors and altered mental status. CT and MRI of the head confirmed a pituitary mass measuring 2.6cm in greatest dimension filling the pituitary fossa, extending into the left cavernous sinus and eroding the clivus. There was minimal displacement of the optic chiasma superiorly. Pituitary functions in the ICU showed FSH 0.7; LH 0.0; TSH 1.48; Free T3 1.6; Free T4 0.62; total testosterone 112.0; free testosterone 3.1; and Prolactin 3.1. All consistent with pituitary dysfunction. The patient received chemotherapy (Bortezomib) and radiation therapy and dexamethasone 40mg weekly, and lenolinamide 25mg every 2 weeks. The patient’s pituitary function was followed as outpatient.

Repeat MRI at one year showed that the pituitary gland had "markedly decreased in size decreasing from 26mm to 14mm height on the coronal images of 3mm. There is only minimal tumor in the pituitary gland and in the clivus." These findings are consistent with response of the tumor to treatment. Plasmocytomas usually present in one of 3 ways: a) sellar plasmocytoma with multiple myeloma at presentation; b) multiple myeloma developing on follow up; and c) as a solitary sellar plasmocytoma without multiple myeloma years after follow up. Cranial nerve involvement with hypofunction of the pituitary with aggressive radiological findings at the time of presentation, strongly suggests this as a possible diagnosis.

131 Sigmoid Volvulus Presenting as Severe Back Pain in an Adolescent Female

Abou Ayash H, Ponnambalam A. University of South Alabama Children and Women’s Hospital, Mobile, AL.

Case Report: Background:
Sigmoid volvulus occurs when a loop of the sigmoid colon twists around its mesentry, leading to intestinal obstruction and subsequent ischemia. It is an uncommon cause of intestinal obstruction, which usually occurs in adults. It is very rarely reported in children. Patients present with sudden onset of severe abdominal pain with gross abdominal distension and failure to pass flatus or stool, which is mistaken for constipation. It is uncommon to present with lower back pain, therefore highlighting the importance of early detection.

Objective: To present a case of sigmoid volvulus in an adolescent female with back pain and highlight the typical radiographic findings.

Case Description: 16-year-old female presented with a four-day history of progressively worsening abdominal and lower back pain. She had no stools for 7 days, and no passage of flatus for 36-48 hours. She sought medical attention on two separate occasions at outside facility for constipation and UTI. A lumbar MRI obtained due to severe back pain was normal. She returned to ER within 12 hours of discharge and was transferred to our institution due to continued severe back pain. On admission, an abdominal x-ray showed distended bowel loops with markedly dilated colon and multiple air fluid levels. An abdominal CT scan was significant for massive distalation of the proximal sigmoid and large bowel with whirl sign. It had characteristic "coffee bean" configuration. Rectal tube was placed for bowel decompression and the volvulus resolved. Anorectal manometry was normal. She subsequently underwent sigmoid resection.

Discussion: Sigmoid volvulus is a surgical emergency in patients of all ages. An accurate history and physical examination followed by prompt assessment is essential. In some instances detorsion of the bowel can be achieved with rectal tube placement alone. Findings of ischemic colonic mucosa would require immediate operative intervention. The case emphasizes the importance of early identification in an atypical patient in order to optimize management.

132 A Type II Choledochal Cyst Presenting as Pancreatic Mass

Geffen JB, Nakshabendi R, Clark M, Munoz J. University of Florida, Jacksonville, FL.

Case Report: A 55 year old male with a history of multiple episodes of recurrent pancreatitis s/p cholecystectomy presented to UF Health’s ED after having a syncopal episode. At admission, the patient was complaining of LUQ and epigastric pain, “gnawing” and mild in nature. Prior labwork included revealed normal LFT’s and Lipase 1184. Multiple, multimodal imaging studies demonstrated a hypodense lesion within the pancreatic head later characterized as a 2 cm septated cystic mass the differential of which was a serious cystadenoma, mucinous cystic neoplasm, or ductal adenocarcinoma. Further studies indicated worsening PD and CBD dilation, and peripancreatic adenopathy. A CA 19-9 was measured and was 119.

The GI service was consulted to manage the patient for acute on chronic pancreatitis. Once stabilized, an ERCP was performed. The entire biliary tree was opacified and exhibited diffusely dilated intra- and extra-hepatic ducts reaching a diameter of 12mm. The length of the common bile duct to the cystic duct junction was <20mm. A large choledochal cyst, likely type II, was noted adjacent to the cystic duct junction.

FIGURE 1. ERCP showing cystic structure maximally opacified and its relationship to CBD.

133 Eosinophilic Gastroenteritis: Not Your Average Diarrhea

Lewis KC, Bryant C, Yu Z, DeLeon S. University of Oklahoma, Oklahoma City, OK.

Case Report: Eosinophilic gastroenteritis is a rare inflammatory disorder characterized by eosinophilic infiltration of the gastrointestinal tract. Pathogenesis is not well understood, but an allergic component is suspected with up to one-half of patients having concurrent atopic disease.

Our patient is a 2 year old male with history of repaired pyloric stenosis who presented with a six week history of cyclical nausea, vomiting, and diarrhea. The patient lived on a farm where he was exposed to livestock, contaminated soil, and well water. On admission, basic labs showed an elevated peripheral eosinophil count of 13.5%. Hyperactive bowel sounds were present on physical exam but all other lab and exam findings were normal. Infectious disease and Gastroenterology were consulted, and cultures testing for bacterial, fungal, viral, and parasitic etiologies were sent. IgE level

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MIRALAX MIX UP: A CASE REPORT ON CAUSTIC INJURY TO THE ESOPHAGUS
Locke A, Phemister J, Carter L, Murthy R. ETSU Johnson City, TN.
Case Report: Ingestion of caustic agents causes a variety of injuries to the esophagus making management difficult. We describe an interesting case of ingestion of an unknown caustic agent and briefly discuss the current recommendations on diagnosis and treatment.

A 68 year old male presented with a two day history of painful swallowing after drinking what he believed was Miralax. The patient denied any recent antibiotic or steroid use. The patient’s past medical history is significant for congestive heart failure, type 2 diabetes mellitus, coronary artery disease, bipolar disorder and polysubstance abuse. On evaluation, the patient’s vital signs were within normal limits. Physical exam was remarkable for scattered white patches on the surface of the oropharynx, which are able for scattered white patches on the surface of the oropharynx, which are

Ingestion of caustic agents occurs most commonly by accident in the pediatric population and as a suicide attempt in adults. These agents include antibiotics. He improved clinically over several days and was discharged on antibiotics. He improved clinically over several days and was discharged on antibiotics.

Eosinophilic gastroenteritis is a rare disorder with less than 300 documented cases and even fewer pediatric cases. Typically presenting in the third to fifth decade, four criteria must be satisfied for diagnosis: presence of gastrointestinal symptoms, eosinophilic infiltration of the gastrointestinal tract, exclusion of parasitic disease, and absence of other systemic involvement. CT and radiographic findings are usually non-specific with diagnosis made on histological evaluation. Dietary therapy and corticosteroids are the mainstay of therapy. Our patient demonstrates a common presentation of an uncommon pediatric illness; practitioners need to be aware of this disease and keep it higher in the differential especially in patients with an elevated eosinophil count.

SLEEP DISTURBANCE AND HEALTH-RELATED QUALITY OF LIFE IN CHILDREN WITH EOSINOPHILIC ESOPHAGITIS
Lynch M2, Dimmit R1, Goodin B2, Avis K1,2,3,4 UAB, Birmingham, AL and 2UAB, Birmingham, AL.

Purpose of Study: The study’s aim is to examine the sleep and health related quality of life (HRQoL) of children with Eosinophilic Esophagitis (EoE). EoE is a gastrointestinal disease with chronic, recurrent symptoms of vomiting, dysphagia, and feeding aversion. Identification of children with EoE is burgeoning with prevalence estimates of up to 5 per 100,000 individuals. Treatments include elimination diets, medication slurreries, and repeat endoscopies to monitor disease progression. As with other childhood chronic illnesses, EoE may cause alterations in HRQoL, mood, and sleep.

Methods Used: 7 children with EoE and 7 healthy comparison children (HC), aged 4-12 years (mean age 7.7), matched by age, sex, and race, were recruited along with their caregivers. Children wore an actigraph for two weeks to evaluate sleep patterns. Parents also completed two measures of the child’s HRQoL, the Pediatric Quality of Life Inventory (PedsQL) and the PedsQL EoE specific module. Scores assessing overall HRQoL; physical, cognitive, emotional, and social functioning, as well as EoE total symptom scores, was obtained.

Summary of Results: Results show children with EoE sleep approximately 1 hour less than HC. Total Sleep Time of children with EoE was 410.80 minutes compared to 527.9 minutes in HC. Sleep Efficiency was lower in EoE (EoE=78% vs HC=87.5%). 71% of EoE were awake for greater than 60 minutes during the night, compared to 0% of HC. There were trends toward significance in the relations between all measures of sleep disturbance and all HRQoL scale scores (p<0.06-0.09).

Conclusions: Children with EoE sleep less, have poor quality sleep, and are awake more often than HC. There is a trend toward sleep disturbances and poorer quality of life. It is possible that education about symptoms related to EoE; including sleep disturbances, will encourage families to seek appropriate treatments faster. In turn, faster treatment may decrease children’s active disease duration and lead to better treatment adherence, faster remission times, and fewer relapses, as well as increase the child’s (and family’s) HRQoL. Data collection is ongoing.

HEPATIC ENCEPHALOPATHY AS THE PRESENTATION OF CONGENITAL SPLENO-RENAL SHUNT IN THE ABSENCE OF HEPATIC CIRRHOSIS
Nakshabendi R1, Clark M2, Hritani A1, Munoz J3, Kanar O1, Nakshabandi A1, Nakshabendi P1. University of Florida Jacksonville, FL;2University of Florida Jacksonville, FL and 3Tampa general, Tampa, FL.
Case Report: A 46 yo white female presents to the hospital with chief complaint of a non-mechanical fall w/o loc. She was not able to fully remember the details of the fall. CT scan of the brain followed by MRI was performed which did not show any intracranial abnormality. Pt’s mental status began to deteriorate with development of slurred speech and asterixis which was not present on outpatient examination. EKG was normal and ammonia level was found to be 127. She was started on lactulose and rifaxamin which improved her symptoms and decreased the ammonia level. Labs including hepatitis A, B, and C were all negative, her ceruloplasmin, ferritin, anti-trypsin, anti-smooth muscle antibody, anti-immunoglobulin were all wnl. Multiphasic helical CT of the abdomen and pelvis with and without contrast revealed a normal liver, splenomegaly and a possible congenital porto-systemic shunt. EGD was performed and did not show any evidence of esophageal varices.

Discussion: Hepatic encephalopathy generally is a reversible impairment of neuropsychiatric function associated with impaired hepatic function. Triggers of hepatic encephalopathy include excessive nitrogen load such as gastrointestinal bleeding, electrolyte or metabolic disturbances, infections, drugs and medication such as; narcotics, benzodiazepine, antipsychotics and etoh intoxication. A thorough history did not reveal any medication or drug as a causative agent, rather the pt is a morbidly obese female with a spleno-renal shunt bypassing some of the portal venous flow to the liver. A spleno-renal shunt refers to an abnormal collateral porto-systemic communication between the splenic vein and left renal vein. It is typically one of the features of portal hypertension. Hepatic encephalopathy due to a congenital porto-systemic shunt is rare and fewer than 10 cases has been reported. This vascular phenomenon has mostly been reported in patients with cirrhosis and is thought to be due to elevations in levels of angiogenic factors. Most reported cases of spleno-renal shunts have been treated with either surgery or, rarely, percutaneous trans hepatic embolization.
AN UNUSUAL CAUSE OF LOWER GASTROINTESTINAL BLEEDING

Panikkath D, Panikkath R, Adiga AG, Mohammed A, Nugent K. Texas Tech University Health Sciences, Lubbock, TX.

Case Report: Ischemic colitis causes 6-18% of cases of acute lower gastrointestinal (GI) bleeding. Drug induced ischemic colitis occasionally causes lower GI bleeding, and amphetamines taken both as prescription drugs and as drugs of abuse can cause ischemic colitis.

A 43-year-old woman with history of hypothyroidism presented with the sudden onset of lower abdominal pain and bloody diarrhea. There was no prior history of GI bleeding or known liver disease. She required blood transfusions from outside hospital. A computerized tomography scan of abdomen showed mesenteric fat stranding. Colonoscopy showed inflammation in the sigmoid colon and features suggestive of ischemic colitis. Microscopic examination of the colonic mucosa also confirmed the diagnosis of mesenteric ischemia. ANA, anti-ds DNA, protein C, protein S, antithrombin III, antiphospholipid antibody, and prothrombin gene mutation tests were negative. Additional history revealed that she snorted cocaine about 2 days prior to admission. Her urine drug screen was positive for amphetamines. A mesenteric Doppler showed normal flow in the mesenteric vessels. She was advised to avoid the use of amphetamines and did not have any events during follow up.

Ischemic colitis develops from hypoperfusion of the colon, and watershed areas are most commonly affected. Mesenteric vascular disease is the most common cause of ischemic colitis, but it also occurs secondary to vasculitis, embolism, hypercoagulable states, colonic obstruction and drugs. Several drugs, including amphetamines, catecholamines, digitalis, cocaine, non-steroidal anti-inflammatory drugs, triptans, pseudoephedrine, and lubiprostone, have been implicated in colonic ischemia. Mechanisms proposed in drug-induced ischemic colitis, include vaso spasms with pseudoephedrine, thromboembolism with oral contraceptives, and hypotension with angiotensin-converting enzyme inhibitors and a sympathomimetic amine. Methamphetamine is a sympathomimetic amine with central nervous system stimulant activity. Its effects include elevation of blood pressure and splanchic vasoconstriction. Consequently, it follows that bowel ischemia may result. Drug abuse is a rare but preventable cause of lower GI bleed. A high index of suspicion is needed for diagnosis as patients often hide this history.

B-CELL LYMPHOMA IN INFLAMMATORY BOWEL DISEASE

Phemister J1, Kapila A2, Carter L1, Khan O1, Murthy R2, Young M1.1 East Tennessee State University, Johnson City, TN and 2 Waters Medical Center, Mountain Home, TN.

Case Report: Adalimumab has been approved for treatment of severely active ulcerative colitis in maintain remission and mucosal healing. Prolonged use of anti-TNF agents like adalimumab has been associated with increased risk of lymphomas by 3 fold. It is challenging to find the exact association of lymphoma with the anti-TNF agents alone as most of these patients are concomitantly on immunomodulators drugs like azathioprine.

We present the case of a 66 year old male with a past medical history significant for left sided ulcerative colitis diagnosed in 2007 and had been maintained on azathioprine (for 5 years) and adalimumab (for 3 years), presented with increasing abdominal cramping and bloody diarrhea with small amounts of mucous for last 3 weeks. On initial presentation vital signs were stable, physical exam was significant for diffuse abdominal tenderness. Blood work revealed mild anemia with hemoglobin - 12.2 g/dL and stool c.diffile was negative. Computed tomography of the abdomen showed distal sigmoid and rectal wall thickening with pericolic fat stranding consistent with active inflammation and lymphadenopathy. Subsequently a flexible sigmoidoscopy was performed, which demonstrated a near complete obstruction due to rectal stricture with inability to pass scope beyond 10 cm. Rectal stricture biopsy revealed diffuse large B-cell lymphoma. No bone marrow involvement was noticed on biopsy. Patient is currently receiving chemotherapy with rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone (R-CHOP), followed by radiation and a subsequent colectomy. The decision to start these agents should made by understanding the potential risks of lymphoma against the improved quality of life when selecting medical therapy for patients with IBD.

A UNIQUE SIZE AND PRESENTATION OF VIPOMA

Pourmorteza M1, Litchfield J2, Borthwick T2.1ETSU Quillen College of Medicine, Johnson City, TN and 2ETSU Quillen College of Medicine, Johnson City, TN.

Case Report: VIPomas are rare pancreatic endocrine tumors that usually present with symptoms of chronic refractory diarrhea, hypokalemia, and achlorhydria (WDHA). We demonstrate a rare glimpse at an unusual small size of VIPoma at its earliest presentation.

Case Report: A 71 Year old male with a past medical history of diabetes mellitus was referred to GI clinic for three month history of worsening diarrhea. Stool output ranged from 5-6 bowel movements daily.1 month following the onset of diarrhea he presented to the ED with upper quadrant abdominal pain as a new 0.4cm calcified lesion was identified on abdominal CT. On examination he was dehydrated with elevated creatinine/BUN (2.1/28mg/dL) from baseline (1.4/21mg/dL) while denying fever, blood in stool, nausea/vomiting. Blood investigations demonstrated Potassium 4.7mmol/L, Sodium 142mmol/L and Chloride 102mmol/L. Blood count and liver enzymes were all within normal limits. MRI exhibited a well circumscribed 13mm solitary lesion within the ventral pancreatic body consistent with pancreatic neuroendocrine tumor. The serum VIP was in excess of 261 pg/ml with a repeat of 245.7 pg/ml. Total body Octreotide Scan failed to demonstrate any evidence of metastasis. Pathology results described a well differentiated pancreatic neuroendocrine neoplasm 1.4cm consistent with VIPoma and margins negative for neoplasm.

Discussion: Perfuse secretory diarrhea is the hallmark of the disease process with 1L to more than 3L of stool output daily. Symptoms include hypokalemia-89%, achlorhydria-43% and weight loss-36%. The majorities of tumors present with metastatic disease and lymph node involvement 78% ranging from 2-6cm in size.

Our patient’s presentation was inconsistent with the WDHA syndrome. To our knowledge this is the smallest reported solitary tumor of its kind. The natural slow progression of the tumor in terms of size and clinical constellation in a setting of diarrhea with newly identified small size pancreatic lesion has raised our suspicion of an early clinical manifestation of VIPoma. The lack of electrolyte abnormalities and metastasis with normal radiologic finding one year prior to the diagnosis has furthermore strengthened our notion of such early presentation.

PANCREAS DIVISUM: AN UNCOMMON CAUSE OF PANCREATITIS

Rasanneehiran S, Wang H, Mulkey Z. Texas Tech University Health Sciences Center, Lubbock, Texas, Lubbock, TX.

Case Report: Background: Pancreas divisum is the most common congenital anatomic variation of the pancreas, which occurs in about 10% of the population. This condition is usually asymptomatic, however, pancreas divisum is found in 20% of cases of idiopathic acute pancreatitis. The patients in this group are more likely to be a young female who presents with recurrent acute pancreatitis. We are reporting an uncommon presentation of pancreas divisum causing acute pancreatitis.

Case report: A 54-year-old man presented with severe epigastric pain associated with nausea and vomiting for 1 day. His past medical history was unremarkable.
He reported moderate alcohol consumption. He was hemodynamically stable. Physical examination revealed mild tenderness over the epigastric area without peritoneal signs. Laboratory findings revealed a mildly elevated l-aspartate aminotransferase level of 133 IU/L, normal amylase level of 29 IU/L, normal triglycerides, urea, and creatinine. He was referred for possible diverticulitis. The endoscopic intervention was done, which revealed a large diverticulum of the left colonic flexure without evidence of acute inflammation. The patient was discharged and referred to the regular gastroenterology clinic for follow-up.

Conclusion:
Diverticulitis is a common condition that can present with abdominal pain, nausea, and vomiting. The endoscopic intervention is a safe and effective treatment for diverticulitis. Treatment with antibiotics and dietary modifications can help prevent recurrence.

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POST-TRANSPLANT LYMPHOPROLIFERATIVE DISORDER PRESENTING WITH SMALL BOWEL OBSTRUCTION 12 YEARS POST-KIDNEY TRANSPLANTATION

Rassameehiran S, Klomjit S, Nugent K.
The Journal of Investigative Medicine

Case Report:
A 59-year-old man with a history of hypertension presented with worsening right-sided chest pain and progressive shortness of breath of two weeks duration. The chest pain was pleuritic in nature. He denied fevers, productive cough, nausea or vomiting. On physical examination, his vital signs were as follows: blood pressure, 114/66 mmHg; heart rate, 123/min; respiratory rate, 21/min; and temperature, 98.7°F. Examination of the lungs revealed decreased breath sounds at right lower lung field, with dullness on percussion. Examination of the heart revealed regular tachycardia without murmur. There was no subcutaneous emphysema. Laboratory revealed the following: WBC, 26,570/μL with 95% polymorphonuclear cells; BUN, 31 mg/dL;; and Cr, 0.9 mg/dL. Chest radiograph revealed a right-sided hydropneumothorax. A thoracotomy tube was placed into the right chest resulting in the drainage of 1 liter of brownish purulent material. Pleural fluid analysis showed a pH of 7.2. Gram stain of the pleural fluid showed few WBCs, many Gram-positive cocci in pairs, chains, and clusters, moderate Gram-positive rods, few Gram-negative rods, and few yeast. Amylase level was not sent at that time. The patient underwent a thoracotomy, which revealed gastrointestinal contents in the pericardial cavity and distal esophagus perforation measuring 2.5 cm. The patient underwent a thoracotomy, which revealed gastrointestinal contents in the pericardial cavity and distal esophagus perforation measuring 2.5 cm. The patient underwent a thoracotomy, which revealed gastrointestinal contents in the pericardial cavity and distal esophagus perforation measuring 2.5 cm. The patient underwent a thoracotomy, which revealed gastrointestinal contents in the pericardial cavity and distal esophagus perforation measuring 2.5 cm.

Conclusion:
Boehrhave’s syndrome is a rare condition that classically presents with severe vomiting and retching followed by severe chest pain and subcutaneous emphysema. However, some reviews point out that the presence of this triad is rare. Unusual presentation should be kept in mind while evaluating the patient with hydropneumothorax even if the right sided.

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ULCERATIVE COLITIS WITH SMALL BOWEL INVOLVEMENT

Spera MA, BOllinger E, Chandamuri BR, Engel LS, Hutchings JJ. LSU Health Sciences Center, New Orleans, LA.

Case Report:
A 21 year old man with a history of ulcerative colitis and recent Clostridium difficile colitis presented with diffuse abdominal pain, anorexia, and hematochezia. The patient had recently discontinued both oral and rectal mesalamine and begun a steroid taper. The patient had abdominal tenderness to palpation. The CBC showed a WBC of 17000 cells/μL with 90% segmented neutrophils, 5% lymphocytes, 1% monocytes, and 1% bands. On day 2 the patient reported worsening abdominal pain with nausea and vomiting. Repeat CBC showed a WBC of 33,000 cells/μL with an increase of bands to 7%. Lactic acid was elevated at 3.4 mmol/L. Empiric antibiotic coverage for Clostridium difficile colitis was initiated. CT of the abdomen and pelvis with IV and oral contrast showed diffuse colitis with dilation of the pericolic vessels. The patient was diagnosed with fulminant colitis secondary to Clostridium difficile colitis versus ulcerative colitis. He was taken emergently for total abdominal colectomy with end ileostomy. Pathology of the total abdominal colon revealed ulcerative colitis with pan colitis and backwash ileitis. On post op day 12, the patient developed abdominal distension and blood in his ileostomy bag as well as from a nasogastric tube that had been placed. EGD and colonoscopic evaluation through the patient’s ileostomy revealed friable mucosa, ulcerations and oozing in the distal duodenal and proximal jejunal. Pathology findings from duodenal ulcer biopsies were consistent with duodenal involvement by ulcerative colitis.

DISCUSSION: Extension of the ulcerative process to the small intestine is not typical of ulcerative colitis. When this finding is present the diagnosis of ulcerative colitis may be questioned in favor of a diagnosis of Crohn’s disease. The etiology of small bowel enteritis in ulcerative colitis may be due to T cell and cytokine mediated inflammatory response induced by severe ulcerative colitis. This inflammatory response, which was previously con- fined to the colon, then goes on to affect the small bowel once the colon is removed. Therefore early initiation of immunosuppression following colectomy may improve outcomes in this patient population.

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A RARE CAUSE OF MASSIVE UPPER GI BLEEDING: PRIMARY AORTOENTERIC FISTULA, A CASE REPORT AND LITERATURE REVIEW
Tantachat Ch, Wondmagegnu NE, Ruthirago D, Rassameehiran S, Tello W.

Purpose of Study: Primary aortoenteric fistula is a rare but fatal cause of upper gastrointestinal bleeding with non-distinctive clinical presentation. This case report aims to remind physicians to consider this clinical entity especially in high risk patients.

Methods Used: A case report and literature review.

Summary of Results: A 61-year-old Hispanic woman presented with massive rectal bleeding and hypotension. Her past medical history included systemic lupus erythematosus, coronary artery disease, diabetes mellitus, hypertension, and colorectal cancer treated with chemotherapy, radiation therapy, and surgery. She subsequently went in to cardiac arrest despite packed red blood cell, platelet, and fresh frozen plasma transfusion. Her initial rhythm was pulseless electrical activity. She regained a pulse after 4 cycles of cardiopulmonary resuscitation and 4 doses of epinephrine. Norepinephrine infusion was started to maintain her blood pressure. Abdominal computed tomography angiography (CTA) revealed 7.2 cm of infrarenal abdominal aortic aneurysm with focal rupture and 5.3 cm hematoma, a small air bubble between hematoma and duodenum without definite contrast extravasation, and numerous clots in stomach. She continued to bleed massively and her clinical status was deteriorating. Her family had refused aggressive treatments including esophagogastroduodenoscopy (EGD) and surgical intervention. The patient passed away after being hospitalized for three days.

Conclusion: Aortoenteric fistulas (AEF) can be categorized into 2 types. Primary AEF is the connection between native aorta and GI tract. Secondary AEF is the connection between native aorta and GI tract. The most valuable diagnostic tool is helical CT scan with intravenous contrast. Implementing imaging for accurate diagnosis would help decrease the mortality of EAF. EGD should be employed in order to exclude other causes of GI bleeding. Mortality rate is 100% if left untreated. Once the diagnosis has been made, surgical intervention should pursue definitely.

A RARE HEPATIC TUMOR MASQUERADING AS FOCAL NODULAR HYPERPLASIA
Turse E1, Pattnana S3, Subramony C2, 1University of Mississippi Medical Center, Jackson, MS; 2University of Mississippi Medical Center, Jackson, MS and 3University of Mississippi Medical Center, Jackson, MS.

Case Report: Focal Nodular Hyperplasia (FNH) is the second most common liver mass and is usually noted as an incidental finding on imaging. It is best characterized on enhanced Computed Tomography (CT) as a well-circumscribed heterogeneously enhancing lesion with a central stellate scar. Here we present a hepatic lesion which appeared classic for FNH on imaging but when biopsied proved otherwise.

An 88-year-old nonsmoker, nondonor black male presented with complaints of dyspnea and wheezing of one-day duration. Past medical history of cerebral meningioma, prostate adenocarcinoma, and Parkinson's disease. Physical exam revealed a chronically ill male. Abdominal exam was unremarkable without tenderness, hepatosplenomegaly or masses. Notable laboratory data included negative HepCAb, HBsAg and positive HBsAb. A CT imaging revealed saddle pulmonary embolism, bilateral pleural effusions, and an incidental finding of a 6cm X 6cm heterogeneously enhancing lesion in the right lobe of the liver with a central stellate scar with no evidence of cirrhosis.AFP, CEA, and Ca 19-9 were unremarkable. Liver biopsy of the mass revealed spindle cells with elongated nuclei and mild atypia between numerous capillaries with thin wall branching vessels in a staghorn configuration. Immunohistochemistry was positive for smooth muscle actin, desmin, alpha-smooth muscle actin, and negative for S100, CD117 and desmin. A diagnosis of primary hepatic hemangiopericytoma (HPC) also known as a solitary fibrous tumor (SFT) was made.

HPC or SFT comprises 1% of all vascular neoplasms. To date there have been forty-two reported primary hepatic SFT cases. It is thought to arise from trauma, steroid use or hypertension. It is generally a benign slow growing tumor, although there have been four reported cases of malignant transformation. Our case is consistent with the radiologic description of a single large heterogeneous mass. However, central stellate scarring is an atypical radiologic feature of this tumor thus mimicking the classic radiographic findings of FNH.
Conclusions: Our hypothesis that patients would not be satisfied with their visit was not supported. There are barriers to getting the wellness visit. We found that patients tend to defer to their primary care providers, even after receiving an explanation of the contents of the AW. If primary care providers recommended the AW, more patients may participate in the future.

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AUTOMATED URINE FLOW RATE AND VOLUME MEASUREMENT: VALIDATION OF A NOVEL SYSTEM
Colombo C1, Stachura M2, Wood E3, George A1, Broughton R1. Dwight Eisenhower Army Medical Center, Ft. Gordon, GA and 2Georgia Regents University, Augusta, GA.
Purpose of Study: Accurate measurement of urine output rate and volume is essential for fluid balance and hydration management in numerous acute and chronic medical conditions in multiple clinical settings. Their current measurement is manual, time consuming, error prone, and costly. When limited numbers of healthcare personnel are responsible for multiple patients with complex medical problems the error inherent in the current manual system becomes immediately apparent. The purpose of this collaborative project was to validate accuracy and precision of a new technology, UroSenseTM, that automates and improves the accuracy of urine output rate and volume measurement.
Methods Used: Ten UroSenseTM containers, their wireless transmitters, computer algorithms used to report results and embedded UroSenseTM container urinometer were tested simultaneously in a laboratory setting. All containers were tested using variable speed pumps drawing from pooled human urine. The study consisted of three 10 hour shifts. One container accumulated urine throughout the three shifts. Nine containers were emptied at the end of each shift in order to test dry vs. wet sensor functions. Sensor sensitivity drift was evaluated by linear regression over the testing period. Total shift output (UroSenseTM reading) was validated using visual graduated cylinder measurement. The main comparison was between UroSenseTM and visual readings, including dry or wet sensors (initial and repetitive use).
Summary of Results: Ten UroSenseTM containers were tested and 201 measurement pairs were used in the analysis. The plotted data revealed clear linearity with UroSenseTM measurements and visual readings strongly correlated, r(201)=0.986. Dry (initial use) and wet (subsequent use) readings were statistically different, based on paired t tests of absolute differences between hourly UroSenseTM and visual readings: mean difference 37.76+30.11 ml with dry and 75.52-73.33 ml with wet containers (P<0.0001) lower in wet condition.
Conclusions: UroSenseTM measurements are strongly correlated with visual readings. Repetitive sensor use (dry vs. wet) exhibits a statistically significant difference; however, clinically this has a minimal impact.

MECHANICAL VENTILATION: RESOURCE UTILIZATION IN 2013
Edris H, Selvan K, Sigler M, Nugent K. TTUHSC, Lubbock, TX.
Purpose of Study: Recent studies in patients with acute respiratory failure have provided information about the best approaches to mechanical ventilation, fluid management, weaning and extubation, and prevention of ventilator-associated pneumonia. This information has the potential to decrease the length of time required for mechanical ventilation and to increase safety. However, intensive care remains very expensive. Our hypothesis was that prospective review of our data could provide useful information about our resource utilization.
Methods Used: We retrospectively reviewed the medical records of 174 patients who required mechanical ventilation for acute respiratory failure in a medical intensive care unit in 2013. We collected information about patient demographics, medical diagnoses, gas exchange, chest x-rays, fluid balances, and complications. We also determined the frequency of common procedures and tests used in ventilator patients.
Summary of Results: This study included 174 patients. The mean age was 57.8 + 16.8 years, 54.02% were men, the mean APACHE II score was 13.8± 6.1, and the overall mortality was 32.2%. The five most frequent diagnoses were pneumonia, septic shock, drug overdose, stroke, and cardiac arrest. The mean initial PaO2/FiO2 was 191.4±109.9. The mean number of ventilator days was 7.5±7.1. These patients had 1.2 ± 0.43 x-rays per day on the ventilator. They had 10.0 ± 9.4 arterial blood gases during hospitalization. They had 0.72 ± 0.74 central lines and 0.30 ± 0.32 arterial lines per patient during mechanical ventilation. The frequency of bronchoscopies and tracheostomies was 0.05 and 0.09 per patient, respectively. These patients had 40.4 ± 70.9 point of care glucose measurements during their hospitalization.
Conclusions: Routine care of patients on mechanical ventilators requires frequent chest x-rays, arterial blood gases, central venous catheters, and arterial lines. Patients often have very frequent point-of-care measurements. These results suggest that we should critically evaluate resource use; the number of arterial lines, arterial blood gases, and point-of-care glucose measurements might be reduced without any change in patient outcomes.

PERITONEAL DIALYSIS CENTER
Vala SK2, NEEMUCHWALA F2, Eldemir O3,4, Burns JF2, Volpe AD1. 1Emory University, Atlanta, GA and 2Emory University, Atlanta, GA.
Purpose of Study: Peritonitis remains a serious complication of peritoneal dialysis (PD). As suggested by International Society of Peritoneal Dialysis (ISPD), on an average, a center’s peritonitis rate should be no more than 1 episode every 18 months (0.67/year at risk). In this study, we aim to evaluate peritonitis and technique failure rates among PD patients at our major inner city PD center.
Methods Used: We conducted a retrospective cohort study of peritonitis episodes among 90 PD patients followed at our PD center over a period of 54 months, between February 2010 and July 2014. Demographic and biochemical variables, clinical outcomes, including peritonitis and technique failure rates were evaluated.
Summary of Results: Demographically our patient population comprised predominantly of 55% Men, with average age of 53.8 years, 64% of African American and 7% Asian ethnicity with 36% having hypertension as their etiology for initiation of dialysis. There were 33 episodes of peritonitis (EOP) among 90 patients during the 54-month period, which equates to a cumulative EOP rate of one episode every 54.67 months (0.22/year at risk), of which 9 were recurrent EOP (10 %, 9/90). Among these EOP, the most common organism accounting for 27% (9/33) EOP was Coagulase-negative staphylococcus (CoNS). Culture-negative peritonitis rate was 27% (9/33). Staphylococcus aureus, other gram positives and negatives accounted for the rest of EOP. While catheters were removed in 24% (8/33) patients, they resumed PD after treatment and remained on PD till end of study period.
Conclusions: Peritonitis rate among our inner city patient population of 1 episode every 54.67 months (0.22/year at risk) is well below the acceptable peritonitis rate as suggested by ISPD. Among the EOP evaluated, one-third of cases were due to CoNS, while other one-third cases were due to culture-negative peritonitis. While peritonitis is still associated with a higher frequency of 24% technique failure among our population, all of our patients following treatment eventually returned back to PD and remained on PD till the end of study period. Frequent education sessions to our prevalent and new PD patients, focused on preventive measures, especially hand hygiene, shall require optimization to reduce future EOP.

EFFECT OF KNOWLEDGE ON COMPLIANCE IN PATIENTS WITH CYSTIC FIBROSIS
Vaia SK2, NEEMUCHWALA F2, Eldemir O3,4, Burns JF2, Volpe AD1. 1Emory Children’s Clinic, Pensacola, FL and 2Florida State University, Pensacola, FL.
Purpose of Study: Despite many advances in the treatment of Cystic Fibrosis (CF), frequent respiratory infections and malabsorption negatively affects the length of survival in these patients. Care of CF includes daily administration of several nebulized medications as well as oral enzymes and supplements which takes 2 to 3 hours a day. The hypothesis for this study is improved knowledge of CF would correlate with higher compliance with health measures.
Methods Used: A cross-sectional study was conducted in patients 6 to 18 years of age with a diagnosis of CF at Nemours Children’s Clinic, Pensacola. A standardized validated CF Questionnaire (CQ) (Chest 2006) was administered to parents of children with CF at routine visits, to assess knowledge and adherence in three domains including lung health, nutrition, physical activity, and medical management. The research assistant administered the questionnaire to the parents in the waiting room before their child’s visit. The data were analyzed using SPSS version 21.0.

Conclusions: Our hypothesis that patients would not be satisfied with their visit was not supported. There are barriers to getting the wellness visit. We found that patients tend to defer to their primary care providers, even after receiving an explanation of the contents of the AW. If primary care providers recommended the AW, more patients may participate in the future.
and CT treatments. Additionally, BMI percentiles and FEV1 were recorded for each patient. The data was analyzed using Spearman correlation in SPSS to correlate measures of knowledge to adherence.

**Summary of Results:** A total of 20 patients were recruited. Total knowledge mean score was 69% and total adherence mean score was 82%. There was no correlation between measures of knowledge and adherence. Nutrition adherence correlated to greater BMI ($r = 0.486$, $p = 0.049$), total adherence similarly correlated to greater BMI percentile ($r = 0.508$, $p = 0.022$). Total adherence approached statistically significant correlation to greater FEV1 ($r = 0.431$, $p = 0.058$).

There were statistically significant inter-correlations among knowledge sub-scores (lung health knowledge vs. nutrition knowledge ($r = 0.474$, $p = 0.035$); lung health knowledge vs. treatment knowledge ($r = 0.594$, $p = 0.006$); nutrition knowledge vs. treatment knowledge ($r = 0.643$, $p = 0.002$)). No inter-correlation was found for adherence sub-scores to other adherence sub-scores.

**Conclusions:** Increased knowledge in CF caregivers does not correlate with adherence in CF patients. Nutritional adherence and total adherence were related to higher BMI. High knowledge in one sub-score correlated to high knowledge in the other two sub-scores.

**Hematology and Oncology**

Joint Plenary Poster Session and Reception

5:00 PM

Thursday, February 26, 2015

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**HEMPAS NEGATIVE CONGENITAL DSYSERYTHROPOIC ANEMIA: A CASE REPORT WITH REVIEW**

Ahmed M1, Begum T1, Iroegbu N2, 1Texas Tech University, Amarillo, TX and 2Saint Joseph Hospital, Chicago, IL.

**Case Report:** Case Report: A 37 year-old female was evaluated in hematology outpatient department for macrocytic anemia. She was not on any home medication, and there was no history of alcohol or drug abuse. Her past medical history was remarkably benign other than non-specific fatigued and unexplained anemia of variable duration diagnosed on different occasions. Her blood work showed Hemoglobin 11.3 gm/dl, MCV-134 fl, MCH-47 pg, WBC-8700/cml with 80% segmented neutrophils, LDH was 845 IU/L. Haptoglobin <26 IU/L. Vitamin B-12 and Folate level were normal. Bone marrow biopsy revealed hyperplastic erythropoiesis characterized by megaloblastosis with extensive dyserythropoietic changes. Cells had large megaloblastic nuclei. Nuclear abnormalities including lobulation were prominent with frequent binucleated cells. A number of erythroblasts had nuclear fragment and maximum 4 nuclei were seen. An acidified serum test (Ham test) was negative. Gene sequencing revealed mutations in the SEC23B gene confirming the diagnosis of congenital dyserythropoietic anemia type-II.

**Discussion:** The congenital dyserythropoietic anemias (CDA) comprise a group of rare hereditary disorders of erythropoiesis, characterized by ineffective erythropoiesis as the predominant mechanism of anemia and by distinct morphologic abnormalities of the majority of erythroblasts in the bone marrow. The leading morphologic abnormality is binuclearity or multinuclearity occurring in 10% to 50% of mature erythroblasts, with equal DNA content in both nuclei. All subtypes present with anaemia, ineffective erythropoiesis, splenomegaly and dyserythropoiesis with bi/multinuclearity in erythrocyte precursors. The frequency, the relevance of complications, and the use of splenectomy are poorly defined. Congenital dyserythropoietic anemia type II (CDA-II), also known as hereditary erythroblastic multinuclearity with a positive acidified-serum test (HEMPSAS), is the most frequently encountered disorder of the CDA group although acidified serum test may be negative. The treatment is mainly conservative. The current recommendation is to consider splenectomy if the anemia compromises patients' performance status, and to manage iron overload according to the guidelines derived from patients with thalassemia.

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**LEPTOMENINGEAL CARCINOMATOSIS FROM ESOPHAGEAL CANCER: A CASE REPORT WITH BRIEF REVIEW**

Ahmed M1, Begum T1, Iroegbu N2, 1Texas Tech University, Amarillo, TX and 2Saint Joseph Hospital, Chicago, IL.

**Case Report:** Case Report: A 47 year-old female presented with headache, neck pain and delirium. She denied any fever. No recent contact with any sick person. She mentioned about 20 lb unintentional weight loss in last 2 months. Upon query, she also mentioned some degree of dysphagia, mostly with solids for last 1 month. She never had such symptoms or gastroesophageal reflux disease in the past. An MRI of the brain and cervical spine revealed meningeal enhancement suggestive of metastatic carcinoma. Cytology from the CSF fluid confirmed adenocarcinoma. An EGD showed a fungating mass at gastro-esophageal junction. A biopsy was taken which revealed moderately differentiated adenocarcinoma. Subsequent CT scan of chest, abdomen and pelvis revealed no obvious metastasis. The patient was started on chemotherapy. She responded poorly with chemotherapy. Her condition deteriorated quickly and died after 4 weeks.

**Discussion:** Leptomeningeal carcinomatosis (LC), also known as neoplastic meningitis, is a serious complication of cancer that carries substantial rate of morbidity and mortality. Approximately 1-8% of patients with cancer are diagnosed with LC. It is present in 1-5% of patients with solid tumors, 5-15% of patients with leukemia, and 1-2% of patients with primary brain tumors. It may occur at any stage of the neoplastic disease, either as a presenting symptom or as a late complication, though it is associated frequently with relapse of cancer elsewhere in the body. Although metastatic involvement of the meninges has been observed in carcinoma of the breast, prostate, and lung, LC arising from esophageal carcinoma is exceedingly rare. Unfortunately, therapeutic options are limited, particularly for patients with chemoresistant tumors. Optimum treatment is controversial and may vary upon a combination of chemotherapy (intrathecal and/or intravenous) and concurrent radiotherapy.

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**DUMPSTER DIVE: A CASE OF BROdifacum POISONING**

Ahmed M1, Begum T1, Iroegbu N2, 1Texas Tech University, Amarillo, TX and 2Saint Joseph Hospital, Chicago, IL.

**Case Report:** Case Report: A 46 year-old man presented with bleeding from a facial wound for two days. He developed the facial lesion from a scratch wound. He had no past medical illness as well as no previous history of abnormal bleeding. Active bleeding from facial wound did not stop after compression for more than 10 minutes. Fresh gum bleeding was also noted. A large ecchymotic lesion on left lower extremity was found. Patient's initial lab showed prothrombin time (PT) 13.4 seconds, INR - 9.7, activated partial thromboplastin time (aPTT) - 101 seconds. Vitamin K dependent factors were markedly reduced. Serum brodifacum level was done which came back 180 ng/ml. Although initially it was not clear how he was exposed to brodifacum. Later on, he mentioned that he often ate leftover food from dumpsters, and most likely ingested brodifacum through rat poison accidentally. He was initially managed with fresh frozen plasma and later on needed prolonged course of oral vitamin K therapy.

**Discussion:** Brodifacum or Superwarfarin is an active ingredient of different rodenticides. The number of exposure is increasing annually, mostly in pediatric age group. Most of the cases are accidental poisoning. Patients usually present with various form of bleeding disorder like hematuria, epistaxis, gingival bleeding, hemoptysis, menorrhagia, hemopterionemia, diffuse alveolar hemorrhage, spontaneous hemorrhax, hematemesis, subarachnoid hemorrhage etc. High level of suspicion is needed when unexplained bleeding is encountered, though number of fatalities is usually rare. Patients usually need high dose of vitamin K ranging from 15 mg to 800 mg/day for a prolong period of time. Some recent studies demonstrated success with treatment of high-dose oral vitamin K1 at a dose of 7 mg/kg per 24 hours divided every 6 hours.

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**TESTOSTERONE SECRETING ADRENAL TUMOR: A CASE REPORT WITH REVIEW OF LITERATURE**

Ahmed M1, Begum T1, Iroegbu N2, 1Texas Tech University, Amarillo, TX and 2Saint Joseph Hospital, Chicago, IL.

**Case Report:** Case Report: A 71 year-old lady came to seek medical care for her recurrent growth of facial hair which did not go away with cosmetic maneuver. There was no change in her muscle mass, deepening of voice or elitoromegally. Abdominal and pelvic examination was normal. Serum
**Case Report:** Case Report: An 82 year-old man presented with proptosis, throbbing pain and blurry vision of right eye. He was diagnosed with pros-tatic cancer, and had a radical prostatectomy nearly 10 years back. After the surgery, his PSA remained normal until recently. Since last one year, his PSA level started rising although no obvious metastatic lesion was found in recent skeletal survey. Over last 3 weeks, he gradually developed progressive throbbing pain in right eye with subsequent blurring of vision and proptosis. A CT scan of the orbit showed a soft tissue mass within the right orbit with bony destruction and optic nerve compression. Patient was started on androgen suppression with leuprolide and steroid. His symptoms improved initially. But a repeat CT scan of the abdomen and pelvis showed diffuse bony metastasis to spine and pelvis. Considering the patient's advanced age and nature of the disease, the patient decided to adopt hospice care. He expired in couple of months.

**Discussion:** Metastasis to orbit from neoplasm of different origin is rare. Among different malignancies, breast cancer is the most common one to metastasize to orbit. Prostate cancer was found to be the second or third most common source on different case series. The mean age of the patients was 70.5 yrs (range: 60 to 90 yrs). Both the left and right orbit was involved at similar frequency with bilateral involvement in some cases. The mean time period between prostate cancer diagnosis and orbital metastasis was 3.7 yrs (range: 1- 8 yrs). On three occasions, orbital metastases were the presenting symptom of prostate cancer. CT scan was the imaging study of choice, and MRI is also very sensitive. Radiotherapy and steroid were used simultaneously as palliative therapy. Except for one case, all the patients expired within one year of metastasis.

**SERUM NILOTINIB LEVEL MONITORING DURING CONCOMITANT USE OF CYP3A4 INHIBITORS**

Ahmed M1, Begum T1, Iroegbu N2, 1Texas Tech University, Amarillo, TX and 2Saint Joseph Hospital, Chicago, IL.

**Case Report:** Case report: A 54 year-old man with HIV and acute myeloid leukemia refractory to conventional chemotherapy presented with overwhelming septicemia. The patient was on Nilotinib for his leukemia as previous chemotherapy regimen failed to show any response. After admission, patient was started on antifungal drug for broad spectrum coverage of his septicemia. Though it is not recommended to use nilotinib along with hepatic microsomal enzyme inhibitors, there was little choice left to choose for patient’s leukemia. During hospital stay, serum nilotinib level was monitored along with EKG for QT interval monitoring. Nilotinib doses were adjusted according to his serum level and QT duration. In this report, we present, for the first time in clinical practice, serum nilotinib level monitoring in a patient during concomitant use of nilotinib and CYP3A4 inhibitors.

**Discussion:** Nilotinib is a new orally active tyrosine kinase inhibitor which is considered a “second-generation” tyrosine kinase inhibitor with greater potency than its 1st generation counter part Imatinib. Serum nilotinib level monitoring during concomitant use of nilotinib and CYP3A4 inhibitors has not been in regular practice. Kinetic experiments showed that CYP3A4 and CYP2C8 enzymes are the main contributors to the metabolism of nilotinib and co-administration with the CYP3A4 inhibitors reduce oxidative metabolism of nilotinib by >95%. If treatment with any of CYP3A4 inhibitors is required, it is recommended that therapy with Nilotinib be interrupted. If interruption of treatment with nilotinib is not possible, patients who require treatment with a drug that prolongs QT should be closely monitored for the QT interval. In our study, this is the first time in clinical practice; serum nilotinib level was monitored in clinical setting with concomitant use of CYP3A4 inhibitor along with regular monitoring of QT interval. As patients who have advance leukemia refractory to other conventional chemotherapy along with HIV or decreased immune status by leukemia, chemotherapy or any other diseases that require antifungal drug use, serum nilotinib monitoring may add additional benefit to patient safety in respect of drug adequacy and efficacy along with prevention of sudden cardiac death by QT prolongation.

**ORIAITAL METASTASIS OF PROSTATE CANCER: A CASE REPORT AND REVIEW OF LITERATURE**

Ahmed M1, Begum T1, Iroegbu N2, 1Texas Tech University, Amarillo, TX and 2Saint Joseph Hospital, Chicago, IL.

**Case Report:** Case Report: An 82 year-old man presented with proptosis, throbbing pain and blurry vision of right eye. He was diagnosed with prostatic cancer, and had a radical prostatectomy nearly 10 years back. After the surgery, his PSA remained normal until recently. Since last one year, his PSA level started rising although no obvious metastatic lesion was found in recent skeletal survey. Over last 3 weeks, he gradually developed progressive throbbing pain in right eye with subsequent blurring of vision and proptosis. A CT scan of the orbit showed a soft tissue mass within the right orbit with bony destruction and optic nerve compression. Patient was started on androgen suppression with leuprolide and steroid. His symptoms improved initially. But a repeat CT scan of the abdomen and pelvis showed diffuse bony metastasis to spine and pelvis. Considering the patient's advanced age and nature of the disease, the patient decided to adopt hospice care. He expired in couple of months.

**Discussion:** Metastasis to orbit from neoplasm of different origin is rare. Among different malignancies, breast cancer is the most common one to metastasize to orbit. Prostate cancer was found to be the second or third most common source on different case series. The mean age of the patients was 70.5 yrs (range: 60 to 90 yrs). Both the left and right orbit was involved at similar frequency with bilateral involvement in some cases. The mean time period between prostate cancer diagnosis and orbital metastasis was 3.7 yrs (range: 1- 8 yrs). On three occasions, orbital metastases were the presenting symptom of prostate cancer. CT scan was the imaging study of choice, and MRI is also very sensitive. Radiotherapy and steroid were used simultaneously as palliative therapy. Except for one case, all the patients expired within one year of metastasis.

**MANAGEMENT OF APLASTIC ANEMIA AND IDIOPATHIC THROMBOCYTOPENIA ASSOCIATED WITH PREGNANCY**

Bennett JA, Milner C. University of Mississippi Medical center, Jackson, MS.

**Case Report:** Aplastic anemia is characterized by diminished or absent hematopoietic precursors in the bone marrow, most often due to injury to the pluripotent stem cell. There have been several reported cases of aplastic anemia associated with pregnancy; however few cases reported pregnancy-associated aplastic anemia and concurrent refractory thrombocytopenia. Those described had unfavorable outcomes. We discuss a patient with both aplastic anemia and successive ITP presenting during pregnancy, with a favorable course and remission of the aplasia following delivery.

A 20-year-old Caucasian woman was referred by her obstetrician to hematology clinic at 28 weeks gestation for assessment and management of aplastic anemia. A local hematologist performed a bone marrow biopsy which was concerning for aplastic anemia and referred the patient to our hematology and transplant center for additional evaluation. Routine laboratory tests showed anemia and thrombocytopenia with a normal white blood cell count and adequate absolute neutrophil count. A second bone marrow biopsy confirmed aplastic anemia. Initially she was treated conservatively, with weekly labs monitoring and supportive care. At 33 weeks, she required hospital admission for shortness of breath, relative hypotension, and decreasing platelets. She exhibited a minimal response to platelet transfusion and there was concern for development of ITP. Intravenous immunoglobulin was administered for 5 days followed by oral prednisone with a subsequent improvement in response to platelet transfusion. After the platelet count achieved the goal of 50,000, the obstetrics service considered it necessary to induce labor and the patient had an uncomplicated vaginal delivery.

Several explanations have been proposed to rationalize the observation of bone marrow aplasia occurring during pregnancy. The associated complications and risk factors are also controversial and outcomes vary from a benign course to severe morbidity and mortality, with most reported complications related to infection and hemorrhage. As of recently, there is no consensus regarding optimal treatment during pregnancy. Our patient was successfully treated with IV immunoglobulin, steroids, and had a favorable hospital course and delivery.
The patient was found to have a moderate Factor VIII inhibitor and was successfully treated with activated prothrombin complex concentrate, steroids, and cyclophosphamide. A 63-year-old male with HIV/AIDS (CD4+ count of 32), as well as ESRD presented with a 3-week history of hematochezia. Patient had no known prior history of bleeding or clotting disorders. The patient initially required multiple transfusions with packed red blood cells and fresh frozen plasma to control the bleeding. His initial aPTT was prolonged at 65.9, and his PT and platelet count were normal. A 1:1 mixing study was performed, which showed an initial correction of the aPTT to 38.3 at room temperature, but at 2 hours incubated at 37 degrees Celsius, the aPTT was prolonged at 60%. Factor VIII activity was less than 1%, and a Factor VIII inhibitor assay was positive with an inhibitor titer of 2.2 (H). Patient was started on activated prothrombin complex concentrate with Factor VIII inhibitor bypassing activity with immediate resolution of his bleeding. He was then started on oral cyclophosphamide and prednisone 1mg/kg. Although patient developed a retroperitoneal hematoma two months later, his bleeding has now completely resolved. His Factor VIII activity five months after diagnosis was >30% with a negative inhibitor titer.

Although there are rare case reports of non-hemophilic patients with HIV who develop acquired Factor VIII inhibitor, the immunopathology that occurs in these patients leading to Factor VIII autoantibody production is poorly understood. Our case highlights the importance of high suspicion of acquired factor inhibitor development in non-hemophilic patients with underlying malignancy. Rapid diagnosis of acquired Factor VIII inhibitor is essential, as these patients can develop life-threatening bleeding without appropriate treatment.

**160 MYELOPROLIFERATIVE NEOPLASM PRESENTING AS SPONTANEOUS HEMORRHAGIC: A RARE AND UNUSUAL PRESENTATION**

Dwary A, Master S, Neppalli AK, Devarakonda S, Munker R, Veillon D, Koshy N. LSUHSC, Shreveport, LA.

**Case Report:**

**ABSTRACT BODY:**

**Case Report:**

A fifty-two-year-old man presented with complaints of worsening dyspnea and weight loss over 2 months. Physical exam was remarkable for moderate splenomegaly. Initial laboratory work up was significant for leukocytosis (110,000/μL) and thrombocytosis (>2 million/μL). Chest X-ray showed large left sided pleural effusion and thoracentesis yielded 1600 mL of bloody fluid. Following thoracentesis, the effusion worsened requiring videoassisted thoracoscopic and drainage of 2.2 liters of bloody fluid and clots. Chest tube was placed to prevent re-accumulation of effusion. Other significant findings on work up were normal PT and on Villebrand factor assay, slightly elevated PTT, bleeding time ~15 minutes, positive JAK-2 V617F mutation and negative bcr-Abl by real time polymerase chain reaction (PCR) assay. Peripheral smear evaluation showed numerous large hypo granular platelets as below. Bone marrow biopsy was consistent with myeloproliferative neoplasm, not otherwise specified. For rapid control of platelet count, platelethrapy was initiated in addition to cytoreduction with hydroxyurea. Normalization of platelet count with above measures led to control of bleeding. Later patient required combination of both hydroxyurea and anagrelide to lower his white blood cell and platelet counts.

**Discussion:**

Hemorrhagic events from myeloproliferative neoplasms like essential thrombocythemia are associated with severe thrombocytosis (platelet count >1.5 million/μL). It commonly results in muco-cutaneous bleeding due to production of dysfunctional megagranular platelets or acquired von Willebrand disease. To the best of our knowledge this is the only case of spontaneous hemorrhaxia secondary to myeloproliferative neoplasm reported so far. Rapid cytoreduction is crucial in the management of these bleeding complications.

**161 REFRACtORY IMMUNE THROMBOCYTOPENIA AS SOLE MANIFESTATION OF ACUTE HEPATITIS A INFECTION**

Evans J, Herrin V. University of Mississippi Medical Center, Jackson, MS.

**Case Report:**

We report a case of refractory immune thrombocytopenia in a 50 year old male associated with acute Hepatitis A infection.

A 50 year old male presented with gum bleeding, hematuria, and melena. He denied recent infections, jaundice, or abdominal pain. He worked as a butcher with a history of chronic alcohol abuse. Initial labs showed a platelet count of 2 x 10^9/L, hemoglobin of 11.4 g/dL, and normal coagulation studies. An acute hepatitis panel was positive for hepatitis A IgM. An abdominal ultrasound was without evidence of hepatosplenomegaly. Liver function tests remained normal throughout the hospitalization. Patient was started on prednisone 1mg/kg/day for empiric treatment of immune thrombocytopenia and given platelet transfusions due to active bleeding. The patient’s platelet count remained less than 5 x 10^9/L and he continued to have active bleeding. He was thus started with intravenous immunoglobulin, and with Rituximab for refractory thrombocytopenia. He developed an acute hemorhage at the base of the tongue that required emergent tracheostomy and transfer to the intensive care unit. While in the ICU the prednisone was changed to intravenous pulse dose decadron and Romiplostim at dose of 5 mcg/kg/wk was initiated. His bleeding improved despite having a platelet count less than 10 x 10^9/L, and was transferred out of the ICU. He then developed acute hypertension, bradycardia, and altered mental status with a CT head showing an acute hemorrhagic lesion in left parietal lobe. He was started on oral Cyclophosphamide and given a dose of Rh(D) immune globulin in addition to other measures (steroids, Rituxan, Romiplostim). After 24 days in the hospital the platelet count improved, and his hemorrhagic complications resolved. At discharge the platelet count was 69 x 10^9/L, and improved to 127 x 10^9/L at first outpatient visit.

Extrapleural manifestations of acute hepatitis A virus infection are uncommon, especially as the sole manifestation. As in our patient they can be severe and independent of liver abnormalities. The etiology of the immune thrombocytopenia has not been clearly defined but thought to be due to various mechanisms, including immune-mediated peripheral destruction, direct bone marrow suppression, and/or viral-associated hemaphagocytic syndrome.
OS, but to our knowledge has not been described in any other type of osteopetrosis. The patient presented with vomiting, secondary to duodenal web and malrotation and was also found to have osteopetrosis, but not the general dysmorphisms associated with OS. This case report may point towards association of gastrointestinal abnormalities and osteopetrosis, although supporting evidence from additional cases is needed.

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RARE ELUSIVE DIAGNOSIS OF NK CELL LYMPHOMA

Gupta E, Minocha V, Landa C. University of Florida, Jacksonville, FL.

Case Report: Malignancies derived from cytotoxic cells of natural killer(NK) cell or T-cell lineages are aggressive neoplasms classified as extranodal NK/T-cell lymphoma (ENNKT)(1,2). Its frequency among all malignant lymphoma is around 3%-10% in East Asia but <1% in western countries(3,4). Due to the rarity of the disease first line treatment has not been established. Even though two thirds of ENNKT patients present with localized disease, prognosis is dismal (5). A 39-year-old male with no past medical illness presented to the hospital with progressive dysphagia and odynophagia of 3 months. He also reported fever and weight loss. He was treated with clindamycin previously for presumed pharyngitis without improvement. On a subsequent ENT visit he was noted to have a right tonsillar/pharyngeal wall mass. He underwent laryngoscopy showing an obvious large mass extending from nasopharynx to the larynx. Biopsies were negative for neoplastic process. Tissue culture grew multiple organisms but symptoms persisted despite broad antimicrobials. CT neck revealed increase in size from previous scan. Patient underwent repeat biopsies and uvulotony. Finally, special staining tested positive for CD3 and CD56 confirming NK cell lymphoma. Tumor cells were positive for EBV. Lymphoma was deemed as Stage III. High risk factors included B symptoms and Ki-67. Patient was started on radiotherapy with 54Gy and concurrent chemotherapy to include 3 cycles of DeVIC (Dexamethasone, etoposide, ifosfamide/mesna, carboplatin). At 6month follow-up, symptoms improved and no recurrence noted. ENNKT mainly affects the nasal cavity and paranasal sinuses. Initial presentation is vague nasal symptoms but with disease progression, necrosis of the nasal mucosa increases. The earlier the biopsy is performed, the greater the chance of obtaining a representative disease sample (6). Historically localized NKTL was treated with anthracycline-based regimens with poor survival (8,9). L-asparaginase-based chemotherapy is more effective and safe for nasal-type NKTL (10). Addition of radiotherapy has shown survival benefit (11). Two recent trials showed that concurrent chemoradiotherapy with DeVIC improved the survival rate to 73% from a historical 45% with radiotherapy (12,13). Hence early diagnosis, risk stratification and multimodality treatment are necessary for improving outcomes.

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PHYSIOLOGIC PARASINAL MASSES IN A PATIENT WITH HEMOGLOBIN C DISEASE

Hansen D, Henegan C, Herrin V. University of Mississippi Medical Center, Brandon, MS.

Case Report: Extramedullary hematopoiesis (EMH) refers to the growth of extramedullary cells of normal non-malignant origin but not of the typical myeloid origin. EMH is a benign condition and usually presents in patients with known hemoglobinopathies. It is localized to the liver and spleen in most cases; however, it may also develop in paravertebral areas. Here, we report a case where a patient was noted to have multiple paraspinal masses on imaging which, upon biopsy, were found to be areas of EMH leading to a further diagnostic evaluation.

A 73-year-old African American man was admitted for resection of an atypical lipomatous tumor of the left thigh. On admission, he underwent Computerized Tomography (CT) of the chest, abdomen and pelvis to follow up a pre-operative Chest x-ray (CXR) concerning for hilar lymphadenopathy. CT was notable for multiple paraspinal masses anterior to the thoracic (T) spine at T8, T9 and T10 as well as a spleen split post a prior partial resection. Biopsy of a paraspinal mass was performed with pathology notable for extramedullary hematopoiesis concerning for a myeloproliferative neoplasm versus a hemoglobinopathy.

Due to the patient’s underlying dementia, an extensive prior medical history was unable to be obtained. He did, however, report a prior abdominal surgery after being “kicked by a mule.” Physical exam demonstrated a midline abdominal vertical scar but no evidence of hepatosplenomegaly. He did not have any palpable masses affecting the thoracic spine or any neurological deficits. Labs demonstrated a normocytic anemia. Patient declined a bone marrow biopsy. Hemoglobin electrophoresis was consistent with Hemoglobin C Disease.

Any paravertebral mass consistent with EMH should prompt a clinician to consider an underlying hemoglobinopathy in addition to myeloproliferative neoplasms. EMH is common in patients with thalassemia and myeloproliferative disorders. However, only one literature case in PubMed reports a patient with Hemoglobin C disease and paravertebral EMH.

Parasinal extramedullary hematopoiesis may cause a variety of neurological symptoms if there is impingement of the spinal cord. Most patients remain asymptomatic and the masses are discovered incidentally by radiologic imaging. Early diagnosis is essential to rule out malignant processes of the posterior mediastinum.

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FIBROMYOBlastic Tumor

Klomjit S1, Verma R2,1, Hosuriezuck N1, Mohamed A1, Cobos E2,1. 1Texas Tech Health Sciences Center, Lubbock, Lubbock, TX and 2Texas Tech Health Sciences Center, Lubbock, TX.

Case Report: Background: Inflammatory myofibroblastic tumor (IMT) of head and neck also known as inflammatory pseudotumor is a rare malignancy. Roughly half of IMT’s carry rearrangements of the anaplastic lymphoma kinase (ALK) locus. The recurrence of the IMT is common. This case study demonstrates an approach to diagnosis and role of chemotherapy, anti-inflammatory drugs and targeted therapy of IMT.

Case: 34-year-old female with no known past medical history presented with known soft tissue neoplasm of nasopharynx since 2011. She had history of recurrent right nasal mass obstruction, epistaxis in 2011. She received complete removal of the masses. Pathological report showed benign inflammatory tumors. Her symptoms were progressive and she underwent repeat transnasal polypectomy in 2012. However, in April of 2014, she developed recurrent right-sided nasopharyngeal mass which was fast-growing and causing right nasal obstruction, progressive right facial pain, difficulty in speech and swallowing, facial disfiguring, and 20 pound weight loss. The biopsy of the right sinonasal mass was performed. The review of the first pathology showed inflammatory fibrososarcoma with anaplastic lymphoma kinase (ALK) receptor positive. She was seen at MD Anderson for second opinion and started on crizotinib which is targeted therapy for ALK positive tumors. Repeat biopsy done at MD Anderson was ALK negative. Her crizotinib was discontinued. She was admitted to our hospital for further management. Her tumor was progressive. Her imaging was suggestive of large mass nearly obliterates the right nasal cavity and right maxillary sinus, extending into the right oral cavity, also submandibular and cervical lymphadenopathy. We treated her with chemotherapy (ifosfamide and dacarbazine) along with radiation, and celecoxib. She responded minimally to steroids. She finished 3 cycles of chemotherapy and COX-2 inhibitors. She had major clinical response.

Discussion: Our patient presented with recurrence of IMT on her maxilla which surgical resection would be curative but more disfiguring. It is generally a benign lesion, but has potential for local invasion and recurrence. Various modalities such as chemotherapy and radiotherapy are tried with limited success in most patients.

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UNUSUAL CAUSE OF DIFFUSE FDG UPTAKE ON PET SCAN

LeTellier SM. University of Mississippi Medical Center, Jackson, MS.

Case Report: A 30 year old male presented with two weeks of right flank pain, shortness of breath, and chest pain. A CT of the abdomen revealed a 0.6 cm area of ...
NAPROXEN INDUCED SEVERE THROMBOCYTOPENIA: A CASE REPORT

Master S1,2, Dwary A1,2, Dembila V1,2, Feist Weiller cancer center, Shreveport, LA and VA Hospital, Shreveport, LA.

Naproxen is widely used over the counter medication for pain, dysmenorrhea, gout and various inflammatory disorders. Here we present a rare case of naproxen induced severe thrombocytopenia.

Case Report: 71 year old Caucasian male with history of coronary artery disease, diabetes, osteoarthritis had symptoms of sore throat and joint aches. He started taking naproxen for symptomatic relief. Patient had two episodes of melena and petechial rash on his arms and legs after starting naproxen. Patient was evaluated at outside hospital and initial platelet count was 18,000. Patient was referred to our VA Hospital. On arrival, his complete blood count showed platelet count of 36,000 with Hb 10.4 gm % and normal WBC count. On exam, patient had petechial rash on upper and lower extremities with no hepato-splenomegaly or lymphadenopathy. Naproxen was stopped and detailed work up for thrombocytopenia was negative. He was on no other offending medicines known to cause thrombocytopenia. Hemoglobin remained stable and blood transfusion was not required for asymptomatic anemia. Platelets counts started to improve in couple of days after stopping naproxen and were normal by day 12. Patient did not have any more melena and his hemoglobin normalized by day 12.

Discussion: Drug-induced thrombocytopenia is a reversible form of thrombocytopenia and it should be suspected in a patient who presents with new onset of thrombocytopenia or recurrent episodes of acute thrombocytopenia, without an obvious alternative etiology. Our patient had a severe thrombocytopenia that resulted in gastrointestinal bleed and petechial rash. The diagnosis of drug-induced thrombocytopenia is made clinically by confirming thrombocytopenia, establishing a temporal relationship to a drug, eliminating other causes of thrombocytopenia and by documenting prompt resolution of thrombocytopenia after discontinuation of the suspected drug. Drug-induced thrombocytopenia is commonly caused by drug-dependent antibodies and less commonly by drug-induced bone marrow suppression. Most patients with drug-induced thrombocytopenia require no specific treatment. In patients with severe thrombocytopenia and/or severe bleeding, the recommendation is to do supportive platelet transfusions.

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CHRONIC MYELOID LEUKEMIA (CML) WITH EXTRA MEDULLARY BLAST CRISIS PRESENTING AS PLEURAL AND JOINT EFFUSIONS

Nagireddy S, Bigelow C, Lam J. UMMC, Jackson, MS.

Case Report: We present a 21 year old AAM who got diagnosed with CML in August’12. He was initiated on Imatinib. In February’14 his treatment was changed to Dasatinib secondary to failure. Around same time he developed severe bilateral knee pain and swelling for which MRI of the knee was performed. It showed diffuse marrow infiltration related to leukemia. He developed severe pain requiring narcotics and even palliative radiation to the knee with minimal improvement. Then he noted to have right pleural effusion. A thoracentesis was performed and pleural fluid analysis showed numerous myeloid precursor cells with many immature forms and 9% blasts. His CBC showed WBC 6.0 TH/cmm, Hb/Hct 9.5/28.8 g/dL and platelets 251 TH/cm. His BM biopsy showed with <10% cellularity and trilineage hematopoiesis with no evidence of acute leukemia. His Bcr/Ab1 (p210) was 71.3% on the International scale. Since his marrow showed no acute leukemia, we believed that his CML converted to blast phase with extra medullary blast crisis. His Dasatinib was held and was initiated on chemotherapy with 7+3 with Idarubicin and Cytarabine. Day 14 pleural fluid cytology and Flow cytometry revealed another BM biopsy at day 29 secondary to poor count recovery, which showed hypocellular bone marrow with <5 % cellularity and marked osteosclerosis. His Bcr/Ab1 after around day 30 dropped to 4.8%. At time of this submission, we still wait for his counts to recover. Currently he is been worked up for allogenic HSCT.

Extra medullary involvement is reported ~ 10-20% of CML cases and commonly involves lymph node and spleen. Extensive involvement of the pleura is very rare and pleural effusion in CML is poorly understood. There are several possible mechanisms of pleural effusion, which include leukemic infiltration of the pleura, bleeding into the pleural cavity, obstruction of pleural capillaries, pleural extra medullary hematopoiesis, and nonmalignant causes including infections, TKIs. Extra medullary hematopoiesis without bone marrow involvement is very rare. Here we present an unusual case of CML with extra medullary blast crisis presenting as a pleural effusion and joint effusion. Involvement pleura should be considered as a poor risk factor and aggressive therapy should be planned.

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BICLONAL GAMMOPATHY OF SIGNIFICANCE - REMISSION FOLLOWING AUTOLOGOUS STEM CELL TRANSPLANTATION

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Introduction: Biclonal gammopathies are characterized by simultaneous appearance of two different M proteins. Biclonal gammopathies occur in at least 5% of patients with clonal gammopathies with two thirds of such patients having undetermined significance. The remainder have multiple myeloma, Waldenstrom's macroglobulinemia, or other lymphoproliferative diseases.

Case report: We report a case of sixty year old white male who presented with complaints of fever, night sweats and weightloss with work up revealing multiple osteolytic lesions and extensive lymphadenopathy. Protein and immunofixation electrophoresis revealed two monoclonal proteins - IgM kappa and IgA lambda. Bone marrow biopsy showed lambda light chain restricted CD138 positive plasma cells.
cells (50%) along with involvement by B cell lymphoproliferative process (30%). Excisional lymph node biopsy was done which showed findings consistent with nodal marginal zone lymphoma.

He was initially treated with bortezomib, liposomal doxorubicin and rituximab for 6 cycles following which his lymphoma went into remission, but achieved only partial remission in regards to his myeloma. Later he was treated with lenalidomide, dexamethasone with very good partial response (VGPR) but after 3 cycles it had to be held due to refractory Clostridium difficile colitis. Two years later his myeloma progressed at which point he was started on cyclophosphamide/bortezomib/dexamethasone. After 8 cycles of this regimen, he attained VGPR again and subsequently underwent autologous stem cell transplant. He has been in stringent complete response post transplant with no disease recurrence so far.

Conclusion:
Bicalutamidic granulopathy is a relatively rare entity with no clear treatment guidelines. Here we present a rare case of bicalutamidic granulopathy / Multiple myeloma coexistent with nodal marginal zone lymphoma which went in to remission post autologous stem cell transplant.

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NOT ALL APPLE CORES ARE FROM AN APPLE: EXTERRMUDARY PLASMACYTOMA MIMICKING COLON CARCINOMA

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Case Report: A 72 yo woman with known IgM kappa multiple myeloma who was showing a very good partial response (VGPR) to 9 cycles of bortezomib, lenalidomide and dexamethasone presented with a week of dark stools, fatigue, dyspea and intermittent abdominal pain. She had no family history of colon cancer, and her colonoscopy three years prior was normal. PE: vitals normal, abdomen soft, non-tender, BS present and no palpable mass. Stool positive for occult blood. Hgb 8 g/dl. Abdominal CT showed a 9 cm mass at the hepatic flexure with an apple core deformity causing a marked narrowing of the lumen. Colonoscopy confirmed a large ulcerative mass in the distal right colon. A hemicolectomy was performed. Histopathology showed ulcerated colonic mucosa infiltrated by sheets of poorly differentiated tumor cells with characteristic staining and markers confirming a plasmacytoma.

Discussion: Physicians encounter both multiple myeloma (MM) and adenocarcinoma of the colon individually on a fairly regular basis. It is uncommon to suspect both in the same patient. Extramedullary plasma cytoma (EMP) is a recognized feature of MM which may occur in 2-20% of myeloma patients. Most often these occur in soft tissue, skin, lymph nodes, liver, spleen and kidneys. Although unusual, GI manifestations of myeloma usually affect the stomach and small intestine. MM involving the colon is truly rare. Our patient’s presentation and clinical picture looked highly suspicious for a typical colon cancer; hence it was surprising when the pathology returned EMP. Not only can a GI EMP clinically mimic a colonic carcinoma, but it may histologically show poorly differentiated cells that resemble a lymphoma, making the diagnosis difficult. Her colonic EMP occurred despite having a VGPR to bortezomib and lenalidomide, which are already the preferred agents for MM with extramedullary involvement. Following surgical resection, our patient’s regimen was changed to bortezomib and lenalidomide, which are already the preferred agents for MM with extramedullary involvement. Following surgical resection, our patient’s regimen was changed to bortezomib and lenalidomide, which are already the preferred agents for MM with extramedullary involvement.

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PITUITARY APOPLEXY A RARE COMPLICATION OF PROSTATE CANCER TREATMENT GEORGES TANIOS MD, DEVARAPIAJAISHANKAR MD

Tanos G, Jaishankar D. East Tennessee State University, Johnson City, TN.

Case Report: An 85 year old male presents with a new onset, severe, pulsating, frontal headache that started four hours after his first dose of intramuscular leuprolide. The headache is associated with left eye pain, photophobia and vomiting. He was recently diagnosed with non-metastatic, high grade, prostate adenocarcinoma. He is in moderate distress. Neurological evaluation is normal. No meningismus noted.

Pertinent labs include TSH 0.131, cortisol 2.5, ACTH 6.2, insulin-like growth factor 78, free testosterone 3.33, prolatin 2.7, arginine vasopressin <0.8, sodium 129 with normal FSH, LH, growth hormone and free T4. MRI confirms pituitary hemorrhage in a 1.9 cm pre-existing adenoma. Findings consistent with pan Hypopituitarism and replacement therapy with hydrocortisone and levothryroxine initiated.

Thunderclap Headaches could be due to subarachnoid hemorrhage, unruptured aneurysm, cerebral venous thrombosis, meningitis, ischemic stroke or pituitary apoplexy. Pituitary apoplexy results from sudden hemorrhage into the pituitary gland often into a pituitary adenoma. Headache, diplopia and reduced vision are common symptoms with associated pituitary hormonal deficiencies. These deficiencies are noted on labs and not readily evident clinically. Thirty percent of pituitary adenomas are nonfunctioning of which the majority (over 80 percent) is hypofunctioning gonadotroph. Pituitary apoplexy may occur spontaneously, during pregnancy, surgery and with anticoagulant and gonadotropin-releasing hormone (GnRH) use.

Androgen deprivation therapy (ADT) with gonadotropin-releasing hormone (GnRH) analogues is standard of care in the treatment of prostate cancer both in the adjuvant (concurrently with radiotherapy) and metastatic setting. The risk of tumor flare, vasomotor symptoms, reduced libido, osteopenosis, metabolic syndrome, and cardiovascular events are well established. The rare risk of pituitary apoplexy is ever-present given the not infrequent incidence of nonfunctioning adenomas in the population. Prompt identification

breast 6 months ago while doing a self-exam. In the last few weeks this had increased in size with associated skin changes and pain. Physical examination showed a 21 cm x 24 cm erythematous right breast mass with serosanguineous drainage. Axilla and nipple was obliterated with underlying ecchymosis. HER-2 over expressing ductal carcinoma responded to five cycles of Trastuzumab, Cyclophosphamide, Docetaxel and Gemcitabine (CEA from 48.5 down to 1.1 ng/ml). A year later chemotherapy was interrupted for mastectomy. ER/PR studies were negative. Subsequent treatments included Gemcitabine Vinorelbine and Nabpaclitaxel with Traztuzumab. 4 years later, she presented with angiography along with dysarthria. An MRI of the head revealed a 4 cm bony mass in the left skull base compressing cranial nerve XII. She received stereotactic radiation treatment plus Doctaxel, Trastuzumab, and Pertuzumab and had improvement. Her breast pathology was then found to be AR positive (>90%), and was placed on therapy with Bicalutamide, Trastuzumab, and Pertuzumab. She remained asymptomatic and tolerated this regimen well. Her CEA has decreased for the last few months.
and intervention of this potentially fatal complication comes from recognition of Pituitary apoplexy as a differential diagnosis of Thunderclap headaches.

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UNUSUAL CAUSE OF THROMBOCYTOPENIA

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Case Report: Merkel cell carcinoma is a rare and highly aggressive neuroendocrine carcinoma of the skin with a high mortality rate. We report a case of thyroidectomy secondary to bone marrow involvement by MCC. 79 yo male with recently diagnosed MCC of the left cheek presented with multiple skin bruises and was found to have profound thrombocytopenia. He had no history of bleeding disorders and did not have any recent administration of heparin. Pertinent lab results (tab 1). Due to persistent thrombocytopenia patient underwent a bone marrow biopsy, which revealed sheets of metastatic cells, consistent with metastatic MCC. Patient decided to proceed with supportive care only and was discharged home with hospice. MCC is a rare, aggressive neuroendocrine tumor of the skin with a high propensity for local, regional and distant spread. The number of cases, reported annually in U.S is quickly rising, secondary to advances in diagnostic technology, growing elderly population and larger population of immunosuppressed patients. Pathogenesis is not clearly understood but involves a double stranded DNA virus with oncogenic features named Merkel cell polyomavirus. Other suggested risk factors include sun exposure, immunosuppression and history of prior malignancy. Diagnosis is made based on immunohistochemistry. AJCC TNM classification system applies for staging. Prognosis is poor with 5 year overall survival rate of 75% for local disease, 59% for locoregional disease and 25% for advanced disease. Treatment options for advanced disease are limited and there is no literature to support specific chemotherapy regimen. Involvement into clinical trial is a preferred option. Our patient had a rapidly progressive course and presented with advanced disease. Due to age, severe thrombocytopenia and poor performance status he was considered a poor candidate for systemic chemotherapy. MCC is a rare and aggressive cancer that might cause infiltrative bone marrow disease. Occasionally patients present with profound thrombocytopenia.

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THYROID NODULE: NOT AS SIMPLE AS IT SEEMS

Bader G1, Puzanov I, CHAKRABORTY K1. 1ETSU-QCOM, Johnson City, TN 2Hendersonville University Medical Center, Nashville, TN.

Case Report: Benign nodules and primary thyroid cancer are most common etiologies of incidental thyroid lesions. Clinically evident metastasis to the thyroid account for 2-3% of thyroid cancer. We report a rare case of clear cell renal cell carcinoma (RCC) with solitary metastasis to thyroid. A 69-year old male presented with complaint of feeling neck nodule on self-palpation. Past history was significant for history of nephrectomy, performance status, hemoglobin, lactate dehydrogenase, calcium and albumin (MSKCC- Memorial Sloan-Kettering Cancer Center prognostic model). In the era of targeted treatment newer algorithm with incorporation of platelet, neutrophil count and subsequently molecular markers (VEGFR-1, p53 etc) further validates MSKCC data. Metastectomy either with or without short course of neo-adjuvant VEGF or TKI helps with proper patient selection. In the case series from Mayo and M. D Anderson cancer center metastectomy led a median survival of 29% over 5 year supporting role of complete resection to improve long term survival. Thyroid metastases is a must differential diagnosis of thyroid nodule. Thus we suggest an expedited pathological diagnosis of occult thyroid nodule especially in individuals with a history of cancer even decades after initial diagnosis.

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A DRUG TO PICK ABOUT A CHEST PAIN

Chakraborty K, Jaishankar D. ETSU-Quillen College of Medicine, Johnson City, TN.

Case Report: Challenges of recognizing toxicities of oral chemotherapy agents have reached beyond oncology to internist’s circle. Cytopenia, diarrhea, nausea and neutropenic fever are well recognized side effects. Here we present a rare side effect of Capecitabine (pro drug of 5Fluorouracil/5Fu). A 50 year old male presented to emergency room with complaint of chest discomfort, anxiety and heartburn. History was significant for remote event of pacemaker placement, new diagnosis of locally advanced rectal cancer needing chemo-radiation but unremarkable for chronic co-morbidities. Examination including cardiac-pulmonary auscultation was unremarkable. Cardiac troponin was normal. Intravenous morphine and Zofran resolved symptoms completely. An EKG showed normal sinus rhythm. Patient again experienced similar events 12 hrs later after receiving scheduled dose of Capecitabine. Telemetry and a repeat EKG confirmed acute ST changes. Repeat troponin was negative. Patient was treated with nitroglycerine patch. A left heart catheterization was normal. Based on the correlation of events Capecitabine related coronary vasospasm was likely etiology. Patient was discharged home on oral diltiazem and Capecitabine was discontinued after discussion with oncology team. Patient completed neo-adjuvant radiation treatment and the subsequent surgery without further events. This conclusively proved Capecitabine as the etiology for coronary vasospasm. Capecitabine needs three step enzymatic conversion mainly in liver and tumor cells to active drug 5-Fu. Preferential presence of thymidine phosphorylase in tumor cells, final enzyme in this conversion allows Capecitabine to reach a higher therapeutic concentration compared to bolus 5Fu. Myocardial injury, immuno-allergic reaction and vasospasm are underlying mechanism of cardiac toxicities from use of 5Fu. Mechanistically toxicities of Capecitabine are similar to infusional 5Fu. Activity of thymidine phosphorylase in atherosclerotic plaques can increase risk but coronary vasospasm can happen even with normal coronary vasculature. Withdrawal of Capecitabine, long acting nitrates and calcium channel blockers are main mode of treatments. Awareness among internists about Capecitabine related chest pain/coronary vasospasm is important in early diagnosis considering frequent use of this drug in different malignancies.

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WHEN HIT STRIKES WITHOUT BEING HIT

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Case Report: Heparin Induced Thrombocytopenia (HIT) is a prothrombotic disorder initiated by heparin exposure that is classically suspected in the setting of thrombocytopenia. However, HIT can present with thrombosis despite normal platelet counts.

A 27-year-old man with a history of extensive venous thromboses over the last 3 years presented with bilateral lower extremity pain. Extensive laboratory work-up for thrombophilia was negative prior to admission. Of
note, the patient had an Inferior Vena Cava filter (IVC) placed for a recent left lower extremity DVT. Imaging demonstrated thrombosis of bilateral femoral veins to the level of the IVC filter. Peripheral vascular intervention was performed (with heparin exposure) with transient improvement in venous blood flow and the patient was started on bivalirudin. However, his clot burden continued to progress and the patient required subsequent interventions (with most interventions involving some heparin exposure) and trials of several anticoagulants, including apixaban and dabigatran because of persistent worsening of his clots despite full dose anticoagulation. Given the progression of his clotting after multiple interventions and history of heparin administration, HIT was suspected despite his normal platelet count of 455 on admission and normal to high values throughout his hospitalization(Fig1). HIT IgG antibody was positive and the diagnosis of HIT was confirmed with a positive serotonin release assay. Once all heparin exposure ceased and the patient was placed back on bivalirudin, he did not experience any additional extension of the venous clots.

The classic presentation of HIT consists of thrombocytopenia and thrombosis following the administration of heparin. However, HIT should be considered in patients who have received heparin with unexplained thrombosis even in the absence of thrombocytopenia.

CD5-NEGATIVE MANTLE CELL LYMPHOMA WITH CCND1 AMPLIFICATION

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Case Report: Mantle cell lymphoma (MCL) is an aggressive B-cell non-Hodgkin lymphoma characterized by Cyclin-D1 overexpression secondary to translocation t(11;14)(q13;q32). Classically, MCL cells express the B-cell markers CD19, CD20 (bright), CD23 (dim to negative), and the T-cell marker CD5. Rare cases of CD5-negative MCL have been reported, where diagnosis has been made based on morphology, immunophenotype, and the presence of Cyclin-D1 overexpression secondary to t(11;14)(q13;q32). Here we report a case of CD5-negative MCL with absence of t(11;14)(q13;q32) diagnosed by Cyclin-D1 overexpression secondary CCND1 duplication.

A 67-year old Caucasian female was diagnosed with ER+/PR+/ Her2- T1cN0M0 invasive ductal carcinoma of the breast, status post lumpectomy and radiation therapy. A sentinel lymph node biopsy showed B-cell lymphoproliferative disorder and was negative for breast cancer. Absolute lymphocyte count was 1070/μL. Peripheral blood and bone marrow flow cytometry showed a kappa-restricted monoclonal B-cell population expressing CD19, CD20 (bright), CD23 (dim to negative), and negative for CD5, CD10, and CD103. Bone marrow biopsy was hyper-cellular for age (90%) mainly consistent of the above described clone that also tested positive for CD21, Pax5, Cyclin-D1 (very bright), BCL2 and Ki67 (10%), and negative for SOX11 and BCL6. Karyotype showed monosomy 12, monosomy X, and 11q deletion. FISH was negative for t(11;14)(q13;q32), however it showed an extra signal of CCND1 consistent with gene amplification.

Diffuse large B-cell lymphoma, follicular lymphoma, and chronic lymphocytic leukemia can express Cyclin-D1 as well, however in most cases this expression is not strong, and their diagnosis can be easily differentiated from MCL based on morphology and immunophenotype. In our case, the diagnosis of MCL was based on morphology, immunophenotype, and Cyclin-D1 overexpression secondary to CCND1 amplification and not t(11;14)(q13;q32).

To the best of our knowledge, this is the first reported case of CD5-negative mantle cell lymphoma with cyclin-D1 overexpression secondary to CCND1 amplification. The absence of CD5, SOX11, and t(11;14)(q13;q32) did not preclude the diagnosis of MCL.

PULMONARY MUCOEPIDERmoid CARCINOMA CAN OFTEN BE MISTAKEN FOR MUCH SINISTER adenocarcinoma: A CASE REPORT

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Case Report: Background: Salivary gland type lung cancers are uncommon. They can be often misdiagnosed as squamous or adenocarcinomas of lung. We present a case of pulmonary mucopidermoid cancer (MEC) which was misdiagnosed as adenocarcinoma of lung based on a bronchoscopic biopsy.

Case: A 34 year old African American man with a past medical history of childhood asthma, bronchitis, and a remote six month history of cigarette smoking was evaluated in 2005 for a left main stem bronchus lesion found on computed tomography (CT). The patient was lost to follow-up. Seven years later the same bronchial lesion was found on CT and a bronchial biopsy of the lesion was performed. Cells from the lesion were found to express cytokeratin 7 and lack the expression of cytokeratin 20 or TTF1, felt to be consistent with adenocarcinoma. Further evaluation and staging with PET/CT showed localized disease without regional spread or metastasis. He underwent a left-sided pneumonectomy with mediastinal lymph node dissection. The tumor measured 1.2 x 0.7 x 0.7 cm and was located just distal to the left bronchial bifurcation. Immunohistochemistry showed that the tumor cells were positive for CK7, p63,EMA, and cytokeratin 14, while negative for S100, TTF1, and Napsin-A. Contrary to the diagnosis of adenocarcinoma made after the bronchoscopy, results from the surgical pathology revealed an immunoprofile and tumor morphology more consistent with MEC. All mediastinal lymph node dissections were negative. The patient recovered well from surgery and surveillance chest CT at 4 months and 12 months showed no evidence of disease.

Discussion: MEC is predominantly a head and neck tumor, however, can be found in lungs, skin and breast among other body parts. Pulmonary MEC represents approximately 9% of all malignant pulmonary cancers in children and only 0.2% of all lung cancers in adults. Patients usually present with respiratory symptoms like cough, wheezing and hemoptysis. Sometimes it is challenging to differentiate MEC from squamous cell or adenocarcinoma especially if the tissue is derived from a small biopsy specimen. Nevertheless it is important to make a correct diagnosis since, MECs have a better overall survival.

TRANSITION OF A PANCREATIC NEUROENDOCRINE TUMOR FROM GHRELINOMA TO INSULINOMA: A CASE REPORT

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Case Report: INTRODUCTION: Pancreatic neuroendocrine tumors (PNETs) have been reported to produce insulin, gastrin, glucagon, vasoactive intestinal peptide (VIP) and more recently, ghrelin. Ghrelin, produced by oxyntic cells in the stomach to regulates appetite, growth hormone, gastric secretion and gut motility has also been found in hypothalamus and other endocrine glands like pituitary, pancreas etc.

CASE: A 60 year old Caucasian man with a history of mitral valve prolapse, type 2 diabetes mellitus and tobacco use was diagnosed with a metastatic pancreatic neuroendocrine tumor in December 2009 after having abdominal pain as his presenting complaint. His initial labs at the time of diagnosis of PNET showed normal urinary 5-HIAA, glucagon, VIP and insulin levels. His initial chromogranin A levels were 3,625 ng/ml in 12/2009 which jumped to 14,000 ng/ml in few months. He was unfit for surgery due to presence of metastatic disease. He was MIBG positive, but he wasn’t considered ideal candidate for MBG therapy either. He was started on Sandostatin LAR 30mg in July 2010. At that point he complained of weight loss, flushing and intermittent severe abdominal pain but no diarrhea. He was started on the combination of capetebit 750mg/m2 and temozolomide 200mg/m2. He clinically responded well to the initial 3 cycles of the chemotherapy with improvement in fatigue and appetite. His ghrelin levels also
trended down from 18,651 pg/ml in December 2013 to 6,342 pg/ml in February 2014. Meanwhile his insulin level went from 33 uiu/ml in 6/2013 to 119 uiu/ml. C-Peptide level was 10 ng/ml. He was symptomatically hypoglycemic and his endocrinologist started continuous glucose monitoring and an increased caloric regimen.

DISCUSSION: Here, we present a case of metastatic functional PNET that initially presented as a ghrelinoma, which later transformed into an insulinoma.None(128,872),(871,917) from being an unusual variant of metastatic PNET, this case demonstrates that changes in functional tumor biology can sometime be more morbid than the metastatic disease itself.

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TROUSSEAU SYNDROME UNMASKING AN ASYMPTOMATIC GALLBLADDER ADENOCARCINOMA

Hall MA1, Chauhan A1, Engel LS1, Wang Y2. 1LSU Health Sciences Center; New Orleans, LA and 2LSU Health Sciences Center, New Orleans, LA.

Case Report: Case: A 61 year old African American woman with a past medical history of insulin dependent diabetes mellitus and hypertension presented to our emergency department with a 5 day history of bluish black discoloration on her left 1st and 2nd toe. On examination, there was dark reddish-brown discoloration of the plantar surface of the left 1st and 2nd distal phalanges. There was no tenderness to palpation, nor purulence, bloody discharge, or cellulitis. Although signs of distal ischemic changes were noted in the 1st and 2nd toes, appropriate capillary refill was noted in the remaining phalanges. The patient had 2+ Femoral, Popliteal, Dorsalis Pedis, and 1+ Posterior Tibial pulses on the affected side. The patient showed no signs of venous insufficiency on further examination, and had no evidence of osteomyelitis. Workup for possible embolic disease incidentally revealed an exophytic lesion arising from a focally thickened and irregular medial wall of the gallbladder body associated with localized peri-portal and aortic lymph node enlargement. Cholecystectomy was performed and pathology revealed a moderately differentiated invasive papillary adenocarcinoma of the gallbladder, stage T2N0M0. Postoperatively, she received no adjuvant chemotherapy, given the extent of her resection and clinical stage. She remains free of local or metastatic recurrence.

Discussion: Trousseau Syndrome first described by Armand Trousseau in 1865 is characterized by presence of subacute hypercoagulability (eg. migratory thrombophlebitis, microangiopathy, arterial emboli, and chronic disseminated intravascular coagulopathy) in association with an underlie malignancy, oftentimes occult. There is strong data to suggest association of pancreatic and pulmonary malignancies (20 and 24% respectively) with Trousseau Syndrome but other malignancies such as prostate, stomach, colon, and hematological malignancies also have been reported.1 There is about 10% chance of detecting an occult malignancy in a patient with symptomatic idiopathic venous thromboembolism. However, gall bladder adenocarcinoma as cause of thrombophlebitis is extremely rare and has been reported only once before.

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A TRANSUDATIVE ASCITES FROM A NOT SO TRANSPARENT STOMACH

DAS D, Spradling E, Jaishankar D, CHAKRABORTY K. ETSU-QCOM, Johnson City, TN.

Case Report: A recurrent transudative ascites can represent varied etiologies including heart failure, liver disease, malignancy and nephrotic syndrome. Refractory ascites in the background of weight loss and bowel obstruction can indicate a malignant process. Here we present a role of endomyelitis. Refractory transudative ascites.

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HIGH GRADE B CELL LYMPHOMA PRESENTING WITH ARTHRHALGIAS AND ISOLATED BONE MARROW INVOLVEMENT

Devani K1, Phemister J1, Sajnani K1, Kulbacki E2, Ginn D1. 1East Tennessee State University, Johnson City, TN and 2Highlands Pathology, Kingsport, TN.

Case Report: Non Hodgkin lymphoma with isolated bone marrow involvement is rare. We present a case of 40 year old female who presented with arthralgias, myalgias, night sweats, fever and unintentional weight loss for last 4 months. Physical exam was unremarkable except for tender lumbar spine. The initial laboratory work was significant for elevated ESR, CRP, alkaline phosphatase and lactate dehydrogenase. CT scan of the chest, abdomen and pelvis did not show any lymphadenopathy or hepatospleno-megaly. Abnormal signals and enhancement were noticed on MRI of lumbar spine, suggestive of a primary bone marrow process (Fig. 1). Subsequently, bone marrow biopsy illustrated intermediate size cells with variable cytopenia, variable 5% of all gastric cancers has a dismal prognosis despite multi-modality treatments including surgery, chemo-therapy and radiation. Thickening and rigidity of the gastric wall are characteristics of this disease. Higher prevalence in younger individuals and late presentation with metastatic disease are common. Loss of expression of E-cadherin leading to defective inter-cellular adhesion accounts for the pathogenesis of diffuse gastric cancer. EUS is critical in diagnosing and staging diffuse gastric cancer. Diffuse gastric cancer is a ‘never miss’ differential to keep in mind while evaluating refractory transudative ascites.

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SPINDLE CELL CARCINOMA OF LUNG: RARE AND AGGRESSIVE TYPE OF NON SMALL CELL LUNG CANCER

Devani K1, Phemister J1, Sajnani K1, Rogers M2, Pierce D1. 1East Tennessee State University, Johnson City, TN and 2V A Medical Center, Mountain Home, TN.
Case Report: Spindle cell carcinoma (SpCC) is a rare type of pulmonary cancer representing only 0.2 to 0.3% of all primary pulmonary malignancies, with increased occurrence in men (5:1 male: female). SpCC bears a strong association with smoking. It is one of five subgroups of pulmonary sarcomatoid carcinoma, an uncommon histologic subtype of non-small cell lung cancer (NSCLC). These tumors are more aggressive than other subtypes of NSCLC and often present with early distant metastasis.

A 76 year old male with a 40 pack year smoking history presented with chest pain, shortness of breath, cough, hemoptysis and unintentional weight loss. Physical exam was unremarkable on initial presentation. Blood work was remarkable for hyponatremia, hypercalcemia and elevated alkaline phosphatase. CT of the chest revealed a large right apical mass measuring 10 x 10 x 11.5 cm with mediastinal lymphadenopathy (Fig. 1). Subsequently a CT guided biopsy of right apical mass demonstrated a malignant spindle cell proliferation (Fig. 2A), thus establishing the diagnosis of SpCC, which was confirmed with immunohistochemical studies (Fig. 2B). PET scan findings were consistent with high grade necrotic right lung mass with metastasis to the mediastinal, right hilar and abdominal lymph nodes, along with diffuse distant metastasis to liver, bilateral adrenals and several bones. Patient is currently being treated with radiation and chemotherapy.

Prognosis of SpCC is poor regardless of surgery, radiation, and chemotherapy. Early diagnosis and aggressive treatment is crucial in order to prolong the life of patients with pulmonary SpCC.
can be secondary to chromosomal abnormalities. The most common is a result of the 4q12 rearrangement that causes the fusion product FIP1L1/PDGFR-A. This rearrangement leads to constitutive tyrosine kinase activity, and thus is associated with a rapid and sustained response to tyrosine kinase inhibitors such as imatinib.

We report the case of a 51-year-old African American male without comorbid conditions who presented with hematemesis and melena. He also noted unintentional weight loss over a two-month period, malaise, and fatigue. His initial laboratory analysis showed leukocytosis of 20,000/mL with 48% eosinophils and an absolute eosinophil count of 9,340/mL. His bone marrow biopsy and aspirate showed marked myeloid predominance with maturation and increased eosinophils without increase in immature blasts. FISH profile probe revealed a deletion of LNXL(4q)(q12q12) resulting in a FIP1L1-PDGFR-A fusion. Findings are compatible with myeloid/lymphoid neoplasms with PDGFR-A rearrangement. FIP1L1-PDGFR fusion is often seen in hypereosinophilic syndrome and chronic eosinophilic leukemia. Other labs were drawn to evaluate for parasitic infections and allergies. IgG: 1251 (680-1530mg/dl), IgM: 60 (40-168mg/dl), and IgE: 31 (10-360IU/mL) were within normal range. The patient tested negative for Strongyloides antibodies. The patient had no evidence of end organ damage, and his echocardiogram showed normal cardiac function. Gastric antrum biopsy showed active, moderate chronic regenerative and follicular gastritis. Imatinib therapy was initiated at 400 mg PO daily resulting in a rapid and progressive reduction in the absolute eosinophil count with normalization of blood counts within 4 weeks.

The incidence of the myeloproliferative variant causing hypereosinophilic syndrome is rare. However, the dramatic response to imatinib emphasizes the need to study the presence of the fusion product FIP1L1/PDGFR in all patients with eosinophilia of unknown etiology.

187 EXTRACAVITARY PRIMARY EFFUSION LYMPHOMA: AN ATYPICAL PRESENTATION OF AN UNUSUAL MALIGNANCY
Henegan JC, Hamilton R. University of Mississippi Medical Center, Jackson, MS.
Case Report: Introduction: Kaposis sarcoma-associated herpesvirus (KSHV) is associated with several human immunodeficiency virus (HIV)-associated diseases including Kaposis Sarcoma (KS) and primary effusion lymphoma (PEL). Here we describe an extracavitary PEL in a patient with HIV.
Case Report: A 47 year old man with HIV/AIDS presents to Medical Oncology clinic for follow-up of KS treated with chemotherapy 7 years ago and squamous cell carcinoma of the anus treated 2 years ago with chemoradiation. Regarding his HIV, he was diagnosed and started on antiretroviral therapy 7 years ago. On physical exam, 2 years had an absolute CD4 count < 250/mL despite an undetectable viral load. A CT Abdomen and Pelvis obtained for anular cancer surveillance demonstrates multiple retroperitoneal and abdominal soft tissue masses. He denied any fever, chills, weight loss, abdominal pain, night sweats, anal pain, hematochezia, hematuria, or difficulty with bowel movements. Physical exam was within normal limits. A biopsy of a soft tissue mass demonstrated a plasmablastic neoplasm reactive with CD138, vimentin, and KSHV that otherwise lacked staining with all other markers tested. Based on these findings, the patient was diagnosed with an extracavitary PEL.
Discussion: PELs are rare, aggressive B-cell neoplasms that usually lack pan-B-cell markers (i.e. CD19, CD20, and CD79a). PEL is caused by KSHV and associated with several human immunodeficiency virus (HIV)-associated diseases including Kaposis Sarcoma (KS) and primary effusion lymphoma (PEL). Here we describe an extracavitary PEL in a patient with HIV.

189 LOOKING AT BONE MARROW THROUGH AN OPHTHALMOSCOPE
Lee D, Spradling E, Shah RD, Chakraborty K. ETSU-Quillen College of Medicine, Johnson City, TN.
Case Report: AML (acute myeloid leukemia) is the most common acute leukemia among adults. The incidence in the United States have ranged 3 to 5 cases per 100,000 population. Fever, dyspnea, gum bleeding, headache, coagulopathy and even focal neurological deficit are common presenting complaints. Here we report a unique clinical symptom which was diagnostic for acute leukemia. A 24 year old female presented with sudden onset of floaters in left eye. She complained of progressive fatigue, shortness of breath, night sweats, weight loss and easy bruising. Examination revealed normal vitals, petechial rash, and no focal neurological deficit. An ophthalmological examination showed 'dot and blot' retinal hemorrhage. This prompted further laboratory studies showing profound anemia, thrombocytopenia and normal leucocyte count. A peripheral blood smear review confirmed presence of leukemic blasts. Subsequently bone marrow biopsy confirmed diagnosis of AML with maturation. Cytogenetic study revealed translocation (8:21) indicating favorable prognosis. Patient received standard induction chemotherapy regimen with doxorubicin and cytarabine. Follow-up bone marrow biopsy confirmed both molecular and hematological remission. Ocular involvement in AML although not uncommon but demands prompt diagnosis. A prospective study of 53 patients with recently diagnosed AML showed retinal or optic nerve involvement among 64 percent of the cases. In the study, involvement was unrelated to the patient's age, AML subtype, white blood cell count, or hematocrit. Platelets tend to be lower among the patients with retinopathy than those without retinopathy. Severe anemia or infiltration of the leukocytes may lead to ischemia of the nerve fibers. The damaged ganglion cells extrude the axoplasm unto the vitreous
**Case Report:**

Griscelli Syndrome is an autosomal recessive disease, which includes mutations in the MYO5A gene, RAB27A gene, and MLPH gene. Griscelli type 2 is associated with RAB27A gene mutation causing immuno deficiency due to impaired cytotoxic lymphocyte function and silvery hair due to impaired transport of melanosomes. We describe a case of a 3 month old female with pancytopenia, hepatosplenomegaly and silvery hair who was found to have Griscelli’s Syndrome.

She presented with fever (temperature 40.2°C), hypofibrinogenemia (137.4mg/dL), hypertriglyceridemia (537mg/dL). Additional workup showed elevated levels of interleukin-2-receptor (20,297unit/mL), ferritin (1,620mg/mL) and hemophagocytosis on bone marrow biopsy. She fulfilled the diagnostic criteria for hemophagocytic lymphohistiocytosis and was started on standard multiagent chemotherapy (etoposide, dexamethasone) with no remission of disease process. AML patients can present identically to those without ocular involvement. The ocular complications usually resolve with remission of disease process. Physical examination showed non pitting edema, pain to palpation and decreased range of motion of the left upper extremity. Initial laboratory studies found anemia and increased inflammatory markers. A computed tomography (CT) scan of the left upper extremity was consistent with humeral osteomyelitis complicated by extensive pyomyositis and abscess. Examination in the operating room found serosanguinous fluid, necrotic tissue and a soft tissue mass not consistent with the presumptive diagnosis of Buerger’s disease. Biopsies of the soft tissue mass and underlying bone were taken. The hematocrit and cosin stain showed a background rich in collagen demonstrating that the soft tissue mass was of fibroblastic differentiation. Electron microscopy revealed elongated accordion-like nuclei verifying that the mass also exhibited muscular differentiation. These histopathological findings confirmed the diagnosis of infantile myofibromatosis. Imaging studies were then used to classify the specific type of myofibromatosis found in our patient. A skeletal survey revealed multiple areas of periosteal reaction. These areas were also noted to have increased activity on the patients bone scan. Ultrasound, CT and magnetic resonance imaging results were negative for visceral involvement. This was a good prognostic factor, however due to the extensive and inoperable nature of our patients lesions chemotherapy was required. Low dose vinblastin and methotrexate are currently being utilized as treatment.

Soft tissue neoplasms are generally rare in children and can be a source of diagnostic confusion for the radiologist, pathologist and pediatrician. The purpose of this report is to review the presenting features, diagnosis and management of this rarely metastasizing tumor.

**Oculo-cutaneous hypopigmentation and hemophagocytic lymphohistiocytosis**

Mack J, Mian A. UAMS, Little Rock, AR

**Case Report:**

Oculo-cutaneous hypopigmentation and hemophagocytic lymphohistiocytosis is a rare autosomal recessive disease that is characterized by accelerated autoantibody-mediated platelet destruction by tissue macrophages located in the spleen and decreased platelet production. Patient is a 63 year-old obese black female with history of ITP, hypertension, and coronary artery disease/CABG 3 years ago who presented with epistaxis and frontal headache for one day. Patient underwent a splenectomy 30 years prior to presentation for ITP and platelet counts had subsequently normalized. The day of admission bleeding ceased spontaneously; however, platelet count was measured at 3,000 × 10^9 per liter. She received two units of platelets and 2 doses of IVIG. Patient was placed on Decadron 40 mg for five days. She had no response to either. Review of peripheral smear revealed no visible Howell Jolly bodies suggesting the presence of an accessory spleen. An ultrasound showed a mass 6.6x2.4 x 2.0 cm which was confirmed to be an accessory spleen by a nuclear medicine liver/spleen. Bone marrow biopsy was performed demonstrating no abnormalities. Due to her morbid obesity and h/o CAD the patient was deemed high risk for post op complications for a laparoscopic splenectomy. Rituximab therapy was started and platelet count responded to 353,000 × 10^3 four weekly treatments. Patient then developed left leg DVT and PE and hence rituximab therapy was started and platelet count responded to 353,000 × 10^3 /C2

**Thrombocytopenia in a splenectomized patient**

Menendez D, Khan YA, Koticha K, Koshy N. LSU Health Shreveport, Shreveport, LA

**Case Report:**

Idiopathic Thrombocytopenic purpura (ITP) is a common autoimmune disorder that is characterized by accelerated autoantibody-mediated platelet destruction by tissue macrophages located in the spleen and decreased platelet production. Patient is a 63 year-old obese black female with history of ITP, hypertension, and coronary artery disease/CABG 3 years ago who presented with epistaxis and frontal headache for one day. Patient underwent a splenectomy 30 years prior to presentation for ITP and platelet counts had subsequently normalized. The day of admission bleeding ceased spontaneously; however, platelet count was measured at 3,000 × 10^9 per liter. She received two units of platelets and 2 doses of IVIG. Patient was placed on Decadron 40 mg for five days. She had no response to either. Review of peripheral smear revealed no visible Howell Jolly bodies suggesting the presence of an accessory spleen. An ultrasound showed a mass 6.6x2.4 x 2.0 cm which was confirmed to be an accessory spleen by a nuclear medicine liver/spleen. Bone marrow biopsy was performed demonstrating no abnormalities. Due to her morbid obesity and h/o CAD the patient was deemed high risk for post op complications for a laparoscopic splenectomy. Rituximab therapy was started and platelet count responded to 353,000 × 10^3 four weekly treatments. Patient then developed left leg DVT and PE and hence rituximab therapy was discontinued. She continues to maintain her platelet counts 7 months after therapy. Splenectomy is the most effective therapy for ITP. This case highlights the importance of evaluating the peripheral smear in every case of ITP. Absence of Howell Jolly bodies on indicates the presence of an accessory spleen or a functional splenic implant left behind after surgery. Removal of accessory spleen or functional splenic implant left behind after surgery. Removal of accessory spleen or functional splenic implant is again curative but may not be always possible. Recent advances in treatment of relapsed/refractory ITP like rituximab and thrombopoietin mimetic agents are very effective but not without potential side effects.
ADENOCARCINOMA OF THE CERVICAL ESOPHAGUS IN AN 88-YEAR-OLD MALE: A CASE REPORT AND REVIEW OF THE LITERATURE
Payne J1, Mobley J2, Radhi S3. 1Texas Tech University Health Science Center, Lubbock, TX; 2Texas Tech University Health Science Center, Lubbock, TX and 3Texas Tech University Health Science Center, Lubbock, TX.

Case Report: Adenocarcinomas of the proximal esophagus are exceedingly rare. In this part squamous cell carcinoma is the usual pathology. Adenocarcinoma can originate from heterotopic gastric mucosa, mucosal and submucosal glands and from an extensive segment of Barrett's epithelium. In this clinical case synopsis an 88-year-old male presented with the chief complaint of dysphagia. Endoscopy revealed a large fungating mass with stigmata of recent bleeding in the upper third of the esophagus. Biopsy showed a moderately differentiated adenocarcinoma with no intestinal metaplasia and negative H. Pylori and Her2. PET/CT showed cervical and axillary lymph node metastasis as well as lung nodules consistent with metastatic disease and a large fungating mass abutting the pleura anterolaterally with rib and chest wall involvement. The patient was treated with palliative radiation therapy to the esophagus with a total dose of 6300 cGy in 35 fractions. He opted not to receive palliative chemotherapy as well. The patient is still alive 10 months after diagnosis. Only 28 prior cases of adenocarcinoma arising in the cervical esophagus have been documented through 2013. Of these, 25 were published in English and were included in the literature review. The mean age at diagnosis was 58.5 years with a range of 38–89 years. Retrospective review in these patients revealed the patient was a 60% survival rate at 1 year.

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Discussion:

T-cell prolymphocytic leukemia represents few leukemia diagnoses, 2% of all small lymphocytic leukemias in adults. Patients primarily present with lymphadenopathy, hepatosplenomegaly or skin lesions. Lymphocytosis with high levels of circulating medium sized lymphocytes along with anemia and thrombocytopenia are common. Dermatologic manifestations include skin nodules, erythroderma, maculopapular lesions, peripheral edema, conjunctival edema. Of note, lymphadenopathy is a common finding in T-PLL. Lymphocytosis with high levels of circulating medium sized lymphocytes along with anemia and thrombocytopenia are common. Dermatologic manifestations include skin nodules, erythroderma, maculopapular lesions, peripheral edema, conjunctival edema. Of note, lymphadenopathy is a common finding in T-PLL. Lymphocytosis with high levels of circulating medium sized lymphocytes along with anemia and thrombocytopenia are common. Dermatologic manifestations include skin nodules, erythroderma, maculopapular lesions, peripheral edema, conjunctival edema. Of note, lymphadenopathy is a common finding in T-PLL.
Conclusion: Hepcidin has been shown as increased in the setting of ACD and elevated ferritin and C-reactive protein in patients with chronic kidney disease. The choice of intravenous iron therapy in patients with concomitant ACD or compromised by iron restriction; would help clarify the best approach to disease management, other influences on erythroid regulation and erythroferron. It is in such individuals we hypothesize, that potential phase II/III clinical trials should be considered with novel Hepcidin inhibitors.

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GRANULOCYTIC SARCOMA (CHLOROMA) OF SKIN: A RARE MANIFESTATION OF CML

Wondimamegnehu NE, Edris H, Nugent K, D’Cunha N. Texas tech University health science Center, Lubbock, TX.

Purpose of Study: Myeloid sarcoma, also known as a chloroma, is a pathologic diagnosis for extramedullary proliferation of blasts of one or more myeloid lineages that disrupt the normal architecture of the tissue in which is found. These tumors can involve any part of the body, either concurrently or sequentially but preferentially involve orbits and subcutaneous tissue. Chronic myelogenous leukemia (CML) presenting with painful skin chloromas is an extremely rare manifestation. We report a patient who presented with large chloroma and was later diagnosed with CML.

Methods Used: Case analysis and literature review

Summary of Results: We report a 55-year-old woman who presented to the emergency center with a large painful scapular mass which extended into the axillary area. Laboratory tests showed WBC of 550,000K/ul and platelet of 776K/ul. Peripheral smear showed 15% myeloblast. Bone marrow biopsy showed Blasts - 0%, Metamyelocytes- 12%, Bands - 5%, Neutrophils- 40%, Eosinophils- 21%, Basophils- 8%. Cytogenetic study showed translocation t(9;22)(q34;q11.2) consistent with CML in accelerated phase. CT showed ill-defined soft tissue density mass in the right upper and mid posterior chest wall. Fine needle aspirate from the mass showed chronic myelogenous leukemia, bcr abl positive. Immunohistochemistry showed 30% of the cells positive for CD34, myeloperoxidase, and lysozyme. The patient was treated with imatinib, a tyrosine kinase inhibitor, and had a significant decrease in the WBC as well as the size of her chloroma.

Conclusions: Chloroma may develop during the course of or as a presenting sign of myelogenous leukemia which is the case in our patient. No prognostic significance exists between acute leukemia patients with choroma and those without. However, patients with choroma who have chronic leukemia or myeloproliferative disorders have a negative prognosis, because these tumors often occur during acute transformation. Chloroma are reported to be sensitive to chemotherapy or radiotherapy but the recurrence rate is about 20%. Our case suggests that CML can present with rare skin manifestations and chloroma must be included in the differential diagnoses of soft tissue mass.

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AN UNUSUAL CAUSE OF BACK PAIN

Aldrete S1, Moanna A2,1. 1Emory University, Atlanta, GA and 2Atlanta VA Medical Center, Decatur, GA.

Case Report: 66 year-old man admitted with worsening low back pain ten days before his presentation. Lumbar MRI revealed a ventral epidural abscess and discitis at L4-L5. He underwent emergent lumbar decompression and abscess evacuation and was started on empiric antibiotic coverage with vancomycin and meropenem. Intraoperative bacterial, fungal and mycobacterial cultures were negative. He underwent revision laminectomy two weeks after his initial surgery and tissue was sent for repeat cultures and pathology. Repeat cultures remained negative and pathology showed reactive inflammation.

He had a remote history of working in a slaughterhouse and given his negative work up, bacterial serologies were ordered. Brucella and bartonella serologies were negative. Finally, coxiella serologies came back: Q fever IgG phase I: positive >1:8192, IgG phase II: positive 1:4096 and phase I and II IgM negative. Doxyxycine and hydroxychloroquine were added to his regimen.

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PIN WORMS INFECTION PRESENTING AS CROHN'S DISEASE

Al-Saffar F1, Ibrahim S1, Clark M2, 1University of Florida, Jacksonville, FL and 2University of Florida, Jacksonville, FL.

Case Report: Case Narrative:

24 YO Caucasian female with PMH of Juvenile RA, Chronic pericarditis, PAH, cholecystitis s/p cholecystectomy presented with intolerable abdominal pain and bloating over the previous 10 days. She also endorsed nausea but no vomiting, fever, or diarrhea. She had similar episodes before and was told she had Crohn’s disease at another facility based on CT scan but never had a colonoscopy. The patient denied sick contacts or recent travel.

A CT abdomen showed wall thickening with submucosal fat at the terminal ileum which is nonspecific but can be seen with chronic inflammatory processes such as ulcerative colitis or Crohn’s disease. She underwent Colonoscopy that showed parasites were found in the transverse colon, the ascending colon and in the cecum. Fluid aspiration was performed through the transillumination channel. Sample(s) were sent for ova and parasites and was positive for Enterobius vermicularis (pinworms). Ilial biopsy was negative.

On subsequent questioning of the patient she endorsed living in a wooded area where it had been rainy with muddy grounds around her house. She also said she and her family usually consume private well water that occasionally dries out. This can be the route of infection (contaminated drinking water) given the lack of pertinent travel history or sick contacts.

The patient received one dose of Albendazole with subsequent improvement of her pain pain and was was discharged with a prescription for Albendazole to take in 2 weeks and instructions to have her children and partner treated.

Discussion: This is a case where a patient had a history of autoimmune disease, was not an immigrant/traveler from a helminths-endemic area, and had radiological features of Crohn’s disease. In this case endoscopy and histopathology yielded a diagnosis of colonic pinworm with the subsequently appropriate treatment of anti-helmintic medications.

We seek to delineate that even in an apparently typical patient inflammatory bowel disease should always be endoscopically-confirmed and biopsy-proven. While it was intuitive to proceed with the management of inflammatory bowel disease in our case, clinicians must keep in mind that colonoscopy is cornerstone in such cases to exclude other causes of the patient’s presentation.

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VESICULAR RASH IN A THREE-WEEK OLD

Baker RA1, Brockman SE2, Wainsscot CE1, Zayas J1, 1 UF College of Medicine Jacksonville, Jacksonville, FL and 2 UF College of Medicine, Jacksonville, FL.

Case Report: Vesicular rashes in the neonatal period can be the manifestation of several conditions ranging from benign to lethal. This case highlights the importance of considering a more common lethal cause of a vesicular rash, Herpes Simplex, while considering other less common and less morbid conditions including Varicella.

A 23 day-old male presented with a five day history of worsening rash that began as “red bumps” on his face (Images can be provided) The rash spread from his face and scalp, to his trunk, and eventually to his extremities. He was afebrile and not fussy. He was born at term via spontaneous vaginal delivery which was only remarkable for meconium-stained fluid. There were no perinatal infections or complications. The neonate was exclusively breastfed and the mother denied any lesions to her breasts. The mother denied any perinatal vesicular rash. The family attended church two weeks prior and were exposed to a missionary group from Africa. On physical exam, vitals signs were normal for age. He was alert and in no distress. The skin revealed crops of vesicles on an erythematous base spread over the face, scalp, trunk, extremities, palms and soles. The lesions were noted in varying stages of healing, ranging from tense vesicular lesions to those that had crusted over. Also, ulcerative lesions were noted on the mucous membranes inside of the mouth.

Our patient undertook a sepsis evaluation and intravenous acyclovir was initiated empirically. Given the vesicular rash specimens were sent for both HSV and VZV. Results confirmed a VZV infection and the presence of transferrered maternal antibodies. Our patient had a more severe course than expected and therefore was prescribed seven days of IV acyclovir followed
Conclusions: In the past decade, United States syphilis cases have increased, particularly in the South. Late and latent syphilis case rates in the New Orleans metropolitan area are 3.5 times the national average for metropolitan areas. Treponema pallidum infection manifests as a chancre, rash, or mucosal lesion in the primary or secondary stages. In late and latent syphilis infections, the triphate may have more severe cardiovascular and central nervous system manifestations. Herein we correlate this public health trend with postmortem findings at autopsy.

Methods Used: To evaluate syphilis infections in the New Orleans decedent population, final autopsy reports for cases occurring between January 2012 and June 2014 were reviewed for postmortem findings consistent with syphilitic infection (n = 495).

Summary of Results: Four cases of tertiary syphilis were identified by autopsy (0.8% cases). All were male with an average age of 60 years. Only one patient had a known positive human immunodeficiency virus status. Manifestations included cerebral vasculitis (n = 3), gummas (n = 2), tabs dorsalis (n = 2), encephalomalacia (n = 1), ependymitis (n = 1), and aortitis (n = 1).

Conclusions: A variety of postmortem pathologic findings were identified in four males with late syphilis. Given our autopsy service’s assistance with medical records for 83% of cases for infectious disease, the number of syphilis cases identified in our decedent population is higher than, but reflective of, increased local syphilis rates. Syphilis remains a major health problem in the American South.

LYMPH NODE LIMBO

Davis LA, Engel LS. LSU Health Sciences Center, New Orleans, LA.

Case Report: A 49 year old woman with a history of HIV (CD4 count 2778 cells/mm3, viral load 1142 copies) presented with complaints of waxing and waning abdominal pain which began six months prior to presentation. Associated symptoms included fatigue, anorexia, a 20 pound weight loss, abdominal fullness, and painless neck swelling. On presentation the patient was febrile and tachycardic. The examination was significant for diffuse bulky lymphadenopathy, tenderness to palpation in the left lower quadrant with a palpable spleen tip. Laboratory data revealed a WBC count elevated above baseline, a mild transaminitis, and hyperuricemia. The patient was empirically treated with vancomycin and piperacillin-tazobactam for sepsis and allopurinol for hyperuricemia, resulting in mild improvement. Previous work up for similar presentations included an axillary lymph node FNA with immunostains positive for paraffoical T cells, and atypical medium to large size B cell population concerning for malignancy. A subsequent axillary lymph node biopsy and bone marrow biopsy were inconclusive. The specimens had been sent to the NIH for a second opinion. Several days post-discharge, further pathological data from the NIH provided a final diagnosis of DLBCL. The patient began cycle one of R-EPOCH shortly thereafter and responded well to chemotherapy.

DISCUSSION: DLBCL is a well-known AIDS-defining malignancy and should be included in the differential for lymphadenopathy in the HIV population. Histology and immunohistochemistry from biopsy specimens, preferably from the lymph nodes, will demonstrate characteristic B cell markers. However, special stains may be required based on the degree of atypical histology.

ACUTE OSTEOMYELITIS DUE TO SALMONELLA HEIDELBERG IN AN IMMUNOCOMPETENT CHILD

Deel MW, Balan A, Ramji F. University of Oklahoma Health Science Center, Oklahoma City, OK.

Purpose of Study: In the past decade, United States syphilis cases have increased, particularly in the South. Late and latent syphilis case rates in the New Orleans metropolitan area are 3.5 times the national average for metropolitan areas. Treponema pallidum infection manifests as a chancre, rash, or mucosal lesion in the primary or secondary stages. In late and latent syphilis infections, the triphate may have more severe cardiovascular and central nervous system manifestations. Herein we correlate this public health trend with postmortem findings at autopsy.

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Conclusions: A variety of postmortem pathologic findings were identified in four males with late syphilis. Given our autopsy service’s assistance with medical records for 83% of cases for infectious disease, the number of syphilis cases identified in our decedent population is higher than, but reflective of, increased local syphilis rates. Syphilis remains a major health problem in the American South.
PSEUDOMONAS STUTZERI PACEMAKER POCKET INFECTION

Fenire M, Rudd D, Patel P. East Tennessee state university, Johnson City, TN.

Purpose of Study: 1- Report a case of Pseudomonas stutzeri (Psutzeri) pacemaker pocket (PP) infection.

2- P. stutzeri is a rare pathogen, however is associated with serious complications.

Methods Used: Case analysis and review of literature

Summary of Results: 76 year old female with history of pacemaker dependent complete heart block and diabetes mellitus presented with increased swelling and redness at her pacemaker site. 4 weeks prior, she had undergone left heart cath with stent placement; prasugrel was added to her medications. A hematoma formed around the PP and prasugrel was subsequently held; however, the associated swelling and erythema did not resolve over the next 4 weeks. Therefore, a PP revision was performed and a diagnosis of PP infection was made after cultures from the wound grew P. stutzeri. Rather than pacemaker removal, the patient elected for antibiotic therapy. Per culture sensitivities, a 2 week course of cefepime was administered with good success.

Discussion: P. stutzeri is a rare pathogen and was first described by Burri and Stutzer in 1895. It is a nonfluorescent, denitrifying, single polar flagellated gram negative rod. In a case report and a review paper, Ronald et al found 29 cases in English literature reported to cause infection between 1966 and 1998. In 23 of the 29 cases, P. stutzeri was isolated in pure cultures - similar to the case above. Freshly isolated colonies are adherent with a characteristic wrinkled appearance (panel 1), resembling craters with elevated ridges that often branch and merge. P. stutzeri infections have been reported in association with bacteremia, joint infection, osteomyelitis, endocarditis, endophthalmitis, meningitis, pneumonia, skin infection, urinary tract infection, and ventriculitis. It is also described in association with infection in synthetic vascular grafts. Goetz et al reported sporadic bacteremia in six hemodialysis patients over a nine month period with further investigation revealing isolation of P. stutzeri from the dialysate and the dialysis machine.

The incidence of more than one infection in the same institution should prompt an infection control work up. P. stutzeri is susceptible to many more antibiotics than P. aeruginosa, however, resistant strains have been isolated, especially in immunosuppressed patients.

Conclusions: P Stutzeri is important pathogen.

STREPTOCOCCUS GALLOLYTICUS NEONATAL SEPSIS AND MENINGITIS

Kavanagh K.1,2, Kennedy G.1,2, Kashinowo I.2,3.1, Cripe P.2,3,1, Steele R.2,3,1, Tuane U. SOM, New Orleans, LA; 1Ochsner Medical Center, New Orleans, LA and 1U. of Queensland SOM, New Orleans, LA.

Case Report: A 4 day old previously healthy term female neonate presented to the emergency department (ED) with lethargy, irritability, decreased urine output and poor feeding of 6 hours duration. ED examination revealed a temperature of 38.1°C and a sepsis workup was initiated. The CSF Gram stain showed Gram positive diplococci and were positive for group D streptococcus identified by RapID STR. Subtyping by 16S rDNA gene sequencing was performed, revealing pansensitive Streptococcus gallolyticus subspecies gallolyticus. The S. bovis/S. equinus complex has undergone a dramatic shift in nomenclature since 2002. The results of 16S rDNA sequencing and DNA-DNA hybridization suggest a more appropriate nomenclature identifying the clinically important pathogens from the S. bovis/S. equinus complex in infective endocarditis (IE), colorectal cancer (CRC) and neonatal meningitis as S. gallolyticus subsps. gallolyticus and S. gallolyticus subsps pasteurianus, which were formerly S. gallolyticus (biotype 1) and S. pasteurianus (biotype II.2), respectively. Retrospective studies on samples previously identified as S. bovis or Group D streptococcus non-enterococci showed a strong association between S. gallolyticus subsps. gallolyticus (biotype 1) with IE and CRC. Only two cases of neonatal meningitis due to S. gallolyticus subsps. pasteurianus were described previously. The vast majority of S. bovis neonatal meningitis cases reported in the literature where subtyping was available were due to S. gallolyticus subsps. pasteurianus (biotype II.2). The 3 neonates with S. gallolyticus subsps. gallolyticus meningitis survived, suggesting the prognosis for neonatal meningitis due to this organism may be better than that of Group B Streptococcus meningitis. The paucity of documented cases of neonatal meningitis caused by S. gallolyticus subsps. pasteurianus may be due to underreporting, as few institutions performed subtyping prior to 2003. Gene sequencing of non-specific S. bovis infections and documenting it in the literature is vital to understanding the antibiotic susceptibility, natural history, morbidity and mortality of neonatal meningitis due to S. gallolyticus subsps. gallolyticus.

ANEMIA IN AN HIV INFECTED PATIENT: PARVO THE CURSE?

Johnh DH, Engel LS. LSU Health Sciences Center, New Orleans, LA.

Case Report: A 29 year old trans-gendered patient with a history of HIV/AIDS (CD4 count 10 cells/mm3, viral load 227,431 copies/mL, not on anti-retroviral therapy) presented with 1-2 weeks of progressive dyspnea on exertion and lightheadedness. No history of orthopnea, chest pain, or palpitations. Physical exam revealed a temperature of 38.1°C, blood pressure was 80/56 and temperature was 100.9°F. She had pale conjunctivae, tachypnea, and tachycardia. Both lung fields were clear on auscultation. She had decreased breath sounds with rales in the left lower lobe. Cardiac exam was remarkable for tachycardia without murmur or gallop. She was admitted to the hospital and a complete blood count was performed which revealed a hemoglobin of 3.0 gm/dl with peripheral smear showing reduced red blood cells but at 1 month for follow up had persistent anemia with hemoglobin of 6. Intravenous immunoglobulin infusions were started at that time, and her hemoglobin has since been stable.

Discussion: Pure red cell aplasia (PRCA) is defined as anemia with reticulocytopenia and absence of normal red cell precursors in the bone marrow. An acute PRCA from direct viral infection, called transient aplastic crisis (TAC), can occur with parvovirus B19 infection. Though rare, Parvovirus B19 infection should be suspected in an immunocompromised host with persistent anemia requiring frequent blood transfusions.

HACEK ORGANISMS AND INFECTIVE ENDOCARDITIS

Rosselot JM, Mansfield JN, Hill CR, Tumarada N, Engel LS, Sanne SE. LSU Health Sciences Center, New Orleans, LA.

Case Report: INTRODUCTION: There are 10,000 to 15,000 new cases of infective endocarditis each year in the US. A rare cause of infective endocarditis is a group of gram-negative organisms, the HACEK group: Haemophilus species, Aggregatibacter (previously Actinobacillus) species, Cardiobacterium hominis, Eikenella corrodens, and Kingella species.

CASE: A 54 year-old woman with diabetes and chronic kidney disease presented to the Emergency Department with fever for four days. Her fevers were associated with chills, wheezes, and intermittent diarrhea. Her vital signs were unremarkable but physical exam revealed a III/VI holosystolic murmur and multiple dental caries. Her laboratory studies revealed a leukocyte count of 18,400 mm3. Blood cultures were positive for Aggregatibacter aphrophilus for which she was treated with Ceftiraxone. Transesophageal echocardiography showed vegetation on the posterior mitral valve leaflet and normal ejection fraction, which was confirmed by transesophageal echocardiography. She underwent a dental extraction to remove a possible source of infection. She improved clinically and was discharged home to complete a course of Ceftiraxone. Follow-up echocardiography demonstrated no further evidence of vegetation.

DISCUSSION: Although HACEK organisms are documented to cause up to 3% of infective endocarditis cases, 60% of individuals with HACEK...
bacteremia were found to have endocarditis as well. Diagnosis of HACEK bacteremia can be easily overlooked because they are fastidious organisms and difficult to isolate. Because of the difficulty with susceptibility testing and increasing beta-lactam resistance, the suggested treatment includes high dose ceftriaxone and ampicillin-sulbactam for 6 months when native valves are involved. Fluoroquinolones have susceptibility in vitro and should be used with patients intolerable to beta-lactams. Regimens are extended to 6 weeks in patients with prosthetic valves. A likely source of infection our patient was her poor dentition. Prognosis is dependent on multiple factors including maintenance of high clinical suspicion. Although HACEK infective endocarditis can result in a high mortality rate, most patients with correctly diagnosed uncomplicated infective endocarditis have an excellent prognosis with therapy.

DISSEMINATED BLASTOMYCOSIS PRESENTING WITH NONHEALING SKIN ULCERS
Mekala V1,2, Khan A1,2, Choudhary MC1,2, Washburn R2,1, Louisiana State University Health Sciences Center, Shreveport, LA and 2Overton Brooks Veterans Affairs Medical Center, Shreveport, LA.
Case Report: A previously healthy 36 year old male was referred to the infectious diseases (ID) clinic for evaluation of enlarging painless nonpruritic left lateral thigh ulcers of insidious onset and eight months’ duration. He denied fever, chills, and weight loss. During the months prior to lesion onset, he had been treated with antibiotics for right middle lobe pneumonia and prostatitis. The patient resided in northern Louisiana with his wife and son, worked as a truck driver for paper mills throughout the continental United States, and had no pets or other animal contacts. Six months prior to ID evaluation, his family physician performed punch biopsies of the ulcers, revealing fibrosis and focal foreign body reaction. Two months before ID evaluation, follow-up physical exam revealed a single coalescent circular pink ulcer (8 x 7 centimeters) having irregular raised crusted margins without pus, bleeding, or cellulitis. The ulcer was widely excised, and treated with wound vacuum therapy. Periodic acid Schiff staining revealed broad-based budding yeast, morphologically consistent with Blastomyces dermatitidis. Fungal culture confirmed blastomycosis, and the patient was referred to ID for management.

Physical exam was normal except for a tender large left lateral thigh ulcer containing beefy red granulation tissue. Chest roentgenogram showed shadowing of the right middle lobe. The patient was treated initially with oral itraconazole 200 mg twice daily, followed by 200 mg once daily with a plan to complete twelve months. After four months, his wound had completely healed.

Discussion: Blastomycosis is a reportable disease in Louisiana. Infection is sporadic in this state, ranging from one to seven cases annually during the past decade. In contrast to direct cutaneous inoculation, this case illustrates probable inhalational acquisition with asymptomatic chronic pulmonary infection followed by prostate and cutaneous dissemination. It also underscores the critical importance of performing appropriate histopathological examination and cultures for establishing the diagnosis.

AN UNEXPECTED GUEST
Phadke VK, Goswami ND. Emory University School of Medicine, Atlanta, GA.
Case Report: A 60-year-old man with systolic heart failure, emphysema, and hepatitis C infection presented with wound dehiscence and two days of purulent drainage from his implantable cardioverter-defibrillator (ICD) pocket site. Two months earlier during a hospitalization for decompensated heart failure he developed ventricular fibrillation leading to cardiac arrest; he recovered and underwent ICD placement. He had persistent edema over the left lateral thigh following implantation, but had not sought medical attention.

On admission blood cultures were collected and he was started empirically on vancomycin. Transthoracic echocardiogram revealed no valvular vegetations; transesophageal echocardiogram could not be performed due to prior intubation-related laryngeal stenosis. The ICD generator and leads were removed and sent for culture. Gram stain of the culture. Gram stain showed beaded gram positive rods. Within five days both blood and pocket exudate cultures were growing an aerobic gram positive rod which was subsequently reported as a rapidly-growing mycobacterium. His antibiotic regimen was empirically changed to clarithromycin, cefoxitin, and ciprofloxacin. He was discharged with a wearable cardioverter-defibrillator (I/C) to use until device reimplantation was felt to be safe. Unfortunately he was lost to follow-up, and two months later he self-discontinued his WCD and died of out-of-hospital cardiac arrest.

The organism was ultimately identified as Mycobacterium fortuitum, with the following susceptibilities: clarithromycin R (MIC 8µg/mL), cefoxitin I (MIC 0.64µg/mL), ciprofloxacin S (MIC 0.12µg/mL).

DISCUSSION: Non-tuberculous mycobacteria (NTM) are ubiquitous, environmental organisms that are being increasingly implicated as causes of surgical site and prosthetic device infections. A high index of suspicion is necessary for accurate diagnosis as they can be frequently misidentified as gram positive contaminants. Cardiac device infections due to NTM are rare, with only about ten cases reported in the literature. Most have been due to rapidly-growing species, with the most commonly reported organism being M. fortuitum. Associated bacteremia and macrolide-resistance - both aspects of this case - are uncommon features. Successful treatment typically involves device extraction accompanied by months of combination antibiotic therapy.

INVASIVE PLEUROPULMONARY INFECTION CAUSED BY CLOSTRIDIUM SORDELLI
Rafeek H1,2, Locati J1,2, Saanani N1,2, Penn R2,1, Louisiana State University Health Sciences Center, Shreveport, LA and 2Overton Brooks Veterans Affairs Medical Center, Shreveport, LA.
Case Report: A 45 year old male was admitted with progressive dyspnea on exertion, edema, and pleuritic left chest pain. Past medical history included dilated cardiomyopathy, ventricular fibrillation with cardiac defibrillator implantation, rheumatoid arthritis, and long term methotrexate therapy. Significant exam findings were normal vital signs, dullness over the lower left chest, and pitting edema. Computerized tomography (CT) showed gas-containing loculated left pleural effusion, collapse of the left lung base, and scattered infiltrates on the right. Lower extremity venous Dopplers were negative for deep vein thrombosis. Abnormal laboratory results included elevated white blood cell count (WBC, 11,600/L), and B-type natriuretic peptide (290 pg/mL). Congestive heart failure and probable pneumonia with a loculated pleural effusion were empirically treated with diuretics and levofloxacin. Thoracentesis revealed pH 7.31, glucose 94 mg/dL, protein 2.4 g/dL, albumin 1.3 g/dL, and LDH 358 IU/L. Gram stain showed rare white blood cells but no organisms. Pleural fluid cultures yielded only a few colonies of Clostridium sordelli. He improved with diuresis and levofloxacin, and was discharged 10 days after admission.

One month later he was readmitted with worsened cough and dyspnea. Between hospitalizations he had been treated with prednisone for Sweet’s syndrome. CT showed loculated left empyema with bilateral infiltrates, and right pulmonary artery thrombosis. He was anticoagulated and given intravenous vancomycin and piperacillin/tazobactam monotherapy, followed by oral amoxicillin/clavulanate.

Discussion: Recovery of C. sordelli from two empyema fluid samples obtained 39 days apart is strong evidence that it was a true pathogen. Although C. sordelli infection is a recognized complication of abortion, it has only rarely been reported as a cause of other infectious syndromes. This case underscores the potential role of C. sordelli in invasive pleuropulmonary infection.

ELIZABETHKINGIA MENOGENEOPTICUM AS A CAUSE OF CELLULITIS AND SEPTICEMIA
Rudd D, Fenire M, Patel P. East Tennessee State University, Johnson City, TN.
Purpose of Study: 1. Report a case of Elizabethkingia meningosepticum (E. meningosepticum) cellulitis and septicemia.
2. Highlight one of the emerging nosocomial infections and summarize some of the unusual characteristics of this bacterium.
Methods Used: Case report and review of literature.
Summary of Results: A 74 year old male nursing home resident with past medical history of diabetes mellitus type 2 and hypertension presented to the Southern Regional Meeting Journal of Investigative Medicine Volume 63, Number 2, February 2015
emergency department with complaints of lower extremity swelling which had progressively worsened over the preceding week. The patient was febrile and tachycardic, but otherwise hemodynamically stable. Physical exam revealed a toxic appearing male with diffusely dry, cracked skin of lower extremities and overlying erythema mildly tender to palpation - right greater than left (panel 1). Laboratory testing revealed leukocytosis. Lower extremity doppler study demonstrated no acute process. Pan cultures were obtained and empiric therapy with vancomycin and piperacillin-tazobactam (pip-tazo) was begun. Blood cultures revealed 2 out of 2 positive growth for E. meningosepticum (panel 2). Antibiotic therapy was de-escalated to monotherapy with pip-tazo per culture sensitivities. Dramatic improvement in clinical status and lower extremity cellulitis subsided. Repeat blood cultures were negative. The patient was discharged home to complete two weeks of intravenous therapy and to follow up in clinic.

Conclusions: E. meningosepticum is a gram negative saprophytic bacilli that was first reported by King in 1959. It is a ubiquitous organism which can survive in chlorine treated municipal water supplies, colonizing sink basins and taps; therefore, leading to colonization of hospital water systems making it a potential pathogen for nosocomial infections. It can cause a variety of infections including endocarditis, soft tissue infections, bacteremia, subdural abscesses, associated peritonitis, catheter associated infections, and endophthalmitis. Though infection with this bacteria is rare, its unique antibiotic profile can make it difficult to treat as it is usually resistant to a wide range of typical gram negative antibiotics; though, it may be susceptible to antibiotics geared toward meningitis prior to lumbar puncture. This patient’s altered mental status further worsened and he began to have respiratory distress that required intubation. Lumbar puncture demonstrated clear, colorless, CSF with 102 WBCs, 453 RBCs, 13% neutrophils, 35% Lymphocytes, 52% Mononuclear cells. CSF glucose was 196mg/dl, and Protein was 64mg/dl. There was no bacterial growth on blood or CSF cultures; however, he had positive IgM West Nile serum antibodies. CSF antibodies were negative and brain MRI obtained was unremarkable. After thirteen days of hospitalization, and no signs of recovery as well as absent to markedly blunted brainstem reflexes, the patient’s family decided to withdraw care.

DISSEMINATED SEPTIC ARTHRITIS IN AN IMMUNOCOMPETENT PATIENT FOLLOWING JOINT SURGERY
Singh MP, Prabha N, Islam A. Texas Tech University HSC, Amarillo, TX.

Case Report: Septic arthritis in an acute polyarthritis. Polyarticular septic arthritis is most likely to occur in patients with overwhelming bacteremia, sepsis, systemic CTD involving the joints. Our case involves a young man presenting with disseminated septic arthritis following joint surgery.

A 35 yr old male presented to the ER with progressively increasing pain,swelling and redness of wrists,shoulder joints and left knee joint. He had low grade fever and was not able to ambulate. He admits smoking amphetamine,denies any IV Drug Abuse.PE: A&D, Moderate distress due to pain,Vitals:WNL,B/L Lungs: Clear. HEENT: Positive sinusitis. Abdomen: Abdominal distension,swelling and erythematous. No associated peritonitis, catheter associated infections, and endophthalmitis. Though infection with this bacteria is rare, its unique antibiotic profile can make it difficult to treat as it is usually resistant to a wide range of typical gram negative antibiotics; though, it may be susceptible to antibiotics geared toward meningitis prior to lumbar puncture. This patient’s altered mental status further worsened and he began to have respiratory distress that required intubation. Lumbar puncture demonstrated clear, colorless, CSF with 102 WBCs, 453 RBCs, 13% neutrophils, 35% Lymphocytes, 52% Mononuclear cells. CSF glucose was 196mg/dl, and Protein was 64mg/dl. There was no bacterial growth on blood or CSF cultures; however, he had positive IgM West Nile serum antibodies. CSF antibodies were negative and brain MRI obtained was unremarkable. After thirteen days of hospitalization, and no signs of recovery as well as absent to markedly blunted brainstem reflexes, the patient’s family decided to withdraw care.

Discussion: Generally, West Nile Virus infections are asymptomatic. Symptoms such as fever, headache and malaise are seen in about 20-40 percent of infected individuals. However, West Nile Virus can present with neuroinvasive disease. These patients present with fever, meningitis or encephalitis. Mortality rate for neuroinvasive West Nile Virus is 10 percent. Our patient had many of the known risk factors for mortality which includes diabetes, cardiovascular disease, muscle weakness, and changes in level of consciousness.

HIV-1 ENVELOPE EPITOPES RECOGNITION IS INFLUENCED BY IMMUNOGLOBULIN DH GENE SEGMENT REPERTIORE
Wang Y1, Schroeder H2,1. 1University of Alabama at Birmingham, Birmingham, AL and 2University of Alabama at Birmingham, Birmingham, AL.

Purpose of Study: Elucidation of mechanisms that underlie the difficulty in generating HIV-1 can be seen as a part of fundamental need to better understand how to distinguish self and non-self receptors that can neutralize pathogenic antigens without self-inflicting dame to the host.

Methods Used: Mice-BALB/c mice cohort limited to the use of single DH gene segments were generated. ΔD-DFL is a human-like repertoire control. ΔD-DLF promotes the use of hydrophobic amino acid CDR-H3. The ΔD-DLF is charged amino acid enriched CDR-H3. Immunization-each strain of 10 mice was immunized with HIV-1 JR-FL gp140 protein. Epitope Identification-Mice in each cohorts were selected (prior to immunization, and after the 2nd and 4th immuniizations) for PEPPERPRINT Chip to detect their epitopes on HIV-1 JR-FL gp140. Serum Assay-Binding ability was examined by HIV-1 envelope protein ELISA.

Summary of Results: We obtained evidence of strong and clear polyclonal responses to immunization with JR-FL gp140. As a general rule, the
heterogeneity of the anti-JR-FL gp140 response varied by DH genotype with ΔD-DFL-Wild Type (WT) $\rightarrow$ ΔD-DuFS, ΔD-Δ ID. Conversely, the intensity of the response was greatest in the ΔD-Δ ID.

2. Linear response showed by PEPperPRINT chip was not identified with the response to natural HIV-1 epitopes, however the ΔD-Δ ID favored charged epitopes that is inconsistent with our hypothesis.

**Conclusions:** The pattern of epitope recognition and antigen binding in the response to HIV-1 JR-FL gp140, in part, is on DH gene segment sequence.

A RARE PRESENTATION OF DISSEMINATED HISTOPLASMA MIMICKING METASTATIC CANCER IN APPARENTLY IMMUNOCOMPETENT INDIVIDUAL

Wondimagegnehu NE, Ruthirago D, Tantrachoti P, Nugent K. Tesh tech University health service center; Lubbock, TX.

**Purpose of Study:** Infection with Histoplasma capsulatum occurs frequently in endemic areas. These infections can cause pulmonary disease in immunocompromised adults, especially if they have chronic lung disease. However, progressive disseminated histoplasmosis in these patients is extremely rare finding. We report an immunocompetent patient who had never been in endemic area but developed disseminated histoplasmosis with multiple brain masses, pulmonary nodules, adrenal nodules, and spine involvement.

**Methods Used:** Case analysis and literature review.

**Summary of Results:** We report a 47-year-old man who was admitted for progressive headache and vomiting. He has had unintentional weight loss, low-grade fever, and night sweats. He has lived all his life in Lubbock, Texas. Physical examination was remarkable for left cerebellar signs, including dysmetria, dysdiadochokinesia, and positive Romberg's sign. MRI of the head showed 5 x 3 x 3 cm rim-enhancing mass in the left cerebellar hemisphere and a 0.6 cm right frontal lobe enhancing mass. Cancer with brain metastases was highly suspected at that time. In an evaluation for a possible primary site, given his history of chronic smoking, CT scan of the chest and abdomen showed bilateral non-calciﬁed pulmonary nodules, multifocal small radiolucent foci in the thoracic and lumbar vertebral, and hypodense nodules in the left kidney and the left adrenal gland. The patient underwent craniotomy with resection of cerebellar mass. Grocott-Gomori's methamine silver stain (GMS) stained slide showed Histoplasma organisms. Work up, including HIV serology and lymphocyte counts, did not reveal any evidence of immunosuppression. Patient was started on voriconazole after resection of the mass.

**Conclusions:** This is the one of the few cases of disseminated histoplasmosis with widespread organ involvement in an apparently immunocompetent person. Multiple brain masses with signs of increased intracranial pressure together with lung, bone, kidney, and adrenal gland involvement represent a rare presentation for this fungal infection. A high degree of clinical suspicion is essential in both immunocompromised and immunocompetent patients, regardless of pulmonary symptoms, and whether in endemic or non-endemic areas.

**Nutrition**

Joint Plenary Poster Session and Reception

5:00 PM Thursday, February 26, 2015

ATTITUDES, BEHAVIORS AND ENVIRONMENTAL PREDICTORS FOR MOTHERS EVER HAVING BREAST FED THEIR INFANTS: A QUESTIONNAIRE STUDY IN A MEDICAID CLINIC POPULATION

Dury A, Burns JJ. FSU, Pensacola, FL.

**Purpose of Study:** To determine factors attributed to mothers having ever breast fed their infant in a predominantly Medicaid clinic population and to measure the age when breast feeding was discontinued.

**Methods Used:** A 24 item questionnaire was administered to mothers of infants presenting to the clinic who were under 12 months old. The relationship between ever having breast fed their current baby to attributes were determined using Chi-Square analysis; multivariate analysis was performed using logistic regression.

**Summary of Results:** Sixty-four percent of mothers ever breast fed their current baby. Percentage of age range groups when mothers discontinued breast feeding were 23% between 0 and 14 days of life, 34% between 15 and 30 days of life, 21% between 31-60 days old and 21% between 61 and 180 days of age.

On multivariate logistic regression controlling for maternal age, race and socioeconomic status, statistically significant relationships were found between ever having breast fed their current baby and planning to breast feed when pregnant (p < 0.001, Odds Ratio 4.73), having previously breast fed a baby (p = 0.018, Odds ratio 4.4) and friends/family supporting breast feeding (p = 0.016, Odds ratio 3.7).

**Conclusions:** Intentions to breast feed while pregnant, previous breast feeding experience, and family/friends who support breast feeding were found to be related to mothers having ever breast fed their baby. This may provide important directions for interventions designed to increase breast feeding rates.

NEW NATIONAL INSTITUTES OF HEALTH DIETARY SUPPLEMENT LABEL DATABASE CAN AID SEARCH FOR INGREDIENT INFORMATION

Costello RB,1 Saldanha L,1 Dwyer J,1 Andrews K2, Baille R,1 Bailey R,1 Betz J,1 Burt V,1 Chang P,1 Emenaker N,3 Gaichje J,2 Hardy C,1 Pehrsom P,1 1National Institutes of Health, Bethesda, MD; 2United States Department of Agriculture, Beltsville, MD; 3National Institutes of Health, Bethesda, MD.

**Purpose of Study:** Over 50,000 different dietary supplement (DS) products are offered for sale in the U.S. increasing the probability of supplement-drug interactions. We sought to develop a public use, searchable database containing label information from virtually all DS products offered for sale in the U.S., with a Web-based user interface providing ready access to the data.

**Methods Used:** An ad hoc Federal Working Group provides guidance to a contractor on the collection, classification, and handling of information from DS labels.

**Summary of Results:** The database information reflects what is printed on product labels and is publicly available through the National Library of Medicine's Dietary Supplement Label Database (DSLD) (http://www.dsld.nlm.nih.gov). The name and form of ingredients, amount(s) of ingredient(s), information about the manufacturer, label claims, warning statements, and percent of daily value are listed along with an image of the label. Simple and advanced search options are available enabling users to locate specific products or specific terms in any DS label field. For example, searching for a supplement by label name identified 53 vitamin K products containing 1.7 to 4.050 mcg corresponding to 2 to 5,063% of the daily value for recommended intake of vitamin K. The retrieved search allows the user to sort by amount of vitamin K in a product, or eliminate products that may contain vitamin K even though they are not labeled as such. Additionally, the advanced search function allows users to search and download information about supplement products collected from the National Health and Nutrition Examination Survey (NHANES).

**Conclusions:** Clinicians, researchers and consumers may find DSLD useful for identifying DS ingredients of potential benefit as well as those of concern.

CHANGES IN MICRONUTRIENT STATUS DURING AND AFTER PULMONARY EXACERBATION IN ADULT CYSTIC FIBROSIS

Lee M, Smith EM, Ziegler TR, Alvarez J, Tangpricha V. Emory University, Atlanta, GA.

**Purpose of Study:** Malnutrition in patients with cystic fibrosis (CF) occurs secondary to periods of inadequate nutrient intake in relation to needs, pancreatic insufficiency-induced malabsorption, and/or catalytic effects of inflammation and is associated with poor clinical outcomes. Patients with CF hospitalized for a pulmonary exacerbation may be at increased risk for nutrient depletion. The purpose of this study was to investigate changes in micronutrient status that occur during and after a pulmonary exacerbation of CF.

**Methods Used:** This was a prospective study of a multicenter study investigating the role of vitamin D supplementation in CF. We measured plasma micronutrient concentrations in subjects during a pulmonary exacerbation at
baseline and 3 months later. Plasma trace element and mineral concentrations were measured using inductively-coupled plasma-atomic emission spectrometry in triplicate to measure calcium (Ca), copper (Cu), iron (Fe), potassium (K), magnesium (Mg), sodium (Na), and sulfur (S) with Seronorm quality controls. Statistical analyses included paired t-tests.

**Summary of Results**: Subjects were 24 hospitalized adults with CF (mean age 28.4 ± 7.4 yr). At baseline, CF subjects exhibited significantly lower mean plasma concentrations of calcium (9.8 ± 1.0 vs. 10.8 ± 1.9 mg/dL, p<0.05), copper (1.4 ± 0.5 vs. 1.6 ± 0.6 μg/mL, p<0.05), and iron (0.8 ± 0.5 vs. 1.3 ± 1.1 μg/mL, p<0.05) compared with 3 months following their exacerbation. CF subjects exhibited similar plasma concentrations of potassium at baseline compared to 3 months later (baseline, 4.9 ± 0.3 vs. 3.5 ± 1.5 mEq/L), magnesium (1.9 ± 0.2 vs. 2.0 ± 0.3 mg/dL), and sulfur (1297.9 ± 325 vs. 1233.8 ± 290 μg/mL) during and after an exacerbation (p>0.05 for all).

**Conclusions**: Adults with CF exhibit significantly altered plasma levels of several micronutrients during an acute pulmonary exacerbation in comparison to 3 months later. Plasma concentrations of calcium, copper and iron increased significantly over time, which may reflect decreased inflammation, improved food intake and increased absorption following recovery. Further investigation into micronutrient status during an exacerbation is warranted to address nutritional challenges during illness in CF.

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**SIMPLE CLINICAL TOOLS TO SUPPLEMENT DIETARY WEIGHT LOSS COUNSELING**

Nantsupawat N, Lipman M, Limas N, Buscemi D, Nugent K. Texas Tech Health Sciences Center, Lubbock, TX, Lubbock, TX.

**Purpose of Study**: Standard approaches to weight loss counseling include dietary education, diet modification and calorie restriction, and increased physical activity. The dietitian typically monitors weight and body mass index (BMI). Additional simple clinical tools could help dietitians and patients understand their health status better with little or no additional time required.

**Methods Used**: We measured blood pressure, gait speed based on the 100 foot walk test, and the Get Up and Go test in subjects in a randomized controlled trial for weight loss testing the effect of sleep counseling on weight loss. In addition, the subjects completed sleep logs and goal calendars.

**Summary of Results**: Forty women entered this randomized controlled trial, and 27 completed a 6 month follow-up. The mean age was 43 ± 10.4 years, the mean BMI was 33.6 ± 4.3 kg/m², and 85% worked full-time. The mean change in BMI was -0.78 ± 1.20 kg/m². The mean initial systolic blood pressure was 117 ± 15 mmHg; the mean initial diastolic blood pressure was 74 ± 7 mmHg. The mean gait speed was 2.56 ± 0.21 m/s; the mean initial Get Up and Go time was 7.93 ± 1.1 seconds. The mean sleep time from sleep logs was 7.36 ± 1.20 hours with a range of 3.4–9.2 hours. 12.5% of the subjects slept for 5–6 hours, and 33.3% of subjects slept for ≥8 hours per night. The overall compliance with maintaining the goal calendars was 60.4%. These calendars indicated that subjects were more compliant with sleep goals than diet or exercise goals.

**Conclusions**: These tools provided quick and reproducible assessments of health (blood pressure), physical performance (gait speed and Get Up and Go test), habits (sleep logs), and compliance (goal calendars). This information provides subjects with an easily understood overview of their basic health and provides dietitians with additional outcome measurements and estimates of compliance based on completion of goal calendars. For example, our subjects had normal fitness levels, but many had abnormal sleep habits based on sleep time records which might improve with counseling.

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**ASSOCIATION OF NEONATAL BACTERIOIDES SP. MENINGITIS AND MATERNAL ULCERATIVE COLITIS**

Abdelmagid K, Estrada B, Weight Sexton LA. University of South Alabama, Mobile, AL.

**Case Report**: Bacteroides sp. are Gram-negative, obligate anaerobic, non-spore forming bacilli. They cause opportunistic infections in immunocompromised patients including CNS, respiratory, abdomen and urinary tract infections. They are fastidious and difficult to isolate, making the infections even more difficult to diagnose. In this report we present an unusual clinical presentation of bacteremia and meningitis in a neonate.

An 11 day old male presented to his pediatrician's office with a 12 hour history of fever, refusal to breastfeed and increased sleeping. Due to his clinical picture and concerns for neonatal sepsis, he was admitted for a full sepsis work-up and intravenous antibiotics. He was ill appearing, mildly hypotonic and irritable. The rest of his physical exam was normal. Initial work-up showed CBC with differential showed WBCs within normal limits. C-reactive protein was 11.7 mg/dl and electrolytes were normal. Blood, urine and cerebrospinal fluid (CSF) were sent for culture. Empiric therapy with ampicillin and cefotaxime was initiated. CSF analysis showed no nucleated cells, protein 40 mg/dl and glucose 55 mg/dl. Blood and urine culture yielded negative results and the patient showed clinical improvement, with return of his activity and feeding to baseline. On further history, it was noted that the patient's mother had been admitted for endometritis, she has history of poorly controlled ulcerative colitis. On day five of admission, beta-lactamase negative Bacteroides species were isolated from CSF culture and therapy with metronidazole was initiated. Cranial ultrasound and CT scan of the brain with and without contrast were normal. A repeat lumbar puncture was done and the CSF culture was sterile. Our patient was discharged after completion a total of 21 days of effective antibiotic therapy against Bacteroides sp. Subsequent follow-up were consistent with normal neurological exam and neurodevelopment.

Anaerobic meningitis in the neonatal period is a very rare entity; with more cases being reported in premature neonates or immunocompromised children. We hypothesize that the development of poorly controlled ulcerative colitis in our patient's mother could have been a predisposing factor for maternal Bacteroides sp. Bacteremia and subsequent transfusional transmission.
PANCREATIC AGENESIS: AN UNCOMMON NEONATAL ANOMALY

Beaulieu CC, Kandasamy J, Black A, Philips J. University of Alabama at Birmingham, Birmingham, AL.

Case Report: Neonatal diabetes mellitus (NDM) is a rare disease with an incidence of 1 in 450,000. Complete pancreatic agenesis, an etiology for NDM, is an extremely rare pathology that results in both endocrine and exocrine pancreatic dysfunction. We report the case of a small for gestational age female infant who presented with persistent hyperglycemia from birth. She also had undetectable plasma C-peptide and fecal pancreatic elastase levels. Imaging studies, first with an ultrasound and later a CT scan of the abdomen, provided preliminary evidence for pancreatic agenesis. Other anomalies that were identified by subsequent evaluations included multiple small ventricular septal defects, a patent ductus arteriosus, and mild pulmonary stenosis. Medical management included continuous insulin infusions initially, followed by intermittent therapy with the aid of a subcutaneous insulin pump. Management with formula rich in medium chain triglycerides, proton pump inhibitors to prevent intestinal acid irritation considering lack of pancreatic bicarbonate production, and pancreatic enzyme replacement as appropriate. Evaluation of the abdomen with plain radiographs after she had repeated episodes of bilious emesis showed intestinal malrotation for which surgical correction was done with a Ladds' procedure. At this time we were able to visually confirm the complete absence of the pancreas as well as the gall bladder. The genetics of pancreatic agenesis is still an area of active research. For this patient, we addressed two significant genes, PDX1, which is associated with both pancreatic and biliary agenesis, and GATA6, which is associated with pancreatic agenesis and congenital heart disease. In the past several years, we have had two other infants with pancreatic agenesis that are currently followed in our clinics. This report discusses the etiology, genetics, associated anomalies, evaluation and management of neonates born with hyperglycemia caused by pancreatic agenesis.

KERION CELSI OF THE MONS PUBIS

Becker LK, Balan A, DeLeon S. University of Oklahoma Health Sciences Center, Oklahoma City, OK.

Case Report: A kerion is a severe fungal infection of the hair follicle associated with a severe inflammatory reaction, often with bacterial superinfection. We present an adolescent with an unusual case of kerion of the mons pubis most likely acquired due to the practice of carrying a cell phone in the suprapubic area. The patient initially presented to her pediatrician with an erythematous rash involving the mons pubis with scattered papules. She was diagnosed with ringworm and started on a topical antifungal. As the infection continued to worsen, clindamycin and TMP/SMX were added due to concern for cellulitis. However, her symptoms progressed to include fever, anorexia, nausea and difficulty walking due to painful inguinal lymphadenopathy.

On admission, she was started on vancomycin due to concern for MRSA cellulitis resistant to both clindamycin and TMP/SMX. Pelvic ultrasound showed reactive inguinal lymph nodes, but no abscess formation or fluid collection. Blood, urine and vaginal cultures showed no growth. Gonorrhea/Chlamydia and HIV were negative. Infectious disease was consulted and the lesion represented a kerion. Although kerions may be found on the scalp and beard area, kerions of the pubic area are rare with few reports in the literature. Griseofulvin and clindamycin were started to provide anti fungal and antibacterial coverage pending final culture results. Systemic steroids were added given reports of greater symptomatic relief, including reduction in pruritus and scaling, when combined with antifungal therapy. The patient was discharged to home to complete a 6 week course of griseofulvin as well as 10 day course of vancomycin and 7 day course of prednisone. Following discharge, wound cultures showed growth of Trichophyton sp. sensitive to fluconazole (2 mcg/ml), griseofulvin (0.06 mcg/ml) and voriconazole (≤0.03 mcg/ml). At follow up, patient reported continued improvement in kerion, with resolution of pain and adenopathy.

The patient was continued on griseofulvin for a total of 2 months of therapy. Upon further inquiry the patient reported she had presented a cow at a cattle show with leg lesions quite similar to her lesions. She also admitted she would wear shorts and place her cell phone in the suprapubic area, making this the most likely mechanism of how she contracted the infection.

WRESTLING WITH A RASH: A CASE OF STAPH SCALDED SKIN SYNDROME

Bradley J, Hudson C, Keene M. ETSU Quillen College of Medicine, Johnson City, TN.

Case Report: A 4 year old male presented from an out side facility for rash and irritability. One week prior to admission, the patient was diagnosed with impetigo of his cheek and forehead, which was treated topically. His rash continued to worsen and he became increasingly irritable, with decreased urine output and lethargy. Social history revealed the patient was a wrestler and his 6 year old brother, his wrestling partner, had been diagnosed with impetigo and treated with topical and systemic antibiotics. Physical exam upon arrival was significant for an ill-appearing male with a diffuse erythroderma. He was in significant pain and vital signs revealed tachycardia and hypertension. Skin exam revealed large bullous lesions on his forehead and erythematous skin lesions in the neck, axilla, and groin areas. The erythematous areas sloughed when
touched, indicating a positive Nikolsky's sign. He was diagnosed clinically with Staph Scalded Skin Syndrome (SSSS) and placed on antibiotic therapy of Vancomycin and Clindamycin along with aggressive fluid resuscitation and wound care. Within hours of admission, the rash and skin sloughing had spread and involved over 50% of the body surface area. He was transferred to a burn center for further treatment and wound care. He made a full recovery.

SSSS includes a range of diseases from localized to systemic symptoms. SSSS is caused by strains of S. Aureus that produce epidermolytic exotoxins A or B. Skin manifestations range from bullous impetigo to diffuse erythroderma leading to sloughing with gentle force, known as Nikolsky's sign. The disease is most common in infants and young children. Treatment is aimed at antibiotic coverage along with wound care. Clindamycin is usually added to the systemic treatment to inhibit the production of the exotoxins. Excessive skin involvement with desquamation needs to be treated similar to a burn and may require treatment at a burn center. Diagnosis is usually made clinically, however a biopsy of a leading edge will reveal a subcorneal, granular layer split and absence of any inflammatory infiltrate. Amateur wrestling runs the risk of many known skin conditions such as herpes gladiatorum, tinea corporis, MRSA, and impetigo. One must not forget about staph scalded skin syndrome when a seemingly simple skin condition worsens quickly.

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**FATAL LATE-ONSET UREA CYCLE DEFECT IN A TEENAGER**

Brown AN, Winkler MK. University of Alabama at Birmingham, Birmingham, AL.

**Case Report:** 13yo previously healthy male presents with altered mental status and seizure activity after complaining of abdominal pain and malaise for two days with no fever or other symptoms. Workup at the referring hospital revealed positive rapid flu and normal CBC, electrolytes, LFTs, head CT, and negative UDS. On arrival to our facility he was intubated for a GCS=7 (E1V2M4) and had generalized hypotonia, rowing eye movements with reactive pupils, with the remainder of the exam normal. Initial management included a lumbar puncture (normal) and empiric treatment with antibiotics.

Day 3 of admission (GCS 7T) he developed sceleral icterus, fever, and significant hypertension. Labs revealed mild transaminists and an ammonia of 300 with cerebral edema on head CT. Due to concern for a late-onset inborn error of metabolism, he was protein-restricted but given glucose infusion and supplemented with ammonia scavengers. Ammonia normalized with these therapies. Protein was slowly reintroduced with close monitoring of ammonia levels which remained normal. During this time, his exam slowly improved to include some spontaneous eye opening as well as withdrawal from painful stimuli.

One week after reaching maximum protein allowance, he developed significant autonomic instability requiring fluid resuscitation and vasopressors, temperature elevation to 109°F, and decline of his neuro exam (GCS 3T). He was found to be severely hyperammonemnic (>1000) with subsequent liver failure (AST 1900, ALT 6200). His hyperammonemic crisis was managed with emergent dialysis, ammonia scavenger therapy, protein restriction, appropriate provision of glucose, and neuroprotective strategies. After three days of maximal therapy he was pronounced brain dead. Definitive diagnosis was eventually made with ornithine transcarbamylase (OTC) gene analysis resulting in a hemizygous mutation (p.R40H). This mutation has been reported in patients with late-onset hyperammonemnia due to partial deficiency of OTC. Males and heterozygous females with partial OTC deficiency can present at any age from infancy to adulthood with hyperammonemic crises regardless of the degree of enzyme activity present.

Dipylidium caninum is a cestode that commonly infects dogs and cats. Humans may be infected by ingestion of the intermediate host, an infected flea or louse. If the person ingests cysticercoids, an adult tapeworm can develop in the intestine. Small proglottids are usually mobile and visible in the stool, resembling a grain of rice. The treatment of choice is single dose praziquantel despite only being indicated for children greater than 4 years old.

History is very important in the evaluation of worm infections in infants. Upon further review in this case, it was determined that the patient’s grandmother had a flea-infected dog which was most likely the source of infection. Most helminth infections occur in persons 2 years or older, but one third of D. caninum infections occur in children less than 6 months of age. The exact incidence of cases in the US is unknown. The infection is typically asymptomatic, but symptoms may mimic that of other tapeworm infections including abdominal pain, diarrhea, and pruritus ani. D. caninum infections can be mistaken for pinworms given the similar presentation and is frequently misdiagnosed. This case exemplifies the importance of history and microscopic evaluation of helminths for appropriate diagnosis and management of neonatal worm infections.

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**PROFUND ANEMIA DUE TO NEW DIAGNOSIS OF ADOLESCENT CROHN’S DISEASE**

Burgess K, Cockrum E. University of Alabama, College of Community Health Sciences, Tuscaloosa, AL.

**Case Report:** An adolescent patient presented with 2 weeks of fatigue, weakness, shortness of breath and an episode of mild chest pain with minimal exertion. Patient was previously healthy and active. Patient also complained of intermittent sore throat, subjective fever and nocturnal sweating during this time. Abdominal pain, nausea, vomiting, diarrhea, dysuria, dysphagia, respiratory distress, hematuria, melena, joint pains, skin rashes, oral ulcers, head- aches, vision changes, jaundice, and weight loss were denied. Patient typically had one yellow-brown, soft stool a day, though recalled seeing bright red blood with stool for a few days about a year ago.

Patient has always been a small, thin child with a poor, selective appetite, but does not eat any unusual items. Patient consumes chicken and green leafy vegetables but no red meat. There were no medical records of any past lab work. No history of blood loss. Patient traveled outside the US a year ago but denied any illness. Patient lives in a rural area in a home less than 10 years old. Two cousins are known to have anemia. On exam patient was a happy, thin child with pulse of 139, BP 108/70 and RR 18. Weight was 84 pounds (7th %ile) and Ht 155 cms (22nd %ile). Exam was completely normal except for marked pallor. On exam a patient was happy, thin child with pulse of 139, BP 108/70 and RR 18. Weight was 84 pounds (7th %ile) and Ht 155 cms (22nd %ile). Exam was completely normal except for marked pallor.

Lab work was remarkable for Hgb of 2.6 Hct 9.3 MCV 48.5 RDW 23.4 Corrected Retic 0.8 WBC 11.9 with normal differential Plt 427 renal and liver panels were normal LDH 138 ESR 25 Iron 6 Ferritin <1. This presentation was unique due to the profound anemia which had not significantly impacted patient’s activities. The cause of iron deficiency must be identified in a child over 2 years old with iron deficiency anemia. Chronic GI blood loss is the most common cause. Obtained stool was hemoccult positive. Differential diagnosis of GI bleeding in this age group includes: infectious colitis, juvenile polyps, inflammatory bowel diseases, HSP, Meckel’s, anal fissures, vascular malformations as well as rare tumors.

The child received PRBC transfusions to assure hemodynamic stability and saw pediatric GI service for EGD and colonoscopy. Findings were consistent with Crohn’s Disease. Confirmatory studies are pending. Patient was not responsive to steroid therapy and is subsequently being treated with infiximab with planned follow up over next months.

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**A PEDIATRIC EMERGENCY ROOM PRESENTATION OF RETINOBLASTOMA**

Butchee RD1, Bogie A2, Siatkowski M3. The University Of Oklahoma Health Science Center, Oklahoma City, OK; The Children’s Hospital at OU Medical Center, Oklahoma City, OK and 3Dean McGee Eye Institute, Oklahoma City, OK.

**Case Report:** Objectives: After presenting a case of retinoblastoma, we will review the pathophysiology, epidemiology, genetics and management of this disease. The techniques that pediatricians and Emergency Care providers can utilize to assess for this disease are invaluable.

**Objectives:** After presenting a case of retinoblastoma, we will review the pathophysiology, epidemiology, genetics and management of this disease. The techniques that pediatricians and Emergency Care providers can utilize to assess for this disease are invaluable.
Background: Retinoblastoma is the most common intraocular tumor in childhood. It affects nearly 4.1 per million children each year. Most bilateral cases are diagnosed in the first year of life, likely due to the genetic component of the disease and a high index of suspicion. However, unilateral cases have a median age of presentation during early childhood, emphasizing the need for a comprehensive eye exam in all patients presenting for opthalmological problems and during well child checks. The most described genetic abnormality is a mutation at 13q14 gene, involving the RB1 protein, which normally functions as a tumor suppressor. The presentation and management differ depending on the genetics of the condition.

Case: A 10 month-old female presented to the pediatric emergency room with a chief complaint of "funny eye movements". On eye exam, leukocoria, a poor red reflex, and exotropia were noted. CT of the head and orbit revealed a calcified mass within the globe concerning for retinoblastoma, which was verified by MRI.

Discussion: It has been estimated that there are 25 million Emergency Room visits by children each year. Families appear to be utilizing Urgent Care clinics and the ER for routine and non-emergent conditions. This high volume of patients further emphasizes the need for review of basic eye exam techniques by non-opthalmological specialists. Evaluation of the red reflex, ocular alignment, and eye motility are valued even in the setting of a busy emergency department.

Table 1. Major and minor criteria for diagnosis of Sweet Syndrome

Major - Abrupt onset of tender or painful erythematous or violaceous plaques or nodules; Predominantly neutrophilic infiltration of the dermis without inkoiciclastic vasculitis

Minor - Illness preceded by fever or infection; Fever accompanied by arthralgia, conjunctivitis, or underlying malignancy; Leukocytosis >10,000/mm³; Good response to systemic corticosteroids and lack of response to antibiotics

FIGURE 1. Neutrophilic infiltration into the dermis in Sweet Syndrome.

FIGURE 1. Example of Sweet Syndrome rash.
Our patient is a 5 year old male with history of cerebral palsy admitted with fever and neck swelling two days after an uncomplicated bilateral excisor extraction. He was noted to be ill appearing with stridor on exam. An emergent CT scan showed RPA with extension into the mediastinum. ORL emergently drained the abscess, and he was started on ampicillin/subactam and vancomycin. Blood and abscess cultures grew GAS. Our patient initially clinically improved, was extubated and sent to the floor on monotherapy. However, fever reoccurred and labs became indicative of a worsening infection; he subsequently required two further I&Ds with drain placements and chest tube placement for a right sided pleural effusion. Although his bacteremia quickly cleared, he was later found to have PA growing from the surgical site central line. He was started on vancomycin and piperacillin tazobactam during this time but developed urinary tract infections; antibiotics were then changed to clindamycin and ceftazidime. Dentistry ruled out an ongoing odontogenic source for the ongoing infection, and our patient eventually improved after final I&D and central line removal. An additional complicating factor was the development of a rapidly spreading fungal dermatitis, cultured to be CA; he was initially started on intravenous fluconazole due to the rapid progression but was later changed to oral antifungal therapy and completed a ten day course. Once discharged, he completed bacteriemia and soft tissue infection treatment with oral clindamycin and ceftazidime until inflammatory markers normalized.

Uncomplicated RPA has a good prognosis with rare long-term consequences. However when complications do occur, they may be life threatening. As demonstrated in this case, patients may have multiple complications requiring an extended hospital course, multiple surgical interventions and medication changes, and input from several sub-specialties.

SEVERE HYPERTENSION AND PROTEINURIA IN A CHILD WITH DYSMORPHIC FEATURES

Crespo-Salgado J1,2, Aviles D1,2, Zambrano R1, LSUHSC, New Orleans, LA; Children’s Hospital, New Orleans, LA; LSUHSC, New Orleans, LA.

Case Report: Proteinuria and edema are the principal clinical manifestations of the nephrotic syndrome. Minimal change disease is the leading cause in children’s, but other etiologies must be considered.

We report of a 4 year old African American boy who was referred due to proteinuria. Pregnancy was uneventful but there is consanguinity. He was found to have a blood pressure of 174/111 and was admitted to the hospital for further management. Laboratory workup showed an elevated BUN and creatinine of 3.1 mg/dL. CBC showed vacuolization of lymphocytes. Complements levels were normal. Urine was notable for protein to creatinine ratio of 8.5 mg/mg.

On physical exam, his weight and height were between 10-25th percentile. He had macrocephaly, ocular hypertelorism and coarse facies. Ears were posteriorly rotated with a right pre-auricular tag. Abdomen was distended with hepatosplenomegaly. He had an uncircumcised penis with hypospadias. Muscular tone was normal and there was no pitting edema.

Renal Doppler ultrasound showed normal size echogenic kidneys without evidence of renal artery stenosis. Echocardiogram was normal.

A renal biopsy showed marked foamy change to the glomerular visceral epithelial cells, proximal tubules, and foamy macrophages within the interstitium, suggestive of nephrosialidosis. Urine total and free sialic acid levels were elevated. Sialidase and Beta-galactosidase activity levels were diminished. Galactosidosis is suspected, but confirmatory genetic test is pending.

Galactosidosis is a rare lysosomal storage disease that can involve the kidney. Patients with infantile form of galactosidosis, can present with nephrotic range proteinuria and share histopathological features matching that of nephrosialidosis. It is characterized by a progressive course of nephropathy leading to renal failure. Nephrosialidosis should be considered in atypical presentations of childhood nephrotic syndrome.

PATIENT WITH BOWEL PERFORATION PRESENTING AS HEART FAILURE

De Vuyst RC. University of Oklahoma, Oklahoma City, OK.

Case Report: Presented here is a case of bowel perforation complicated by heart failure in a patient with Hx of hypoplastic left heart. Patients with single ventricle physiology have a poor cardiopulmonary reserve, because of this major conditions can be overlooked due to severity of cardiovascular symptoms. This case demonstrates the importance of having a high index of suspicion for inciting pathology in any of these patients presenting in heart failure.

A 2mo female with a Hx of hypoplastic left heart syndrome s/p Norwood, Sano, atrial septectomy presented to the ED due to inconstistiousness. AUS was done showing small bowel small bowel intussusception. Patient became calm and tolerated PO intake in the ED, at which point the parents took the patient home, as the intussusception was presumed to be spontaneously reduced. After returning home the patient started having emesis with every feed. Patient started acting lethargic, had increased WOB, greatly decreased UOP and was taken to the ED. Lab results were significant for an anion gap of 23. Patient had a 100cc/kg bolus of NS and was admitted to the floor for further hydration. There she developed increased urticaria; antibiotics were then changed to clindamycin and cefepime, and was transferred to the PICU. Lactate was found to be 19.48 on POC testing. Patient was started on Milrinone and given additional fluid resuscitation. Patient was then intubated for progressive respiratory failure. After stabilization the patient was found to have blood in stool. A KUB was performed showing peritoneal free air. Laparotomy patient was found to have multiple perforations of the proximal jejenum requiring resection of 20 cm of bowel. Patients hemodynamic status improved and she was progressivley weaned off of the ventilator and sedation.

BARAKAT SYNDROME PRESENTING AS SYNCPE

Dodd A, Schmit EO. University of Alabama at Birmingham, Birmingham, AL.

Case Report: Barakat Syndrome, or HDR Syndrome (hypoparathyroidism, sensorineural deafness, and renal dysplasia) was first identified in 1977 by Barakat et al. It is caused by haploinsufficiency of the GATA3 gene on the distal 10p chromosome. GATA3 is a transcription factor expressed in the developing inner ear, kidneys, parathyroid glands, thymus and central nervous system. Phenotypic expression of HDR syndrome is variable. Typically, patients present with symptoms of hypocalcemia such as myalgias, parathesias, tetaury, or convulsions. Hearing loss is the most penetrant and characteristic and ranges from mild to profound. The degree of renal involvement ranges from dysplasia to agenesis and can involve renal failure.

Our patient is an 11 year old female with a past medical history of sensorineural hearing loss diagnosed at birth who presented with fever and abdominal pain. The abdominal pain was generalized without vomiting or diarrhea. She also reported several pre-syncopal episodes and one syncopal episode in the two days prior to presentation. Her EKG demonstrated a borderline QTc of 450 msec. Initial laboratory evaluation showed an ionized calcium of 6.65 mmol/L, serum calcium of 5.2 mg/dL, serum magnesium of 1.5 mg/dL, and serum phosphorus of 8 mg/dL. Her hearing loss was congenital and reported to be moderate. Her family history was significant for renal disease on the paternal side of the family; father was started on dialysis at age 33, paternal grandmother had a solitary kidney and renal stent, and paternal grandfather was on dialysis starting in his mid-50’s. Her father also has mild hearing loss, but no other family members reported hearing loss. Patient was admitted and given calcium and magnesium, which improved her pre-syncpe and normalized her QTc interval. She was also diagnosed with UTI given too numerous to count white blood cells on her urinalysis, for which she was treated with cefadolin. A renal ultrasound to evaluate for dysplastic kidneys found two cysts in the right kidney. Her PTH hormone level was normal but inappropriately low for her degree of hypocalcemia, at 13 pg/mL. She was also found to have low vitamin D 25-OH at 13 ng/mL. She was discharged home on calcium carbonate, calcium acetate, and calcitriol. Ultimately, her suspected diagnosis of Barakat Syndrome was confirmed through CGH array demonstrating 10p14 deletion.

TESTICULAR MIXED GERM CELL TUMOR PRESENTING AS MANDIBULAR MASS WITH BRAIN LESIONS

Dubey V1, Sims K1, Meyer W1, Yu Z2, Pokaia H1. OU Health Science Center, Oklahoma City, OK and OU Health Science Center, Oklahoma City, OK.

Case Report: Introduction: Testicular germ cell tumors (GCTs) are a common malignancy in males 15-19 years of age. GCTs can metastasize to the lungs, viscera, and brain. Elevated serum human chorionic gonadotropin (HCG) is seen in GCTs with elements of choriocarcinoma. Choriocarcinoma is particularly aggressive, often presenting with distant metastases despite...
minimal symptoms from the primary tumor. Treatment includes orchietomy, dissection of local lymph nodes, and chemotherapy.

Case Presentation: A 16-year-old previously healthy male presented with 4 days of worsening right facial swelling unresponsive to oral steroids. Imaging at referring hospital illustrated a right parotid mass and 2 ring enhancing brain lesions. Review of systems was significant for unintentional 30 pound weight loss over last 2 months and nonspecific visual changes. Physical exam was remarkable for a 3x3cm non-fluctuant right parotid mass. Patient initially declined genital exam.

Initially an infectious disease workup was pursued. However, on further questioning the patient admitted to noticing testicular enlargement over the prior 3 weeks. Exam revealed that the right testicle was markedly enlarged, which was concerning for testicular malignancy. Screening labs for germ cell tumor revealed a markedly elevated HCG of 165,047. A radical orchiectomy was performed with pathology consistent with mixed germ cell tumor consisting of primarily choriocarcinoma in addition to components of yolk sac tumor and teratoma. Imaging was remarkable for multiple well-circumscribed lesions throughout the brain parenchyma, a well-circumscribed lesion in the right parotid gland, numerous pulmonary nodules, several liver lesions and enlarged abdominal lymph nodes. Final diagnosis was mixed germ cell tumor of the right testicle.

Discussion: Metastatic germ cell tumor must be considered in the differential in patients with a history concerning for malignancy and imaging consistent for CNS metastases. Tumors that include choriocarcinoma often metastasize rapidly with patients frequently having diffuse disease at diagnosis. A parotid mass is an unusual presenting symptom for germ cell tumor. This case reinforces the need for a thorough history and exam when concerned for malignancy.

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HAEMOPHILUS INFLUENZA TYPE A Meningitis WITH PROLONGED HOSPITAL COURSE
Farooq MA, Balan A, Ramji F. University of Oklahoma Health Science Center, Oklahoma City, Oklahoma, OK.

Case Report: Little is known about clinical course of infants with meningitis due to non-type B Haemophilus influenza. We present a case of Haemophilus influenza type A associated meningitis with a prolonged fever for more than 2 weeks with cerebritis and subdural empyema. A 4-month old term female infant presented with high fever, increased fussiness and bulging anterior fontanelle. CSF exam was consistent with elevated WBC 669, RBC 517, neutrophils 85 %, lymphocytes 4 %, glucose <5, protein 85.8 mg/dL, spinal fluid cultures were positive for Haemophilus influenzae type A. She had intermittent fevers during her hospital course which was also complicated by multiple seizures. Repeat head imaging showed widespread menigitis and frontal parafalcine subdural empyema. She continued to have daily fevers. Other sources of fever explored before another MRI was obtained which showed persistent subdural empyema, parenchymal destruction and communicating hydrocephalus. Her inflammatory markers remained elevated with high WBC and CRP, hence on hospital day 18, she underwent subdural empyema washout. Subsequent imaging showed resolution of empyema. Shortly after washout she improved tremendously. She was ultimately discharged after a prolonged hospital stay of 22 days.

Haemophilus influenza type B meningitis can cause fevers for longer period of time which can prolong patient’s hospital course. As described in a retrospective study conducted by Martin et al., 8 out of 86 patients with type B meningitis demonstrated an unusual clinical course characterized by persistent fevers (duration: greater than 10 days), but little is known about the clinical course of non-type B illnesses, which can significantly affect patient’s prognosis.

Infants with Haemophilus influenza type A meningitis may have a complicated course due to cerebritis and subdural empyema. In infants with prolonged fevers, wash-out of empyema should be considered as it may shorten duration of fever and hospitalization.

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RECURRENT HIVES AND THE ROAD TO A MYSTERY DIAGNOSIS: A CASE OF FAMILIAL COLD AUTO-INFLAMMATORY SYNDROME
Favier LA, Schikker K. University of Louisville, Louisville, KY.

Case Report: Familial Cold Auto-inflammatory Syndrome (FCAS) is one of the three cryopyrin-associated periodic syndromes involving a constellation of inflammatory symptoms spurred by exposure to cold weather. This case discusses the presentation of a pediatric patient with bouts systemic inflammation for the preceding six years of his life, and the final episode of arthritis which led to his diagnosis.

An 8-year-old male presented to our center with a one day history of right knee pain and swelling, and was found both clinically and radiographically to have arthritis of the affected joint. Inflammatory markers including CRP and ESR were only mildly elevated. Joint aspirate fluid excluded the likelihood of septic arthritis. It was discovered that the patient had a prolonged history of recurrent urticaria spurred by both exposure to cold temperatures and physical stress. His past medical history also yielded two prior musculoskeletal hospitalizations for myositis and a history of intermittent non-infectious conjunctivitis. This combination of symptoms in addition to a father with similar complaints, led to genetic testing for NLRP3 gene gain of function mutation. Testing yielded positive results for heterozygocity and the patient was started on monoclonal Interleukin-1 beta blockade therapy. With the exceedingly rare nature of this syndrome, with an incidence of 1:1,000,000 children annually, this patient’s diagnosis was delayed for six years. Since starting therapy, the patient and his father have each had dramatic increases in quality of life.

Cryopyrin-associated periodic syndromes include Familial Cold Auto-inflammatory Syndrome, Muckle Well Syndrome and Neonatal Onset Multisystem Inflammatory Disease. These syndromes represent a spectrum of disease severity in which the innate immune system is spontaneously activated to release Interleukin-1 causing downstream systemic inflammation. The release of pro-inflammatory cytokines is spurred by a dominant genetically inherited derangement in the intracellular organelle, known as the inflammasome. This case serves to spotlight this genetically rare and exceedingly interesting disease spectrum, but also to exemplify the importance of a thorough history of physical exam when dealing with symptoms of a nebulous nature.

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PNEUMOMEDIASTINUM AND EXTENSIVE CREPI TUS SECONDARY TO ASPIRATION OF RADIO LUCENT FOREIGN BODY
Ferrin SN. The University of Oklahoma, Oklahoma City, OK.

Case Report: In the following case, I present a patient with pneumomediastinum and subcutaneous emphysema with multiple possible etiologies including trauma, asthma, and foreign body aspiration. This case emphasizes the importance of obtaining careful history, adapting a working list of differential diagnoses as further information is obtained and patient status evolves, and maintaining a high index of suspicion for radiolucent foreign body aspiration.

A 7 yo male with Hx of asthma presents from an outside facility with complaint of cough, shortness of breath, chest pain, and a CT scan of the chest demonstrating pneumomediastinum and questionable esophageal abnormality. He had a recent Hx of trauma when another child collided with him on a bicycle prior to presentation, and also mentioned he had possibly swallowed pencil graphite although mother believed that was false. Pt had a Hx of “coughing attacks” with his asthma per mother, and one such attack had preceded his chest pain and shortness of breath at home, prompting a visit to their local ER. Physical exam findings on admission to our facility were significant for diminished breath sounds in the right lung field and subcutaneous emphysema over the mid and right chest, tracking into the neck to the angle of the mandible and into the axilla. Pt was started on asthma protocol. Radiology was consulted and pneumomediastinum was felt secondary to Macklin effect associated with asthma exacerbation, however increased density was noted on radiographic imaging in RLL of lung with no pneumothorax or foreign body identified. Esophageal perforation was considered to be unlikely after consultation with Pediatric GI and Surgery, and was ruled out with esophagram. Pt was continued on asthma protocol and showed initial improvement in ease of breathing with decreased cough and wean from supplemental oxygen, however his crepitus continued to progress into his scrotum and up to his scalp causing periorbital crepitus and closure of his right eye. Serial CXR demonstrated new hydro pneumothorax and pt's respiratory status did not improve; therefore pt was taken to the OR for rigid bronchoscopy where the plastic tip of a mechanical pencil was extracted from the right mainstem bronchus and chest tube was placed. Pt continued on to full recovery.

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BILATERAL MYOSITIS OSSIFICANS TRAUMATICA IN A CHILD
Eleenor TT. University of Alabama at Birmingham, Birmingham, AL.

Case Report: Myositis ossificans traumatica (MOT) is an unusual occurrence in children under 10 years of age. This non-neoplastic heterotropic bone...
formation can be secondary to severe blunt trauma or repetitive minor trauma. A unique case of a 6 year old found with bilateral myositis ossificans traumatica of the biceps will be reviewed in this case report. The patient was a victim of child abuse and found restrained at the wrists for an unknown period of time. The gruesome origin of the pathology was determined to be the repetitive attempts to escape. These distinctive findings will be reviewed along with diagnostic modalities and treatment.

AN UNUSUAL CASE OF PEDIATRIC IBW WITH CAVITATING PULMONARY NODULES

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Purpose of Study: Report a case of an interesting case of pediatric IBW with cavitary pulmonary nodules.

Methods Used: Case analysis and review of literature.

Summary of Results: Pulmonary manifestations of inflammatory bowel disease are a rare extra intestinal finding in children, with only 17 cases reported in the literature. We report a case of a 16 year old female who presented with colitis, asymptomatic pulmonary nodules and SIADH.

A 30 pound weight loss, along with intermittent bloody diarrhea, prompted her initial workup.

The cavitary nodules were initially discovered on a routine X-ray for tachycardia. Cavitary pulmonary nodules in an adolescent patient would more likely be associated with infectious etiologies including mycobacteria. An extensive infectious, rheumatologic, and immunologic work up failed to reveal an infectious diagnosis. A tissue lung biopsy was reported as chronic active bronchiolitis with negative viral stains. Bronchiectasis and bronchitis are common findings with IBW, but not bronchiolitis. Fistulizing lung disease and eosinophilic pneumonias are seen in Crohn’s disease (CD), while pulmonary vasculitis has been associated with ulcerative colitis (UC); however, cavitary pulmonary nodules are not a usual finding.

Her serum sodium consistently ranged from 128-132 despite fluid restriction. A workup revealed chronic SIADH secondary to her pulmonary lesions. SIADH has not been previously reported as a manifestation of pulmonary nodules related to IBW in children.

Diffuse thickening of the entire colon and terminal ileal narrowing was noted on CT imaging. Colonoscopy showed extensive superficial ulcers without any skip lesions in the colon and edema around the terminal ileum. Biopsies revealed chronic active colitis but no granulomas. Serum IBW SGI showed a pattern consistent but indeterminate for UC or CD.

Conclusions: The diagnosis of inflammatory bowel disease in this pediatric patient could only be made after clinical improvement of hematochezia and regression of pulmonary nodules after initiating treatment with infliximab.

HYPERINFLAMMATORY SYNDROME-RARE COMPLICATION OF EBSTEIN-BARR VIRUS

gavrineni M, Bidot L, Dankhara N, Popescu M. ETSU, Johnson city, TN.

Case Report: We report the case of a 17-year old white female with fevers, body aches, nausea, vomiting and swollen glands.

Past history includes Tumor Necrosis Factor Receptor Associated Periodic Syndrome diagnosed at age 11 responsive to steroid therapy and she’s been in remission for 2 years.

Family history is significant for brother having recurrent “monon” infection.

On examination, she was febrile, tachycardic, ill appearing adolescent with enlarged bilateral cervical lymph nodes, hepatosplenomegaly and edema of hands and feet.

Initial laboratory showed elevated AST and ALT to 599 and 232, respectively. GGT 62, ammonia 47, albumin 2.5, LDH >2700; uric acid was normal. CRP was 66.8, ESR was 18. WBC 1.7, neutrophils 31%, monocytes 62%, lymphocytes 7%, hemoglobin 9.5, reticulocytes 1.7%, platelets 55000. Periperal smear showed reactive lymphocytes, few giant platelets. Bone marrow exam showed hypercellular marrow with trilineage hematopoiesis and no hemophagocytosis.

Blood, urine and stool cultures were negative. Patient was started on Cefepine due to fever and neutropenia. Infectious disease team tested for EBV, CMV, Parvovirus, Histoplasma, Bartonella which were all negative except serum EBV-PCR showing 19,800 copy/ml.

Ultrasound abdomen showed hepatosplenomegaly. MRCP showed enlarged spleen, periperal edema and small bilateral pleural effusions. Ferritin was markedly elevated at 8,412. Fibrinogen was low at 103. Hepatitis panel, ANA and RF were negative; immunoglobulin levels were normal. Circulating soluble IL2 receptor was not obtained.

At this time, patient was transferred to a higher center to be evaluated for HLH. Lymph node biopsy revealed EBV-positive T-cell lymphoproliferative disorder with marked hemophagocytosis and she was started on Dexamethasone with fevers resolving shortly thereafter.

Discussion: Hemophagocytic Lymphohistiocytosis(HLH) is characterized by a reactive process resulting from excessive activation of antigen-presenting cells and CD8+ T cells, and excessive proliferation and ectopic migration of T cells. The patient met HLH diagnostic criteria, which include: fever, splenomegaly, cytopenias, markedly elevated ferritin, hypofibrogenemia. Since HLH can be rapidly fatal, it is recommended that treatment be started when there is a high clinical suspicion.

CVID - HIT BY HIDS

gavrineni M, Macariola D. ETSU, Johnson city, TN.

Case Report: We present a 9 year old white female with recurrent fevers relapsing every 2-4 weeks lasting for 7-10 days for the past 2 years associated with lymphadenopathy, fatigue and muscle aches. She does not have joint pains, skin rashes or weight loss.

Past history is significant for sinus and ear infections, pneumonia, asthma and MRSA cellulitis. She received antibiotics on several occasions but her fevers kept relapsing.

She is of half Dutch ancestry and has no family history of immunodeficiency or autoimmune disease.

Physical examination showed a slightly ill appearing female with no other abnormal findings.

Initial results included a normal comprehensive metabolic panel, WBC of 11.3, hemoglobin 14.6 gm/dl, platelets 360,000, neutrophils 38%, lymphocytes 59%, no bands. Negative blood culture, negative EBV titers, negative respiratory viral panel, and normal thyroid panel. Normal vitamin B12 and folate level. CXR was normal.

Immunoglobulin levels showed a low IgG level of 559mg/dl, normal IgM and IgA level after which an Infectious Disease consult was obtained.

Anti dsDNA and anti-CCP level were negative, to rule out autoimmune etiology. To rule out chronic granulomatous disease, a neutrophil oxidative burst test was done which was normal. Immunoglobulin levels were repeated with a low IgG of 57mg/dl, normal IgM, IgA and IgE and an elevated IgD of 22.5mg/dl. Repeat IgD level was elevated at 35.6mg/dl. Based on the above labs, patient was diagnosed with Common Variable Immunodeficiency and Hyper IgD syndrome.

For CVID, she was started on IVIG infusions and for Hyper IgD syndrome, was started on simvastatin with clinical improvement noted.

Discussion:

Hyperimmunoglobulin D Syndrome (HIDS) is a rare auto inflammatory disease characterized by periodic attacks of fever and a systemic inflammatory reaction (cervical lymphadenopathy, abdominal pain, vomiting, and diarrhea, arthralgia and skin signs). Majority patients with HIDS are Caucasians with 60 percent of them being either Dutch or French ancestry. Over 80% of HIDS patients also have high IgA levels. During an attack, leukocytosis and elevated C-reactive protein are noted with occasional mevalonate aciduria. Here, we report a patient with an unusual combined hypergammaglobulinemia and hypergammaglobulinemia syndrome. Clinicians should therefore, be cognizant that this situation can occur in patients with hypogammaglobulinemia.
prothrombin G20210A mutations, are noted to have a high prevalence at 5% and 2% respectively among Caucasians.

A 15-year-old previously healthy but obese Caucasian female presented to the ED with a 3-day history of right leg pain and pleuritic right chest pain. She had tachycardia on presentation and ECG changes suggestive of right heart strain. She endorsed oral contraceptive pill use for the last few months. Given her presenting symptoms a pulmonary embolism was suspected. A chest CT-angiogram was performed which showed an infarct in the right lung consistent with pulmonary embolus to the right pulmonary branch artery. She was started on low molecular weight heparin and later changed to warfarin when her INR was between 2-3. Evaluation for hypercoagulability risk factors was negative. Despite being on warfarin and having a supratherapeutic INR of 7.5, the patient presented again after 6 days with progression of pulmonary embolus to the left side in the left pulmonary branch artery. Her warfarin was held for 3 days until her INR was therapeutic at 3. Further genetic evaluation revealed her being heterozygous for factor V Leiden and homozygous for prothrombin G20210A.

The prevalence of individuals with both of the factor V Leiden and prothrombin G20210A mutations is rare. In people with factor V Leiden mutations, factor V cannot be inactivated normally, resulting in a prolonged clotting process. Individuals with the prothrombin G20210A mutation have an increase in the normal amount of prothrombin protein that increases the risk for clotting. Since pulmonary embolism is a thrombotic condition with a high mortality rate in young patients with recurrent pulmonary emboli, a hypercoagulability screening including testing for factor V Leiden and prothrombin G20210A mutations, along with an evaluation of acquired risk factors for thrombosis should be performed.

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RED CHROMONYCHIA IN KAWASAKI DISEASE

George AP1, Gaby C2, Broughton R3. 1University of Kentucky, Lexington, CA; 2University of Kentucky, Lexington, KY; 3University of Kentucky, Lexington, KY.

Case Report: Kawasaki disease in an acute, self-limited vasculitis that primarily affects infants and young children. A literature review reveals that nail changes occasionally occur in Kawasaki disease. The most common nail alteration is transverse leukonychia, also known as Beau's lines. Chromonychia, a change in nail color, was first reported in Kawasaki disease in 1984. Chromonychia remains a rare clinical finding and has been reported only once in the pediatric literature in the last 20 years.

Our patient is a previously healthy 2-year-old female who presented with a 6-day history of fever and diminished activity. Her pediatrician diagnosed the patient with otitis media and prescribed cefdinir. She returned to her pediatrician after 2 days with persistent fever along with red, cracked lips, and non-exudative conjunctival injection; this constellation of findings raised the suspicion for Kawasaki disease. The patient also had red, transverse lines across the bed of her fingernails. Laboratory studies included a normal blood count, serum electrolytes, and urinalysis. A transthoracic echocardiogram revealed left main coronary artery ectasia. Due to the high clinical suspicion that she had Kawasaki disease, the patient was treated with intravenous immunoglobulin and high dose aspirin.

Chromonychia has been seen in association with many drugs,thermal injury, environmental exposure, and several infections. Red transverse chromonychia in Kawasaki disease was first described in four patients by Lindsay. Our patient, like others reported in the literature, illustrates that nail bed changes, when considered in conjunction with other clinical manifestations of Kawasaki disease, should lead the clinician to strongly consider Kawasaki disease.

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CMV COLITIS AND STRICUTURE IN A PRETERM INFANT WITH FEEDING INTOLERANCE: CASE REPORT

Rivera D, Gerber Z, Begue R, Champagne K, Bond B. LSUHSC, Kenner, LA.

Purpose of Study: Cytomegalovirus (CMV) remains the most important cause of congenital viral infection and neonates born to mothers who are primarily infected during pregnancy are the most vulnerable. Localized gastrointestinal manifestations of the disease are rare with only a few case reports described. We report a case of CMV in a pre-term infant who presented with feeding intolerance and CMV associated stricture.

Methods Used: A chart review was performed.

Summary of Results: A 1144mg male was born at 27 4/7 weeks of gestation via Cesarean section to a 43 year old G3P0 mother. Feeds were tolerated until day of life 17 when he was noted to have bloody stools, suspected pneumonitis inteninals and leukocytosis. He completed 10 days of antibiotics and bowel rest for suspected necrotizing enterocolitis. All cultures were negative. The infant had repeated episodes of emesis and abdominal distension despite changes to elemental formulas. He was transferred to our institution on DOL 71 for further evaluation. A contrast barium enema demonstrated strictures in the left hemicolon. A hemicolectomy with an end-to-end anastomosis was performed. Histopathology showed areas of ulceration demonstrating active colitis. Immunohistochemical analysis confirmed the presence of CMV inclusions. Urine CMV PCR and blood CMV DNA were both positive. Eye exam, hearing screen, head ultrasound and liver function tests were normal. The infant was treated with Gancyclovir for 6 weeks. Postoperatively, he tolerated feeds without further issues.Mother was found to be seropositive for CMV and her breastmilk was restricted due to suspected mode of transmission.

Conclusions: It is well documented that postnatal transfer of the virus can occur through the breastmilk of seropositive mothers. There may be value to screening mothers for CMV and discussing potential risks associated with breastmilk transmission. This case also highlights CMV presenting with only localized gastrointestinal manifestations. It is important to recognize CMV as a potential etiology in a premature infant with feeding intolerance and NEC-like illness prior to the presentation of associated late complications such as stricture.

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GROUP A STREPTOCOCCUS CAUSING PRIMARY PERITONITIS AND SEPTIC SHOCK IN A HEALTHY CHILD: AN UNCOMMON PRESENTATION OF A COMMON PATHOGEN

Graves KS1, Rutledge CL1,2,3. 1University of Alabama Birmingham, Birmingham, AL; 2Children’s of Alabama, Birmingham, AL.; 3Children’s of Alabama, Birmingham, AL.

Case Report: We report the case of a previously healthy 8 year old African American female who developed septic shock following three days of abdominal pain. She initially presented to the emergency room with abdominal pain, fever, and CT findings consistent with terminal ileitis; she was discharged with a diagnosis of viral gastroenteritis. Within 24 hours, the child returned to the ER with septic shock and evidence of abdominal compartment syndrome. There was initial concern for appendicitis, intestinal perforation, or pelvic infection secondary to vaginal foreign body. She required intubation, pressor support, and emergent exploratory laparotomy. Surgical findings were inconsistent with all suspected etiologies excepting primary peritonitis. Infection with streptococcus pyogenes was confirmed by cultures of blood and ascitic fluid. Despite a dire initial presentation, the patient recovered rapidly and completely with targeted antimicrobial therapy and supportive treatment. Immunologic workup has revealed no immunodeficiency. Though primary peritonitis with streptococcus has been described, nearly all cases have been in children two years or younger, or in children with co-commitent medical conditions such as hepatic failure or nephrotic syndrome.
CENTRAL NERVOUS SYSTEM PHAEOHYPOMYCOSIS MIMICKING TUMEFACTIVE MULTIPLE SCLEROSIS IN AN IMMUNOCOMPETENT PEDIATRIC PATIENT

Henson SE, Balan A. Oklahoma University, Oklahoma City, OK.

Case Report: Introduction: Fungal infections of the central nervous system in immunocompetent patients are rare. We report a case of a Bipolaris s. brain abscess in a previously healthy 14 year old male. His brain MRI showed findings consistent with tumefactive multiple sclerosis (MS). He was ultimately diagnosed with Bipolaris s. based on repeat CSF studies and brain biopsy. This case report illustrates the importance of considering fungal infection as a differential diagnosis in immunocompetent patients with MRI findings suggestive of tumefactive MS.

Presentation: A 14 year old male presented with a one month history of headaches. On the day of admission he developed altered mental status, fever and seizures. He underwent a head CT which was concerning for an underlying mass. CSF studies revealed pleocytosis, protein of 150, glucose of 57 and a negative calcofluor stain. Neurology and radiology considered the MRI findings to be suggestive of tumefactive MS and high dose steroids were started. He improved neurologically and the seizures resolved, but he continued to have diplopia and headache. After receiving 4 doses of steroids he rapidly deteriorated and developed cerebral edema. CSF studies were repeated revealing a glucose of 22 and calcofluor stain positive for fungal elements. He was started on Ambisome with no improvement and a brain biopsy was consistent with invasive fungal elements. He was declared brain dead 9 days after initial presentation.

Discussion: Phaeohyphomycosis is a rare infection caused by fungi that produce melanin in their cell walls giving them a dark appearance. From 1966-2002 only 101 cases of culture-proven CNS phaeohyphomycosis infections were reported in the English-language literature. Only 4 of those cases were caused by Bipolaris s. Mortality rate in immunocompetent patients was 74%. Bipolaris s. infections have been misdiagnosed as other CNS conditions making diagnosis difficult. While brain biopsy allows early diagnosis in CNS fungal infections, it can be contraindicated in conditions mimicking fungal infections. In conclusion, Phaeohyphomycosis should be considered in immunocompetent patients with CNS lesions, as early intervention with brain biopsy, surgical excision and medical management may improve outcomes.

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FAMILIAL PRIMARY HYPERTROPHIC OSTEOARTHROPATHY AND ELEVATED SWEAT CHLORIDE

Hothi JS, Stokes D, Srinivasan S. University of Tennessee Health Science Center, Memphis, TN.

Case Report: Two brothers with digital clubbing were found to have elevated sweat chloride, suggesting a possible diagnosis of cystic fibrosis (CF) (Table). Although one sibling had mild asthma, other clinical features were atypical for CF; including a lack of bronchiectasis, sinusitis, or gastrointestinal findings. Genetic evaluation revealed several mutations in CFTR, but only one possibly associated with cystic fibrosis (c.1209+20T>G). The bipolaric father of both siblings also has digital clubbing, but no history of lung or gastrointestinal disease. He also showed elevated sweat chloride levels, although less elevated than the siblings. Based on the family history of clubbing the diagnosis of primary hypertrophic osteoarthropathy (PHO, or congenital clubbing) is likely, but this disorder has not previously been associated with elevated sweat chloride. In 2008, the genetic basis for PHO was found in two mutations in the prostaglandin metabolism pathway, 15-hydroxyprostaglandin dehydrogenase, a major enzyme involved in the degradation of prostaglandin, and SLC20A1, the gene encoding the prostaglandin transporter (1,2). In several families with these disorders, elevations of prostaglandins and prostaglandin metabolites have been demonstrated. A potential link between PHO, elevated prostaglandin levels and elevated sweat chloride values is suggested by a study showing elevated sweat Cl- values in an infant receiving exogenous prostaglandin, as well as studies in vitro showing that prostaglandins alter Cl- transport in cell models (3,4). The higher sweat chloride levels in younger patients fits with observations of falling prostaglandin levels in PHO with age (2). Clubbing and elevated sweat chloride levels due to alterations in genes associated with PHO could lead to the false diagnosis of CF at any age and may explain some cases of atypical CF without identified mutations in CFTR or minimal sino-pulmonary findings.

<table>
<thead>
<tr>
<th>Age (years), relationship</th>
<th>Sweat chloride (mmol/L)</th>
<th>Notes</th>
</tr>
</thead>
<tbody>
<tr>
<td>15, sibling</td>
<td>110, 113</td>
<td>Mild sweat, normal spirometry, chest X-ray, sinus films, stool elastase, CFTR genotype: negative for c.1209+20T&gt;G, Clubbing</td>
</tr>
<tr>
<td>17, sibling</td>
<td>Initial: 68, 62 Repeat: 81, 64</td>
<td>Normal spirometry, normal stool elastase, chest x-ray/CT chest: old granulomatosus disease, CFTR genotype: c.1209+20T&gt;G, Clubbing</td>
</tr>
<tr>
<td>49, Father</td>
<td>46, 53</td>
<td>Clubbing</td>
</tr>
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JAPANESE ENCEPHALITIS IN AN AMERICAN ADOLESCENT AFTER TRAVEL TO ASIA

Huang MZ, Novara SC, Hough-Telford C, Oliver SE, Wu CL, Pinninti S. University of Alabama at Birmingham, Birmingham, AL.

Case Report: Japanese encephalitis (JE) is a rare cause of CNS infection in the United States. Those diagnosed in the U.S. have traveled to endemic regions. We present the eighth reported case of JE in the U.S. since licensure of the JE vaccine in 1992 and the first case since 2010.

In July 2014, a 17 year-old male presented with altered mental status 6 days after returning from a 3-week trip to Cambodia and Vietnam. He received the typhoid vaccine prior to travel and incomplete malaria prophylaxis. During the trip, he had diarrhea, vomiting, fever, and headache. The symptoms resolved but returned 5 days after arrival in the U.S. He was admitted to a local hospital, and labs revealed 12,000 WBCs/µL with 78% neutrophils, an elevated CRP, and CSF notable for 618 WBCs/mm3, 87 mg/dL protein, and 49 mg/dL glucose. He was transferred to our facility for increasing confusion that progressed to irritability, difficulty following commands, mask-like facies, and diffuse abdominal tenderness. Brain MRI showed swelling and increased T2/FLAIR signal in the left temporal lobe cortex, limbic system, basal ganglia, and thalamus. An EEG showed slowing without epileptiform activity. His travel history placed JE high on the differential; HSV, enterovirus, malaria, rabies, and arboviral encephalitides were also considered. Empiric vancomycin, cefotaxime, and acyclovir were given while awaiting studies. CSF, blood and stool cultures, and labs for enterovirus, HSV, HIV, and malaria were negative. Serologic studies for JE virus were sent to the CDC. At time of abstract submission, preliminary studies had evidence of IgM and neutralizing antibodies against JE virus in the CSF and serum. JE virus is transmitted to humans by infected Culex mosquitoes. Prevention focuses on minimizing exposure and vaccinating high-risk travelers. CDC guidelines recommend vaccination for those traveling to endemic areas for >1 month during the summer/fall seasons in temperate zones and year-round in the tropics. Travelers to rural areas or areas of active outbreak, and those engaging in frequent outdoor activities, should be vaccinated regardless of travel time. Ixaro is a 2-dose vaccine series administered 28 days apart, to be completed >1 week prior to travel and licensed in the U.S. for children >2 months old.

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ATYPICAL MICROBIOLOGIC PRESENTATION OF LEMIERRE SYNDROME WITH M ethillin-resistant STAPHYLOCOCCUS AUREUS AND LITERATURE REVIEW OF PEDIATRIC CASES

Huang MZ, Boppara SB. University of Alabama at Birmingham, Birmingham, AL.

Case Report: Lemierre syndrome (LS), or jugular vein suppurrative thrombophlebitis, is well-described in literature and oftentimes caused by Fusobacterium necrophorum or anaerobic flora. We present a case of LS caused by methillin-
resistant Staphylococcus aureus (MRSA), the first such case at our institution, and review the literature on pediatric MRSA-derived LS. A 13-month-old girl with no significant past medical history presented with five days of fever and enlarging neck mass which had not responded to outpatient antibiotics. Parents also reported increasing cough and a weak, high-pitched cry. Her exam revealed a large fluctuant mass that extended from the left posterior triangle of the neck to the sternal notch and tracheal deviation. A CT scan revealed a loculated abscess and thrombosed left internal jugular (IJ) vein. The abscess was emergently drained in the OR and cultures grew MRSA. She was treated with vancomycin and completed an 8-week course of antimiobiotics. The patient did not develop any complications, was not bacteremic, and did not develop any disseminated septic emboli. Repeat imaging at follow-up revealed no residual abscess but absence of the left IJ was noted with multiple collateral and normal flow.

A review of the literature revealed only six pediatric cases of LS caused by MRSA. Three patients were teenagers, one an 8-year-old girl, and two infants. All six patients were bacteremic. One child had bilateral IJ thrombi, three had septic pulmonary emboli, one developed paracarditis with cardiac tamponade, two had dural sinus involvement, two had orbital involvement, and one patient had significant neurologic symptoms including vision loss and hemiparesis. All six patients were bacteremic. One child had bilateral IJ thrombi, three had septic pulmonary emboli, one developed paracarditis with cardiac tamponade, two had dural sinus involvement, two had orbital involvement, and one patient had significant neurologic symptoms including vision loss and hemiparesis. All six patients were bacteremic.

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Case Report: Oklahoma City, OK. Khawar U, DeLeon S.

PERSISTENCE PAYS OFF

This data indicates that many genes in the Xq28 region are characterized by intellectual disability, absent or limited speech, frequent infections, neuropathy, and seizures. There are no consensus breakpoints in this syndrome but most reported duplications are larger than 500kb and contain at least the MeCP2 gene. MeCP2 under expression is causative in Rett syndrome and patients display a similar phenotype of intellectual disability, autistic behaviors, neuropathy, regression, and seizures. Because of the clinical overlap, MeCP2 is usually considered to be the critical gene in Xq28 duplication syndrome. For this reason we designed a study to investigate the pathogenicity of other genes in the Xq28 region and hope to further elucidate their functions.

Methods Used: Array comparative genomic hybridization (aCGH) was used to screen over 1,200 patients for sub microscopic genomic alterations. Patients in this group were referred to the Hayward Genetics Center for intellectual disability, developmental delay, or a suspected genomic disorder between the years 2010 and 2014.

Summary of Results: Duplications in the Xq28 region not containing the MeCP2 gene were identified in ten patients. The clinical phenotype of these patients varied some but all of the hallmark traits of Xq28 duplication were present in this group. Intellectual disability and developmental delay were identified in five cases, absent or limited speech and frequent infections in two cases, and seizures in one case. Additionally no other genomic alterations were identified in any of these ten patients. Five of the cases were represented in two families. In one family the duplication was inherited by a affected boy from his unaffected mother while in the other family the duplication was identified in both the son (affected) and daughter (mildly affected) of a mildly affected mother.

Conclusions: This data indicates that many genes in the Xq28 region are dosage sensitive, play a key role in early development and that Xq28 duplication syndrome should not be thought of only as MeCP2 duplication. Furthermore, aCGH technology is a very powerful tool for identifying sub-microscopic genetic alterations.

ACGH IS A SENSITIVE TOOL FOR IDENTIFYING SUBMICROSCOPIC XQ DUPLICATION IN PATIENTS WITH INTELLECTUAL DISABILITY

Janssen AT, Chen TG. Tulane University, New Orleans, LA.

Purpose of Study: The clinical phenotype of Xq28 duplication syndrome is characterized by intellectual disability, absent or limited speech, frequent infections, neuropathy, and seizures. There are no consensus breakpoints in this syndrome but most reported duplications are larger than 500kb and contain at least the MeCP2 gene. MeCP2 under expression is causative in Rett syndrome and patients display a similar phenotype of intellectual disability, autistic behaviors, neuropathy, regression, and seizures. Because of the clinical overlap, MeCP2 is usually considered to be the critical gene in Xq28 duplication syndrome. For this reason we designed a study to investigate the pathogenicity of other genes in the Xq28 region and hope to further elucidate their functions.

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UNEXPLAINED ASYMPTOMATIC HYPOXEMIA: PERSISTENCE PAYS OFF

Khawar U, DeLeon S. University of Oklahoma Health Sciences Center, Oklahoma City, OK.

Case Report: Isolated right superior vena cava (R SVC) drainage into the left atrium is a rare cardiac anomaly in the absence of other cardiac malformations. We describe an unusual case of an infant with hypoxemia who was found to have this defect after an extensive workup.

A previously healthy 11-month-old female was admitted for the evaluation of room air hypoxemia in the absence of respiratory symptoms. Initial evaluation revealed oxygen saturations (SaO2) of 95%, which responded to supplemental oxygen via nasal cannula with the SaO2 increasing to 98%. Initial workup of her hypoxemia was negative, including a chest X-ray, respiratory viral panel, and levels of carboxyhemoglobin and methemoglobin. Meanwhile, hemoglobin electrophoresis done to rule out underlying hemoglobinopathies showed a normal hemoglobin variant. Room air arterial blood gas revealed a low PO2 and elevated alveolar to arterial oxygen gradient. A thoracic echocardiogram (TTE) showed drainage of left superior vena cava (LSVC) to coronary sinus and an incompletely visualized R SVC. High-resolution chest computed tomography did not reveal any pulmonary pathology. TTE with injection of agitated contrast saline in left arm revealed a LSVC draining to coronary sinus. A third TTE with injection of agitated contrast saline in right arm revealed immediate microcavitations in the left side of the heart concerning for R SVC draining into left atrium. Cardiac magnetic resonance imaging (MRI) confirmed this rare defect. Our patient underwent surgical correction of the systemic anomalous return without any complications and was discharged home on room air.

The most commonly encountered anomaly of systemic venous return in the presence of bilateral SVCs is a persistent LSVC draining into the right atrium or coronary sinus. However, R SVC draining into the left atrium causing hypoxemia in the absence of clinically detectable abnormalities represents a rare congenital malformation. Isolated R SVC-to-left atrium should be considered in the differential diagnosis of hypoxemia. Given potential complications from hypoxia and paradoxical emboli, surgical correction is indicated even in asymptomatic patients. Isolated hypoxemia without an overt cardiac defect should always be thoroughly investigated by contrast echocardiography and MRI.

CARDIAC SURGERY IN AN ADOLESCENT WITH PREKALLIKREIN (FLETCHER FACTOR) DEFICIENCY

Kiefer AC, Provo J, Carollo D, Caspi J, Lotteman C, Warrier R. Tulane University School of Medicine, New Orleans, LA; Ochsner Clinic Foundation, New Orleans, LA and LSUHH/Children’s Hospital, New Orleans, LA.

Case Report: Prolonged partial thromboplastin time (P TT) is a frequent cause for pre-surgical referral of children to hematology with resultant postponement. We report the case of an adolescent with abnormal activated coagulation time (ACT) performed by anesthesia just prior to surgery leading to an emergency consult.

A previously healthy 15-year-old African American male football player was noted to have a significantly prolonged ACT of 424 (three times) after the induction of anesthesia and arterial line placement. He had negative personal and family history for bleeding disorders. His physical exam was significant only for the Grade 2/6 ejection systolic murmur secondary to his atrial septal defect (ASD). Work up showed mildly elevated PTT with a weak lupus anticoagulant (LAC). In spite of the disappearance of the LAC, the PTT remained prolonged. Mixing studies were inconsistent. Prothrombin time (PT), international normalized ratio (INR), Factor XII, IV, XI, XII and XIII, Fitzgerald factor, liver function, and fibrinogen were all normal. Plasma prekallikrein (PK) activity was <5% (normal 63-135%) and confirmed with repeated testing. We recommended family genetic testing to determine if any siblings have this rare disorder and fresh frozen plasma (FFP) 1 hour prior to surgery to normalize his PK and allow for easier monitoring during surgery. Since his ASD was too large to be repaired by interventional methods, he underwent an open cardiac surgery repair without any significant or unusual bleeding.

PK deficiency is a very rare heritable disorder with reduced or absent functional PK in the plasma. Since coagulation in vivo does not require PK, there is no risk of bleeding in the proband. However, the interference with ACT and PTT could cause significant problems in the postoperative monitoring of anticoagulation routinely used for cardiac surgery. Our patient had no intraoperative or postoperative difficulties and did not require any supplemental support.

Sponsored by: Dr. Russell W. Steele, Pediatric Infectious Disease, Ochsner Clinic Foundation

CANCELLED
Case Report: Oklahoma City, OK.
Lowe G, Williams RS.
OF SURVIV AL
A HEALTHY TWO YEAR OLD FEMALE: A STORY

Introduction: Epitrochlear lymphadenopathy should always be considered abnormal. The differential diagnosis includes malignancy and infections including bartonellosis, tuberculosis, HIV and tick-borne illness. We present a pediatric patient with epitrochlear lymphadenopathy due to cat-scratch disease.

Case Presentation: The patient is a 6-year-old previously healthy boy presenting with a 5-week history of right arm pain and swelling near his elbow. He had no history of significant trauma or systemic symptoms including fever. Physical exam revealed a well-appearing child, with a small mass visible on the medial side of the elbow. On palpation there was a 1.5-cm tender mobile mass present just proximal to the elbow joint. A faint healing scratch was noted to the nail bed on the 4th digit of his right hand. Ultrasound of the arm revealed multiple necrotic hyperemic lymph nodes in his medial right arm suggestive of lymphadenitis. CBC and CRP were within normal limits. Bartonella IgM reticular tests were positive at 1:320. Serology was repeated 4 weeks later and showed further increase in IgG to 1:640. Since the patient was otherwise asymptomatic with minimal discomfort caused by the enlarged epitrochlear lymph node, decision was made to monitor him without antimicrobial therapy.

Discussion: Cat-scratch disease is the most common cause of chronic unilateral regional lymphadenitis in children in the United States. Lymphadenitis usually involves nodes that drain the site of inoculation. Nearly 43% of cases lymph nodes of the upper extremity are involved and there seems to be a predilection for medial epitrochlear nodes. Demonstration of rising IgG titer provides the best evidence of infection. The disease is usually self-limited, resolving spontaneously in 2-4 months.

Conclusion: Epitrochlear lymphadenopathy should always be considered abnormal. The differential diagnosis includes lymphoma, soft tissue sarcoma, and infectious causes: local pyogenic infection, cat-scratch disease, sporotrichosis. Our case illustrates that cat scratch disease must be considered in the differential diagnosis as positive serology would allow diagnosis and avoidance of invasive procedures.

A UNIQUE MECHANISM OF COMMOTIO CORDIS IN A HEALTHY TWO YEAR OLD FEMALE: A STORY OF SURVIVAL
Lowe G, Williams RS. Oklahoma University Health Sciences Center, Oklahoma City, OK.

Case Report: Blunt trauma to the chest in children, even in the absence of musculoskeletal injury, is still cause for concern. Elasticity of the pediatric skeleton allows for the possibility of vital organ contusion or internal hemorrhage. Rare but important to consider, a direct blow to the precordium can lead to cardiac arrhythmia and collapse. Commotio cordis is a primary arrhythmic event, usually ventricular fibrillation, triggered by a blunt, non-penetrating blow to chest without thoracic or cardiac injury in the absence of underlying cardiovascular disease and is often fatal.

Our patient is a 2 year 9 month old previously healthy female with no underlying medical conditions who presented to a tertiary children’s ER after blunt trauma to the chest with subsequent collapse requiring bystander resuscitation. Our patient was jumping on a trampoline while her father was mowing the lawn. The lawn mower ran over a golf ball, propelled it across the yard and struck her in the left chest. She cried briefly, gasped, and collapsed. Within 60 seconds she was not breathing with perioral cyanosis. After failed arousal attempts, her father performed cardiopulmonary resuscitation and transported her to the hospital via private car. He reports she awakened in the car after about 2 minutes and began crying. She was confused but able to answer questions. On arrival to the tertiary children’s ER the patient was awake, alert, tearful, but in no distress. She was tachycardic with a small circular bruise on her left chest but otherwise a normal physical exam. Chest radiograph revealed no fracture or underlying medical conditions which presented to a tertiary children’s ER after blunt trauma to the chest with subsequent collapse requiring bystander resuscitation. Our patient was jumping on a trampoline while her father was mowing the lawn. The lawn mower ran over a golf ball, propelled it across the yard and struck her in the left chest. She cried briefly, gasped, and collapsed. Within 60 seconds she was not breathing with perioral cyanosis. After failed arousal attempts, her father performed cardiopulmonary resuscitation and transported her to the hospital via private car. He reports she awakened in the car after about 2 minutes and began crying. She was confused but able to answer questions. On arrival to the tertiary children’s ER the patient was awake, alert, tearful, but in no distress. She was tachycardic with a small circular bruise on her left chest but otherwise a normal physical exam. Chest radiograph revealed no fracture or abnormality. Laboratory tests showed significantly elevated cardiac enzymes and she was admitted for observation on telemetry. Repeat cardiac enzymes trended down, and she had a normal echocardiogram with no cardiac injury. Vital signs normalized and she was discharged home in good condition.

Commotio cordis has been described primarily in adolescents in competitive sports. Victims are usually hit by a projectile, occasionally by another body in a collision, while events have been reported in non-sport settings. This case highlights that any direct blow to the chest at a critical moment in the cardiac cycle is a risk for commotio cordis, even in younger children, and prompt resuscitation increases chances of survival.

MISSED DIAGNOSIS OF BENIGN RECURRENT INTRAHEPATIC CHELASTISIS IN A 17 YEAR OLD CAUCASIAN MALE
Merchant S1, George D2, McGoogan K2. University of Florida College of Medicine, Jacksonville, FL and 2Nemours Children's Clinic, Jacksonville, FL

Case Report: Our patient is a 17 year old previously healthy Caucasian male who presented with a 3 week history of sclaim icterus, jaundice and severe pruritus. He also had intermittent bilateral lower quadrant abdominal pain, diarrhea, nausea and non-bloodly, non-bilious vomiting. There were no recent travels, exotic foods or sick contacts. No family history of liver disease. Recent medications included doxycycline for acne and benadryl for pruritus. Due to his age and recent doxycycline use, he was previously diagnosed at an outside community hospital with drug induced liver injury by a Gastroenterologist. When his symptoms persisted, his parents requested a second opinion and he was referred to our institution. Upon evaluation, he was severely jaundiced. His abdomen was mildly tender to deep palpation without rebound or guarding in the right upper and lower quadrant as well as the left lower quadrant. No hepatosplenomegaly was noted. Laboratory studies showed total bilirubin 32.3mg/dL, conjugated bilirubin 14.4mg/dL with AST 68 IUL and ALT 84 IUL. Serum GGT, total protein, A/G ratio, PT, INR and cephalosporin were all within normal range. Viral studies for hepatitis A and B, EBV and CMV were negative as were thyroid studies, serum alpha 1 Antitrypsin level and anti-mitochondrial antibody. A liver biopsy was performed and showed coarse granular bile without fibrosis and he was thought to have a benign form of Familial Intrahepatic Cholestasis. Gene mutation analysis showed a nonsense mutation in ATP8B1 confirming the diagnosis. BRIC is a rare autosomal recessive disease caused by homozygous or compound heterozygous mutation in the ATP8B1 gene on chromosome 18q. The progressive form of this condition usually presents in infancy with recurrent episodes of cholestasis without extra hepatic bile duct obstruction. Jaundice and excessive pruritus without development of chronic liver damage can be indicative of BRIC. It is important for physicians to recognize the late presenters of these genetic diseases, as adolescents and adults with BRIC are often misdiagnosed due to the rarity of presentation past infancy.

HIV: THE GREAT MIMICKER
Murphy M, Anson E, Paccione R, Beatty K. LSU/HSC, New Orleans, LA

Case Report: A 15 year old male presented with progressive bilateral cervical lymphadenopathy for three months. Prior to admission he had been treated with multiple antibiotics and had a tonsillectomy, which did not improve his lymphadenopathy. He also complained of a dry cough that he noticed three days prior to admission. He denied sexual activity, exposure to cats, and recent travel. He did endorse recent exposure to prisoners and immigrants.

On examination, the patient was afebrile and had diffuse lymphadenopathy which consisted of bilateral anterior cervical lymphadenopathy measuring 7.5 x 5cm on the right and 6 x 5cm on the left. They were non-tender, fluctuant and soft. Bilateral posterior cervical lymphadenopathy was also noted and was tender to palpation. Non-tender right axillary and inguinal lymphadenopathy were also present. Mild splenomegaly was present on exam. The remainder of his exam was normal.

Laboratory evaluation included a CBC, CMP, EBV/CMV titers and Bartonella antibody titers and were normal. CT of the head and neck showed extensive lymph node enlargement throughout the cervical and supraclavicular regions bilaterally. Due to high suspicion of malignancy, a CT of the abdomen and pelvis was obtained. This also showed diffuse lymphadenopathy concerning...
for lymphoma. PPD was placed and was negative. Inguinal node biopsy was obtained and showed lymphoid hyperplasia. Acid fast staining of the lymph node was negative. Rapid HIV and confirmatory testing returned positive.

This case demonstrates the importance of considering HIV in the differential for long standing lymphadenopathy. Our patient presented with the diagnosis of chronic lymphadenitis but had never been tested for HIV because he denied all risk factors. It is imperative to include HIV in the differential in the adolescent population despite a negative HEADOSS test.

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HYPOKALEMIC PERIODIC PARALYSIS IN THE CONTEXT OF ALBUTEROL USE AND HYPERINSULINEMIA

Nortman J, Fogarty M, Hahn D. The University of Oklahoma Health Sciences Center, Department of Pediatrics, Oklahoma City, OK.

Case Report: A 10 year old female presented to the hospital with complaint of cough, wheezing, and bilateral lower extremity numbness and weakness. Parents report that her first wheezing episode occurred following an appendectomy six months prior to presentation. Since that time, she has had multiple admissions, during which she was treated with albuterol as well as oral and inhaled corticosteroids. Upon physical examination, lungs were clear to auscultation bilaterally and she exhibited no signs of respiratory distress, seven hours after her last albuterol treatment. Neurological exam was significant for decreased strength of upper and lower extremities and deficits of light touch sensation and temperature discrimination bilaterally. She exhibited a wide-based gait and required support to ambulate. Laboratory data was remarkable for hypokalemia (3.0 mEq/L), hyperglycemia (242 mg/dL), and euthyroidism. Later laboratory results revealed concomitant hyperinsulinemia (309 uU/mL). Within twenty four hours of symptom onset, hypokalemia resolved (3.6 mEq/L) and the patient returned to her baseline neurological function.

It has been shown that episodic muscle weakness in hypokalemic periodic paralysis may be induced by stress, exercise, or a high carbohydrate meal, all of which increase the release of epinephrine or insulin, which drive potassium intracellularity, decreasing plasma levels. Albuterol is known to have the same effect. This case illustrates a unique presentation of hypokalemic periodic paralysis whereby both albuterol and hyperinsulinemia may have contributed to hypokalemia, resulting in paralysis.

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ACUTE HEPATIC FAILURE IN A PREVIOUSLY HEALTHY CHILD

Neill L. UAB, Birmingham, AL.

Case Report: A (p.R69H) mutation in the TRPS1 gene. Our patient was discharged on DOL 2 and re-admitted on DOL 4 with hyperbilirubinemia, poor feeding, weight loss, and severe hypotonia. Laboratory work-up demonstrated elevated calcium (33.7-36.8 mg/dL) and PTH (867 pg/mL) levels.

The patient was transferred to CHONLA NICU. A skeletal survey demonstrated diffuse osteopenia. Renal and neck ultrasounds, CT scan of the neck, and a sestamibi scan were normal. Initial treatment included IV saline, furosemide, and bisphosphonate therapy. Cinacalcet, a calcimimetic agent, was trialed. Despite titration of cinacalcet to maximum dosages and additional bisphosphonate infusions, she required a total parathyroidectomy and has been managed with supplemental calcium postoperatively.

The oxime data previously obtained on the patient's parents was re-analyzed showing that both parents were heterozygotes for a mutation of the CASR gene: c.206G>A (p.R69H). Sanger sequencing confirmed our patient was a homozygote for the same mutation.

Medical treatment of NSHPT has been recently described but is not always successful. Success is related to specific receptor mutations. Often, these mutations reduce the sensitivity of the CASR to serum calcium leading to elevated PTH levels. In our patient's case, the c.206G>A (p.R69H) mutation may account for medical management failure. One other patient has been reported with the same homozygous mutation and has responded to cinacalcet only after parathyroidectomy. This patient was suspected to have miliary spread of the parathyroid cells and required maximum dosages of cinacalcet. This case demonstrates the critical role molecular testing has in guiding therapy and underscores the importance of reporting incidental oxime sequencing findings when both parents are heterozygotes for a variant causing severe neonatal disease.

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WHEN HERPES ISN'T HERPES: A CASE REPORT

Qasimyar H1, Belt E1, Lawrence H2. 1Oklahoma University Health Sciences Center, Oklahoma City, OK; 2The University of Oklahoma Health Sciences Center, Oklahoma City, OK.

Case Report: A 2 year old female presented with worsening vesiculopustular lesions to her head and neck. The lesions were clinically concerning for herpes simplex virus versus...
transient myeloproliferative disorder. After persistently negative serum, wound, and CSF studies as well as clinical stability otherwise, a skin biopsy was obtained. Histology confirmed features consistent with allergic contact dermatitis, a surprising finding given the patient’s age and the character of the lesions. Upon re-questioning, the child’s mother recalled applying a popular baby oil lotion only to the affected areas of skin prior to symptom onset. Although rare in this age group, allergic contact dermatitis should be included in the differential diagnosis for a young infant who presents with acute onset rash.

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RECURRENT LATE-ONSET GBS SEPSIS IN THE NICU
Rietsch C, Phillips J. University of Alabama at Birmingham, Birmingham, AL. Case Report: Group B streptococcus (Streptococcus agalactiae, or GBS) is a major pathogen in infants causing significant morbidity and mortality, particularly in those born prematurely. The incidence of early-onset GBS disease has been greatly decreased by screening mothers prior to birth and treating with intrapartum antibiotics. However, the incidence of late-onset GBS infection in the US has remained unchanged, at approximately 0.4 per 1000 live births since 1990. Late-onset GBS is often associated with serious illness and can be resistant to initial treatment. We report two cases of recurrent late-onset GBS infection in premature infants that were particularly difficult to manage. Initial presentations included increased apneas and bradycardic events and/or respiratory distress requiring positive pressure ventilation. One infant was born at 30 weeks gestation, was diagnosed with late-onset GBS sepsis on day of life 24 and completed a 14-day course of ampicillin. One week later, she developed apneic events which prompted an infection work-up that revealed GBS sepsis and meningitis. After a 21-day course with penicillin G, including 5 days of gentamicin for synergism, repeat CSF and blood cultures were negative. A second 26 week gestational age infant developed late-onset GBS meningitis at day of life 12 and completed 21 days of ampicillin. On the last day of treatment, CSF and blood cultures were obtained and had no growth. Subsequently, she developed recurrent GBS meningitis two weeks later and completed another 21 days of ampicillin. Repeat blood and CSF cultures were negative after the second course of treatment. This infant’s mother had received intrapartum ampicillin for preterm labor. Both infants had extensive immunologic evaluations and no defects were identified. The first infant had globally depressed immunoglobulins (lgG <200, IgM <25, IgE 4, IgA <6) during the first septic shock event. His CH50 was low at 47, likely secondary to inflammation and complement activation. He had normal distribution of T and B cells as well. The second infant had normal IgG (227) but low IgM (7) and IgA (4). AH50 was slightly low at 64 (normal 75-170), and she had normal mitogen stimulation of T cells. This report should alert clinicians to the possibility of recurrent late-onset GBS sepsis and/or meningitis in the convalescing preterm infant.

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INCOMPLETE KAWASAKI DISEASE IN A 5 MONTH OLD
Rojas Gallegos MB, Gremse D, Custudio H. University of South Alabama, Mobile, AL. Case Report: Gastrointestinal symptoms of abdominal pain, vomiting and/or diarrhea occur in association with Kawasaki disease (KD) and usually are secondary to the main clinical diagnostic criteria. ‘Atypical KD’ is more common in children <6 months or >5 years of age, although their risk of developing coronary arteritis higher. We present a case of an infant with KD presenting with predominantly fever and diarrhea. Case: A 5 month old African-American female presented with 3 days of diarrhea (5-6 stools per day) and fever. White blood cell count was 14.8 k/cL, CRP 18 mg/dL, ESR 64 mm/hr, sodium 134 mEq/dL, and albumin 2.3 g/dL. Uricanalysis was positive for leukocytes and nitrates but urine culture was negative. Blood culture and lumbar tap were negative. Clostridium difficile toxins were detected in the stool. Despite use of intravenous ceftriaxone and oral metronidazole, she continued to have fever. She had extreme irritability and developed dryness and reddening of the lips and buccal mucosa, as well as edema of her face, hands and feet. A diffuse erythematous maculopapular eruption in her diaper area as well as conjunctival injections appeared. Progressive abdominal distention was noted and ultrasound showed nonspecific ascites. Echocardiogram showed a 2-3mm hemodynamically insignificant posterior pericardial fluid collection without evidence of coronary aneurysms. The diagnosis of KD was made on day 7 and high dose immunoglobulin was given along with high dose aspirin. She dramatically improved with complete resolution of fever and diarrhea, and was sent home on low dose aspirin 48 hours later.

Conclusion Although our patient did not meet diagnostic criteria for KD on admission, the diagnosis became apparent during the course of the illness. The constellation of symptoms of diarrhea, diaper rash, and edema from low albumin may be seen in gastroenteritis. However, the persistent fever, and evolution of clinical and laboratory findings pointed to the diagnosis. We conclude that Kawasaki disease should be considered in the differential diagnosis of infants with gastrointestinal symptoms such as diarrhea with fever.

FIGURE 1. Peripheral blood smear showing monocytosis, nucleated red blood cells, and thrombocytopenia.
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UNILATERAL ECTOPIC TESTIS: A RARE CONGENITAL GENITOURINARY ANOMALY

Shah N, Barche A, Nirgiotis J. Texas Tech University Health Sciences Center, Amarillo, TX.

Case Report: A term large for gestational age male was born to a 23-year-old G2 P1 A1 L1 mother at 40 weeks of gestation via spontaneous vaginal delivery. His birth weight was 4015 grams and Apgar scores at 1 and 5 minutes were 9. Mother had an event free pregnancy with normal prenatal laboratory tests. There was no family history of any genitourinary disorders.

On physical exam, patient had a 4 x 3 cm swelling in the left perineal region with loss of volume in ipsilateral scrotal sac, while the right testis was palpable in right scrotal sac. Rest of the exam at birth was normal for age. He was followed up regularly by pediatric surgeon and a left orchidopexy was performed at 6 months of age under general anesthesia. Left testicle was mobilized at inguinal ring and then gently brought down towards scrotum. A new scrotal sac was created on left side, the testicle was fixed to the wall, and skin was closed at incision sites. Patient tolerated the surgery well and was discharged on the same day.

Discussion: The testes develop at 7-8 weeks of gestation and its descent is regulated by mechanical and hormonal factors. Absence of testes from its normal location is the most common congenital genitourinary anomaly, seen in 0.2% - 1% of men. It may be due to undescended testes, arrested descent of testes in its normal pathway or due to ectopic testes. Ectopic testes are seen only in about 5% of absent testes cases, making it extremely rare. The exact etiology and pathogenesis of ectopic testes still remain a controversy. Ectopic testes are prone to trauma, testicular torsion, infertility, epididymo-orchitis and malignancy.

A thorough physical exam is paramount and absence of testes in its normal location should involve examination of ectopic testes sites. The timing of surgery should be based on preserving normal physiological function, preventing complications from the condition itself and anesthesia considerations.

FIGURE 1. 1) At birth, 2) At 6 months of age (prior to surgery).

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46,XY,t(1;15)(Q24.2;Q13): PHENOTYPIC IN A PEDIATRIC PATIENT

Sheth S, Thakore PS, Vasylyeva T. Texas Tech University Health Sciences Center, Amarillo, TX.

Case Report: A 2-year-old boy presented with a 3-months history of non-bilious and non-bloody vomiting associated with watery and foul smelling diarrhea. He also had a history of poor appetite and significant weight loss of 20 pounds since last 3 months. He had significant past medical history of congenital lung hypoplasia, hip dysplasia, hearing defect, posterior urethral valve and polycystic kidney disease. He underwent renal transplant 5 years ago and was on immunosuppressive therapy (Prograf, CellCept, and Prednisone).

On admission, he had watery diarrhea following any oral intake. His complete blood counts, complete metabolic panel, urine analysis, tissue transglutaminase immunoglobulin A, zinc level, vitamin A, vitamin D, vitamin E, total and free carnitine, and stool studies for cryptosporidium and giardia, 72 hours fecal fat, reducing substance, pancreatic lipase were normal. Concerning for his weight loss, free T4, thyroid stimulating hormone, Epstein Barr virus, BK virus, and HIV test were performed with the negative finding. Chest computed tomography and ultrasound of abdomen were also normal. Patient continued to have loose, watery and foul smelling diarrhea till pancreatic enzymes were added to the treatment. After initiation and dose adjustments of the pancreatic enzymes, the patient showed dramatic improvement with normal stool and weight gain.

The constellation of his symptoms promoted the interest to look into his past genetic studies. His genetic study had showed: 46,XY,t(1;15)(q24.2;q13), indicating an abnormal male karyotype with an apparently balanced translocation involving the long arm of one chromosome 1 and the long arm of one chromosome 15 with a breakpoint. This mutation has not been reported in the past. The patient’s parental genetic studies were normal indicating this chromosomal abnormality developed as a de novo cytogenetic event.

Discussion: Phenotypical features of 46,XY,t(1;15)(q24.2;q13) include association of lung hypoplasia, hip dysplasia, hearing defect, posterior urethral valve, polycystic kidney disease and pancreatic insufficiency in our patient. Further studies might identify precise molecular pathways leading to these linked characteristics.

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COMPARING PRIOR RADIOGRAPHS AIDS IN THE DIAGNOSIS OF BRONCHIECTASIS

Sheth S1, Kanu A2, Turner C1. 1Texas Tech University Health Sciences Center, Amarillo, TX and 2Texas Tech University Health Sciences Center, Lubbock, TX.

Case Report: A 2-year-old female presented with one-day history of high-grade fever, cough and rhinorrhea. She was lethargic and had decreased oral intake. There were no sick contacts or choking episodes. On admission, she was febrile with 104F, tachycardic, tachypneic with moderate respiratory distress. Physical exam revealed bilateral inspiratory wheezes with decreased air entry on right upper chest. She was hospitalized 3 times in the past for presumptive aspiration pneumonia beginning at 1-month of age. At 15-months of age, modified barium swallow study showed severe aspiration and so she underwent a gastrostomy tube placement. Home medicines included albuterol as needed and budesonide twice a day. Parenteral clindamycin was started for presumptive aspiration pneumonia of the right upper lobe. However, all her prior radiographs were reviewed and a computed tomography of the chest was performed, which unveiled bronchiectasis. Further workup revealed normal quantitative sweat test and immune studies.

Bronchiectasis is injury to bronchi resulting in dilatation. Chronic aspiration pneumonia is a frequent cause of bronchiectasis. High resolution computed tomography (HRCT) is the most sensitive imaging method for the diagnosis. The initial management in aspiration related bronchiectasis is to diagnose and treat the cause of aspiration. HRCT should be considered earlier when there is a clinical suspicion of aspiration especially on chest x-rays. Comparing the previous chest x-rays can aid in the early identification of bronchiectasis and alters the management and prognosis. Omitting comparison of prior radiographs may delay the diagnosis of bronchiectasis.

FIGURE 1. A, B, C) Chest Xray, D) HRCT.
THORACOSCHISIS ASSOCIATED WITH DIAPHRAGMATIC EVENTRATION
Travers C1, Klima D2, Huber J1, Chen M2, Anderson S2, Philips J1
1University of Alabama at Birmingham, Birmingham, AL; 2Children’s of Alabama, Birmingham, AL
Case Report: A female infant was delivered at 39 weeks by spontaneous vaginal delivery weighing 2665g. Mother was a 24 year old para 3, term 1, preterm 2. She had a previous child with dilated cardiomyopathy who died of pneumonia at 18 months. Prenatal medications included escitalopram (discontinued in the first trimester), trazodone and topiramate (discontinued in the second trimester), oxytocin/acetaminophen and prenatal multivitamins. An anomaly scan at 20 weeks gestation was normal. At birth she was noted to have tissue protruding from a left sided thoracic defect (stills and movies will be presented). This tissue was red/purple color, 3cm long, and tubular in appearance. There was an accessory nipple on the left side of the chest. The defect was situated between the two nipples. She required oxygen via nasal cannula. There were no other dysmorphic features or congenital anomalies noted on physical exam. The stomach bubble was displaced superiorly on radiographs. At surgery she had a left sided congenital diaphragmatic eventration. The diaphragm was intact with a superiorly displaced attachment to the anterior costal margin. The left lateral segment of the liver and omentum appeared adherent to the diaphragm. There was tissue protruding through an intercostal defect below the diaphragm. This tubular tissue was attached to the omentum and to the superior extent of the left lateral lobe of the liver. There was medial rib aplasia of the two overlying ribs. The tissue was resected and the defect closed. The post-operative course was uneventful. Tissue pathology report is currently pending. Echocardiogram showed a moderate sized patent ductus arteriosus, patent foramen ovale, and dextroposition of the heart. She was discharged home on day 6 on full feeds, breathing room air with no support. This is the first case report of thoracoschisis associated with diaphragmatic eventration. Thoracoschisis is an extremely rare congenital anomaly. It has been associated with diaphragmatic hernia and limb-body wall complex. Limb abnormalities were not present in this infant. The pathogenesis of thoracoschisis is not fully understood.

LATE-ONSET POSTOPERATIVE PROPIONIBACTERIUM ACNES SPINE INFECTION IN ADOLESCENTS
Wafadari D1, Estrada B1, Nimityongskul P2, Custodio H1
1University of South Alabama, Mobile, AL and 2University of South Alabama, Mobile, AL
Case Report: We present 2 cases to highlight Propionibacterium acnes (P. acnes) as a cause of spine infection among adolescents with hardware and the difficulty in establishing its role as a pathogen.

Case 1
A 14-year-old female with scoliosis had posterior spinal fusion and instrumentation. She presented 18 months later with a 1 week history of painless swelling at the incisional site. Aspiration of the swelling was done and culture was negative after 5 days of incubation. Intraoperatively, purulent material was noted and the hardware was removed. Wound vacuum-assisted closure (VAC) and incision and drainage (I & D) were performed. All 8 cultures sent in 5 days grew P. acnes after 5-7 days of incubation.

Case 2
A 17-year-old female with history of scoliosis, status-post posterior spinal internal fixation 3 years ago, presented with 3 weeks of pain and discharge from the surgical site. She took trimethoprim-sulfamethoxazole with no improvement. As in the first case, removal of the hardware, wound VAC placement and repeated I & D were performed. Out of 7 cultures in 3 days, 5 cultures grew P. acnes at 7-9 days post-incubation.

Both patients received Clindamycin for 4 weeks and remained infection free at 1 year visit.
AN ADOLESCENT PATIENT PRESENTS WITH PYODERMA GANGLRENSUM AS AN EXTRANTESTINAL MANIFESTATION OF CROHN’S DISEASE
Wisner E, Sandlin C. LSUHSC New Orleans, New Orleans, LA.

**Case Report:** Pyoderma gangrenosum is a neutrophilic dermatosis associated with systemic disease in up to 50% of patients. Inflammatory bowel disease (IBD) is associated with extraintestinal manifestations, and pyoderma gangrenosum is observed in only 0.5 to 5% of such patients. Only 4% of patients with pyoderma gangrenosum are younger than 15 years at diagnosis. Here we describe the case of an adolescent patient presenting with pyoderma gangrenosum as a cutaneous manifestation of Crohn’s disease.

A 14-year-old female with a past history of chronic anemia and eczema presented with a one month history of bilateral anterior leg lesions described as erythematous nodules, which evolved into draining and ulcerating lesions. She reported having similar lesions in the past treated with systemic antibiotics for presumed cellulitis. Two weeks prior to admission, oral prednisone resulted in slight improvement of the skin ulcers. The patient also endorsed intermittent joint pain and swelling as well as several watery bowel movements daily. She denied fever, oral lesions, night sweats, and weight loss. Prior rheumatologic work-up revealed a positive ANCA (1:80), although a thorough evaluation for systemic vasculitis was negative.

On physical exam, the left leg contained a 12x14 cm hypopigmented area with multiple painful, punched-out healing ulcers ranging from 3-4 cm. The right leg contained similar skin findings as well as a 3x2.5 cm nontender shin nodule. The ulcerations were erythematous with purulent, bloody drainage. Pertinent laboratory evaluation revealed anemia of chronic disease, hemocult positive stool, ESR 79, CRP 11, and negative wound cultures. A skin biopsy was consistent with pyoderma gangrenosum. Colonoscopy confirmed the diagnosis of Crohn’s disease. The patient was started on IV steroids and infliximab. At follow-up one week after discharge, the patient showed considerable improvement.

Although rare, neutrophilic dermatoses should be considered when presumed cellulitis does not improve with systemic antibiotics. While PG can be an isolated finding, roughly half of cases are associated with systemic disease. This case highlights the association between extraintestinal manifestations and IBD and should urge the practitioner to maintain a high index of suspicion.

**Discussion**

The role of *P. acnes* as a pathogen among adolescents with spine manipulation and hardware placement is under recognized and establishing true infection can be challenging. *Propionibacteria* are anaerobic bacilli and part of the skin flora, so their isolation is often dismissed as contamination. Since they are slow growing and may require up to 19 days of incubation, absence of growth after routine 5 days of incubation may be misleading. Also, clinical and laboratory features of infections due to *P. acnes* are nonspecific. Nevertheless, due to its low virulence and slow growth, *P. acnes* infection should be suspected in cases of delayed onset of symptoms ranging from months to years, and clinicians should send multiple specimens and request prolonged incubation to establish true infection. As *P. acnes* form biofilm, removal of the hardware and debridement of tissues are vital in the management. Penicillin, tetracyclines, clindamycin and vancomycin are often used, usually for 4 weeks.

**Case Report:** A 6 month old female was admitted with the chief complaint of worsening eczema-like scaly rash for 2 weeks. She had a history of intolerance to multiple cow’s milk formulas including partially hydrolyzed ones. As a result of restricting her diet, she was being fed a regimen of rice cereal, banana based baby food and apple juice with occasional fruit. She was maintained on this regimen for months while being followed by her pediatrician as well as being seen at local WIC offices. She also had progressive swelling in her distal extremities.

On physical examination, her weight was 2nd %ile, height was < 2nd %ile and weight-for-height was 98th %ile. There was an erythematous, blanching, scaly, patchy and in certain areas papular rash predominantly on lower extremities and trunk without crusting or punch out lesions. There was significant generalized edema most prominent on the lower extremities. Her hair was sparse and light reddish. She had a moon face appearance.

The lab work revealed elevated transaminases, decreased levels of serum albumin, protein, prealbumin, BUN and zinc. Patient had a normocytic anemia. Analysis of urine organic acid showed elevated levels of pyruvic acid, fumaric acid, 2-ketoglutaric acid, 4-OH phenyllactic acid and 4-OH phenylpyruvic acid. Cow’s milk protein IgE level was 0.63 kU/L (Normal: < 0.34 kU/L). Patient was diagnosed with cow’s milk protein allergy and started on amino acid based formula after which she showed clinical improvement.

Kwashiorkor is a type of protein-energy malnutrition mainly seen in developing countries. There are only 19 cases with kwashiorkor reported over the past 30 years in US due to maladaptive nutritional practices such as nutritional ignorance, food faddism or food allergen avoidance. Physicians should consider nutritional deficiencies and should take a thorough social and dietary history particularly in patients who present with refractory dermatitis. Additionally, milk alternative beverages should carry more prominent warning label as to their inappropriateness to serve as food source for children.
**Perinatal Medicine**

Joint Plenary Poster Session and Reception

5:00 PM

Thursday, February 26, 2015

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**IMPORTANCE OF MULTIPLE SITE SWABS FOR DIAGNOSING NEONATAL HERPES**

Aboaziza A, Macarida D, Shah D, Devoe M, Gibson J. East Tennessee State University, Johnson City, TN.

**Case Report:** A 4-day-old female term infant was admitted for neonatal fever. Patient had struggled with breastfeeding yet otherwise asymptomatic. While at Primary Care Provider (PCP) for a routine newborn care she was found to have a temperature of 102.5 °F. The PCP sent her to a nearby ER facility, and then was transferred for further management & evaluation.

Mother received intrapartum antibiotic prophylaxis for positive GBS status. Birth weight was 3430 grams. Dad with close contact to this newborn had oral blisters few days before her symptoms.

Initial laboratory tests included Sodium of 154mEq/L, BUN 41mg/dl, Creatinine 1.3mg/dl, Glucose 45 gm/dl, WBC of 14,300 [68% neutrophils; 27% lymphocytes], hemoglobin 17.1 gm/dl, hematocrit 49.6 %, and platelet count 144,000. CSF analysis revealed RBC of 11, WBC of 2, glucose 40 gm/dl, & protein 52.

Pertinent Physical Exam (PE) findings on admission include; vital signs: blood pressure of 81/36 mm/Hg, heart rate 156 bpm, respiratory rate 66 bpm, oxygen saturation 100% on 2 L/minute nasal cannula, temperature 102.6 °F, weight of 2.9 kg. Anterior fontanel was sunken. She had no skin or oro-pharyngeal lesions. A complete PE was otherwise unremarkable.

She was treated initially with intravenous ampicillin, cefotaxime, vancomycin & acyclovir. She developed multiple episodes of apnea & seizures. A complete head CT was normal. She had oral blisters few days before her symptoms. She was treated with intravenous ampicillin, cefotaxime, vancomycin & acyclovir. She developed multiple episodes of apnea & seizures along with lower body & respiratory viral panel was negative. Rectal & CSF PCR were negative while Nasal swab PCR was positive for HSV. Her status gradually improved as she completed a 21-day course of Acyclovir.

Neonatal herpes occurs in 1 in every 3,000 to 20,000 live births affecting 1,500 to 2,000 infants per year in the United States. Neonatal herpes infection is usually an infection from the mother. Father in this case was the possible source of infection. Our patient despite having severe infections had no skin or mucous membranes lesions. Typically, Neonatal HSV CNS infection presents with positive CSF PCR. In our case, CSF PCR was negative despite elevated CSF RBC & pleocytosis. We would like to highlight the peculiar presentation of HSV infection, & emphasize the need to perform viral studies from different sites such as the naso PCR.

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**A STUDY OF WITHDRAWAL OF UMBILICAL CATHETER AND ITS ASSOCIATION WITH SEPSIS IN VERY LOW BIRTH WEIGHT INFANTS**

Ali M, Eyal F. University of South Alabama, Mobile, AL.

**Purpose of Study:** Umbilical lines are widely used in the care of premature and ill infants. Catheter related infections are common in these infants. Our objective is to study whether there is an association between umbilical catheter withdrawal and sepsis; and if the factors which may impact the same, blood pressure of 81.36 mm/Hg, heart rate 156 bpm, respiratory rate 66 bpm, oxygen saturation 100% on 2 L/minute nasal cannula, temperature 102.6 °F, weight of 2.9 kg. Anterior fontanel was sunken. She had no skin or oro-pharyngeal lesions. A complete PE was otherwise unremarkable.

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**Summary of Results:** During 1990 to 2013, 3459 babies were identified as having received a UAC; all of these infants were given antibiotics. Of the total babies, 2576 (74%) did not develop sepsis post removal of UAC. The remaining 883 babies were divided into three groups for the purpose of the study; the first being, sepsis before UAC removal or within 1 day of removal, these were 8% of the total; the second group consisted of babies with sepsis more than 4 days after removal of UAC, 15% of the total; the final group was babies who developed sepsis within 2 - 4 days of removal of UAC, these constituted 2% of the total. A logistic regression considering the factors affecting sepsis in group 3 was run; the variables taken into account were discontinuation of antibiotic before or at the time of removal of UAC, the duration of UAC and the gestational age (GA). The discontinuation of antibiotic and the gestational age were found to be statistically significant as seen in Table

**Conclusions:** In babies who developed sepsis within 2-4 days of UAC removal, it was found that there was an association between the discontinuation of antibiotics and development of sepsis. Since this group represents only 2% of the total, hence we do not recommend prophylactic antibiotic post UAC removal. However, a close follow up of these babies is recommended.

**Table**

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<th>Sepsis within 2-4 days of UAC removal</th>
<th>Odds Ratio</th>
<th>Std Error</th>
<th>Z</th>
<th>P</th>
<th>95% CI</th>
</tr>
</thead>
<tbody>
<tr>
<td>Antibiotics stopped before UAC removal</td>
<td>2.18</td>
<td>0.82</td>
<td>2.06</td>
<td>0.039</td>
<td>1.059-4.5591</td>
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</tbody>
</table>

**Duration of UAC**

- 0.95
- 0.04
- 1.18
- 0.168
- 0.8787-1.0227

**Gestational Age**

- 0.75
- 0.07
- 3.15
- 0.0022
- 0.6262-0.8966

**Constant**

- 142.1039
- 355.28
- 2.1
- 0.036
- 1.3955-14475.14

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**INHALED PROSTACYCLIN TO IMPROVE PULMONARY HYPERTENSION REFRAC TORY TO NITRIC OXIDE**

Bashamba MT, Bhatia J. Georgia Regents University/MCG, Augusta, GA.

**Case Report:** Pulmonary hypertension is refractory to nitric oxide in 40% of NICU patients. We report the use of inhaled epoprostenol sodium in a full-term infant with left-sided diaphragmatic hernia who developed severe pulmonary hypertension subsequent to ECMO. By using iPGI2, we were able to wean NO and significantly reduce oxygen support.

Infant was born at 39 weeks completed gestation via scheduled C-section for left-sided congenital diaphragmatic hernia at the Children's Hospital of Georgia. Despite high-frequency oscillation and NO, the infant continued to deteriorate. Echocardiogram confirmed severe PPHN prompting cannulation for veno-venous ECMO at 5 hours of age. After multiple attempts to wean ECMO support, infant underwent surgery on day of life 22 followed by discontinuation of ECMO support by post-operative day 1. However, despite minimal ventilator support, extubation attempts on day of life 33, 45, and 49, were unsuccessful. Sildenafil was initiated on day of life 42 followed by NO due to persistent need for 100% oxygen. Multiple attempts to wean NO were unsuccessful. On day of life 64, after obtaining informed parental consent, infant was placed on continuous iPGI2 at a dose of 30 ng/kg/minute by aerosolizing the intravenous formulation of epoprostenol sodium. By 12 hours after initiation, NO was discontinued and the FiO2 decreased from 100% to 40% by 24 hours, to 25%. However, 3 days after the iPGI2 was discontinued, infant’s oxygen requirement returned to 50% and by 4 days to 100%. iNO was restarted at 20 ppm but infant was unable to be weaned from 100% oxygen. At this time, a discussion was held with the family deciding to withdraw support for the infant.

We used epoprostenol sodium in an infant refractory to NO and sildenafil to significantly reduce FiO2. Although there is limited information on the effectiveness and safety of prostacyclin in infants, we did not observe any significant adverse effects in our patient. Though our patient received iPGI2 after ECMO and repair, the indication for its use to avoid ECMO particularly in those infants with less extensive lung disease and who do not respond to NO is reasonable. When cost is considered, iPGI2 offers a low cost alternative to NO particularly in resource-limited settings.
APLASIA CUTIS CONGENITA ASSOCIATED WITH FETUS PAPYRACEOUS: A CASE REPORT

Bidot L, Dankhara N, Shah D. ETSU, Johnson City, TN.

Case Report: 30 weeks preterm female was transferred to NICU soon after delivery due to prematurity, respiratory distress and symmetrical skin abnormality on anterior abdominal wall noted at birth. Mother is a 34-year-old, G2P1, Caucasian female, who presented with imminent delivery and cord prolapsed.

After emergent C-section, infant was delivered, Apgar scores were 2, 4 & 6 at 1, 5 & 10 minutes of life. Resuscitation required included positive pressure ventilation, intubation and 2 doses of epinephrine. Due to persistent respiratory distress, transport team intubated again and gave a dose of surfactant.

Normal saline bolus was given followed by IV antibiotics. Birth weight was 1,191 grams.

Pregnancy was complicated by fetal demise of one of the twin pregnancies at 14 weeks of gestation. Mother denied smoking, alcohol and other drugs. Mother took prenatal vitamins. Maternal labs were negative. No known family history of similar condition.

Examination revealed large bilateral symmetrical, almost mirror image, skin defects involving lateral aspects of her trunk. The affected areas were covered with a transparent membrane through which the underlying blood vessels were clearly seen.

Plastic surgeon was consulted and patient was managed conservatively with occlusive petrolatum gauze and antiseptic ointment to prevent dryness and infection.

Aplasia Cutis Congenita (ACC) also known as congenital cutis aplasia is a rare disorder that affects bone and skin. ACC is defined as localized absence of (normal) skin at the time of birth. Skin appears thinner and underlying structures are visible. Commonly, it manifests as a solitary defect on the scalp, but it may occur as multiple lesions and other than scalp area as in our case.

Based on classification suggested in recent literature this case would fall under group five which is aplasia cutis congenita associated with fetus papyraceous or placental infarct. Disseminated intravascular coagulation may be the cause of fetal demise and may cause selective hypoperfusion of mesodermal tissue of the skin and trunk.

Management varies depending upon location and depth of skin involvement. Superficial involvement may heal well with conservative treatment. Deep lesions may require skin grafting or reconstructive surgery.

DETECTION RATE OF FETAL TRICUSPID REGURGITATION IN NEW VERSUS OLD ULTRASOUND TECHNOLOGY AND ITS ASSOCIATION WITH OTHER HEART ABNORMALITIES

Deighan T1, Smith M1, Samples S2, Lutin W2, Hiles H1. 1Medical College of Georgia, Georgia Regents University, Augusta, GA; and 2Medical College of Georgia, Georgia Regents University, Augusta, GA.

Purpose of Study: Pediatric cardiologists use several tools to evaluate and predict the risk of prenatal heart abnormalities. One tool used is reviewing fetal echocardiograms for the presence of tricuspid regurgitation (TR). Conventional wisdom has been that the presence of TR is a strong indicator of other fetal heart abnormalities. However, advancements in ultrasound technology have brought this theory into question.

Methods Used: Pediatric cardiologists use several tools to evaluate and predict the risk of prenatal heart abnormalities. One tool used is reviewing fetal echocardiograms for the presence of tricuspid regurgitation (TR). Conventional wisdom has been that the presence of TR is a strong indicator of other fetal heart abnormalities. However, advancements in ultrasound technology have brought this theory into question. Predicting that TR detection is higher today due to technological advances, the aim of this study was to quantify and compare the detection rate of TR in old (e.g. Philips IE33 software 4.02.13) and new (e.g. Philips IE33 software update 6.3.3.134) echocardiographic technology. A secondary aim of this study was to quantify detected TR on a severity scale and to assess the association between TR severity and the presence of other heart abnormalities.

Summary of Results: Biostatistical analysis demonstrated that TR detection rates were significantly higher in newer technology than in older technology (p=0.0118, OR 1.72). Analysis also demonstrated that patients with TR had a significantly higher likelihood (p=0.0001, OR 2.6) of having other heart abnormalities such as pericardial effusion, restrictive ductus arteriosus, and right ventricular hypertrophy than those without TR. There was no correlation between higher TR severity and heart abnormality (p=0.068).

Conclusions: With improved technology, detection of TR is increasing. In addition, TR has been shown to be linked to other heart abnormalities. This knowledge will be useful for obstetricians and pediatricians for understanding the significance of prenatal tricuspid regurgitation.

PROSTAGLANDIN E2 AND METABOLITE LEVELS IN PRETERM INFANTS BEFORE AND AFTER ADMINISTRATION OF INDOMETHACIN

Fischer HR1, Matheson PD2, Walker SK2, Garrison RN2, Downard CD2. 1University of Louisville, Louisville, KY; and 2University of Louisville, Louisville, KY.

Purpose of Study: Indomethacin is administered to preterm infants to induce closure of a patent ductus arteriosus (PDA), and has been associated with intestinal complications such as necrotizing enterocolitis (NEC) and intestinal perforation. Prostaglandin E2 (PGE2) is produced in the gastrointestinal tract and functions to maintain the integrity of the gastrointestinal lining and regulate mucosal blood flow. In an established animal model of NEC, application of PGE2 receptor agonists and antagonists resulted in altered ileal blood flow, suggesting that inhibition of PGE2 formation by indomethacin may alter intestinal blood flow in preterm infants leading to intestinal complications. Our study was designed to evaluate the relationship between indomethacin administration and the serum levels of PGE2 and its metabolites in preterm infants.

Methods Used: Infants were eligible if they had a PDA amenable to indomethacin treatment, weighed greater than 650 grams on the day of enrollment, and had no prior exposure to indomethacin or ibuprofen. Serum samples were drawn before and after the first dose of indomethacin. Serum PGE2 and metabolite levels were determined by ELISA. Results were analyzed using paired t-test and 2-way ANOVA with Tukey test.

Summary of Results: This study evaluated ten infants, and found a statistically significant difference in mean (± SEM) serum levels of PGE2 (708.1 ± 117.4 vs. 552.7 ± 102.1 pg/mL, p-value 0.026) and PGE2 metabolite (710.2 ± 104.2 vs. 572.9 ± 96.7 pg/mL, p-value < 0.007) before and after administration of indomethacin. The magnitude of change for individual patient PGE2 and metabolite levels ranged from a 27% increase to an 86% decrease. No infants in this study developed intestinal complications.

Conclusions: This study showed a significant decrease in both PGE2 and metabolite levels after indomethacin administration in this group of preterm infants. There was variability in baseline levels and in the magnitude of change after indomethacin administration. Further studies with a larger population of infants will determine if there is an increased risk of intestinal complications in preterm infants correlated with PGE2 levels after indomethacin administration.

CONGENITAL CMV INFECTION WITH INTRAPULMONARY CALCIFICATIONS ON PRENATAL SONOGRAM

Fort P1, Chang M1, Boppana SB1, Davis R2, Owen J1, Carlo W1, Philips J1. 1University of Alabama at Birmingham, Birmingham, AL and 2University of Alabama at Birmingham, Birmingham, AL.

Case Report: A 19 yo African-American woman was referred for a detailed sonogram at 31 wks due to a left club foot and prominent stomach. The club foot was confirmed but mild intracranial ventriculomegaly and
intracranial and pulmonary calcifications were also seen. Aneclopyrid screening was negative. CMV IgG was positive and IgM was negative. Scan 4 weeks later showed poor interval growth with an estimated fetal weight <5th percentile. At 37 weeks, a biophysical profile was ≥10 and labor was induced. Delivery was vaginal and Apgar scores were 4 and 8. The infant was hypotonic with minimal respiratory drive. She required bag-mask ventilation for ~ one minute and was transferred to the NICU. Birth weight was 2.46 kg (12th %tile), head circumference was 29.5 cm (1st %tile) and length was 44 cm (3rd %tile). Physical examination showed mild respiratory distress, a palpable liver edge, and mildly decreased tone. Chest x-ray showed low lung volumes with interstitial densities. The infant received ampicillin and gentamicin. Congenital CMV infection was confirmed with a positive saliva rapid culture. Cranial US showed immature gyral and sulcal pattern with calcifications, prominent third and lateral ventricles, and prominent cystic space in the posterior fossa. MRI revealed microcephaly, simplified gyral pattern, cerebro calcifications, and dilatation of third and lateral ventricles with a small posterior fossa arachnoid cyst. There was no evidence of CMV retinitis. Auditory brainstem response was normal. The placenta weighed 336g and had signs of chronic villitis. Imaging showed placenta (low weight for gestation, lymphophasmacytic villitis, villous sclerosis and hemosiderin with scattered dystrophic mineralization). Valgancyclovir was started on day 3. The infant had persistent respiratory distress requiring oxygen since birth to maintain saturations above 90%, consistent with CMV pneumonia. The infant was sent home on day 17 on valgancyclovir and a nasal cannula at 0.1 L/min and 100% FiO2. On one-month follow up visit, she continued to require oxygen. To our knowledge, this is the first case report of a live-born infant with pulmonary calcifications due to CMV noted on prenatal sonogram.

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RANDOMIZED TRIAL OF EARLY ORAL FEEDING IN VERY PREMATURE INFANTS
Gerges A, Kennedy K. University of Texas Health Science Center, Houston, TX.

Purpose of Study: To determine if earlier initiation of oral feeding attempts in infants born at <29 weeks gestation leads to earlier attainment of full oral feedings and/or hospital discharge.

Methods Used:
Design: Randomized clinical trial.
Inclusion Criteria: Infants born at <29 weeks gestation and tolerating enteral feedings.
Exclusion Criteria: Respiratory contraindications to oral feedings (i.e. PPV, CPAP, persistent tachypnea) at the time of randomization, craniofacial anomalies, defects, neuromuscular or genetic disorders, CV disease on medications for CHF, congenital GI malformations or history of GI surgery.

Intervention: Eligible infants were enrolled and randomized at 30 weeks postmenstrual age (PMA). The Earlier Oral Feeding group initiated oral feedings at 30 weeks PMA. The Later Oral Feeding group initiated oral feedings at 33 weeks PMA. Oral feeding attempts were advanced according to an Oral Feeding Progression Algorithm for both groups. Breastfeeding attempts were also incorporated into the algorithm. All infants were monitored for adverse events during oral feedings.

Analysis: Our primary outcomes are time to full oral feedings and time to hospital discharge. We will perform an intention-to-treat analysis on all enrolled patients. A non-parametric analysis will be performed on all enrolled patients so infants who never achieve full oral feedings prior to discharge will be included in the analysis.

Summary of Results: To date, 40 infants have been randomized and 35 discharged. 80% of eligible infants have been enrolled. 0 patients have withdrawn after randomization. We are continuing to actively recruit patients until enrollment ends on 12/31/14. Most (or all) outcome data will be available at the time of the meeting. Preliminary data and secondary outcomes are outlined in the table below.

Conclusions: If our hypothesis is confirmed, we would conclude that clinicians should consider initiating oral feedings in premature infants at an earlier PMA than is typical in current practice.

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MINING BEDSIDE DATA FOR EFFECTS OF CAFFEINE ON HEART RATE VARIABILITY IN PRETERM INFANTS
Gill S1, Duncan S1, Radmacher PG1, Wang Y4, Sokhadze E2, Bosley K1, Mellen N.1 University of Louisville School of Medicine, Louisville, KY; 2University of Louisville School of Medicine, Louisville, KY; 3Kosair Children’s Hospital, Louisville, KY and 4University of Louisville School of Medicine, Louisville, KY.

Purpose of Study: Apnea of prematurity has been observed in up to 70% of infants < 34 wks gestation and is associated with neurodevelopmental impairment. Caffeine is administered prophylactically to improve respiratory function, reducing apnea of prematurity. Caffeine inhibits purinergic receptors, which are ubiquitous in the cardiovascular, autonomic and central nervous systems, yet its effect on the autonomic system in preterm infants has not been studied. Well being can be determined by heart rate variability and is reflected both by parasympathetic and sympathetic drive, which is detectable as high and low frequency variability.

Aim: To test whether caffeine modulates heart rate variability in preterm infants via an increase in sympathetic drive.

Methods Used: Eligible infants were ≥32 wks gestational age, 3-14 days of age, receiving IV caffeine at maintenance dosage (5-10 mg/kg body weight IV q 24 hrs). Cardiorespiratory signals were downloaded from Intellivue monitors via RS 232 port using ixTrend software for 1 hr prior to, during and 3 hrs after drug administration.

Demographic data included birth weight, gestational age at birth, sex, postnatal age and weight at time of recording.

Clinical data included time/dose of drug administration.

Summary of Results: 25 subjects were enrolled; data from 5 subjects were analyzed. Ten subjects were dropped from the study due to changes in caffeine dosing. Time series of respiratory activity and heartbeat were obtained in the form of successive respiratory cycle periods and heart beat-to-beat intervals. One way ANOVAs statistical analysis was used to study the effects of caffeine on heart rate variability modulation in preterm infants who were given caffeine for apnea of prematurity. Caffeine was found to have significant increases in heart rate directly. There was also a near-significant decrease in low frequency heart rate variability, consistent with a decrease in sympathetic drive.

Conclusions: Caffeine’s effects on heart rate and respiratory drive is via direct effects on CNS and cardiac purinergic receptors, rather than via up regulation of sympathetic drive.

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ASSOCIATION OF EXCLUSIVE HUMAN MILK DIETS (EHM) USING COMMERCIAL DONOR HUMAN MILK (CDHM) WITH REDUCED ESTIMATED HOSPITAL CHARGES FOR NECROTIZING ENTEROCOLITIS (NEC) AND LATE-ONSET SEPSIS (LOS)
Huff ML, Shattuck K. University of Texas Medical Branch, Galveston, TX.

Purpose of Study: EHM diets have been shown to reduce the incidence and hospital costs of NEC and LOS in very low birth weight (VLBW) neonates. A
2012 analysis of UTMB data showed that the charges to provide EHM using CDHM would achieve cost neutrality if the incidence of NEC was reduced by 67%. In March 2013, a policy of providing a diet of CDHM fortified with liquid human milk fortifier (CDFHM) was instituted for VLBW infants. Our aim was to determine the cost-effectiveness of this strategy.

Methods Used: Incidences of LOS (>72 hours, positive blood culture), medical NEC and surgical NEC were ascertained for the years 2009-2011 (period of providing preterm formula) in VLBW babies. The comparison period was the 12 month period June 2013-May 2014, to allow a 3 month equilibration after EHM policy implementation. Cost estimates for NEC and LOS (Ganapathy 2012) were used to calculate annual hospital charges for the periods before and after the change in policy.

Summary of Results: During 2009-2011, there were 8, 17 and 9 cases of NEC annually (3y mean=SD 11.3±4.9, mean population annual incidence 12.5%, range 8.8-19.1%) and 9, 7 and 14 cases of LOS (13.3±6.0, incidence 14.6%, range 7.9-20.9%). There was one case of surgical NEC in each year. During the June 2013-May 2014 period, there were 73% and 77% reductions in NEC (3) and LOS (3). There was no surgical NEC. This amounted to a total reduction in morbidity-associated charges of $918,608. When the hospital charges of CFDHM purchase was subtracted, the adjusted annual savings estimate associated with an EHM diet remained a robust $461,371.

Conclusions: By reducing the incidence of medical NEC by 73%, an EHM diet consisting of CDHM resulted in substantially decreased hospital charges in a Level III NICU (cost neutrality at 67%). When fewer cases of LOS and surgical NEC were also considered, the total reduction in estimated hospital charges was $918,608. Ongoing changes in unit practice such as increased supply of maternal milk and reduced LOS rates may have a mixed effect on hospital savings when the cost of CDHM is considered. This is the focus of ongoing studies.

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DECISION-MAKING ON BEHALF OF A FETUS WITH A SERIOUS ILLNESS AND ITS EFFECT ON MATERNAL MENTAL HEALTH

Hutchens A, Jones P. University of Texas Health Science Center, Houston, TX

Purpose of Study: To determine if discordance between a mother’s preferred and perceived role in decision-making on behalf of her fetus with a serious medical condition is associated with higher symptom scoring for depression or PTSD at six months post-partum.

Methods Used: Design: Prospective cohort study.

Inclusion Criteria: Pregnant women referred to the Texas Fetal Center at Children’s Memorial Hermann Hospital for consultation and to make treatment decisions on behalf of a fetus with a serious medical condition.

Exclusion Criteria: Not fluent in the English language.

Exposure: Discordance between a person’s preferred role in decision-making for a fetus and the role that person experienced.

Measurements: Upon enrollment, subjects were administered the Survey on End-of-Life Decision Making. The score for “preferred role” was subtracted from the score for “perceived role” to determine the presence and level of discordance. Tools to measure symptoms of depression (Patient Health Questionnaire) and PTSD (PTSD Checklist-Civilian Version) were administered six months post estimated date of delivery.

Analysis: Unadjusted linear regression will be performed to evaluate the association between discordance scores at six months post partum and depression and PTSD with all values treated as continuous outcome variables.

Summary of Results: A total of 35 subjects have been recruited, with 10 (29%) displaying discordance in preferred and perceived decision-making roles. Six-month scoring for depression and PTSD is being collected, and this data will be available at the time of presentation.

Conclusions: If there is an association between discordance scores and PTSD or depression discord scores at six months post partum, we would conclude that interventions to decrease discordance in decision-making roles could have a positive effect on maternal post-partum mental health.
preoperative endotracheal intubation. 87% had a 1-step surgery and 72% of the infants had at least 1 other procedure, most commonly a g-tube placement. 3 infants developed pneumothorax; 1 developed a chylothorax and empyema and 4 had anastomotic leak. 6 infants developed strictures, and empyema and 4 had anastomotic leak. 7/27 had a long gap atresia (> 3 cm), 2 had primary repair and, and 3/7 developed strictures. Median length of stay was 33 days. A feeding tube was needed in 72% at discharge. Overall survival to discharge was 85%, while infants <1500gm BW (BW) had significantly higher mortality (50% vs 5%, p = 0.02).

Conclusions: Our survival-to-discharge rate is comparable to those reported in the literature. Mortality was higher in <1500gm BW infants. A majority of infants needed g-tube feedings for feeding problems. Although the postoperative complication rate was low, 22% of patients developed strictures that required dilatation.

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DOXAPRAM DOES NOT ADVERSELY AFFECT NEURODEVELOPMENTAL OUTCOME OF EXTREMELY LOW BIRTH WEIGHT INFANTS AT 12-18 MONTHS OF AGE

Patel S, Eyal F. University of South Alabama, Mobile, AL.

Purpose of Study: Doxapram, a neurorespiratory stimulant is used for prevention of mechanical ventilation (IPPV) in apneic infants. The aim of our study was to assess whether this central neurostimulatory effect may adversely affect neurodevelopmental (ND) outcome.

Methods Used: Retrospective review of NICU medical records of all extremely low birth weight infants (ELBWI); gestational age (GA) <29 weeks admitted to the NICU at USA CW Hospital between 1998-2012. The ND outcome was assessed by the Psychomotor Development Index (PDI) and Bayley Scales of Infant Development scored by Mental Developmental Index (MDI). MDI or PDI <70 are associated with neurodevelopmental impairment (NDI). Data were analyzed by bivariate analysis (chi square or t test) and logistic regression.

Summary of Results: Amongst the surviving 956 infants followed, 500 infants received treatment with Doxapram (DOX) and 456 control infants did not (CNT). By bivariate analysis, the frequency of infants with an MDI<70 was higher in the DOX group (p=0.009). Rates of PDI<70 and cerebral palsy were similar between both groups. Patients treated with DOX were younger (GA : 24.6±1.5 weeks vs. 25.8±1.6, p=0.0001) and had a higher incidence of white matter injury (WMI, p=0.03). Logistic regression (TABLE 1) negated the detrimental effect of DOX upon MDI after inclusion of GA, WMI and other perinatal factors known to affect MDI (OR: 1.5 CI: 0.9-2.6). However, MDI was adversely and significantly affected among infants who received and failed DOX treatment (i.e. required IPPV within 8 days of treatment, OR: 1.9 CI: 1.1-3.4).

Conclusions: Doxapram did not adversely affect ND outcome of ELBWI. The failure of Doxapram treatment rather than Doxapram itself is a risk factor for NDI. This failure of treatment may underline a common pathology associated with NDI or be the result of the need for a more prolonged duration of mechanical ventilation.

<table>
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<tr>
<th></th>
<th>MDI&lt;70</th>
<th>MDI&lt;70 &amp; NO CP</th>
<th>MDI&lt;70, NO CP &amp; NO WMI</th>
<th>MDI&lt;70, NO CP, NO WMI &amp; DOX FAILURE</th>
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<tr>
<td><strong>AA race</strong></td>
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<tr>
<td>Female</td>
<td>0.7 (0.2-2.1)**</td>
<td>0.6 (0.4-0.9)**</td>
<td>0.6 (0.4-0.9)**</td>
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<td>WMI</td>
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<td>6.0 (2.1-8.3)**</td>
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<td>GA</td>
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<td>1.3 (0.8-2.2)</td>
<td>1.5 (0.9-2.6)</td>
<td>1.9 (1.1-3.4)**</td>
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MATERNAL LABETOLOL TREATMENT AND ITS EFFECTS ON A PRETERM INFANT

Shah SD, Apostolakis-Kyrus K, Talati AJ1,2 University of Tennessee, Health Science Center, Memphis, TN and 1University of Tennessee, Health Science Center, Memphis, TN.

Case Report:

BACKGROUND: Hypertension during pregnancy can cause severe mortality and morbidity in neonates. Antihypertensive medications such as beta-blockers, calcium channel blockers etc are commonly used and considered safe and effective. These medications can cross the placenta and affect the fetus. Several case reports have described significant neonatal side effects such as bradycardia, hypotension and hypoglycemia after use of beta-blockers. Half-life of maternal labetolol in the premature infant could be as long as 24 hour. We present a case of premature infant who received multiple antihypertensive medications in a short period of time that may have resulted in fetal bradycardia and persistent bradycardia after delivery along with hypoglycemia and hypotension.

CASE REPORT: A 29 wk gestation male, with a BW of 1340 gm, was born by emergency caesarean section (CS) to a 39-year-old woman G3P2 woman secondary to fetal bradycardia. Her prenatal screening tests were unremarkable. Pregnancy was complicated by severe hypertension, morbid obesity, and preterm labor. She received 300 mg of intravenous labetolol prior to CS, (within 2.5 hr) and also received 1 dose of nifedipine and hydralazine. Umbilical cord venous pH was 7.13. He had persistent bradycardia (HR 70-80 bpm) following delivery, which required 2 doses of epinephrine at 8-10 min leading to HR >100. The infant’s Apgar scores were 1, 2 and 5 at 1, 5 and 10 min respectively. In the NICU, his heart rate was 110-120 bpm with blood pressure of 35/15 mmHg inspite of vasopressors (Dopamine and dobutamine) and saline bolus. Initial glucose was undetectable, required two glucose boluses and glucose infusion rate of 7.3 mg/kg/min to maintain serum glucose levels 50 mg/dl. HR and BP normalized after 36-48 hr. Sepsis work up was negative. At 24 hr, he developed severe hypoxemia and renal failure that subsequently resolved in 2 days. Head ultrasound on 15th day showed moderate ventriculomegaly with grade 3 IVH.

CONCLUSIONS: This report provides clinical description of possible severe effects of maternal labetolol treatment in a premature infant. The severity of these side effects is not predictable and should be considered by clinicians.
extrauterine growth restriction (weight for gestational age, WGA) often experience growth faltering during their NICU stay, resulting in ex-

Conclusions: Though NEC is of main concern in extreme preterm infants, late preterm and term infants can also have NEC. As previously reported, NEC appears to develop earlier in infants ≥24 weeks. They have a better survival and less severe NEC. The differences in other characteristics are probably related to prematurity.

Pulmonary and Critical Care Medicine
Joint Plenary Poster Session and Reception

5:00 PM
Thursday, February 26, 2015

PROPOFOL INFUSION SYNDROME: A CONCERNING CAUSE OF NEW ONSET METABOLIC ACIDOSIS AND RENAL FAILURE IN ICU PATIENTS

Adiga AG, Panikkath D, Mohamed A, Nugent K. Texas Tech University Health Sciences Center, Lubbock, TX.

Case Report: Introduction:

Propofol infusion syndrome (PRIS) is a rare but fatal syndrome observed more commonly in young obese men receiving high dose (usually >4mg/kg/hr) or long term propofol (>48 hrs) [1]. It is more common in critically ill patients receiving catecholamines or steroids, and it presents as severe unexplained metabolic acidosis, renal failure, rhabdomyolysis, hyperkalemia and cardiac failure[2,3]. Incidence of PRIS is yet unclear and is solely based on case reports in last two decades.

We report a case of possible PRIS in 24 year old obese Latin American man admitted for acute severe asthma who developed PRIS in less than 12 hours of lower doses of propofol (3mg/kg/hr tapered within 3 hrs). He also received concurrent steroids.

Case Report: 24 year old Latin American obese gentleman with history of asthma since childhood presented with complaints of shortness of breath and chest tightness for 1 day. He was found to be in hypercapnic respiratory failure due to acute severe asthma. He was intubated and put on ventilator support and was started on propofol, fentanyl, methylprednisolone (100mg/day), bronchodilators, azithromycin, enoxaparin. His labs showed increased WBC count and serum potassium levels but normal renal function test. His urine was positive for cannabinoids screen. CAT scan of thorax showed a suspected pulmonary embolism which was later ruled out by VQ scan. On day 2 of admission patient started developing red colored urine and deterioration in renal function (creatinine increased from 0.7mg/dl to 2.3mg/dl). ABG showed metabolic acidosis with pH of 7.178, triglyceride level increased from 138mg/dl to 223mg/dl and creatinine kinase (CK) levels increased to 4168IU/L. A diagnosis of propofol infusion syn-
drome (PRIS) was entertained, and propofol (total 2.937gm of propofol infused over 12hr) was stopped, he was switched to dexmetomidine. He was treated with IV fluids (normal saline, 1/2 normal saline with bicarbonate) with a goal to maintain urine output of >150ml/hr and pH 6.5 with which his CK started downward trend and within 4 days renal function returned to the baseline. He was extubated on day 4 of admission and discharged home in a stable state on day 8.

DIAGNOSIS OF PSEUDO-PULMONARY EMBOLI WITH ENDOBRONCHIAL ULTRASOUND

Al-SaffarF1, IbrahimS1, SeeramV2, ShujaatA2. 1University of Florida, Jacksonville, FL and 2University of Florida - Jacksonville, Jacksonville, FL.

Purpose of Study: A systematic literature review to identify the usefulness and accuracy of endobronchial ultrasound in detecting endovascular lesions mistaken for pulmonary emboli.

Methods Used: PubMed, Cochrane, and Google Scholar were searched for: endobronchial ultrasound, EBUS, embolism, embolus, embolic, thromboembolic, thorboembolism, emboli, and thromboemboli

Summary of Results: Six female and 1 male cases. EBUS done to evaluate persistent or progressive filling defects in the pulmonary artery (PA) despite anticoagulation (n =2), mass involving PA (n = 2) or a significant uptake on FDG-PET without corresponding lymphadenopathy (LAD) in the region (n = 1). An endovascular lesion was identified in the EBUS-LAD (n=1). EBUS-TBNA was diagnostic in 4 of the 5 cases in which it was done. The final diagnoses were: lung cancer (n = 2), sarcoma (n =2), thyroid cancer (n =1), renal cell cancer (n =1) and melanoma (n =1). The cancer was a recurrence in 4 of the 5 cases with history of cancer.

Conclusions: EBUS should be considered as a possible method for evaluating endovascular lesions when pulmonary artery sarcoma or tumor embolism is suspected, particularly when there is a persistent filling defect despite anticoagulation and history of a cancer known to be associated with pulmonary tumor embolism.
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ACUTE STROKE FROM INTERNAL CAROTID ARTERY OCLUSION SECONDARY TO CAVERNOUS SINUS THROMBOSIS AND ORBITAL CELLULITIS

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**Case Report:** Cavernous sinus thrombosis (CST) affects the nervous system structures in the vicinity, but there are few reports of involvement of the internal carotid artery (ICA). We present a case of a 14-yr old girl with right orbital cellulitis, who developed bilateral superior ophthalmic vein thrombosis (SVOT), bilateral CST, occlusion of the right ICA resulting in right MCA distribution ischemic stroke.

A previously healthy 14 yr female presented to the ED with right orbital protopisis, pain with extraocular movements, and fever. A CT scan confirmed right orbital cellulitis with right SOVT. She was started on levofloxacin, vancomycin and ceftriaxone. After a blood culture grew Methicillin Sensitive Staphylococcus Aureus, she was converted to naftisol. She continued to have fevers and worsening protopisis. Repeat imaging with MRI showed bilateral SOVT and bilateral CST with narrowing of bilateral ICA. She also developed hypotension and was transferred to the PICU on hospital day 11. Daptomycin and meropenem were added. In the PICU, she developed left hand weakness, which progressed to left-sided weakness. Repeat MRI/MRA demonstrated complete occlusion of right ICA and ischemic stroke in the right MCA distribution. She began high dose steroids and her mean arterial pressure was maintained supratherapeutic (MAP therapy) with dopamine and norepinephrine infusions.

She improved on steroids and MAP therapy. Dexamethasone and norepinephrine were discontinued and the steroids were tapered. She was transferred to a rehabilitation facility on hospital day 22. At time of transfer, she had regained strength in her left side, but had persistent weakness of the left arm.

CST is well described and known to cause significant neurologic sequelae but there are no case reports of CST secondary to orbital cellulitis in pediatrics. Antibiotic coverage is the mainstay of treatment, with steroids and anticoagulants being more controversial. In our case we believe inflammation resulted in occlusion of the right ICA and was the cause of the subsequent ischemic event. With prompt recognition of CST with carotid artery involvement, early initiation of steroid therapy may help improve neurologic outcomes.

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INVASIVE VS. NON-INVASIVE BLOOD PRESSURE MONITORING IN MEDICAL INTENSIVE CARE PATIENTS

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**Purpose of Study:** Invasive arterial catheters (AC) are frequently used to monitor blood pressure and draw blood for laboratory evaluation with little evidence to support that an AC actually provides clinical benefit to patients. The study hypothesized that AC would improve weaning from vasopressors in medical ICU patients.

**Methods Used:** We evaluated severe sepsis patients requiring vasopressors for a minimum of 24 hours with & without an AC who were identified by random selection from an existing database of sepsis patients. The primary outcome of the study was the duration (days) of vasopressors therapy. Secondary outcomes were the number of blood transfusions, ICU days, days of mechanical ventilation, fluid volume received while in the ICU and the initial 24 hours post admission, average daily hemoglobin, multi-organ dysfunction score (MODS), and urine output.

**Summary of Results:** The 31 patients with AC were sicker (by APACHE II) than the 23 patients without AC, even though there were no significant differences in age, gender or type of infection. Survival of AC patients (45%) was similar to no AC patients (52%, p=0.76). AC patients had more days of vasopressor use, longer ICU stay and more days on mechanical ventilation. The AC group received more fluid during ICU admission, urine output, blood products transfused or in the average daily hemoglobin between AC and no AC groups.

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PREDICTING THE NEED FOR UPFRONT DUAL THERAPY IN PULMONARY ARTERIAL HYPERTENSION

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**Purpose of Study:** Combination therapy is commonly used for pulmonary arterial hypertension (PAH) after initial monotherapy as it improves six minute test (6MWT) distance, pulmonary hemodynamics and functional class. There is increasing interest to evaluate the value of treating patients with combination therapy up front instead of staggering it. We thus aimed to identify factors that may predict the need for upfront combination therapy.

**Methods Used:** Retrospective review of PAH patients from July 2007 to July 2012. 68 patients were included. Pulmonary vascular resistance (PVR) minute test (6MWT) distance, pulmonary hemodynamics and functional class. There is increasing interest to evaluate the value of treating patients with combination therapy up front instead of staggering it. We thus aimed to identify factors that may predict the need for upfront combination therapy.

**Summary of Results:** Six out of 23 patients with low pulmonary vascular resistance (PVR) were on dual therapy compared to 23 out of 29 patients who had high PVR (OR 10.86 (2.98 - 39.59)). 62.8% patients with low values and 21/30 patients with high ones were on dual therapy (OR 8.56 (2.59 - 28.22)). 9/39 with normal PAM compared to 23/29 with high PAM were on combination treatment (OR 12.78 (3.98 - 41.05)). Risk score: 21 of 54 patients with high PVR (OR 10.86 (2.98 - 9.59)). RVSP: 6/28 patients with low values on dual treatment (OR 0.36 (0.13-1.00)). We evaluated severe sepsis patients requiring vasopressors for a minimum of 24 hours with & without an AC who were identified by random selection from an existing database of sepsis patients. The primary outcome of the study was the duration (days) of vasopressors therapy. Secondary outcomes were the number of blood transfusions, ICU days, days of mechanical ventilation, fluid volume received while in the ICU and the initial 24 hours post admission, average daily hemoglobin, multi-organ dysfunction score (MODS), and urine output.

**Summary of Results:** The 31 patients with AC were sicker (by APACHE II) than the 23 patients without AC, even though there were no significant differences in age, gender or type of infection. Survival of AC patients (45%) was similar to no AC patients (52%, p=0.76). AC patients had more days of vasopressor use, longer ICU stay and more days on mechanical ventilation. The AC group received more fluid during ICU admission, urine output, blood products transfused or in the average daily hemoglobin between AC and no AC groups.
A COMPARISON OF PROPOFOL VERSUS HYPERVENTILATION FOR INDUCTION OF APNEA IN LUNG FUNCTION TESTING OF INFANT OLIVE BABOONS (PAPIO ANUBIS)

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Purpose of Study: Olive baboons (OB) are attractive animals to model pulmonary diseases of humans. For quantitative evaluation of lung function, sedation and induction of apnea are required. The consistent induction of apnea with the standard method of hyperventilation is not always possible in subjects with severe lung disease and high level of spontaneous ventilation. We investigated if using propofol to induce apnea in infant OB influences pulmonary function measures (PFT) in comparison with data gained with hyperventilation-induced apnea.

Methods Used: A month old OB (n=9) were sedated, intubated and placed on an Aeva ventilator equipped with a hot wire pneumotachometer. Initially the OB were allowed to breathe spontaneously with zero end-expiratory pressure. Then the animals were mildly hyperventilated with respiratory rate 60 breaths/minute, inhalation time of 0.5 sec. and tidal volume 8 ml/kg until cessation of spontaneous breaths, when PFT were measured. Next, the animals were allowed to return to spontaneous breathing, after which apnea was induced by IV administration of propofol (~10 mg/kg). Apneic OB were ventilated with the same parameters and the same measurements were done.

Summary of Results: Values obtained using propofol and hyperventilation, respectively, were: static compliance 0.897 ± 0.09 vs. 0.899 ± 0.17 ml/cm H2O/kg, dynamic compliance 1.08 ± 0.22 ml/cm H2O/kg, work of breathing 0.46 ± 0.09 vs. 0.47 ± 0.12 J/L, peak expiratory flow rate 2.6 ± 0.60 vs. 2.7 ± 0.64 L/min, resistance at peak pressure 49 ± 7.4 vs. 46 ± 10 cm H2O/L/sec, compliance ratio (C20°C) 2.20 ± 0.24 vs. 2.23 ± 0.20 (mean ± S.D.). All the differences were not statistically significant in paired two-tailed t-test.

Conclusions: Results of PFT in OB were similar whether apnea was induced by hyperventilation or by IV propofol. However, measurements obtained under propofol showed a trend to lesser variability than those obtained during hyperventilation. This enables use of propofol for apnea induction in animals with high background ventilation without loss of data quality. The reduced variance in data obtained with propofol may allow a more accurate evaluation for differences in PFT.

PERSISTENT TACHYPNEA AND HYPOXIA IN AN INFANT: IS IT NEUROENDOCRINE HYPERPLASIA OF INFANCY (NEHI)?

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Case Report: Introduction: Neuroendocrine hyperplasia of infancy (NEHI) remains an unique form of childhood Interstitial Lung disease (ILD). Here we present a case of persistent tachypnea and room air hypoxia in an infant.

Case Report: A 4 month old male presented with persistent tachypnea and hypoxia since birth which persisted despite antibiotics and steroids. He required hospitalizations at 3, 4 and 5 months age for respiratory distress and tachypnea. Physical exam was significant for subcostal retractions, clear lungs, and persistent hypoxemia requiring supplemental oxygen. Extensive workup was done including: negative PPD; nonreactive HIV; negative sweat chloride test; CXR with hyperinflation and atelectasis; normal echocardiogram; normal immune workup; fluoroscopy without paresis or paralysis of the diaphragm; negative surfactant protein mutations; mildly low percentage of CD2, CD3, CD4, CD8 cells; and low complement. Work up was positive for acid reflux on upper GI. HRCT of the chest showed a smaller volume right lung and bronchiolitis along with a mosaic pattern with geographic ground glass opacity and air trapping in the right lung and the left infrahilar region. He had a bronchial alveolar lavage (BAL) with negative AFB culture, negative tuberculosis PCR, and culture that grew mixed bacteria. He subsequently underwent lung biopsy of the right middle lobe at 7 months old with staining for bombesin cells. Pathology showed prominent increase in neuroendocrine cells and large neuroepithelial bodies, which was consistent with NEHI. He was treated with supportive care (supplemental oxygen and reflux therapy) and has since been stable from a respiratory standpoint.

Discussion: We present a case of NEHI. NEHI should be considered in the differential diagnosis for persistent tachypnea and hypoxia with an otherwise negative workup. Diagnostic evaluation should include HRCT and confirmatory lung biopsy with immunostaining for bombesin showing neuroendocrine cells within the bronchioles and increased size and number of neuroepithelial bodies in the lobules. Treatment is primarily supportive with optimizing nutrition, supplemental oxygen, treating reflux, and preventive medical care, including immunizations.
A CASE OF EVOLVING PULMONARY HYPERTENSION WHO GROUP CLASSIFICATION

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Introduction: Pulmonary hypertension can be caused by a variety of different pathologies. The WHO classification of pulmonary hypertension can be useful for classifying the disease and treatment options though there are still diagnostic challenges within those groups. The case below can illuminate some of the difficulties when trying to find the etiology of pulmonary hypertension in the setting of interstitial lung disease.

Case: A 35 year old Hispanic man originally from Honduras was first seen in 2011 when he presented to the emergency department with symptoms of progressive dyspnea, 25 pound weight gain, and chronic cough. At the time of diagnosis via right heart cath his mean PA pressures were 58mm Hg and wedge pressure was 15mm Hg. He was tentatively diagnosed with WHO group 1 pulmonary hypertension and was started on Bosentan, coumadin, lasix, and oxygen. Eventually the patient had worsening dyspnea and hypoxia and the patient had a port placed and IV treprostinil therapy was started. He continued to have worsening symptoms and repeat CT showed worsening of the patient’s ground glass opacities. The patient was referred to CT surgery for an open lung biopsy which showed a UIP pattern. Repeat right cath showed PAP of 94/40 mm Hg (mean ~58) despite IV prostinoid therapy. Rheumatologic work up revealed a high SSA antibody profile. Lip biopsy was done and negative. Despite this there was still concern that he had a connective tissue disease and the patient received a dose of rituximab. Given his significant lung disease it was now thought that there may be the etiology of his pulmonary hypertension and he was transferred to the regional lung transplant facility for evaluation for transplant.

Discussion: Pulmonary Hypertension can be diagnostically difficult. The WHO group classification is useful for categorizing and treating pulmonary hypertension. In brief: Group 1 - idiopathic; Group 2 - left heart disease; Group 3 - lung disease and/or hypoxia; Group 4 - chronic thromboembolic pulmonary hypertension; Group 5 - unclear multifactorial mechanisms. The evaluation of patients can be complicated by overlapping and evolving disease process which can initially lead to the wrong classification of the disease.

USE OF BETAINE ANHYDROUS IN THE TREATMENT OF HYPOCHLOREMIC METABOLIC ALKALOSIS - A NOVEL APPROACH

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Purpose of Study: Metabolic alkalosis (MA) is a common complication in the pediatric intensive care unit (PICU) that can lead to cardiovascular, pulmonary and metabolic dysfunction. These complications may lead to difficulty in weaning mechanical ventilation and a prolonged PICU stay. In the pediatric intensive care unit (PICU) that can lead to cardiovascular, pulmonary and metabolic dysfunction. These complications may lead to difficulty in weaning mechanical ventilation and a prolonged PICU stay. Due to a lack of and potential toxicity of currently used therapies, pediatric critical care physicians at The Children’s Hospital at OU Medical Center have utilized betaine anhydrous (BA) for the treatment of hypochloremic MA. The purpose of this study was to describe the effectiveness of BA in correcting hypochloremic MA.

Methods Used: This was a descriptive, retrospective study of children, 0-18 years of age (YOA) receiving BA during hospitalization from October 1, 2010 to October 31, 2012. Data collection included baseline demographics, laboratory data [e.g., serum chloride (Cl-) and bicarbonate (HCO3-)], and the BA regimen. Children had to have pre-treatment and 72 hours post-treatment serum Cl- and HCO3- values to be included for analysis. Other agents used to treat MA, use of diuretics and use of mechanical ventilation were also collected. The primary objective was the mean change in HCO3- and Cl- from baseline. Secondary objectives included description of adverse events (AE). Descriptive statistics were utilized. Student’s paired t-test was used to evaluate continuous data.

Summary of Results: Ten patients received BA, but only four met inclusion criteria for analysis. Two (50%) were male, and the median age was 3.6 years (range 0.04-14). The majority (75%) of patients received mechanical ventilation, all were receiving diuretics, and all were concomitantly receiving acetazolamide. There was a statistically significant decrease in HCO3- from baseline (36.8 ± 30.8 mEq/L, p = 0.0434). There was noticeable but not statistically significant increase in Cl- from baseline (88 ± 99.5 mEq/L, p = 0.0634). No AE were noted.

Conclusions: BA was associated with a statistically significant decrease in HCO3- and a noticeable, but not significant increase in Cl-. However, it is difficult to ascertain the potential effectiveness of BA as all children were concomitantly receiving acetazolamide. Further research comparing the effectiveness of BA versus acetazolamide is warranted.

PRIMARY PULMONARY MULLERIAN TUMOR PRESENTING IN A YOUNG WOMAN

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Case Report: A 35 year old female, with no significant past medical history, was referred to our center after work-up for dyspnea revealed a pleural effusion that recurred after thoracentesis. The initial thoracentesis showed negative cytology on the six liters drained. After recurrence of the effusion the patient underwent VATS and talc pleurodesis at an outside facility. At that time a thick, white pleural process encompassing the visceral and parietal pleura was noted and biopsies were performed. Pathology initially confirmed epithelioid neoplasm, suspicious for mesothelioma; however, tissue pathology and the immunohistochemistry stains were discordant.

Upon presentation at OU Medical Center, the patient was evaluated for extrapleural pneumonectomy (EPP) to further control her neoplastic process. The patient’s PET-CT scan revealed diffuse activity along the visceral and parietal pleural surfaces without evidence of hypermetabolic activity elsewhere including in the abdominal cavity or pelvis. After negative findings on both breast MRI and pelvic ultrasound, the patient was taken for surgery for EPP with diaphragmatic and pericardial reconstruction which she tolerated well. Surgical pathology revealed malignancy consistent with the initial biopsy showing a fibrotic layer with numerous small nests of neoplastic glands with occasional micropapillary formations. The immunohistochemical profile of the tissue did not provide strong evidence for the diagnosis of a mesothelioma. The calretinin and thyroid transcription factor 1 were negative. Pax-8, shown to be positive in Mullerian tumors and negative in mesothelioma, as well as Ber-EP4, ER, and PR were positive in the tumor cells. Lymph nodes at stations 5, 6, 7, 9, and 10L were positive for tumor but tumor margins were negative.

This is an unusual case of a primary mullerian tumor presenting as a pleural neoplasm. There is one previous case report of a primary pleural mullerian tumor in a 23 year old female. Both patients were treated with EPP and subsequent chemotherapy with a platinum/pemetrexed based therapy.
A 38 year old female was diagnosed with idiopathic bronchiectasis in 2010. Cystic fibrosis and alpha 1 antitrypsin deficiency were excluded. Immunoglobulins were normal. The patient refused lung biopsy. She had been diagnosed with AIH in 2007 and treated with azathioprine and mycophenolate mofetil. Treatment was stopped in 2008 due to medication side-effects. A repeat liver biopsy in 2014 showed cholestatic hepatitis consistent with anti-mitochondrial antibody-negative PBC versus primary sclerosing cholangitis (Fig 1). Computed tomography of the chest showed worsening chronic bronchiectasis (Fig. 2).

Alteration of lung parenchyma secondary to chronic liver disease of autoimmune etiology, such as PBC or AIH, may be related to autoimmune antibodies that activate neutrophils and alveolar macrophages. IgG and IgM immune complexes act as a contributing factor in the formation of the interstitial fibrotic reaction and thickening of the basement membrane. Fibroblast growth factor produced by lymphocytes, macrophages, and platelets may initiate collagen production. The clinician should recognize that autoimmune liver disease may result in concomitant autoimmune lung disease.

FIGURE 1

PRIMARY LEIOMYOSARCOMA OF INFERIOR VENA CAVA EXTENDING INTO THE RIGHT ATRIUM AND VENTRICLE
Samuel J. UF Health, Jacksonville, FL.
Case Report: A 38 year old female presented to the ER with worsening symptoms of shortness of breath on exertion and dysphagia. On physical exam she was tachypneic with decreased air entry in all lung fields. On admission she was normotensive, afebrile with no other acute complaints. She had bilateral jugular venous distention with massive hepatomegaly. She had a resected astrocytoma when she was 8 and had residual seizure disorder. She had bilateral jugular venous distention with massive hepatomegaly. She was admitted to the medical intensive care unit in 2013. We collected information about patient demographics, medical diagnoses, gas exchange, chest x-ray abnormalities, fluid balances, and complications, including pneumothoraces, ventilator-associated events, self-extubation, and failed extubation.

Methods Used: We retrospectively reviewed the medical records of 174 patients who required mechanical ventilation for acute respiratory failure in a medical intensive care unit in 2013. We collected information about patient demographics, medical diagnoses, gas exchange, chest x-ray abnormalities, fluid balances, and complications, including pneumothoraces, ventilator-associated events, self-extubation, and failed extubation.

Summary of Results: This study included 174 patients. The mean age was 57.8 ± 16.8 years, 54.02% were men, the mean APACHE 2 score was professional guidelines on the management of IVC leiomyosarcomas, but the consensus among surgeons and best prognosis is aggressive surgical removal in combination with chemotherapy and radiation.

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THE MEDICAL PARADOX - WHEN A GOOD DRUG TURNS BAD (AMIODARONE)
Satyavada S, Singh M, Yalamanchili K, Smalligan R. Texas Tech University Health Sciences Center. Amarillo, TX.
Case Report: A 57-year-old non-smoking male came to the ER with complaints of progressive shortness of breath, runny nose, and productive cough over a 3 week period that had not responded to oral antibiotics. He did not have fever or leukocytosis and tested negative for the flu. PMH: atrial fibrillation, hyperlipidemia, history of MI in 2010 with stent placement. Medications: amiodarone, simvastatin, aspirin, clopidogrel, losartan, carvedilol, and ranitidine. Physical exam: BP 113/73, R 18, P 60, T 97.5, O2 sat 98% on 3L NC, no JVD, lungs: crackles bilaterally with good air movement, heart: irregularly irregular, no murmur, remainder of exam normal. Labs: WBC 5,200, 75% neutrophils, HCT 40, CMP normal, BNP 142. Blood cx neg and sputum with moderate gram (+) cocci. CXR: patchy infiltrates in lower lung zones. A dx of CAP was made and ceftriaxone and doxycycline were started. By the third day his hypoxemia and respiratory rate worsened and his CXR showed increased infiltrates and he required mechanical ventilation. Amiodarone-induced pulmonary toxicity was suspected, the drug was discontinued and high dose steroids begun. A CT of the chest showed ground-glass and consolidative opacities and diffuse inflammation. Despite meticulous care in the ICU the patient developed ARDS and died approximately 3 weeks later.

Discussion: Pulmonary fibrosis is a well-known potential toxicity of chronic amiodarone use. Making the diagnosis of amiodarone-induced pulmonary toxicity is challenging because its presentation is not always overt fibrosis, but rather it can present as an acute or subacute pneumonitis with diffuse inflammation as in our case or as a more localized form involving the pleura, or as multiple nodules. It can occur after any dose or length of treatment with amiodarone. In our case it was felt that based on the patient's history of 4 years of use, negative lab work-up for other causes of ARDS and a CT image consistent with the diagnosis that his fulminant clinical course was due to amiodarone-induced pulmonary toxicity. It is crucial for physicians to consider this diagnosis early in the course because patients may respond to discontinuation of the medication and high dose corticosteroids. Despite best available care mortality is up to 50% in patients with amiodarone induced toxicity that develop ARDS.
Patients frequently develop increasing infiltrates and/or pleural effusions. The frequency of pneumothorax, frequently in a heterogeneous group of patients requiring mechanical ventilation was 1539 patients and effusions in 19 patients. The mean fluid balance during mechanical ventilation was 1721 mL per day. Three patients (1.7%; 95% CI=0.22% to 3.62%) developed a pneumothorax; 5 patients required chest tube placement. Twenty-five patients (14.4%; 95% CI= 9.2% to 19.6%) had a ventilator-associated event. Ten patients had an episode of self-extubation; 11 had an episode of failed extubation. Chest x-rays showed new or increasing infiltrates in 113 patients and new or increasing pleural effusions in 29 patients.

**Conclusions:** This study indicates that complications occur relatively infrequently in a heterogeneous group of patients requiring mechanical ventilation for acute respiratory failure. The frequency of pneumothorax, ventilator-associated events, and self-extubation was low. However, these patients frequently develop increasing infiltrates and/or pleural effusions. These radiographic changes may represent progression of underlying disease or the development of a complication and require clinical investigation.

**Case Report:** Introduction: Pulmonary hemorrhage is rare but potentially life threatening in children. Some cases that previously would have been diagnosed as idiopathic pulmonary hemosiderosis have now been shown to be due to pulmonary capillaritis, a small vessel vasculitis. This is a unique case presentation of pulmonary capillaritis in a 2 year old male with no identifiable underlying cause.

Case Presentation: The patient is an 8 year old male with a history of asthma who presented initially at 2 years of age with three episodes of hemoptysis, and severe anemia. He had a two month history of malaise, cough, and pallor. On physical exam his lungs were clear and he had no respiratory distress, but required supplemental oxygen. Initial hemoglobin was 4.4, hematocrit of 15.4 with an MCV of 72. Chest x-ray revealed bilateral infiltrates, and chest CT which showed bilateral multifocal air space disease with ground glass opacity. Infectious and cardiovascular causes were ruled out. He underwent bronchial lavage which revealed Hemosiderin-laden macrophages, and lung biopsy showed abundant hemosiderin-laden macrophages and patchy acute hemorrhage with rare intestinal neutrophils and focal lymphoid aggregates suggestive of acute capillaritis. Antibody levels were normal except for IgE of 70, celiac panel was negative. ANCA, p-ANCA, Anti-Neutrophil Ab, and ANA screen were all negative. He was successfully treated initially with 2mg/kg of IV corticosteroids daily, then 1mg/kg of oral corticosteroids for one month, followed by 1mg/kg every other day. He has recently been changed to hydroxychloroquine and the steroids discontinued with no further episodes of hemoptysis.

**Discussion:** Here we present a unique case of pulmonary capillaritis a rare condition in children. Pulmonary capillaritis should be considered in the differential diagnosis for any patient who presents with alveolar hemorrhage. Diagnostic evaluation should include the presence of hemosiderin laden macrophages and a lung biopsy. Treatment options include; high dose IV pulsed steroids up to 30mg/kg daily for three days. Cytotoxic drugs such as cyclophosphamide, azathioprine and hydroxychloroquine should be considered in severe cases.
Minimal Change Disease is very common in children but significantly less common in adults. MCD is more likely to occur from malignancy or medications (such as nonsteroidal anti-inflammatory drugs) but should also be considered on individuals with systemic diseases. To date, there have been 27 cases per million of autoimmune disease related MCD reported in the USA, although it is rare to see MCD with GBS especially in adults. Clinical features of minimal change glomerulopathy in adults tend to be somewhat different. Patients may or may not have typical clinical manifestations such as edema, ascites, hypertension and renal failure. This case highlights the impact of systemic diseases to the development and pathogenesis of MCD and to consider the MCD as a cause of nephrotic syndrome in all adults with any autoimmune disorder.

Case Report: Hyponatremia in West Nile Encephalitis

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**Case Report:** The exact pathogenesis of hyponatremia in patients with West Nile (WN) infection remains unknown; however it might resemble that of the Syndrome of Inappropriate Antidiuretic Hormone Secretion (SIADH) or cerebral salt-wasting syndrome. In consideration of these patterns, we analyze the following case report of WN encephalitis (WNE) complicated with hyponatremia.

**Case-history:** A 67-year-old Hispanic male was admitted for a severe generalized weakness, headache, nausea, and intermittent fever. Physical examination had no significant findings. On Day 2, the patient complained of headache and neck pain, and he was confused. A CT scan of the head revealed no intracranial process. Cerebrospinal fluid was negative for viral and bacterial antigens. Immunoglobulin M for WN was positive. On Day 6, sodium level began to gradually decrease and reached its lowest reading on Day 11. The patient appeared euolemic. His serum osmolality was 254 mOsm, urine osmolality was 754 mOsm, and urine sodium was 153 mmol/L. Thyroid function, glucose and cortisol levels were within normal limits. A presumptive diagnosis of SIADH was made and the patient was placed on a fluid restriction, and sodium chloride tablets. Patient’s neurologic status was improving steadily, and fully recovered on Day 18.

**Discussion:** Hyponatremia accounts for 30% to 42% of cases and is more commonly associated with WNE rather than meningitis, and its mechanism resembles that of SIADH. In our patient, assessment of hyponatremia was complicated by the use of a diuretic, which was discontinued immediately after sodium level started to decrease. Patient’s euolemic status invalidated the cerebral salt wasting mechanism. Impaired mental status preceded the decreasing of sodium level could be attributed to WNE, rather than hyponatremia. Further investigation is needed to understand the precise mechanism of hyponatremia in WNE.

**FIGURE 1**

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**TRIPLE RIPPLE: TRIPLE ACID BASE DISTURBANCE FROM HOMEMADE MOONSHINE AND ISOPROPYL ALCOHOL**

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**Case Report:** 48 year old Hispanic male with no past medical history who presented with weakness, altered mental status, epigastric abdominal pain, vomiting, diaphoresis, tachycardia, and tachypnea. He was well until four days before presentation when he started celebrating his father’s birthday with an alcohol binging vacation. His unnamed friend supplied him with “homemade moonshine” of unknown ingredients, brewing equipment, and age as well as beer. He described the alcohol as “fiuere.” It was thought to have rubbing alcohol (isopropyl) added into the mixture. He could not quantify the amount but was drinking it throughout the three days prior to presentation. He was not feeling any different from a normal alcohol ingestion until one day before admission when he developed emesis with increased epigastric pain. He came to the hospital when he became weak enough he could not stand and was confused with memory loss of the prior events. Mixed triple acid base was revealed by and arterial blood gas, osmolar gap and chemistries which included a respiratory alkalosis (tachypnea), anion gap metabolic acidosis (ketones from ethanol, and lactate), and metabolic alkalosis (emesis and volume contraction). A high osmolar gap was seen due to isopropyl alcohol and its conversion to acetone which does not cause a metabolic acidosis. No crystals were seen in the urine and there were no visual disturbances. While ruling out other ingestions and etiologies, he was treated with aggressive fluid replacement and symptomatically leading to resolution of his metabolic disturbances and acetone level decline. A leukocytosis resolved and was thought to be from a stress reaction. Ingestion of ethanol concomitantly with isopropyl alcohol can have a protective effect as they both compete for alcohol dehydrogenase for metabolization. Isopropyl alcohol is otherwise metabolized quickly into acetone which is metabolized slowly and is detrimental. It was important in our case to rule out other ingestions considering the unknown ingredients of the moonshine; therefore, we ruled out ethylene glycol (microscopic urine analysis), methanol (thorough eye examination), and salicylates (salicylate level) which would have changed management to the need for likely dialysis instead of fluid replacement and supportive care.

**MINIMAL CHANGE DISEASE IN SYSTEMIC LUPUS ERYTHEMATOSUS**

Syed SZ, Siriki R, Kidd L, Teran F, Simon EE. Tulane University School of Medicine, New Orleans, LA.

**Case Report:** We report a patient with nephrotic range proteinuria due to Minimal Change Disease (MCD) with active Systemic Lupus Erythematosus (SLE).

We were consulted for significant nephrotic range proteinuria in a 22 y/o African American female with history of SLE admitted to medical service for abdominal pain secondary to acute pancreatitis with active lupus. On physical examination, the patient had typical malar facial rash and mild pedal edema. Pertinent laboratory abnormalities included 16 gm of proteinuria (via a spot protein/creatinine ratio), hypoalbuminemia of 2.2g/dL, positive ANA, and decreased complement (C3 and C4) but with a normal creatinine of 0.45. Renal biopsy showed changes that were consistent with mesangial proliferative lupus nephritis, ISN/RPS Class II with no active lesions seen and a “full house” immunofluorescence-staining pattern and on electron microscopy there were glomeruli with diffuse podocyte foot process effacement consistent with minimal change nephropathy. Patient was being treated with an intravenous methylprednisolone for active lupus. The steroids also treated there were glomeruli with diffuse podocyte foot process effacement consistent with minimal change nephropathy. Patient was being treated with intravenous methylprednisolone for active lupus. The steroids also treated there were glomeruli with diffuse podocyte foot process effacement consistent with minimal change nephropathy. Patient was being treated with intravenous methylprednisolone for active lupus. The steroids also treated
confined to the mesangial regions of the glomeruli, which have mild or minimal clinical renal findings. Clinically, these patients rarely have nephrotic range proteinuria unless there is a superimposed podocytopathy. MCD may occur as superimposed on mild mesangial proliferative lupus nephritis, this entity may be misinterpreted as an atypical presentation of lupus nephritis class II. Therefore, it is important to emphasize the recognition of this entity requires careful integration of the renal biopsy immunofluorescence and electron microscopic findings.

** Purpose of Study:** Congestive heart failure (CHF) has its pathophysiologic origins rooted in neurohormonal activation. Hormone-mediated elevations in cytosolic free [Ca\textsuperscript{2+}] and subcellareomembran mitochondria [Ca\textsuperscript{2+}]m are coupled to the induction of oxidative stress and opening of the mitochondrial (mt) inner membrane permeability pore (mPTP) with ensuing structural degeneration and consequent myocyte necrosis. We hypothesized the selective sequestration and removal of minimally defective mitochondria (mt) (mitophagy) and their replacement by fusion and fission (mitogenesis) would favor the prosurvival cardiomyocyte phenotype.

**Methods Used:** 8-week old male Sprague-Dawley rats received 4 wks aldosterone/salt treatment (ALDOST). Cellular/subcellular and molecular events were monitored in cardiac tissue, cardiomyocytes and subsarcolemmal mitochondria (SSM) harvested weekly during the prosurvival (wks 1-3) stage of ALDOST and with the appearance of cardiac pathology at wk 4. A separate group received nebivolol, a 3 adrenergic receptor agonist, cotreatment while untreated, age-sex-matched rats served as controls.

**Summary of Results:** Prosurvival phenotype: increased mRNA and protein expression of beclin-1 and LC3, each integral to phagophore genesis; and expression of mitophagy and mitogenesis-related genes to eliminate degenerative mt and preserve mt mass, together with morphologic evidence of autophagosome formation, mt fusion and fission; microscopic scarring scattered throughout the right and left heart; and neivobil cotreatment augmented PGC-1α and Drp1 transcription to enhance mitogenesis and prevent necrosis and scarring.

**Conclusions:** Homeostatic mechanisms preserve myocyte viability early during ALDOST. Intracellular self-repair with removal of minimally defective mt and their replacement (mitogenesis) preserves mt mass and avoids mPTP opening. Through NO-based upregulation of mitogenesis, nebivolol sustains the prosurvival phenotype.

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**TARGETED DELIVERY OF HUMAN INDUCED PLURIPOTENT-ENDOTHELIAL CELLS OVEREXPRESSING INTERLEUKIN-8 RECEPTORS INHIBITS NEOINTIMAL AND INFLAMMATORY RESPONSES TO ENDOLUMINAL INJURY OF THE RAT CAROTID ARTERY**

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**Purpose of Study:** Interleukin-8 (IL8) receptors A and B (IL8RA and IL8RB) on neutrophils membranes bind to IL8 and play a critical role in neutrophil recruitment to sites of injury and/or inflammation. We have generated targeted based cell therapy using adenoviral vectors that contain full length IL8RA or IL8RB DNAs and the green fluorescent protein (GFP) and transduced endothelial cells differentiated from human induced pluripotent stem cells (HIPS-IL8RA/B-Ecs) and controls. In this in vivo study, we hypothesize that transduction of HIPS-EC overexpressing IL8RA/B into rats with balloon injury of the carotid artery will target to the area of injury, decrease inflammatory response, and inhibit neointima formation.

**Methods Used:** 12 wk old male Sprague-Dawley rats received balloon injury of the right carotid artery and were immediately administered i.v. transfusion of HIPS-IL8RA/B into rats with balloon injury of the carotid artery; i.v. transduction of HiPS-ECs equipped with the IL8RA/B homing device mimic the behavior of neutrophils that target to injured arteries; i.v. transduction of HIPS-IL8RA/B/Ecs inhibited inflammatory cells infiltration and decreased pro-inflammatory cytokine levels in injured artery 24 hr post injury; and inhibited neointima formation after endoluminal injury; and human iPSC-Ecs could be used for targeted cell therapy in rats (xenotransplantation).

**Conclusions:** These findings indicate that acute i.v. transduction of HIPS-IL8RA/B/Ecs mimic the behavior of neutrophils that target to the injured blood vessel and provide a novel strategy for the treatment of vascular injury.

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**THE TRANSITIONAL B CELL CHECKPOINT INCOMPLETELY SELECTS CELLS THAT LACKED BONE MARROW PRE BCR EXPRESSION**

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**Purpose of Study:** Expression of the preB cell receptor (preBCR), which is created by the association between a nascent Mu heavy chain (HC) and surrogate light chain proteins (SLC), marks an early bone marrow check point during B cell development. Loss of any of the pre BCR components causes a block in B cell development, allowing only a few B cells expressing an abnormal repertoire to survive and enter the peripheral compartments, a state mimicking many autoimmune diseases. In unpublished studies, we have found that preBCR selection adjusts the composition of third complementary region of the H chain (CDR-H3), thereby limiting autoreactivity and optimizing H chain amino acid content. The extent to which subsequent selection steps can be used to adjust the CDR-H3 repertoire in mice lacking the preBCR is unknown.

**Methods Used:** To test whether CDR-H3 content in mature B cells could be properly regulated in the absence of the preBCR, we sorted mature B cell subsets in the spleen and the peritoneal cavity from SLC-deficient (A5 KO) BALB/c mice and then amplified and cloned their CDR-H3 sequences.

**Summary of Results:** We found that the splenic T1 transitional checkpoint was able to limit the passage of B cells expressing Mu HCs with CDR-H3s using reading frame (RF)2 that would otherwise have been re-selected. Therefore, these A5 KO mice produced IgM dsDNA binding antibodies, suggesting that this T1 selection step was incomplete and failed to control the repertoire.

**Conclusions:** Our findings suggest that the preBCR selection step plays an essential role in controlling autoimmune by controlling repertoire content.
GLUCOCORTICOID LEVELS IN RESISTANT HYPERTENSIVE PATIENTS WITH AND WITHOUT ALDOSTERONE EXCESS

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Purpose of Study: Resistant hypertension (RHTN) is a prevalent and growing clinical problem. Aldosterone excess is common in patients with RHTN. Recently, cortisol (C), cortisone (Cn) levels and the Cn-C ratio have been described as potential additional factors that may contribute to RHTN. Our aim in this study was to evaluate cortisol, cortisone levels and the urinary cortisone to urinary cortisol ratio in resistant hypertension patients with and without aldosterone excess.

Methods Used: We retrospectively analyzed 77 patients seen at the referral Hypertension Clinic at the University of Birmingham Alabama who were evaluated for RHTN. Tests included blood pressure measurement, physical exam, complete metabolic profile, plasma aldosterone, plasma renin activity, and 24 hour urinary aldosterone (UAldo,ug/24 hr), sodium (UNa+, mEq/24 hr), potassium (UK+, mEq/24 hr), urinary cortisone (UC, ug/24 hr), and urinary cortisone (UCn, ug/24 hr) levels.

Summary of Results: In this study, 30 patients had aldosterone excess and 47 had no biochemical evidence of aldosteronism. Patients with aldosterone excess were significantly younger (51.3±11.5 vs 58.6±14.4 yrs, p<0.018) and had more males (63.3% vs 34.05 %, p=0.012). There was no difference for race, BMI, or duration of hypertension. The biochemical evaluation revealed that for patients with aldosterone excess; UAldo (23±12.6 vs. 6.96±3.17, p<0.001), U-C (17.94±4.18 vs 11.52 ±7.65, p= 0.037), U-CN (88.4±47.4 vs 58.5±29.0, p= 0.0038), U-Na+ (223.7±121.4 vs 164.2±76.5, p =0.025), and U-K+ (85±45.3 vs 49.1±24.2, p=0.00033) values were higher than in patients without aldosteronism. The U-CN- to U-C ratio was not significantly different in patients with aldosterone excess and without aldosteronism. However, there was a wide range of U-CN to U-C ratios in patients with aldosterone excess (2.9-18.2) and in patients without aldosteronism (2.25-15.3). Similarly, preliminary analysis revealed that Cn-C ratio was not associated with U-Na+ and U-Aldo levels.

Conclusions: Further analysis is needed to characterize the phenotypes of resistant hypertension patients with and without aldosterone excess and different U-CN-U-C ratios and its effects on blood pressure and its control.

THE SPECTRUM OF ANTIBODY DEFICIENCY ACCORDING TO B CELLS

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Purpose of Study: We routinely characterize B cell subsets of study subjects with recurrent infections who present to the immunodeficiency clinic for evaluation. We focused on subjects with Classical CVID and patients with Recurrent SinoPulmonary Infections (RESPI) with normal immunoglobulin levels. In this report, we present B cell subset data from subjects with hypogammaglobulinemia, or ICR compared to unrelated controls. In screening family members, we found 38% with low immunoglobulin levels compared to 7.8% of controls.

Methods Used: Of the enrolled, 71 subjects were classical CVID, 80 were RESPI, 25 had hypogammaglobulimemia, 77 had ICR, 64 were unrelated controls, and 50 were related controls. The percentage and absolute numbers of IgM memory and class switched memory cells were significantly lowered in CVID compared to unrelated controls (p =0.0081 and p=0.0001), whereas hypogammaglobulimemia subjects had significantly lower percentage of IgM memory cells compared to controls (p=0.0252), and ICR subjects had lower absolute number of IgM memory cells and absolute class number of switch memory B cells compared to controls (p=0.0029 and p=0.0204). Naive cells were significantly lower in subjects with CVID, hypogammaglobulimemia, or ICR compared to unrelated controls. In screening family members, we found 38% with low immunoglobulin levels compared to 7.8% of controls.

Conclusions: We identified a new cohort of patients with recurrent infections with subnormal immunoglobulin levels that do not meet CVID criteria, termed ICR, and this cohort has lower IgM memory and naive B cells compared to controls. By examining B cell subsets, we may be able to delineate subtle differences in patients with antibody deficiencies and treat these patients earlier to decrease infectious complications.

SUPPLEMENTATION IN EXTREMELY PRETERM INFANTS

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Purpose of Study: Vitamin D is critical for normal lung development, function, and immunity. Our Aims were:

1. To evaluate the feasibility and safety of enteral vitamin D supplementation in extremely preterm infants.
2. To determine whether enteral vitamin D supplementation decreases respiratory morbidity in extremely preterm infants.

The primary pharmacologic outcome was serum vitamin D concentration on day 28, and primary clinical outcome was number of days alive and off respiratory support in the first 28 days.

Methods Used: One hundred infants 23/0-27/6 wks gestational age at a regional NICU were randomized to either a placebo group (routine vitamin D supplementation in TPN or feeds; 200 IU/day), a low-dose group (additional 200 IU/day vitamin D for a total of 400 IU: AAP recommendation), or a high-dose group (additional 800 IU/day: ESPGHAN recommendation of 800-1000 IU/day). The study drug was continued for 28 days. Serum 25 (OH) vitamin D was measured by ELISA on days 1, 14, and 28.

Summary of Results: 1) 74% of the infants were vitamin D insufficient at birth (25(OH) vit D<20 ng/mL).
2) Infants in the 800 IU group had higher vit D levels on day 28 than the other two groups (25(OH) vit D in ng/mL: Median [25-75th centiles]: 85 (52-99) vs. placebo group: 22 (13-47) and 200 IU group: 39 (26-57), p=0.05). 3) No differences were noted in days alive off respiratory support (Median [25-75th centiles], days: placebo: 1 (0-11), 200 IU: 0 (0-8), 800 IU: 0.5 (0-22), p=0.63) or other respiratory outcomes among the three groups. Death or BPD by 36w PMA were not different (47% placebo, 56% in 200 IU, 37% in 800 IU, p=0.63). O2 requirement at 36w PMA was not different (42% in placebo, 35% in 200 IU, and 50% in 800 IU, p=0.51). A trend toward lower late onset sepsis was noted in the 800 IU group (placebo: 36%, 200 IU: 32%, 800 IU: 17%, p=0.19). Other outcomes (ROP, proven NEC, IVH) were not different.

Conclusions: The majority of extremely preterm infants were vitamin D insufficient at birth. The ESPGHAN recommendation of 800-1000 IU/day led to serum 25(OH) vit D above recommended concentrations (30-50 ng/mL) in many infants, while routine intake+200 IU/day led to more infants being in recommended range. Larger multicenter trials are required to determine clinical outcomes of vitamin D supplementation in extremely preterm infants.
URINARY NT-PROBNP LEVELS AND ECHOCARDIOGRAPHIC PARAMETERS FOR PATENT DUCTUS ARTERIOSUS
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Purpose of Study: B type natriuretic peptide (BNP) is released from ventricles in response to volume and pressure overload. We hypothesized that urinary NT-proBNP levels correlate with ductal diameter and left atrium to aortic root ratio (LA/Ao). Specific aims of the study included, 1) to determine the correlation between urinary NT-proBNP and echocardiographic findings (ductal diameter, LA/Ao ratio, blood flow velocity) in term infants with PDA, 2) to compare levels of urinary NT-proBNP in infants with PDA and those with no PDA.

Methods Used: Infants with birth weight less than 1000 grams were enrolled prospectively. Ductal diameter, LA/Ao ratio and blood flow velocity were determined by echocardiography when clinically indicated. Urine was collected from infants’ cotton lined diaper (1) 1. Urinary NT-proBNP and urinary creatinine levels were measured simultaneously. Infants were divided into two groups. Group 1 included infants with PDA who required pharmacological treatment; group 2 consisted of infants with no PDA. Urinary NT-proBNP/creatinine ratios were compared between the two groups. A single pediatric cardiologist was assigned to interpret echocardiographic findings and was blinded to urinary NT-proBNP levels.

Results: To date 66 infants were enrolled. Echocardiography was performed on 49 infants. There was no significant difference in mean gestational age (GA) or birth weight (BW) between the two groups, GA: (25.6 ± 0.5 vs 26 ± 1.2 weeks), BW: (833.3 ± 194.2 vs 860.4 ± 94.6 grams). Urinary NT-proBNP/creatinine ratios were significantly higher in babies with PDA compared to those with no PDA (mean 2284 ± 2046 vs 591 ± 527 pg/mg p=0.003). Urinary NT-proBNP/creatinine levels were positively correlated with ductal diameter (0.42, p=0.002) and LA/Ao ratio (0.44, p=0.002) on Pearson correlation. However, there was no statistically significant correlation between Urinary NT-proBNP/creatinine levels and ductal blood flow velocity (0.23, p=0.1).

Conclusions: Preliminary findings indicate that there is a positive correlation between ductal diameter and urinary NT-proBNP. It is a non-invasive and simple method and may be a useful adjunct to echocardiography.

RANDOMIZED TRIAL OF SKIN-TO-SKIN CONTACT TO PREVENT HYPOThERMIA IN TERM NEONATES
Ramani M, Choe EA, Major M, Newton R, Carlo W. UAB, Birmingham, AL

Purpose of Study: To prevent neonatal hypothermia, the World Health Organization (WHO) recommends the thermoregulation care protocol that includes warm delivery rooms, immediate drying, skin-to-skin contact (SSC) as continuously as possible, breastfeeding, delayed bathing and weighing, and training and awareness raising. All elements of the thermoregulation care protocol according to WHO recommendation are practiced in term infants born at the University Teaching Hospital (UTH), Lusaka, Zambia. Our objective is to test the hypothesis that in term neonates SSC as continuously as possible along with the WHO thermoregulation protocol as practiced reduces moderate or severe hypothermia at (a) one hour after birth and (b) at discharge more than the WHO thermoregulation protocol as practiced alone.

Methods Used: Term neonates (gestational age ≥37 weeks) born at UTH were randomized in two phases (Phase 1: birth to 1 hr, Phase 2: 1 hr to discharge) to either SSC as continuously as possible along with the thermoregulation protocol or a control group (Phase 1 were re-randomized at 1hr for Phase 2 of the study). The primary outcome was moderate or severe (<36.0°C, axillary temperature) hypothermia at (a) one hour after birth or (b) at discharge.

Summary of Results: Neonates randomized to the SSC groups (birth to 1 hr, N=101; 1hr to discharge, N=90), or to the control groups (birth to 1 hr, N=102; 1hr to discharge, N=90) did not differ in their baseline characteristics. Ambient temperatures averaged 27.7±1.0 oC: 82.0±1.9 oF (mean±SD) and did not differ between the groups. Duration of SSC in the SSC group in Phase 1 was 36/15 minutes (mean±SD) and in Phase 2 was 55±25% (mean±SD) of the length of hospital stay. The proportion of neonates with moderate or severe or moderate hypothermia in the SSC group did not differ compared to the control group at 1 hr after birth (25% versus 27%, RR=0.93, CI=0.79-1.4, p=0.78) or at discharge (7% versus 2%, RR=2.8, CI=0.6-13.9, p=0.16).

Conclusions: In facilities where ambient temperature is maintained appropriately, increased duration of SSC either at birth or at 1hr after birth does not reduce moderate or severe hypothermia in term infants compared to the practiced WHO thermoregulation protocol.

BASEAL AND MAXIMAL OXYGEN CONSUMPTION RATES OF HUMAN UMBILICAL VENOUS ENDOTHELIAL CELLS (HUVEC) ARE DECREASED IN INFANTS WHO DIE OR DEVELOP BRONCHOPULMONARY DYSPLASIA (BPD)
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Purpose of Study: Mitochondrial bioenergetics play a critical role in the pathogenesis of many disorders characterized by increased oxidative stress. Our aims were:

1. To identify possible differences in bioenergetic profiles of HUVECs obtained from extremely preterm infants that could be associated with increased risk for death or BPD.
2. To identify clinical risk factors that could alter HUVEC bioenergetic profiles.

Methods Used: HUVECs were isolated from cord segments of 24 infants between 23-32 weeks gestation and grown in 5% CO2 at 37°C. Mitochondrial bioenergetics were assessed using a Seahorse Bioscience XF Analyzer. After baseline oxygen consumption rates (OCR) were obtained, mitochondrial stress tests were performed using Oligomycin (ATPase inhibitor), FCCP (protonophore that makes mitochondria respire at maximal rates), and rotenone/antimycin A (stop oxidative phosphorylation at complexes I and III), NICHD physiologic classification was used to define BPD.

Summary of Results: 1) Infants who developed BPD or died had lower baseline and maximal OCR (Median [25-75th percentiles]) than infants that survived without BPD: 145.3 [66.4-189.8] and 155.4 [63.8-248.7], p<0.005 and p<0.05 respectively. A trend towards lower maximal OCR for African-American infants compared to Caucasian infants was noted: 55.9 [48.9 - 55.9] vs. 66.4 [55.8-110.5], p<0.05 for both.

2) Infants who developed BPD had a higher O2 fraction consumed for purposes other than ATP synthesis; Median [25-75th percentiles]: 0.75[0.54-0.87] vs. infants that survived without BPD: 0.51[0.45-0.56], p<0.05.

3) Infants exposed to chorioamnionitis had lower baseline and maximal OCR; Median [25-75th centiles]: 55.9[47.8-81.1] and 60.4[50.5-92.3] vs. Infants not exposed to chorioamnionitis: 133.8 [65.7-169.2], p=0.06.

Conclusions: With more O2 used for purposes other than ATP synthesis, vascular endothelial mitochondria of infants at risk for BPD may produce increased amounts of free radicals leading to mtDNA damage and reduced mitochondrial function that may contribute to BPD pathogenesis. Measures of ROS production and mtDNA damage in the HUVEC are ongoing.
Purpose of Study: The balance between nephron progenitor cell renewal and differentiation determines the ultimate nephron number, and consequently, susceptibility to renal hypoplasia, hypertension, and chronic kidney disease. In spite of emerging studies implicating chromatin-based mechanisms in the regulation of nephron endowment, little is known regarding the epigenetic factors that orchestrate nephrogenesis. Dot1L, the only known H3K79 methyltransferase, is essential for early embryogenesis, but its role in organogenesis is largely unknown. This study aims to characterize the nephron developmental defects in mice with targeted disruption of Dot1L in nephron progenitor cells.

Methods Used: 1. Six2Cre-GFP transgenic mice were crossed to conditionally Dot1L-flox mice to generate nephron progenitor-specific deletion of the catalytic domain of the Dot1L gene. 2. Kidneys harvested from Six2Cre-Dot1Lfox and wild-type mice at embryonic day E14.5, E17.5, and postnatal day 20 and P35 were subjected to morphological and histological studies, section HE staining, and nephron counts. 3. Immunofluorescence staining was performed to characterize the phenotypic alterations at the molecular level.

Summary of Results: Dot1L mutant kidneys displayed an aberrant phenotype as early as E17.5, which can be summarized as follows: 1. A significant reduction in kidney size and surface petechial hemorrhagic spots. 2. Gene dosage-dependent reduction in multiple stages of nephrogenesis (comma, S-shaped and capillary loop glomeruli) but not renal vesicles. 3. Intraglomerular hemorrhage involving 25% of capillary loop glomeruli, and 4. Premature depletion of H3K79me2-deficient nephron progenitor cells, poorly developed nephrogenic zone, and less complex renal tubular system. Conclusions: Dot1L deficiency in nephron progenitor cells causes nephron deficit and abnormal glomerulogenesis, eventually leading to renal hypoplasia. The relative sparing of renal vesicles in Dot1L mutant kidneys suggests that histone methylation on lysine 79 targets the gene-regulatory networks controlling nephron progenitor differentiation rather than induction.

NEUROFIBROMIN REGULATES MONOCYTE/MACROPHAGE FUNCTION

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Purpose of Study: Neurofibromin results from mutations in the NF1 gene and functions as a negative regulator of Ras activity. Loss of neurofibromin, as observed in persons with neurofibromatosis type 1 (NF1), sensitizes cells to growth factor stimulation and aberrant Ras activation. Some NF1 patients have developmental deficits and abnormal glomerulogenesis, eventually leading to renal hypoplasia. The relative sparing of renal vesicles in Dot1L mutant kidneys suggests that histone methylation on lysine 79 targets the gene-regulatory networks controlling nephron progenitor differentiation rather than induction.

Methods Used: We utilized a carotid artery ligation model to induce neointima formation. The left common carotid artery was ligated proximal to the bifurcation and mice were allowed to recover for 28 days. Control and ligated arteries were analyzed for neointima formation. 

Summary of Results: The balance between nephron progenitor cell renewal and differentiation determines the ultimate nephron number, and consequently, susceptibility to renal hypoplasia, hypertension, and chronic kidney disease. In spite of emerging studies implicating chromatin-based mechanisms in the regulation of nephron endowment, little is known regarding the epigenetic factors that orchestrate nephrogenesis. Dot1L, the only known H3K79 methyltransferase, is essential for early embryogenesis, but its role in organogenesis is largely unknown. This study aims to characterize the nephron developmental defects in mice with targeted disruption of Dot1L in nephron progenitor cells.

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inner membrane permeability pore (mPTP) with ensuing structural degeneration and consequent myocyte necrosis. We hypothesized the selective sequestration and removal of minimally defective mitochondria (mt) (mitophagy) and their replacement by fusion and fission (mitogenesis) would favor the prosurvival cardiac myocyte phenotype.

**Methods Used:** 8-wk-old male Sprague-Dawley rats received 4 wks aldosterone/salt treatment (ALDOST). Cellular/subcellular and molecular events were monitored in cardiac tissue, cardiomyocytes and subarcosomal mitochondrial (SSM) harvested weekly during the prosurvival (wks 1-3) stage of ALDOST and with the appearance of cardiac pathology at wk 4. A separate group received nebivolol, a β3 adrenergic receptor agonist, cotreatment while untreated age/sex-match rats served as controls.

**Summary of Results:** Prosurvival phenotype: increased mRNA and protein expression of beclin-1 and LC3, each integral to phagophore genesis; and autophagosome formation, mt fusion and fission; microscopic scarring scattered throughout the right and left heart; and nebivolol cotreatment augmented PGC-1α and Drp1 transcription to enhance mitogenesis and prevent necrosis and scarring.

**Conclusions:** Homeostatic mechanisms preserve myocyte viability early during ALDOST. Intracellular self-repair with removal of minimally defective mt (mitophagy) and their replacement by fusion and fission (mitogenesis) preserves mt mass and avoids mPTP opening. Prosarcotic phenotype: mPTP opening with persistent expression of mitophagy and mitogenesis-related genes to eliminate degenerative mt and preserve mt mass, together with morphologic evidence of autophagosome formation, mt fusion and fission; microscopic scarring scattered throughout the right and left heart; and nebivolol cotreatment augmented PGC-1α and Drp1 transcription to enhance mitogenesis and prevent necrosis and scarring.

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**PREDICTORS OF SURVIVAL OF THERAPEUTIC HYPOTHERMIA BASED ON ANALYSIS OF A CONSECUTIVE AMERICAN INNER CITY POPULATION OVER 4 YEARS**

Roberts BA, Toca H, Martinez J. LSU Health Sciences Center, New Orleans, LA.

**Purpose of Study:** Therapeutic hypothermia (TH) is the international standard of care for infants born asphyxiated. Patients with near cardiac arrest, but criticism focuses on poor outcomes. We sought to develop criteria to identify American urban patients more likely to benefit from TH.

**Methods Used:** Retrospective chart review of 107 consecutive adults undergoing TH in downtown New Orleans from 2010-2014 yielded records for 99 patients with all 44 survivors or families contacted up to four years.

**Summary of Results:** 69 males and 38 females with a mean age of 60.2 ± 13.6, 20 minutes ROSC in non-shockable rhythms had 19 patients with 2 survivors (8%, p=.001). Survivors of shockable rhythms showed 19 (100%) living post TH. 15 survivors (79%, 20 minutes were 21 patients with 15 (71%) survivors (p=.001). Time >20 minutes until ROSC in shockable rhythms had 5 patients with 3 survivors (78%, p=.001). Presenting in shockable rhythms with ROSC <20 minutes were 54 patients with 18 survivors (33%, p=.001). ROSC <20 minutes in non-shockable rhythms had 19 patients with 2 survivors (8%, p=.001). Survivors of shockable rhythms showed 19 (100%) living post TH. 15 survivors (79%, n=19, p=.001) had CPC score 1 or 2 with 4 survivors (21%, n=19) having a CPC score of 3. A total of 25 survived non-shockable rhythm. Acute survival of patients with non-shockable rhythm showed 18 expired <72 hours (72%, n=25) with long term survival of 4 patients (5%, n=74) and CPC scores of 1 or 2 (p=.001). Interestingly, patients with time to ROSC <20 minutes exhibiting more than one loss of sustained ROSC showed 100% mortality (p=.001). Patients presenting with shockable <20 minutes ROSC had overall survival of 70% (p=.001), but those undergoing >3 cardiac rhythm changes had 100% mortality (p=.001).

**Conclusions:** Patients presenting with shockable rhythms undergoing TH had overall acute survival of 70% followed by long term survival of 100% after 4 years. In contrast, patients presenting with non-shockable rhythm had long term survival of 5%. TH is not recommended for patients presenting with non-shockable rhythm and requiring greater than 20 minutes for restoration of ROSC.

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**UNDERLYING CAUSES OF HEART FAILURE WITH REDUCED EJECTION FRACTION IN A HISPANIC POPULATION**

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**Purpose of Study:** The objective of this study was to determine the underlying causes of heart failure with reduced ejection fraction (HFrEF) in Puerto Rican patients referred for heart transplant.

**Methods Used:** We analyzed the medical records of 300 patients with heart failure referred to the Heart Failure and Transplant Clinics of the University of Puerto Rico School of Medicine. We evaluated the clinical history and physical examination, laboratory data, electrocardiogram, echocardiogram, radionuclear ventriculography, and cardiac catheterization. After reviewing the records the etiology for HFrEF was identified in each patient.

**Summary of Results:** The patients were grouped into the following categories according to underlying cause of HFrEF: idiopathic dilated cardiomyopathy 52%, ischemic heart disease: 33%, peripartum cardiomyopathy: 2%, cardiomyopathy due to myoccarditis: 2%, end-stage hypertrophic cardiomyopathy: 2%, hypertension: 2%, substance abuse: 2%, chemotherapy associated: 2%, and other causes (Left Ventricle Non-Compaction Cardiomyopathy, Stress Induced Cardiomyopathy, Congenital Heart Disease): 3%. A total of 157 cases of idiopathic dilated cardiomyopathy were identified (64% males, 36% females) and 97 cases of ischemic heart disease causing HFrEF (80% males, 20% females).

**Conclusions:** Idiopathic dilated cardiomyopathy is the leading cause of HFrEF in Puerto Rican patients referred for heart transplant, followed by ischemic heart disease in both males and females. This study is, to our knowledge, the first to report the different causes of HFrEF in the Puerto Rican. Literature on heart failure in Hispanics contains scanty primarily restricted to subgroup analysis of HF clinical trials and Puerto Ricans have accounted for less than 10% of the population in those studies. We feel this study is relevant since our population has not been well represented in previous studies and it will provide further understanding of HFrEF in Puerto Rico.

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**THE EFFECT OF PEDIATRIC EARLY WARNING SYSTEM SCORING UPON ADMISSION FROM THE PEDIATRIC EMERGENCY DEPARTMENT ON EMERGENCY RESPONSE CALLS**

Frascogna MN1, Merkle E2, Dowdy K1, Seals S3, 1University of Mississippi Medical Center, Jackson, MS; 2University of Mississippi Medical Center, Jackson, MS and 3University of Mississippi Medical Center, Jackson, MS.

**Purpose of Study:** Pediatric Early Warning System (PEWS) scores are an easy to use diagnostic tool to detect patient deterioration. The goal of this study was to determine if using PEWS scores in the pediatric emergency department (PED) at the time of admission helped to ensure placement in the appropriate inpatient unit, as determined by a decrease in the number of emergency response calls within six hours of admission.

**Methods Used:** A retrospective chart review of the six months before and after the initiation of PEWS scoring upon admission from our urban, tertiary-care PED was conducted to determine the number of patients who had emergency response calls within six hours of admission. Data was analyzed using logistic regression.
Summary of Results: 1978 patients were admitted from the PED in the six months before and 2027 patients in the six months after the initiation of PEWS scoring. The percentage of patients admitted from the PED who required an emergency response call within six hours of admission dropped from 1.77% before to 0.79% after PEWS scoring began, a 55% reduction (p=0.0070). The odds of being placed in the appropriate inpatient unit post-PEWS scoring were 127% greater than before PEWS scoring was initiated.

Conclusions: Assigning PEWS scores to patients admitted from the PED reduced the number of emergency response calls in the period immediately following admission, indicating more accurate patient placement.

A CASE-CONTROLLED STUDY OF ROTAVIRUS VACCINE EFFECTIVENESS
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Purpose of Study: Since the implementation of rotavirus (RV) vaccines, RV disease in children has decreased dramatically. Clinical trials of RV vaccines have demonstrated high efficacy against severe outcomes such as hospitalization. Vaccine effectiveness (VE) is usually lower due to heterogeneity in the population and differences in outcomes measured. We evaluated RV VE using a case-control methodology in children after implementation of RV vaccination.

Methods Used: Stool samples submitted to Children's Healthcare of Atlanta microbiology laboratory were tested by Remel RV Xpect from July 2007 - June 2014. Subjects were excluded if duplicate, >8 months of age, birth before 1 January 2006, partially vaccinated for RV, or if vaccination could not be verified in the electric Georgia vaccine registry. Controls were RV-negative children with stool sent for RV testing. Controls were matched if both date of birth (DOB) and test date were +/- 30d from those of cases. VE was calculated among children with available controls as 1 - (vaccination rate of cases/vaccination rate of controls). For subjects without matched controls, a separate analysis of unmatched subjects was conducted using +/-90d from DOB and test date.

Summary of Results: RV was identified in 724 (12%) of 5,840 stools. Of these 724 RV-positive stools, 336 (46%) were not eligible (107, age >8 months; 143, birth before 2006; 62, partial vaccination; 24, unavailable vaccine information). Among the 388 eligible children, 71 (18%) fully RV vaccinated children were diagnosed with RV gastroenteritis (mean age=1.9 yrs). A total of 892 controls (range 1-11 per case) of +/-30d were identified for 321 eligible subjects (83%). Overall VE for 2008-2014 was 56% and was similar among children 23 yrs of age (n=59; VE 55%). VE appeared higher in 2008-2010 (73%) than in 2011-2014 (45%). Among the 67 subjects who did not have +/-30d matched controls, 46 (69%) were matched to a control with expansion to +/-90d for DOB and test date. VE in these 46 subjects was 44% and mean age was 3.8 yrs.

Conclusions: VE was lower in this study of outpatients, emergency room visits, and hospitalizations as compared to other studies that have enrolled primarily hospitalized children. VE did not appear to decline in children 3 years of age.

IMPACT OF A TRIAGE TOOL AND STANDARDIZED ORDER SET FOR EARLY RECOGNITION AND GOAL-DIRECTED THERAPY IN PEDIATRIC SEPSIS
Pruitt CM1, Haggerty S2,1University of Alabama at Birmingham, Birmingham, AL and 2University of Alabama at Birmingham, Birmingham, AL.

Purpose of Study: Sepsis is a leading cause of pediatric mortality worldwide. Despite widely accepted clinical parameters for its recognition and management, and in stark contrast to the adult literature, scant evidence exists on early prediction and management of sepsis in children.

Methods Used: As part of a collaborative effort through the Children's Hospital Association and the American Academy of Pediatrics, our emergency department (ED) implemented both a tool for recognition of possible sepsis at triage, and a standardized order set for management. We retrospectively compared subjects with likely sepsis in the 2 months prior to implementation of this “pathway” with those during the first 2 months of its utilization. To ensure full capture of subjects in the pre-pathway group, as well as those not detected by the pathway post-implementation (thus reducing bias), both ED and inpatient charts were extensively reviewed using various strategies to detect all subjects treated for presumed sepsis. Primary endpoints were time to physician assessment; time to first isotonic fluid bolus; time to antibiotics; and time to third fluid bolus. Descriptive statistics are presented as medians. Data were analyzed with a two-tailed Mann-Whitney U test, with P<0.05 considered statistically significant.

Summary of Results: Thirteen subjects were treated for likely sepsis during the 2 months prior to pathway implementation. Following rollout of the pathway, 44 subjects were treated for likely sepsis (26 of which were recognized through the pathway). Time to physician assessment did not differ before or after pathway implementation (6 vs. 3 minutes, P=0.199); however, when only comparing subjects detected by the pathway, assessment time was improved (9 vs. 3 minutes, P=0.014) in the latter 2 months. Times to first fluid bolus (73 vs. 24 minutes, P=0.001), antibiotics (59 vs. 34 minutes, P=0.042), and third fluid bolus (124 vs. 64 minutes, P=0.016) were significantly decreased after pathway implementation.

Conclusions: Adoption of a triage trigger tool and standardized order set for pediatric sepsis led to decreased times for goal-directed therapies for these patients. Further research is needed to gauge how these measures impact patient outcomes.

PARENTAL PERCEPTIONS OF SAFETY, EFFICACY AND LEVEL OF CONFUSION REGARDING OVER THE COUNTER COUGH AND COLD MEDICATIONS
MacGeorge C, McElligott J, Carter E, Hinton E, Basco W, Roberts JR, Medical University of South Carolina, Charleston, SC.

Purpose of Study: The AAP does not recommend over the counter (OTC) cough and cold medications for any child less than 6 years due to confusion about safety and a lack of effectiveness in this age group. The objectives of this study were to determine parents’ perceptions of the safety and efficacy of OTC cough and cold medications and the frequency that they administer them to their children. A secondary aim was to identify the level of parental concern about selecting appropriate OTC medications.

Methods Used: We distributed 30 surveys to parents who brought a child aged 1-6 years for a well check to each of 16 practices in the South Carolina Pediatric Practice Research Network. An additional 60 Spanish surveys were sent to 2 practices with large Spanish-speaking populations. The survey consisted of questions with yes or no and 5 point Likert scales responses. We compared responses between various demographic subgroups (gender, race, insurance type and highest level of education) using univariate analysis and logistic regression.

Summary of Results: Of the 540 surveys distributed, we received 450 (83 %). 35 surveys were excluded for not having a child aged 1-5 years leaving 395 surveys for analysis. 76% of parents reported using a cough and cold medication in their child age 1-6 years over the last year. 36% felt they were safe including 50% of African American parents and 29% of all other races surveyed. Multiple logistic regression results revealed that African Americans were more likely to view medications as safe; (OR = 2.2, 95% CI 1.3, 3.7, p=0.003) as were parents with less than a high school education (OR = 2.3, 95% CI 1.1,4.9, p=0.03). Parents with less than a high school education were also more likely to report that selecting a cough and cold medication was confusing although this model did not reach statistical significance (model p=0.0675). The model evaluating factors related to perception of efficacy of medications did not converge.

Conclusions: Most parents gave their child OTC medications despite AAP recommendations. Although some racial and educational differences exist, a large proportion of parents view OTC medications as safe. Interventions are needed to educate parents regarding the lack of safety and efficacy of these medications to decrease their usage.
### PROVIDER AND PRACTICE PEDIATRIC ANTIBIOTIC PRESCRIBING VARIATION IN THE TREATMENT OF RESPIRATORY TRACT INFECTIONS

Doshi P, Onukwube J, Robbins J, Goudie A, Ray O. University of Arkansas for Medical Sciences, Little Rock, AR.

**Purpose of Study:** Antibiotics are among the most frequently prescribed classes of medications in the pediatric population. Inappropriate prescribing is a significant reason for antibiotic resistance. Evidence-based clinical guidelines for the appropriate prescribing of antibiotics have been widely endorsed and disseminated. The purpose of this study was to identify provider and practice characteristics where inappropriate pediatric prescribing rates are high in the treatment of viral respiratory tract infections (RTI) and study variations to be able to target quality improvement interventions.

**Methods Used:** National Ambulatory Medical Care Survey & National Hospital Ambulatory Medical Care Survey data for years 2006-2010 were used. The focus was on prescribing practices RTI in the office-based (OB) & emergency department (ED) settings. Analyses were restricted to OB setting between 1-17 yrs. Statistical analyses accounted for the complex survey design.

**Summary of Results:** Inappropriate antibiotic prescribing rates for viral RTIs increased from 28.6% in 2006 to a high of 37.5% in 2009 before declining to 33.8% in 2010. Inappropriate antibiotic prescribing differences are observed across several provider and practice characteristics: Doctors of Osteopathy compared to Doctors of Medicine (50% vs. 34%; p<0.05), General or Family Practice physicians compared to Pediatrics (47% vs. 30%; p<0.05); Urgent Care Centers compared to Federally Qualified Health Centers (43% vs. 30%; p<0.05); and practices seeing <25% Medicaid patients compared to those seeing >75% Medicaid patients (39% vs. 17%; p<0.05).

**Conclusions:** More than one in three children is inappropriately prescribed an antibiotic to treat a viral respiratory tract infection. Significant variation is evident across provider and practice settings. Based on these results, specific provider and practice characteristics can be targeted for educational interventions to reduce inappropriate antibiotic prescribing. Overall, inappropriate prescribing rates are too high and increased adherence to prescribing guidelines is needed.

### CELL PHONE USE WHILE DRIVING: A SURVEY OF ALABAMA TEENAGERS

Monroe K1, Hardwick WE2, King WD1. 1Univ of Alabama, Birmingham, AL and 2Auburn University, Auburn, AL.

**Purpose of Study:** Despite the passage of a 2005 Graduated Teen Driver’s License Law and a 2012 law banning texting and driving, Alabama continues to have one of the highest death rates for teen drivers in the country. Alabama teens were surveyed to assess cell phone use while driving.

**Methods Used:** A 2013 convenience sample survey of Alabama teens self reported driving behaviors over the preceding thirty days was conducted. The group of teens responding “yes” to cell phone use while driving was compared to the group responding “no.” Other comparison points included their knowledge of the Alabama driving laws, participation in driver’s education classes, observation of distracted driving behavior in peers or parents and involvement in an automobile crash as a driver.

**Summary of Results:** A total of 1024 teens participated. 298 teens were excluded (20 did not have cell phones, 235 did not drive, and 43 did not answer). Of the 726 valid respondents, 372 (51.2%) answered yes to using a cell phone while driving.

White teenagers had 1.8 (95%CI (1.3, 2.5)) times the odds of using a cell phone while driving compared to non-white teens. Teenagers who observed their parents use a cell phone while driving had 2.1 (95%CI (1.5, 2.9)) times those whose parents did not use cell phone. Teens who had been passengers with friend drivers who texted had 3.2 (95%CI (2.2, 4.6)) times the odds of using a cell phone and driving. Teenagers who answered yes to using a cell phone while driving had 2.8 (95%CI (1.8, 4.2)) times the odds of being involved in a car crash as a driver.

There was no significant difference between the two groups in regard to having taken a driver’s education class or having signed a driver safety contract. Also there was no significant difference between the two groups in regard to having discussed the dangers of texting and driving with a parent or their physician.

**Conclusions:** Cell phone use while driving place teenage drivers, their passengers and potentially others on the road at risk for crash-related injury. The behavior of peers and parents appears to have an influence on teens and their choice to use cell phones while driving. Unfortunately educational interventions such as driver’s education classes and driver’s safety contracts were not shown to be as effective in this study in influencing teen behavior.

### SHORTENING NEONATAL INTENSIVE CARE UNIT STAY AND IMPROVING MEDICAL OUTCOMES FOR PREMATURE INFANTS USING EVIDENCE-BASED MUSIC THERAPY PROTOCOLS

Standley JM. Florida State University, Tallahassee, FL.

**Purpose of Study:** The purpose of this presentation is to improve NICU Quality of Care by educating physicians on research demonstrating benefits of four reimbursable NICU-MT protocols (music therapy provided by a Board Certified Music Therapist with a Special Certificate for training in NICU care). Research shows that these protocols shorten NICU length of stay, promote neurological maturation, and increase oxygen saturation of premature infants.

**Methods Used:** The developmental and medical benefits of evidence-based music therapy for preterm infants in the NICU were identified by meta-analysis and research. Four highly beneficial effects (overall large, significant effect size, Cohen’s d=8.2, p<0.00). Research shows that when selected protocols from this study are clinically implemented with individual, developmentally appropriate referral by physicians, clinical results mirror research findings with greatly shortened NICU stays for infants born <1000g. These protocols include music listening for neural protection that reduces stress, increases oxygen saturation, and masks aversive auditory stimuli; contingent music to reinforce non-nutritive sucking and to promote independent nipple feeding; music to maintain homeostasis during developmentally appropriate, layered stimulation to reduce over-stimulation and promote neurological tolerance; and contingent music to promote patient triggered ventilation and shorten time to extubation.

**Summary of Results:** NICU-MT clinical protocols consistently contribute to earlier discharge of premature infants, reduced time for gavage feeding with faster transition to independent nipple feeding, reduced time on ventilator, and reduced medical costs. NICU-MT improves quality of care, increases patient satisfaction and is highly economical with an average savings of $10,000-$20,000/infant.

**Conclusions:** These consistently significant results justify consideration for the inclusion of evidence-based NICU-MT protocols in best practice standards for treatment of preterm infants.

### DOES BULLYING CORRELATE TO DEPRESSION IN CHILDREN/ADOLESCENTS?

Fosseca A2, Burns JJ1. 1FSU, Pensacola, FL and 2Medical University of South Carolina, Charleston, SC.

**Purpose of Study:** To determine if there is a correlation between bullying and depression in children/adolescents.

**Methods Used:** Two validated self-report questionnaires were administered to middle and high school students at an urban city Medicaid teaching clinic.

Questionnaire 1: Clinical Depression Inventory (CDI). This is a 12 item checklist which helps assess depressive syndrome in children.

**Conclusions:**
Questionnaire 2: Multidimensional Peer Victimization Score (MPVS): This is a 16 item survey which measures bullying. This includes a total bullying score and 4 bullying subtypes: Social, Verbal, Physical and attacks on property.

The correlation between depressive symptoms as measured on CDI vs. bullying total and subscores from the MPVS was determined using Spearman correlation in SPSS.

Summary of Results: Statistically significant correlation was found between bullying total score on MPVS and depression score on CDI (Spearman Correlation Coefficient of 0.501; p = 0.007). Also statistically significant correlation was found between physical, verbal and social bullying subscores and depression.

Conclusions: A correlation between bullying and depressive symptoms was found. This requires further assessment using longitudinal study design.

### Table: Summary of Results

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<th>Social Bullying</th>
<th>Property Bullying</th>
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**PERFORATED GASTRIC ULCER IN A PEDIATRIC PATIENT WITH HYPERMOBILITY SYNDROME**

Jamgade A, Kielbasa J, Fits S, Hayes J, Singh N. University of Oklahoma HSC (tulsa), Tulsa, OK.

**Case Report:** Introduction: Peptic Ulcer Disease (PUD) is uncommon in children, and perforation is rarely considered in the differential diagnosis of abdominal pain. We report a case of a perforated gastric ulcer in a 16-year-old female with a hypermobility syndrome (HMS).

Case Description: The patient presented to the emergency department (ED) with a 3-week history of intermittent left upper abdominal pain. Two weeks prior she was prescribed ibuprofen for costochondritis. She reported one episode of hematochezia, but no vomiting or hematemesis. Family history is significant only for endocrine tumors. She was admitted for evaluation of abdominal pain and discharged the next day with polyethylene glycol and ibuprofen. She returned to the ED within 24 hours after experiencing severe abdominal pain.

In the ED, she was tachycardic and in severe distress. Examination revealed diffuse abdominal tenderness with guarding. Labs were remarkable only for leukocytosis (24,000 cells/mm3) and bandemia. CT of the abdomen showed pneumoperitoneum with an edematous stomach.

Emergent exploratory laparotomy revealed a 1.5 mm perforation of the anterior wall of the stomach, which was closed. The patient was discharged on a proton pump inhibitor for 6 weeks.

Upper endoscopy 8 weeks later showed a well-healed ulcer. Biopsy for Helicobacter Pylori infection, serum gastrin, and parathyroid level were negative. The patient diagnosed by her pediatrician with a HMS.

Discussion: Peptic disorders account for less than 5% of children presenting with abdominal pain. Only four cases of perforated peptic ulcer in children have been reported in North America, most commonly perforated duodenal ulcers, as opposed to our patient with a gastric ulcer perforation.

We postulate an increased risk for perforation due to abnormal collagen structure secondary to her HMS. Nonsteroidal anti-inflammatory (NSAID) use may have increased the risk of perforation.

Conclusion: PUD should be considered in the differential diagnosis of epigastric pain, especially in the setting of NSAID use. HMS may increase the risk of perforation. Further studies are needed to demonstrate if patients with HMS are at increased risk for perforation.

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**PAIN AND PAIN IMPACT IN YOUTH WITH SICKLE CELL DISEASE UNDERGOING CHRONIC TRANSFUSION THERAPY**

Maxwell SL1, Schlenz A2, Kanter J2,3. 1Medical University of South Carolina, Charleston, SC and 2Medical University of South Carolina, Charleston, SC.

**Purpose of Study:** Sickle cell disease (SCD), the most common inherited blood disease in the US, is often associated with recurrent pain and stroke. Chronic transfusion therapy is used for primary and secondary prevention of stroke in SCD; however, few prior studies have evaluated the effects of transfusion therapy on pain and its impact. Transfusion therapy has already been shown to be neuroprotective and may also protect other organs, resulting in less vaso-occlusion, decreased pain, and reduced healthcare utilization. The goal of this study was to describe the experience of pain and its impact in youth undergoing chronic transfusion therapy.

**Methods Used:** The Pediatric Quality of Life Sickle Cell Disease module (PedsQL SCD) was completed by youth with SCD as part of clinical care. Chart reviews were conducted to identify 20 youth above age 10 (M age = 16.2, SD age = 4.88; 11 male, 9 female) who had been receiving chronic transfusion therapy for at least 1 year and had completed the PedsQL SCD. The Pain and Hurt domain, measuring overall pain, and Pain Impact domain, measuring its functional impact, were calculated from this measure. Clinical cutoffs were used to describe functioning in these domains, with higher scores indicating better quality of life.

**Summary of Results:** On average, youth fell in the intermediate range of functioning for the Pain and Hurt (M = 76.67, SD = 25.00) and Pain Impact (M = 70.68, SD = 23.54) domains. For Pain and Hurt, 55% fell in the high functioning range versus 25% in the intermediate and 20% in the low ranges. For Pain Impact, 40% fell in the high functioning range versus 20% in the intermediate and 40% in the low ranges.

**Conclusions:** Our ratings of pain and its functional impact in this population undergoing chronic transfusion therapy were higher on the PedsQL SCD compared to a previous national study of youth with SCD. The ratings in our sample were 10 points higher for Pain and Hurt and 15 points higher for Pain Impact. The youth in our sample were also twice as likely to fall in the high functioning range in both domains. Whereas research has demonstrated the benefits of transfusion therapy for stroke prevention, this study suggests that it may also have positive impacts on pain and pain impact.
Summary of Results: Paired sample t-tests explored changes in BASC-2 T-scores from intake to follow-up. On average, there were decreases in mental and behavioral health problems. At follow-up, students reported significantly lower social stress (t(24)=2.44, p=.022), depression (t(25)=2.18, p=.039), somatization (t(25)=2.70, p=.012), issues with locus of control (t(25)=2.57, p=.017), attention problems (t(25)=2.69, p=.012), and inattention/ hyperactivity (t(24)=2.10, p=.047). For parent-reports, there was a significant increase in perception of the child's resilience (t(9)=3.41, p=.014). In addition, there was a significant reduction in the number of behavioral incidents cited by the school (t(23)=2.16, p=.042). Academic grades were initially unchanged.

Conclusions: Initial findings support the effectiveness of focused mental health treatment in decreasing mental and behavioral problems that disrupt schoolwork. This treatment may increase the likelihood of high school graduation or attainment of a GED.

Allergy, Immunology, and Rheumatology I
Concurrent Session
2:00 PM
Friday, February 27, 2015
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THE SPECTRUM OF ANTIBODY DEFICIENCY ACCORDING TO B CELLS
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Purpose of Study: We routinely characterize B cell subsets of study subjects with recurrent infections who present to the immunodeficiency clinic for evaluation. We focused on subjects with Classical CVID and patients with Recurrent SinoPulmonary Infections (RESPi) with normal immunoglobulin levels in this report. We present B cell subset data from subjects with hypogammaglobulinemia, subjects with an intermediate CVID phenotype termed ICR, and healthy family members who were tested for immunoglobulin deficiency.

Methods Used: Whole blood was collected from over 420 study subjects from 2008 to 2013. Marginal zone-like B cells, IgM+ memory B cells, and class-switched memory B cells were examined from the start of the study; transitional B cells, naive B cells, and immature B cells were analyzed starting in 2012 by flow cytometry. Immunoglobulin levels and HLA haplotype were also determined. Unpaired t-test was used to analyze the means.

Summary of Results: Of the enrolled, 71 subjects were characterized as classical CVID, 80 were RESPi, 25 had hypogammaglobulinemia, 77 had ICR, 64 were unrelated controls. The percentage and absolute numbers of IgM memory and class-switched memory cells were significantly lowered in CVID compared to unrelated controls (p =0.0081 and p =0.0001), whereas hypogammaglobulinemia subjects had significantly lower percentage of IgM memory cells compared to controls (p =0.0252), and ICR subjects had lower absolute number of IgM memory cells and absolute number of class switch memory B cells compared to controls (p =0.0029 and p =0.0204). Naive cells were significantly lower in patients with CVID, hypogammaglobulinemia, or ICR compared to unrelated controls. In screening family members, we found 38% with low immunoglobulin levels compared to 7.8% of controls.

Conclusions: We identified a new cohort of patients with recurrent infections with subnormal immunoglobulin levels that do not meet CVID criteria, termed ICR, and this cohort has lower IgM memory and naive B cells compared to controls. By examining B cell subsets, we may be able to delineate subtle differences in patients with antibody deficiencies and treat these patients earlier to decrease infectious complications.

Purpose of Study: Diversity of the T cell receptor is mainly developed through V(DJ) rearrangement and N addition during TCR development. The product of V(DJ) rearrangement is the CDR3, a region of high variability that recognizes antigen and includes all of the D gene. The Dβ sequence is highly conserved across various species, from trout to mouse to humans. This suggests that there are some natural constraints on the TCR; these constraints are thought to limit deleterious T cells from reaching the periphery. We hypothesize that altering the D region will have an effect on the development of thymocytes.

Methods Used: In order to test our hypothesis, mice were generated with an alteration in their Dβ gene segments. The alterations are a Dβ2 KO (Dβ1), a replacement of the Dβ5 locus with a charged Dβ (DβDKRQ), and a co-placement of the Dβ locus with a hydrophobic DH (DβYTL). Both developing thymocytes and mature splenic T cells were sorted from these mice by flow cytometry based on their expression of either CD44, CD25 and CD28 or CD4 and CD8. RNA from the thymocytes was extracted and the VDJ-beta genes were sequenced using primers to the VB13-1 and to the VBC1. In frame sequences were analyzed using IMGT junction analysis program.

Summary of Results: We have data comparing the T cell repertoires of WT and altered Dβ mutants. When compared to WT mice, the mutant Dβ mice have an altered T cell repertoire in both CDR3B amino acid composition and CDR3B length and these differences can be attributed to the changes in the germline sequence. Changing the Dβ also changes the total T cell number in both developing and mature T cells, with an altered Dβ being selected against. Functional data has also shown an effect of altered responses to both viral infection, as well as self-antigen.

Conclusions: The Dβ germline sequence is affecting the TCR repertoire. In DβDKRQ mice, the repertoire is skewed toward a shorter, more charged distribution. In DβYTL mice, CDR3B repertoire appears skewed toward a more hydrophobic, longer, BCR-like distribution. Ongoing experiments using varied TCR Dβ locus mutants will elucidate the role of the germline sequence on the development of thymocytes as well as function of mature T cells.

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HYDROXYCHLOROQUINE USE IN LUPUS PATIENTS WITH END STAGE RENAL DISEASE
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Purpose of Study: Hydroxychloroquine (HCQ) is considered to be a cornerstone in the management of patients with systemic lupus erythematosus (SLE). However, the use of HCQ in patients with SLE and end stage renal disease (ESRD) has not been well described. The purpose of this study was to assess the use and examine predictors of HCQ use in lupus patients with ESRD.

Methods Used: A retrospective analysis of 9681 cases of lupus in the 2011 United States Renal Data System (USRDS) was conducted. The ICD-9 code 710 was used to identify patients having SLE. All SLE patients in the USRDS were included in the analysis. HCQ use was identified by the generic name. The daily dose was calculated as the product of the strength (200mg) and the quantity dispensed divided by the days’ supply. The overall prevalence of HCQ use among the cohort as well as the mean, mode and range of daily doses was determined. Additionally, the following predictor HCQ use were examined: age, race, gender, type of dialysis, and Medicaid coverage prior to entrance into the USRDS.

Summary of Results: There were 3488 lupus patients (36.0%) who used HCQ. The mean daily dose was 319.5mg (Range [28.1mg, 333.3mg]). The two most frequently found daily doses were 200mg daily (n=1114 [32.9%]) and 400mg daily (n=1981 [53.4%]). Prior Medicaid coverage was associated with a significantly decreased likelihood of HCQ use (OR, 0.53 [95%CI, 0.48-0.58]). Compared to those on hemodialysis, patients using continuous ambulatory peritoneal dialysis were more likely to use HCQ (OR, 1.37 [95%CI, 1.09-1.73]). Conversely, patients on continuous cycling peritoneal dialysis were less likely to be on HCQ (OR, 0.80 [95%CI, 0.68-0.96]). The likelihood of HCQ use increased with age. Compared to those less than 20 years of age, patients between age 20 and 45 (OR, 1.26 [95%CI, 1.04-1.51]), between age 46 and 65 (OR, 1.38 [95%CI, 1.20-1.59]) and between age 66 and 75 (OR, 2.31 [95%CI, 1.83-2.93]), and older than 75 (OR, 2.48 [95%CI, 1.77-3.48]) were all more likely to use HCQ.

ROLE OF Dβ GERMINE SEQUENCE ON CONSTRAINING TCR CDR3 DIVERSITY
Levinson M, Silva Sanchez A, Zhuang Y, Schroeder H. University of Alabama at Birmingham, Birmingham, AL.

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Conclusions: HCQ is used in a minority of patients with SLE who have ESRD on dialysis. Dose adjustment for ESRD is not done in the majority of these patients. Future studies should address the efficacy and toxicity of HCQ dosing strategies in this patient population.

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IDIOPATHIC CD4 LYMPHOPENIA PRESENTING AS PROFOUND HEMOLYTIC ANEMIA

Favier BJ, Huecker M. University of Louisville, Louisville, KY.

Case Report: Introduction: A rare and poorly understood disease, Idiopathic CD4 Lymphopenia, most commonly presents with opportunistic infection. We present the case of an unusual manifestation of Idiopathic CD4 Lymphopenia in a man with profound hemolytic anemia. The patient is a 32 year old male with a 1-month history of worsening back pain and weakness. He initially presented to an outside hospital where CT scan of his chest revealed 4 spiculated masses in the central right lung and a left pleural effusion. Additional studies were as follows; Reticulocyte count 19.01%, LDH 1404 U/L, total bilirubin 5.2 mg/dl (0.8 mg/dl direct), serum haptoglobin <10 mg/dl, ferritin 574 mg/ml, iron 260 mcg/dl. Toxicology and HIV tests were negative. Intensive care and hematology were consulted and packed red blood cells were transfused. Despite receiving 6 units of pRBCs the patient's repeat hemoglobin was 3.5. A direct antiglobulin test was positive for warm and cold autoantibodies and anti-kell antibodies. Serology for Lupus, Sjogren's, and other autoimmune causes was negative. Flow cytometry showed CD4 count of 31, CD4 8% and CD8 56%, and a CD4/CD8 ratio of 15%

The CDC diagnostic criteria for Idiopathic CD4 Lymphopenia include a CD4 count less than 300, CD4 cells less than 20% of T-lymphocytes, lack of HIV infection, and absence of other cause for CD4 cell depletion. Our patient was further managed with steroids and continued transfusion of pRBCs. His anemia improved and he was discharged on hospital day two in good condition.

Discussion: The CDC first described Idiopathic CD4 Lymphocytopenia in 1992 as a form of HIV negative Immunodeficiency. Although the cause of Idiopathic CD4 Lymphopenia remains unknown, it has been commonly associated with opportunistic infections due to the immunocompromised state in 40% of those affected. Hemolytic anemia has been reported in 10% of those affected. Our case demonstrates that profound symptomatic anemia may be the initial presenting symptom in a patient with Idiopathic CD4 Lymphopenia.

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ORGANIZING PNEUMONIA AS PULMONARY MANIFESTATION OF GRANULOMATOSIS WITH POLYANGIITIS

Fowler AH1, Senitko M1, Haynes D1, Majithia V1,2. 1University of Mississippi Medical Center, Jackson, MS and 2G.V. Sonny Montgomery VA Medical Center, Jackson, MS.

Case Report: The classic histologic features of granulomatosis with polyangiitis(GPA) are necrotizing granulomatous inflammation and necrotizing vasculitis in a sino-pulmonary-renal distribution, but other variants have been reported. We report a case of organizing pneumonia variant of GPA on lung biopsy with classic histologic findings in the kidney.

The patient is a 60-year-old male with a 1-month history of worsening shortness of breath, nasal congestion, and hemoptysis along with 20-pound weight loss, subjective fevers and night sweats. Social history is significant for 15 pack year smoking history as well as occasional marijuana use. He initially presented to an outside hospital where CT scan of his chest revealed 4 spiculated masses in the central right lung and a left pleural effusion. On transfer to our hospital, physical exam was significant for hypoxia, tachycardia, diffuse rales in bilateral lung fields and thick blood tinged nasal discharge in bilateral nares.

Basic metabolic panel revealed normal electrolytes with serum creatinine of 1.97 mg/dL. Complete blood count showed a white cell count of 19,000, hemoglobin of 10.4, hematocrit of 31.8 and platelets of 475,000. Urinalysis showed proteinuria, large blood with 25 RBCs and no casts. Urine drug screen was negative. Erythrocyte sedimentation rate was 78 mm/hr. C-ANCA and proteinase 3 antibodies were positive.

Bronchoscopy revealed alveolar hemorrhage and fine needle aspiration of lung nodules was nondiagnostic. Open lung biopsy revealed an organizing pneumonia and respiratory bronchiolitis without evidence of granuloma formation or vasculitis. Renal biopsy showed pauci-immune necrotizing glomerulonephritis with crescents and acute necrotizing vasculitis. The patient was treated with pulse dose steroids and rituximab and discharged home in improved condition.

Granulomatosis with polyangiitis is characterized by necrotizing granulomas with typical sinopulmonary-renal involvement. A variant of lung involvement has been reported to include organizing pneumonia. It is important to recognize these histologic variants as the diagnosis of granulomatosis with polyangiitis may otherwise be missed and appropriate aggressive immunosuppression withheld inadvertently.

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PARAPLEGIA AS INITIAL PRESENTATION OF SARCOIDOSIS WITH NEUROLOGICAL INVOLVEMENT

Jarrell SA, Fowler AH, Majithia V. University of Mississippi Medical Center, Jackson, MS.

Case Report: Sarcoidosis primarily presents with pulmonary involvement. It has rarely been reported to present with isolated neurological disease. We report a 25-year-old African American male with no past medical history, who presented with progressive three week history of bilateral lower extremity weakness, which he noticed after jumping off a porch. He also had urge incontinence and an unintentional 15 pound weight loss. He denied rashes, pulmonary or cardiac symptoms, or family history of autoimmune diseases. The weakness progressed and he could not ambulate and was unable to move his legs. Evaluation revealed an elevated CSF protein and angiotensin-converting enzyme levels, normal multiple sclerosis panels, serum inflammatory makers, TSH, ACTH and ANA, and negative cultures for infections. Chest X-ray revealed bilateral hilar adenopathy. MRI of the brain, lumbar and cervical spine revealed multiple enhancing intramedullary lesions throughout his spinal cord without brain lesions. Bronchoscopy with biopsy of the perihilar lymph nodes was significant for non-caseating granulomatous inflammation consistent with sarcoidosis. He was diagnosed with neurological sarcoidosis due to biopsy results, the appearance of spinal imaging, and the absence of another etiology. Treatment consisted of methylprednisolone 1mg IV daily for five days and subsequently prednisone one mg/kg/day with a slow taper. He responded well to treatment and regained back most of his lower extremity function. Neurologic sarcoidosis should be a consideration in young patients with paraplegia as neurologic complications occur in approximately five percent of patients with sarcoidosis. In our case, it was important get a prompt diagnosis of sarcoidosis to avoid unnecessary interventions and to start empiric steroids in order to prevent worsening of spinal cord impingement.

FIGURE 1
MALIGNANCY MAY NOT BE ASSOCIATED WITH OSTEOPOROSIS IN MALE: RESULTS OF COHORT ANALYSIS AND LITERATURE REVIEW

Chiang E, Aujla K, Majithia V. University of Mississippi Medical Center, Jackson, MS.

Purpose of Study: Malignancy has been considered a secondary risk factor for osteoporosis (OP). Data has shown that female patients with cancers have higher risk to develop OP. The relationship of malignancy and OP in males is unclear. The objective was to examine if malignancy is associated with increased risk of OP in males.

Methods Used: 585 men who underwent DXA performed at UMC from 2005-2012 were included in the analysis of retrospective cohort. Furthermore, a PubMed literature search retrieved a total of 1859 articles using keywords: Male, OP, and Malignancy (limiting to English, pub year 2000 or later, 1655 articles excluded with abstract review, full review due to confounders). Only two articles were found with relevant data and included in analysis. The statistical significance of these results was assessed using T-test and Odds ratio as appropriate.

Summary of Results: Table

<table>
<thead>
<tr>
<th>Malignancy N=60/291</th>
<th>Normal DXA</th>
<th>Osteoporosis</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>18.1% (36/198)</td>
<td>22.9% (24/105)</td>
<td>NS</td>
<td></td>
</tr>
</tbody>
</table>

Cross-sectional, self-reported data survey PMID 17638108

Conclusions: There was no increase in the prevalence of malignancy in male patients with normal DXA vs those with OP in UMC cohort. Similar results were found upon the review of recent literature, where no increased risk seen in a cross-sectional case control study as well as upon multivariate cohort analysis. Two findings stand out in this review: A) very limited data is available to analyze the association of malignancy with OP in males, & B) Available data suggests that malignancy may not increase the risk of developing OP in males. An increased risk of OP with malignancy has been shown in multiple studies in females and is felt to be a significant contributor to morbidity. This difference may be due to the type of underlying malignancy, hormonal factors, or other unknown factors. These findings suggest male sex may be a protective factor against OP in malignancy and warrant further investigation.

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A CASE OF STILLS

Green RM, Engel LS. LSU Health Sciences Center, New Orleans, LA.

Case Report: A 30 year old African American woman with no past medical history presented to the emergency department with a 3-week history of subjective fevers, night sweats, chest tightness, bilateral PIP pain and swelling, right knee arthralgia, and a mildly productive cough. Three weeks prior to admission, the patient completed a 5-day course of azithromycin for right acute otitis media. The ear pain completely resolved, but her other symptoms persisted. Her temperature was 101.3°F and her heart rate was 128 bpm. Physical exam revealed right third PIP edema with decreased flexion due to pain. Initial labs revealed a WBC count of 19,800/μL with neutrophilia and a normocytic anemia with a hemoglobin and hematocrit of 9.6 g/dL and 28.4%, respectively. A urine toxicology screen revealed caffeine and tobacco metabolites. Although the patient fulfilled SIRS criteria, the patient looked remarkably well. Hence, the patient was not started on antibiotics. Blood and urine cultures were negative. On the second hospital day, examination of her bilateral medial thighs revealed a subtle salmon-colored urticarial rash although the patient denied pruritus, pain, or scratching the rash. The patient continued to spike temperatures at least three times per day up to 103°F with a maximum WBC count of 30,300/μL, yet the patient appeared and felt well. CRP was 8.9 mg/dL and ESR was 115 mm/hr. HIV, rheumatoid factor, ANA, hepatitis panel, TPO antibody, parvovirus IgM, Mycoplasma IgM, Monospot, C3 complement, and CMV DNA probe were negative. C4 was elevated most likely secondary to systemic inflammatory response. Her ferritin was 6127.3 ng/mL. The medial thigh rash had fully resolved the next day. Rheumatology consult diagnosed the patient with adult-onset Still's disease based on the constellation of symptoms and abnormal lab results. The patient was then started on prednisone 40mg po daily with resolution of her fever within 24 hours.

DISCUSSION: We present a case of a young patient with a septic clinical picture. When infectious causes were ruled out, rheumatologic etiologies were pursued. The patient met all major criteria and 3 minor criteria from the Yamaguchi scale for diagnosis of Still's disease. This case highlights the importance of maintaining a wide differential when a patient's clinical course is not improving with standard therapy.

14.6 %, CRP 51 IU/L. X-rays of the right shoulder and left hip were both negative for effusion, fractures, or any sign of septic arthritis. MRI of the hips showed small hip joint effusion and synovial synovitis changes bilaterally, along with a focal area of inflammatory degenerative change of right femoral head. Her left hip joint synovial fluid analysis showed WBCs of 86,000/μL and was negative for crystals. Synovial fluid and blood cultures were negative. Rheumatoid factor, anti-centromere antibodies, anti-DNA antibodies, and antinuclear antibodies were negative as well. She showed clinical improvement with Naproxen. The patient was diagnosed with Parry-Romberg Syndrome (PRS) as an infant. At age five, she had a graft of abdominal soft tissue to augment the atrophic tissue. PRS is a rare disorder characterized by facial hemiatrophy of skin and soft tissue. It has been hypothesized that PRS is an immune-mediated process due to the frequent presence of autoantibodies. No definitive pathogenesis has been discovered, and the course of the disease is incredibly variable, thus presenting a significant challenge when managing patients with PRS. Recently, extracutaneous involvement has been described in literature, which supports the theory that PRS is not only a cutaneous disease. The clinical features could include hematologic department with a 3-week history of trunk or leg, atrophy of the tongue, dental and ocular abnormalities, enophthalmos, uveitis, episcleritis and ocular myopathy. Patients may present with inflammatory polyarthritides, and auto-immune antibodies may be present. It is likely that this patient had recurrent inflammatory arthropathy, which went unrecognized prior to this admission. Although the autoantibodies were negative, it is likely that an autoimmune process was responsible for her inflammatory arthropathy.
ONCHOPHAGIA LINKED TO LÖFFLER SYNDROME ON A LOUISIANA PIG FARM
Gipson K1, Avery RH1, Malone JB2, Wall LA2,1, Shah HA2,1. 1LSU Health Sciences Center, New Orleans, New Orleans, LA; 2LSU School of Veterinary Medicine, Baton Rouge, LA.

Case Report: Löffler syndrome, fulminant eosinophilic pneumonitis associated with the larval migratory phase of human parasites, is rarely reported in the United States. Similarly, zoonotic transmission of helminths is an infrequently encountered entity in developed nations.

A previously healthy 8-year-old male was hospitalized with tachypnea, cough, hypoxemia, and fever of one week’s duration. Systemic corticosteroids were initiated for his acute respiratory insufficiency and produced rapid clinical improvement. History revealed exposure to pigs on his family’s farm in southernmost Louisiana, no travel, and no asthma or atopy. The patient cleans the pig pens frequently and has severe onychophagia (fingernail biting).

He demonstrated peripheral eosinophilia (39%), pulmonary eosinophilia (86%), high IgE, diffuse reticulonodular lung opacities, and mixed obstructive and restrictive pulmonary function pattern. Serum Ascaris-specific IgE was profoundly elevated, and he was treated with albendazole. Strongyloides serology was negative, as was an extensive evaluation for other infectious and allergic etiologies. A site visit and laboratory investigation was coordinated with the Louisiana Animal Disease Diagnostic Laboratory at LSU. Ascaris eggs were detected in the pig feces and in the soil immediately surrounding the pens. Ascariasis should be considered even in the absence of travel history, especially in endemic areas such as the southeastern United States. Onychophagia is a highly probable mechanism of zoonotic fecal-oral transmission in this case, and such habits should raise suspicion for soil-transmitted helmint infections. Human transmission of Ascaris suum, the species which commonly infects pigs, has rarely been reported in the United States. Of note, systemic corticosteroids were effective in treating the patient’s acute respiratory compromise due to Löffler syndrome, despite concern for lethal dissemination of the parasite which has been documented with strongyloidiasis in immunocompromised hosts.

UNDERESTIMATING MPA: A GI BLEED MISTAKEN
Patel MM, Patel AR, Engel LS. LSU Health Sciences Center, New Orleans, LA.

Case Report: A 74 year old woman with history of ESRD secondary to pauci-immune crescentic glomerulonephritis and microscopic polyangiitis (MPA) presented with fatigue and shortness of breath and chest pain. She had described menopause and hematemesis prior to her admission. She was found to be anemic with a hemoglobin of 5.1 mg/dl. Chest CT at that time showed patchy rounded ground glass opacities in both lungs. EGD and colonoscopy did not define a source of bleeding. The patient was transfused 2 units packed red blood cells and was discharged with follow up. Two days after discharge, she developed dyspnea on exertion, along with hemoptysis. She denied fevers/chills, weight loss, sick contacts, or history of travel. She was noted to have a temperature of 102.3°F, stable blood pressure and heart rate, oxygen saturation 95% on 2 liters/minute nasal cannula. On physical exam, she had crackles in the right lower lobe. Her hemoglobin was 6.2 mg/dl, hematocrit 18.7%. Her ESR and CRP were both markedly elevated and p-ANCA titer was 1:320. Chest x-ray demonstrated airspace disease within the right lung base, lateral portion of right upper lobe, and lateral portion of middle left lung. Bronchoalveolar lavage was consistent with hemorrhage. She was immediately started on high dose methylprednisolone for 3 days, with the plan to switch to Rituximab thereafter. On day 3, patient had a drop in hemoglobin from 8.3 to 7.4. Chest x-ray now showed worsening airspace. Her oxygen requirements rapidly increased. She was switched to oral prednisone, and underwent five days of plasmapheresis, resulting in mild radiographic improvement; however we were unable to wean the oxygen. She was then started on azathioprine and rituximab. After the administration of rituximab, the patient slowly improved and oxygen requirements were weaned. The patient’s final diagnosis was microscopic polyangiitis (MPA) with diffuse alveolar hemorrhage.

Discussion: MPA is an ANCA-associated vasculitis characterized by profound constitutional symptoms and majority of patients have glomerulonephritis. Diffuse alveolar hemorrhage is associated with 10-50% of cases. Treatment is guided by disease severity. The presence of alveolar hemorrhage is a hallmark of severe disease and should be treated with plasmapheresis, high dose steroids and rituximab or cyclophosphamide.
Conclusions: We considered this abnormal elevation of HDL-C be due to an elevation of dysfunctional HDL-C. We think this rapid change and constant elevation of this lipoprotein can be considered a biomarker of rejection. This change can be used as a marker to change the rejection protocol to a more aggressive one or find a new method to intervene with HDL-C to avoid this elevation of dysfunctional lipoprotein. This will avoid the massive accumulation of atheroclerotic material in the aorta and coronaries, and in this way avoid this rejection abnormality and death.

HEMOCONECTRATION AS A MARKER OF DECONGESTION IN THE MANAGEMENT OF ACUTE DECOMPENSADED HEART FAILURE AND ITS ROLE AS A PREDICTOR FOR READMISSION

Ibrahim S, Al-Saffar E. University of Florida - Jacksonville, Jacksonville, FL.

Purpose of Study: Congestive heart failure (CHF), is the leading cause of hospitalizations, and death in developed countries. Effective decongestion may reduce intravascular volume leading to hemocoencentration, with improved survival. It is unknown whether hemocoencentration results in decreased rates of readmission and/or delayed times from discharge to readmission. We hypothesize that effective acute decompensated heart failure treatment results in a measurable rise in hemoglobin/hematocrit that is associated with reduced rate and/or delayed time to readmission rates.

Methods Used: The study was a single center, retrospective, non-randomized study. Patients with an ejection fraction of less than or equals to 40% and documented hemoglobin and hematocrit both at the time of admission and discharge were analyzed. Excluded were those with stage III chronic kidney disease (eGFR <59 ml/min/m2) or acute blood loss/anemia requiring transfusion. Pearson chi-square tests and multivariable logistic regression were fitted using univariable and multivariable models.

Summary of Results: 121 patients (mean age was 58.9 ± 14.1 years) of 399 patients screened met inclusion criteria; 65% were male and 68% were non-white. Significant predictors of readmission in univariate analyses were admission use of beta blockers (p=0.001) ACE-I (p=0.018), diuretics (p=0.04), or Biventricular Implantable Cardioverter Defibrillator (p=0.005), Coronary Artery Disease (CAD) (p=0.02), and Cerebrovascular Accident (CVA) (p=0.013). In those readmitted there were no differences in any of the characteristics. In a multi-variable model, only CAD (p=0.002), weight loss (p=0.008), and CVA (p=0.013) were significant predictors of readmission. ADHF conveys a poor prognosis for subsequent readmission.

Conclusions: Despite greater in-hospital weight loss, and more intensive HF therapy, readmitted patients did not have evidence of hemocoenctration, suggesting that weight loss was perhaps due to extravascular fluid losses. AD and CVA were shown to be predictors of readmission.

ALTERATIONS IN CARDIAC AMP KINASE AND NITRIC OXIDE SYNTHASE IN A RAT MODEL OF METABOLIC SYNDROME

Mandaloju S, Vego C, Prabhakar S. TTUHSC School of Medicine, Lubbock, TX.

Purpose of Study: Metabolic syndrome increases the risk for cardiovascular morbidity and mortality. While the constituent phenotypic features (hypertension, glucose intolerance, obesity, hyperlipidemia) independently and collectively contribute to increased cardiac risk, the molecular mechanisms that lead to cardiac disease and heart failure in metabolic syndrome remain unclear. We postulate that alterations in nitric oxide and energy regulator, AMP kinase play a role in the heart disease in metabolic syndrome.

Methods Used: To examine this hypothesis, we studied ZSF rats, a well-established model of metabolic syndrome. Obese ZSF rats are normal at birth but develop all features of progressive metabolic syndrome starting from 8th week, and die around 50 weeks of age from severe cardiac failure. Sprague Dawley (SD) rats which do not exhibit metabolic syndrome were used as controls. All rats were obtained at 7th week and were studied from 8th week till 32 weeks. Rats were sacrificed either at 16 weeks or 32 weeks and hearts harvested. Urine and blood was obtained at the start and end of the study for evaluation of renal function and lipids. The homogenates of cardiac tissue were examined for protein expression of total and phosphorylated AMP kinase and eNOS (NOS 3) using immunoblot techniques. Comparisons were sought between the expression of these enzymes in the cardiac tissue of obese ZSF and SD rats.

Summary of Results: At 32 weeks, ZSF rats exhibited marked obesity (796 ±38 gms vs SD 573 ± 29 gms), hypertension, hyperglycemia and hyperlipidemia. In particular there was very severe hypertriglyceridemia (6.45± 1.9 gms/dl), renal functional impairment and heart failure in ZSF rats. Examination of immunoblots revealed no differences in total eNOS and total AMP kinase between the two rat groups. However there was a significant and progressive reduction of phosphorylated eNOS expression on Western in the myocardium of ZSF rats while the expression of phosphorylated AMP kinase was increased but not significantly.

Conclusions: We conclude that ZSF rats exhibit severe heart failure as a consequence of metabolic syndrome and that decreased phosphorylation of eNOS in myocardium may play a role in the pathogenesis of heart failure. Further investigation is needed to understand the signalling pathways by which reduced NO production leads to heart failure.

VISUALIZATION OF INTERNALIZATION AND INTRACELLULAR TRAFFICKING OF GUANYLYL CYCLASE/ Natriuretic Peptide Receptor-A WITH CONCURRENT GENERATION OF CGMP

Mani I, Pandey KN. Tulane University, New Orleans, LA.

Purpose of Study: Binding of cardiac hormone, atrial natriuretic peptide (ANP) to the guanylyl cyclase/natriuretic peptide receptor-A (GC-A/NPRA), induces the rapid internalization and subcellular trafficking via the endosomal compartments with concurrent generation of cGMP. The objective of this study was to visualize the internalization and intracellular trafficking of NPRA in the subcellular compartments of intact cells.

Methods Used: To study the internalization and intracellular trafficking of NPRA, we utilized immunofluorescence staining and co-immunoprecipitation (co-IP) of plasma membrane, endosomal, lysosomal, and Rab 11 markers to follow trafficking and signaling by confocal immunofluorescence microscopy and immunoblotting. A chimeric construct of enhanced green fluorescence protein (eGFP) and NPRA (eGFP-NPRA) was used to study internalization and trafficking of receptor in human embryonic kidney-293 (HEK-293) cells.

Summary of Results: The treatment of cells with ANP at different times accelerated the internalization of receptor from cell surface to cell interior. Colocalization of eGFP-NPRA with pan-Cadherin indicated that internalization of receptor accounted for 58.4%, 69.0%, 70.8%, and 75.6% at 5, 10, 15, and 30 min, respectively, compared with untreated cells. Colocalization of eGFP-NPRA with early endosome antigen-1 (EEA-1) marker was maximum at 5 min (63.3%), and gradually decreased at 10 min (42.8%), 15 min (30.9%) and at 30 min (23.2%) respectively. Similarly, colocalization of NPRA with lysosome-associated membrane protein-1 (LAMP-1) gradually increased at 5 min (13.0%), 10 min (35.6%), 15 min (42.5%), and at 30 min (44.5%). Rab 11 was used as a recycling endosome marker, which showed that 20% receptor recycled back on plasma membrane. Immunofluorescence and enzyme-linked immunosorbent assay (ELISA) analyses showed a marked increase in the accumulation of intracellular cGMP concurrent with receptor internalization.

Conclusions: Our study suggests that after ligand binding, receptor internalized and trafficked into subcellular compartments with concurrent generation of intracellular cGMP, which regulates hypertension and cardio-vascular homeostasis.

HEPATITIS C INFECTION AND CORONARY ARTERY DISEASE BURDEN

Pothineni NK, Rocchiani Y, Vallurupalli S, Kovelamudi S, Hakeem A, Mehta JL. University of Arkansas for Medical Sciences, Little Rock, AR.
Purpose of Study: Hepatitis C (HCV) infection is thought to be associated with an increased risk of coronary artery disease (CAD) events. Increased inflammation is believed to be the mechanism of accelerated atherosclerosis. We sought to examine the angiographic severity of CAD in patients with active HCV (RNA positive) infection compared to HCV negative patients.

Methods Used: All consecutive HCV RNA positive patients (n=61) who underwent coronary angiography (CA) at the University of Arkansas for Medical Sciences from 2001-2012 were identified. A parallel group of HCV negative controls (n=61), matched for age, sex and indication for CA, served as control. Angiographic burden of CAD was assessed by computing Gensini scores. Statistical analysis were performed using SPSS 21.0.

Summary of Results: HCV patients had significantly lower levels of total and LDL cholesterol. Incidence of obstructive CAD (stenosis >50%) and angiographic Gensini score were similar in both groups. There was no correlation between HCV RNA titer and Gensini score (p=0.9, analysis of variance).

Conclusions: Patients with active HCV infection have similar angiographic CAD burden as do HCV negative patients despite lower total and LDL cholesterol. Further, viral load measured by HCV RNA titer does not appear to correlate with coronary atherosclerosis burden. Further studies are needed to study the basis of association of HCV with coronary atherosclerosis and CAD events.

<table>
<thead>
<tr>
<th>Age (years) (±SD)</th>
<th>HCV C RNA positive (n=61)</th>
<th>HCV C negative (n=61)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Female sex [%]</td>
<td>53.27</td>
<td>52.7</td>
</tr>
<tr>
<td>Diabetes mellitus [%]</td>
<td>54.4 (21)</td>
<td>42.4 (26)</td>
</tr>
<tr>
<td>Hypertension [%]</td>
<td>63.9 (39)</td>
<td>77 (47)</td>
</tr>
<tr>
<td>Smoking [%]</td>
<td>39.3 (24)</td>
<td>43.6 (26)</td>
</tr>
<tr>
<td>Body mass index, kg/m² (±SD)</td>
<td>29.7</td>
<td>31.9</td>
</tr>
<tr>
<td>Lipid panel (mg/dl) (±SD)</td>
<td>145±41</td>
<td>180±50</td>
</tr>
<tr>
<td>TC<em>LDL</em> HDL* TG</td>
<td>15±8</td>
<td>15±7</td>
</tr>
<tr>
<td>Acute coronary syndrome [%]</td>
<td>9.8 (6)</td>
<td>13.1 (8)</td>
</tr>
<tr>
<td>Gensini score [%]</td>
<td>22±27</td>
<td>23±24</td>
</tr>
</tbody>
</table>

FIGURE 1

Impact of Cigarette Smoking on the Development of Vulnerable Coronary Artery Plaque: An In Vivo Virtual Histology Intravascular Ultrasound Study

Bolorunduro OB, Kapoor D, Giri S, Robinson V, Ibebuogu UN. 1 UAB, Birmingham, AL and 2LSUHSC, Baton Rouge, LA.

Purpose of Study: Cigarette smoking is a major risk factor in the progression of atherosclerosis and has been shown to cause endothelial dysfunction, inflammation and modification of lipid profile. However, its role in the pathogenesis of a vulnerable coronary artery plaque remains unknown. We investigated the relationship between smoking and the development of a vulnerable plaque (prominent necrotic core) using virtual histology intravascular ultrasound (VH-IVUS).

Methods Used: Data from consecutive patients who underwent clinically indicated cardiac catheterization at Georgia Regents University over a 2-year period was analyzed. VH-IVUS assessment of the native coronary artery stenotic lesions was performed. Baseline demographic and study characteristics were collected on all patients. Coronary plaque compositions of the culprit lesion were compared on bivariate and multivariate analysis.

Summary of Results: 160 patients (60±11 years) in whom 69% were admitted for acute coronary syndrome, 31% were smokers and the mean plaque burden was 66%. The average plaque composition was 58% fibrous, 19% fibro-fatty, 18.3% necrotic core and 5.4% dense calcium. Cigarette smokers had a higher burden of necrotic core (20.7% vs. 17.2%, p<0.05). On multivariate analysis, cigarette smoking was independently associated with a 4.62% increase in the burden of necrotic core (p=0.05). Older age (>65 years) was also a predictor of higher necrotic core burden (p=0.05).

Conclusions: Cigarette smoking is associated with a higher burden of necrotic core in coronary atherosclerotic plaques and may represent a mechanism for increased cardiovascular events in cigarette smokers.

Inflammation and Arterial Compliance: A Paradox? The Bogalusa Heart Study

Fernandez-Alonso C, 1Bashorp RP, Sun D, 1Li S, Chen W, 1Berenson GS. 1 Tulane University, New Orleans, LA; 2Tulane University, New Orleans, LA and 3Tulane University, New Orleans, LA.

Purpose of Study: Cigarette smoking is a major risk factor in the progression of atherosclerosis and has been shown to cause endothelial dysfunction, inflammation and modification of lipid profile. However, its role in the pathogenesis of a vulnerable coronary artery plaque remains unknown. We investigated the relationship between smoking and the development of a vulnerable plaque (prominent necrotic core) using virtual histology intravascular ultrasound (VH-IVUS).

Methods Used: Data from consecutive patients who underwent clinically indicated cardiac catheterization at Georgia Regents University over a 2-year period was analyzed. VH-IVUS assessment of the native coronary artery stenotic lesions was performed. Baseline demographic and study characteristics were collected on all patients. Coronary plaque compositions of the culprit lesion were compared on bivariate and multivariate analysis.

Summary of Results: 160 patients (60±11 years) in whom 69% were admitted for acute coronary syndrome, 31% were smokers and the mean plaque burden was 66%. The average plaque composition was 58% fibrous, 19% fibro-fatty, 18.3% necrotic core and 5.4% dense calcium. Cigarette smokers had a higher burden of necrotic core (20.7% vs. 17.2%, p<0.05). On multivariate analysis, cigarette smoking was independently associated with a 4.62% increase in the burden of necrotic core (p=0.05). Older age (>65 years) was also a predictor of higher necrotic core burden (p=0.05).

Conclusions: Cigarette smoking is associated with a higher burden of necrotic core in coronary atherosclerotic plaques and may represent a mechanism for increased cardiovascular events in cigarette smokers.
**Case Report:** Systemic inflammation -as measured by high-sensitivity C-reactive protein (hs-CRP)- is adversely associated with arterial compliance. However, information is scant on whether this association in consistent throughout the available non-invasive measurements of arterial compliance. The purpose of this study is to assess racial (black-white) divergences in the association between hs-CRP and measurements of arterial compliance, in relatively young and healthy adults.

**Methods Used:** Measurements of hsCRP and non-invasive arterial compliance –large-artery elasticity index (C1), small-artery elasticity index (C2), brachial-ankle pulse-wave velocity (ba-PWV) and augmentation index (AI@75) – were assessed in 702 non-institutionalized participants with a mean age of 43.5 years (29.4-51.1 years); 70.8% whites and 43.7% males, as a part of the Bogalusa Heart Study. Race-specific independent associations were tested through multivariable-adjusted linear regression analyses.

**Summary of Results:** Black vs white participants had higher hs-CRP, baPWV and AI@75 (p<0.01); whereas C1 and C2 were higher in whites (p<0.01). In multivariable-adjusted linear regression analyses, controlling for age, sex, high-density lipoprotein cholesterol (HDL-C), low-density lipoprotein cholesterol (LDL-C), body mass index (BMI), smoking status and other traditional cardiovascular risk factors, hs-CRP was significantly and independently associated with AI@75 (β=0.11, p<0.01) and C1 (β=-0.08, p<0.04), in whites only. In contrast, black participants did not show any significant associations among these parameters in the statistical models. C2 and baPWV did not exhibit association with either race group.

**Conclusions:** These findings help enhance the concept that the association of inflammation and arterial compliance is seemingly relative, as it is dependent on the measurement used to assess the latter; with its impact varying by race (black-white). Further, these observations may aid in revising existing methodologies used in the diagnosis of inflammation-mediated structural and functional damage, in addition to enhance race-specific approaches for screening and prevention of cardio-metabolic risk factors.

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**WITHDREW**

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**FALSE POSITIVE TROPONINS DUE TO HETEROPHILE ANTIBODIES**

Paccione R, Amin AN, Leong KG, Engel LS, Jain N. LSU Health Sciences Center, New Orleans, LA.

**Case Report:** INTRODUCTION: Cardiac troponin I is a marker of myocardial necrosis and is often utilized to determine the need for intervention. Elevated troponins, with an otherwise negative cardiac work-up, can create a dilemma for the physician.

CASE: A 59 year old African American man with a past medical history of hypertension and cocaine induced NSTEMI presented with chest pain after reportedly using cocaine. Vital signs were: temperature 98.8°F, blood pressure 168/78 mmHg, heart rate 82 beats per minute, respirations 14 per minute. EKG revealed normal sinus rhythm. Pertinent laboratory values include Troponin I (cTnI) 2.61 ng/ml, 2.58 ng/ml, 2.62 ng/ml, and 2.44 ng/ml. He had multiple admissions and emergency room visits with the same complaint and his cTnI averaged 6 ng/ml. A left heart catheterization in 2009 revealed normal coronaries. A stress echocardiogram from a month prior was negative for stress-induced ischemia and an echocardiogram from this admission showed normal ejection fraction and no wall motion abnormalities. His troponin I elevations over time and it was noted that he had a several year period when his troponins were negative, though he presented with the same complaint of chest pain. A plausible explanation for this was that the troponin assay was creating false positive results due to antibody production. His troponin was sent to an outside facility to confirm that this was the case. The troponin from the outside facility was negative and we concluded that heterophile antibodies to the assay at our institution were responsible for the repeated troponin elevations.

DISCUSSION: Cardiac troponin I (cTnI) is specific to cardiac tissue and is released into serum after myocardial necrosis. cTnI has become a marker of choice for detecting myocardial damage because of its specificity to cardiac tissue. Heterophile antibodies can cause false-positive cTnI results. There are a number of causes of troponin assay interference which includes heterophile antibody, rheumatoid factor, albumin and plasmin, excess fibrin, high concentration for alkaline phosphatase and analyzer malfunction. Heterophile antibodies should be included in the differential when faced with repeatedly high troponins in the setting of normal coronaries.

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**SERUM PHOSPHORUS IS ASSOCIATED WITH CYSTATIN C IN WHITES BUT NOT IN BLACKS: THE BOGALUSA HEART STUDY**

Thakkar BG, Fernandez C, Chen W, Srinivasan SR, Berenson GS. Tulane University, New Orleans, LA.

**Purpose of Study:** Serum phosphorus is a novel cardiovascular risk factor in people with and without renal dysfuction. Serum cystatin C is a marker for renal function, and it is also an independent predictor for cardiovascular morbidity and mortality. Both of these biomarkers have been shown to be significantly associated with various cardiovascular risk factors in past studies. However, the information is scant on whether there is any association between these two biomarkers. The aim of this study is to determine the relationship between serum phosphorus and cystatin C in a bi-racial (blacks-whites) population consisting mainly of young adults.

**Methods Used:** A cross-sectional study was carried out on 794 asymptomatic predominantly healthy subjects enrolled in the Bogalusa Heart Study from year 2007 to 2010. Multivariable linear regression analysis was performed to determine significant association between serum phosphorus and cystatin C.

**Summary of Results:** The mean age of the study cohort was 43.4 years; it included 41.7% males and 62.8% whites. After adjusting for traditional cardiovascular risk factors, serum phosphorus was found to be significantly associated with cystatin C in whites (β=-0.2, p<0.04) but not in blacks (β=-0.06, p=0.29).

**Conclusions:** Serum phosphorus is significantly associated with cystatin C in asymptomatic predominantly healthy young adults, and there is a racial (blacks-whites) divergence in this relationship. Further studies should be undertaken to determine its clinical implication as it would be beneficial considering the ready availability and low cost of measuring serum phosphorus levels.

**Gastroenterology**

Concurrent Session

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Friday, February 27, 2015

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**EOSINOPHILIC ESOPHAGITIS: ANALYZING THE ESOPHAGEAL AND COLONIC MICROBIOME**

Smith E, Paeck T, Kumar R, Morrow C, Atkinson P, Dinmitt R. 1University of Alabama, Birmingham, AL; 2University of Alabama, Birmingham, AL; 3University of Alabama, Birmingham, AL and 4University of Alabama, Birmingham, AL.

**Purpose of Study:** Eosinophilic Esophagitis (EoE) is a T-helper 2 cell mediated disease resulting from genetic and environmental factors. Patients with other atopic diseases have intestinal dysbiosis but little is known about the esophageal and intestinal microbiota in EoE patients. We hypothesized that the microbiome in EoE would be different than control or other GI diseases.

**Methods Used:** Following IRB approval, data was collected from September, 2013 to August, 2014 from patients 2 to 18 years old. Categories included: controls, reflux esophagitis, untreated EoE, treated EoE,
untreated IBD, and treated IBD. Samples were obtained with a cytology brush. If a colonoscopy was not performed, a rectal exam was done. DNA was extracted and PCR used to amplify the V4 region of the 16S rRNA gene. PCR products were sequenced using Illumina MiSeq. The data was processed, integrated, and analyzed using the QIIME program. Taxonomy was assigned to 16S sequences using RDP Classifier against the Greengenes 16S rRNA database. PERMANOVA was used as our statistical test with a significance of p < 0.05.

**Summary of Results:** The microbiomes were analyzed from 8 controls, 3 reflux esophagitis, 4 untreated EoE patients, 7 treated EoE, 5 untreated IBD, and 4 treated IBD. As expected, there was a significant difference between esophageal and colonic microbiomes. The esophageal microbiomes from both control and EoE patients were similar. However, in the colonic microbiomes, we saw a significant difference between untreated EoE and controls. In particular, untreated EoE had over twice the abundance of Firmicutes compared to controls (57% vs 26%). We also found controls had five times the number of Bacteroides than untreated EoE patients (48% vs 9% respectively, p < 0.001), while untreated EoE patients showed a higher proportion of Bacteriodes (48% vs 9%).

**Conclusions:** Patients with EoE had colonic dysbiosis compared to healthy controls. Further studies with a larger cohort are needed to determine specific microbiome differences and provide better insight into the influence of the microbiome on the pathogenesis of EoE.

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**PRELIMINARY REPORT OF OUR EXPERIENCE WITH DEXTRANOMER IN STABILIZED SODIUM HYALURONATE (SOLESTA®) INJECTIONS IN ADULTS WITH FECAL INCONTINENCE REFRACTORY TO MEDICAL THERAPY**

Saadi M, Elhanafi S, McCallum RW. Texas Tech University Health Sciences center, El paso, TX.

**Purpose of Study:** 1) To provide a preliminary report of our experience with Solesta injections in patients with FI who were refractory to medical therapy; 2) To assess their HRAM post Solesta injection. 3) To compare the HRAM and EUS in the assessment of patients with FI.

**Methods Used:** The study includes 12 patients with mean ages of 57 ± 3 years, with FI, 83% were female 81% Hispanic population. All patients underwent both HRAM and EUS examination. All patients were defined as normal who had <20cc or less of air in the rectal balloon was the threshold for recognition. A peak voluntary squeeze pressure increase of >100 mm Hg above the baseline internal anal sphincter (IAS) resting pressure was considered an adequate response. All measures are described as mean ± SD. 7/12 patients who had <25% in anal sphincter defect documented by EUS had not responded to medical therapy based on high fiber diet and physical exam. In 7/12 patients, rectal sensation was defined as normal when >50% symptoms improvement and <25% in anal sphincter defect documented by EUS. All 7 patients receiving Solesta injection had <25% decrease in resting pressure. Two patients had post Solesta injection HRAM.

**Summary of Results:** 8 of the 12 patients undergoing both HRAM and EUS had decreased external sphincter squeeze pressure responses of < 60 mm Hg rise from the baseline sphincter pressure. Eleven of the 12 had impaired rectal sensation, and a mean balloon volume of 40.9 ± 7 cc of air was their threshold sensation. All 7 patients receiving Solesta injection had <25% decrease in squeeze pressure. In 7/12, HRAM and EUS demonstrated improvement. 2/7 patients had post injection HRAM and balloon sensation decreased from 45 to 25 cc in one and 30 cc to 20 cc in the other.

**Conclusions:** 1) In FI patients HRAM showed both a decreased squeeze pressure response and impaired rectal sensation while EAS sphincter defects were demonstrated in 75% on EUS exam. 2) All 7 patients who received Solesta injections responded with ≥50% reduction in FI episodes. 3) Threshold for the balloon was decreased in 2 of the 7. Further Solesta has therapeutic efficacy in FI and further studies to objectively assess are needed.

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**VALIDATING THE INFANT/TODDLER ROME III CRITERIA**


**Purpose of Study:** Functional gastrointestinal disorders (FGIDs) are diagnosed by symptom-based criteria. The Rome III Questionnaire for Pediatric Gastrointestinal symptoms (QPGS-RIII) was validated for children and adolescents, but there is no comparable diagnostic questionnaire for infants and toddlers. The purpose of the current study was to validate a questionnaire for FGIDs in infants and toddlers <4 years old.

**Methods Used:** We developed a questionnaire based on symptom-based diagnostic criteria for the 7 infant/toddler FGIDs. We tested face validity by asking 8 pediatric FGID experts and 30 parents to review the questionnaire for content, understandability and completeness. Next we tested content validity by comparing parents’ questionnaire results with physician diagnoses. In a sample of 332 new infants 1 month to 4 years of age, we found 172 subjects with a FGID by parent responses to the questionnaire. We asked clinicians who had seen these subjects to provide a functional Rome III diagnosis for each of these children (without access to the questionnaire data).

**Summary of Results:** Face validity was good. Parents understood all items and experts suggested small changes in item wording. No major changes or problems were identified. Of all 172 subjects, everyone had a functional diagnosis by doctor report. Most common diagnoses were: functional constipation (47.9% doctor, 55% parent report) and infant regurgitation (24.3% doctor and 26% parent report). Agreement between parent and doctor was poor for EUS examination syndrome (kappa [k] = 0.04), fair for cyclic vomiting syndrome and functional diarrhea (k=0.25 and 0.32), moderate for infant colic and regurgitation (k=0.44 and 0.58) and substantial for functional constipation and infant dyschezia (k = 0.72 and 0.78).

**Conclusions:** The new questionnaire had good face and content validity. Overlap between questionnaire and physician diagnoses was reasonable for all but one disorder and much higher than similar data reported for the child/ adolescent version of the QPGS-RIII (k ranging from 0.04 to 0.34 for the latter).

The new questionnaire will be an important addition to clinical care and research of infant/toddler FGIDs.
RISK FACTORS AFFECTING COLONIZATION OF LACTOBACILLUS IN NEONATES

Sunkara M1, Eyal F2, 1University of South Alabama, Mobile, AL and 2VA Medical Center, Mountain Home, TN.

Purpose of Study: To review the efficacy of probiotics supplementation by looking at the rate of LB colonization as determined by serial stool cultures of premature infants admitted to the NICU. Possible risk factors that may affect the rate of GI colonization will be investigated.

Methods Used: This was a retrospective study of 872 babies in the NICU at University of South Alabama from 2006-2012. The mean gestational age (GA) was 27 ± 2.6 weeks and mean birth weight was 939 ± 303 grams. For the first month of life, stool cultures were obtained weekly and analyzed by the USAMC microbiology lab.

Summary of Results: By the first week of age, only 4% of patients had LB identified in stool cultures. This number rose to 30% by the second week and 59% by the third week. At the end of the fourth week, 83% of the infants were colonized. The overall rate of NEC was 3%; 50% of these cases occurred in the first 28 days. The rate of NEC at 2 weeks of age was 1.5% in those patients not colonized, and 2.2% in those patients who were colonized (not statistically significant). The rate of NEC at 4 weeks was 4.6% in patients not colonized and 1.5% in those who were (P=0.05). The presence of early LB colonization by 2 weeks of age was affected by the age of initial feeding (OR=0.8, CI:0.71-0.89) and the duration of antibiotic administration (OR=0.95, CI:0.87-0.99). The absence of LB colonization by 4 weeks of age increased for every additional day of antibiotics given (OR=1.07, CI:1.01-1.15) and increased for every additional day kept NPO (OR=1.21, CI:1.09-1.34).

Conclusions: LB GI colonization in premature infants receiving probiotics from birth is not universal or is often delayed up to the age of one month. Prolonged NPO status and antibiotic treatment decrease successful LB colonization and may account for the higher rate of NEC observed in the noncolonized infants.

HEYDES SYNDROME: A COMMON BUT FORGOTTEN ENTITY

Robichaud FN1, Phemister J1, Carter L1, Murthy R2, 1Quillen College of Medicine, Johnson City, TN and 2VA Medical Center, Mountain Home, TN.

Case Report: The triad of angiodyplasia, aortic stenosis (AS) and acquired von Willebrand factor (vWF) deficiency describes Heydes syndrome. Turbulent flow around stenotic valves creates high shear stress whereby multimeric vWF is cleaved by plasma metalloprotease leading to an acquired vWF deficiency with subsequent gastrointestinal bleeding.

71-year-old male with history significant for severe aortic stenosis and total colectomy presented with recurrent bleeding in ostomy bag, increasing pallor, and anemia. Physical examination revealed a II/VI systolic ejection murmur heard best at the right upper sternal. Upper gastrointestinal endoscopy (EGD) revealed bleeding due to colonic angiodyplasia. TTE showed severe AS. Subsequently, he underwent three endoscopic treatments and mesenteric angiogram followed by embolization of gastroduodenal artery, which failed to resolve bleeding. Cardiovascular surgery proceeded with aortic valve replacement (AVR) which resulted in stabilization of hemoglobin.

Anemia secondary to colonic angiodyplasia and severe AS is indicative of Heydes syndrome. Angiodyplasia is caused due to decreased gastrointestinal perfusion leading to vasodilation induced by hypoxia. Degradation of vWF multimers by high shear stress across the stenotic aortic valve leads to an abnormality of coagulopathy. Colonoscopy and EGD are the initial diagnostic modalities of choice. Mesenteric angiography is a diagnostic technique for angiodyplasia in patients with massive bleeding, whereas small bowel capsule endoscopy is helpful in diagnosing small bowel angiodyplasia. Treatment of choice is for Heydes syndrome is AVR.

AGE OF ONSET OF FUNCTIONAL CONSTIPATION

Malowitz SM, Rosenberg A, Hyman P, LSUHSC-NO, New Orleans, LA.

Purpose of Study: Children with functional constipation (FC) have large, hard, painful, infrequent stools as a result of stool withholding. FC results from failure to relax the pelvic floor to allow defecation after an episode of painful or frightening defecation, a maladaptive behavior. Children under age 7 lack the logical thought processes that would enable them to cope with painful or frightening defecation. We hypothesized that children with FC were <7 y at onset of symptoms and that children with a later age of onset have a developmental or behavioral abnormality that would predispose them to developing FC.

Methods Used: We performed a chart review of children evaluated in the pediatric gastroenterology clinic from January 2012 to June 2014 who were diagnosed with constipation at their initial clinic visit. We identified 983 children with constipation, 570 of whom met Rome III criteria for FC based on history recorded by the clinician. In these 570 children we analyzed age of presentation, age of onset, and duration of symptoms. We noted if the records documented behavioral and developmental problems: ADHD, autism spectrum disorders, depression, anxiety, conduct disorder, and oppositional defiant disorder.

Summary of Results: Of 570 FC patients, 491 (86%) had onset of symptoms from age 0-6 y and 79 (14%) had onset from 7-18 y. The overall mean age of onset was 3.3 ± 3.4 y with a median of 2.3 y. The mean duration of symptoms prior to presentation was 3 y in the younger group and 1.4 y in older children (p=0.001), with medians of 2.0 and 0.6 y respectively. We found developmental or behavioral disorders in 47 (10%) of children 0-6 years compared to 18 (22.5%) of children 7-18 years at onset (p=0.001).
Conclusions: The majority of children with FC have age of onset prior to age 7. Children whose onset is later than age 7 are more likely to have developmental or behavioral disorders. Children with onset at age 7 or greater are referred to GI specialists more rapidly than children younger than 7.

PROFILE OF INTESTINAL MICROBIOME IN INFANTS WITH SHORT BOWEL SYNDROME (SBS)
Premkumar MH, Ajami N, Petrosino J, Lee B. Baylor College of Medicine, Bellaire, TX; Baylor College of Medicine, Houston, TX and Baylor College of Medicine, Houston, TX.

Profile of Study: SBS is a common cause of gastrointestinal mortality and morbidity in newborn infants. Though microbiome-altering interventions such as antibiotics or probiotics are commonly used, the exact profile of intestinal microbiome in infants with SBS is unknown. We describe the profile and evolution of intestinal microbiome in infants with SBS.

Methods Used: SBS included those infants who underwent an abdominal surgical intervention and if they needed total parenteral nutrition for over 4 weeks. Controls were gestationally age-matched infants without gastrointestinal morbidities. Stool samples from the diapers or from the stoma were collected once a week. DNA was extracted using PowerMag Microbiome isolation kit (MoBio) and the bacterial V4 region of the 16s rRNA gene was amplified by PCR and sequenced on the MiSeq (Illumina) platform. Sequencing reads were analyzed using QIBME, and the SILVA database was used for operational taxonomic unit (OTU) classification.

Summary of Results: 255 stool samples were analyzed from 16 cases and 21 controls. Cases included 4 gastoschisis, 5 small intestinal perforations, 4 with NEC & perforation, 1 NEC, 1 volvulus and 1 jejunal atresia. Infants with SBS showed decreased diversity and abundance of microbiome. Decreased diversity of microbiome was associated with cesarean section, absence of oral feeds and use of antibiotics. Infants fed with artificial formula demonstrated higher diversity compared to breast fed infants. Infants with complicated gastrochisis and NEC demonstrated the least diversity. In SBS there were decreased lachnospiraceae, lactobacillus, while pseudomonas, Escherichia, Shigella were more abundant.

Conclusions: This is the first ever description of intestinal microbiome profile in infants with SBS. We have demonstrated that infants with SBS have decreased abundance, diversity and evenness of intestinal microbiome. More significantly, within the group of SBS, the diversity, evenness and abundance decreased with complications. Defining the microbiome in infants with SBS, will refine existing gut microbiome altering therapies and also promote development of new strategies.

EVALUATION OF THE FREQUENCY OF NUTCRACKER AND JACKHAMMER ESOPHAGUS AS THE EXTREME PHENOTYPES OF ESOPHAGEAL HYPERCONTRACTILITY: A SINGLE CENTER EXPERIENCE
Jia Y, Arenas J, hejazi RA, Elhamani S, Saadi M, McCallum RW. Texas Tech University Health Sciences Center, Paul L. Foster School of Medicine, El Paso, TX.

Purpose of Study: Hypertensive esophageal peristaltic contractions are generally referred to as Nutcracker Esophagus while the Jackhammer Esophagus is defined as extremely elevated peristaltic contractions accompanying within the normal range. We conducted a retrospective study to identify the frequency of esophageal hypercontractility and the clinical characteristics.

Methods Used: Medical records of patients referred for manometric study at a tertiary care center were reviewed from January 2012 to June 2014. Data were collected and the threshold for hypercontractility was defined as at least a single contraction from 15 wet swallows with a distal contractile integral (DCI, mmHg-s-cm) >5000, while a DCI >8000 met the criteria for Jackhammer Esophagus. Normal DCI<5000.

Summary of Results: 25 patients were identified with Nutcracker or Jackhammer Esophagus from a total of 205 (127 F/77 M) patients referred.

A CASE OF COLONIC GANGLIONEUROMAS
Spera MA, BOllinger E, Gupta M, Engel LS, Hutchings JI. LSU Health Sciences Center, New Orleans, LA.

Case Report: INTRODUCTION: Ganglioneuromas found in the gastrointestinal tract are rare tumors made up of nerve ganglion cells, nerve fibers and other supporting cells of the enteric nervous system. These tumors may be associated with von Recklinghausen’s disease, MEN lib, neurofibromatosis, familial adenommatosis coli, Cowden’s disease, tuberous sclerosis, colonic adenocarcinoma and juvenile polyposis.

CASE: A 26 year old man with past medical history of angiopoma diagnosed at the age of 21 with previous multiple cutaneous angiopoma resections presented with new masses in his neck, axilla, forehead, finger, forearm and flank. CT neck showed large, complex multinodular goiter with compression and tracheal deviation. CT abdomen and pelvis showed innumerable small 2-4 mm polyps throughout the ileum and colon. Polyp biopsies were consistent with a pathologic diagnosis of ganglioneuromas. Immunostains S-100 andNSE to confirmed the diagnosis of ganglioneuromas.

DISCUSSION: Ganglioneuromas (GN) located in the GI tract have been divided into three groups: polypoid GNs, ganglioneuromatous polyposis and diffuse ganglioneuromatosis. Ganglioneuromatous polyposis has a similar presentation as our patient and is characterized by greater than 20 sessile, pedunculated or submucosal lesions. Patients with ganglioneuromatous polyposis also exhibit extraintestinal findings such as multiple cutaneous lipomas and skin tags. A previous case report describes GI tract ganglioneuromatosis with associated cutaneous lipomas, adrenal myelolipomas, pancreatic telangiectasias and a multinodular goiter that were identified at autopsy; possibly similar to findings in our patient. These findings may represent a hypothetical MEN variant or an unrecognized syndrome.

A NEW MODEL FOR EXPLORING CENTRAL AND PERIPHERAL MECHANISMS FOR NAUSEA: EFFECTS OF TRANSCUTANEOUS ACUPUNCTURE THERAPY IN PATIENTS WITH DIABETIC GASTROPARESIS
Song G, Sun Y, Sandoval H, Sands S, sarosek I, McCallum RW. Texas Tech University Health Sciences Center, Paul L. Foster School of Medicine, El Paso, TX.
Purpose of Study: Nausea of diabetic gastroparesis (DMGP) can be alleviated by needleless transcutaneous electro-acupuncture (TEA). Central nervous control of nausea are well recognized. The right inferior frontal lobe is more active during unpleasant stimuli e.g. nausea while in contrast the left inferior frontal is more active during pleasant stimuli. Our goals were to: 1) Investigate the central and peripheral mechanisms of nausea by performing simultaneous recordings of electroencephalography (EEG) and electro-gastrography (EGG); 2) Correlate grading of nausea with findings on EEG and EGG both before and after TEA therapy in nauseated DMGP patients.

Methods Used: Eleven nauseated DMGP patients underwent concurrent EEG and EGG recordings while also grading the severity of nausea during the following sequence of experiments: 30-min baseline, 30-min visual stimulation (by a flashing/rotating checkerboard), 30-min visual stimulation plus TEA therapy at PC6 and ST36 acupoints, 30-min with TEA alone, and 15-min post TEA.

Summary of Results: 1) The nausea score was increased with visual stimuli, and then decreased during TEA and Post TEA (All post baseline results were P<0.05, vs. visual stimuli without TEA). 2) The mean percentage of normal gastric slow waves was decreased with visual stimuli, then improved during TEA and sustained post TEA (All significant, P<0.05, vs. visual stimuli without TEA). 3) During initial visual stimulation, right inferior frontal activity was prominent but when the visual stimuli was presented simultaneously with TEA, left inferior frontal activity predominated and this effect was maintained even after stopping TEA suggesting a carryover effect from the treatment.

Conclusion: In DMGP: 1) Nausea is correlated with EEG and EGG abnormalities. 2) TEA reduces nausea and improves EGG abnormalities exacerbated by visual stimulation, and results in a change of dominance from right to left inferior frontal lobe activity based on reconstruction of EEG data. 3) This model for studying nausea in humans may provide new insights into mechanisms and treatment targets.

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EARLIER PSA TESTING IN AFRICAN-AMERICAN MEN - CLINICAL SUPPORT FOR THE RECOMMENDATION
Saltzman A1,2, Luo S1,3, Hudson M4, 1Ochsner Clinic Foundation, New Orleans, LA; 2Louisiana State University, New Orleans, LA; 3John Cochran Veterans Affairs Medical Center, St. Louis, MO; 4Washington University School of Medicine, St. Louis, MO and 5University of Kentucky, Lexington, KY.

Purpose of Study: To determine whether prostate-specific antigen (PSA) testing in African American (AA) veterans ages 40-54 years (y) is associated with high-risk prostate cancer characteristics compared to AA veterans ages 55-70 y or white veterans ages 40-54 y when prostate cancer screening and detection tools are applied without racial bias.

Methods Used: From 231,174 healthy veterans ages 40-70 y without clinical evidence of prostate cancer who underwent PSA testing between 1/1/2000 and 9/30/2007, clinico-pathologic tumor characteristics were available on 1,044/1,059 AA veterans and 1,006/1,971 age-matched white veterans diagnosed with prostate cancer after a PSA >4 ng/mL triggered prostate biopsy. Clinico-pathologic tumor characteristics of AA veterans ages 40-54 y were compared to AA veterans 55-70 y, 40-54 y white veterans ages 40-54 y when prostate cancer screening and detection tools are applied without racial bias.

Methods Used: From 231,174 healthy veterans ages 40-70 y without clinical evidence of prostate cancer who underwent PSA testing between 1/1/2000 and 9/30/2007, clinico-pathologic tumor characteristics were available on 1,044/1,059 AA veterans and 1,006/1,971 age-matched white veterans diagnosed with prostate cancer after a PSA >4 ng/mL triggered prostate biopsy. Clinico-pathologic tumor characteristics of AA veterans ages 40-54 y were compared to AA veterans 55-70 y, 40-54 y white veterans ages 40-54 y when prostate cancer screening and detection tools are applied without racial bias.

Summary of Results: Of PSA-tested veterans ages 40-70 y diagnosed with prostate cancer, >90% had clinically localized disease. AA veterans ages 55-70 y were more likely to have high grade prostate cancer than AA veterans ages 40-54 y in this cohort (p<0.0204). Although AA veterans ages 40-54 y were more likely to have higher pre-biopsy PSA levels (p=0.0364) and ≥3 positive cores (p=0.0229) and were less likely to be active surveillance candidates (p=0.0340) compared to white veterans ages 40-54 y within this cohort, no racial differences in prostate cancer grade or clinical stage at diagnosis were observed.

Conclusions: This is the first large, national cohort to suggest that PSA testing at an earlier age for AA men may allow diagnosis of lower risk prostate cancer, potentially reducing outcome disparities.

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IN VITRO CHEMOTHERAPY PROFILING OF WELL-DIFFERENTIATED MIDGUT NEUROENDOCRINE TUMORS (NETS) BASED ON INDIVIDUAL PATIENT TUMOR BIOMARKERS ANALYSIS

Wang Y2, Chauhan A1, Boudreaux JP2, Carrasquillo JP2, Ramirez RA1, Woltering E2, Anthony LB3, LSU Health Sciences Center, New Orleans, LA; LSU/HSC, New Orleans, LA and 3University of Kentucky, Lexington, KY.

Purpose of Study: Midgut neuroendocrine tumors (NETs) are rare malignancies with indolent clinical courses. In general, they are well differentiated with most tumor cells in the G0 phase of the cell cycle, consistent with the poor response rate of NETs to chemotherapy in vivo. We hypothesize that insults, such as surgery, can drive NET cells from G0 into S phase and that biomarker analysis of individual patient tumors harvested and grown in the lab will provide useful practical guide for future intra and post operative adjuvant therapy.

Methods Used: 97 well-differentiated midgut NET patients underwent cytoreductive surgery. Our institution between 5/2012 and 10/2012. 148 surgical specimens were collected and submitted to a single commercial lab for processing. Primary tumors, lymph nodes and liver metastases were harvested and cultured. Their RNAs were then extracted to analyze the expressivity, a total of 88 different biomarkers. Based on our patients specific tumor biomarker expressivity and known correlations between 36 anti-neoplastic agents with their linked biomarkers, recommendations were reported as clinically benefit or lacking such benefit.

Summary of Results: A total of 148 specimens from 97 patients were tested. In four of the 97 patients, no clinically beneficial chemotherapy agent could be identified. Among the remaining 93 patients, the top three agents that are most likely to be clinically beneficial were: Fluorouracil, Cisplatin and Carboplatin. These were reported to be clinically beneficial in 135/148 (91.2%), 103/148 (69.6%), and 103/148 (69.6%) patients respectively.

Conclusions: Midgut NETs are slow growing tumors which are chemotherapeutically inert owing to the fact that most of the tumor cells are in G0 cell cycle. Surgical insult drives NET cells into active synthetic phase where they begin to express biomarkers specific to their tumor cells. Analysis of these biomarkers may guide further potential beneficial therapy.

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WITHDREW

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ROTEM: MANAGING BLOOD PRODUCTS BEFORE INVASIVE PROCEDURES IN LIVER DISEASE

Meserve J, Vaughn CD, Reuben A, Squires JE, Greenberg C. Medical University of South Carolina, Charleston, SC.

Purpose of Study: Controversy exists over the significance of abnormal coagulation tests in patients with liver disease. There is a need to identify patients with significant risk and target therapy to reduce risks. Currently, standard coagulation tests are used outside their scope to determine bleeding risk. ROTEM measures clotting in whole blood and consists of Extrem, Interm, and Fibtem assays; measuring extrinsic, intrinsic, and fibrinogen contribution

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to clotting, respectively. The measures obtained using these assays are Clotting Time, Clot Formation Time, and Maximum Clot Firmness. The measures obtained using these assays are

### Summary of Results:

Patients with normal PT/PTT had normal CT. Many had normal CT despite abnormal PT/PTT (81% to 52%). 5-9% of patients had prolonged CT that correlated with low Factors. Platelet counts less than 100K correlated with abnormal Extem (35%) and Intem (20%) CFT.

### Conclusions:

ROTEM can distinguish liver disease patients that do not have any defect in whole blood coagulation that from those that have. Our algorithm successfully managed over 40 patients. Randomized clinical studies are needed to identify at-risk patients and target therapy.

### Methods Used:

A retrospective study via chart review from January 2010-December 2013.

### Summary of Results:

127 patients were included in the study. In-hospital mortality rate was 17.3% and 30-day mortality rate was 20.5%. The independent risk factors for 30-day mortality rate were abnormal liver function tests (OR=4.23; p=0.022) and fever duration more than 4 days (OR=4.23; p=0.022). The study revealed that diagnosis of hematologic malignancies (OR=4.46; p=0.002) and neutropenia duration more than 5 days (OR=3.42; p=0.004) were associated with LOS more than 8 days.

### Conclusions:

FN in hospitalized patients results in significant mortality rate. Factors associated with increased mortality include ICU admission and abnormal liver function tests.

### Results:

7-year old male Jehovah’s Witness presented with fatigue and poor exercise tolerance. Examination showed conjunctival pallor and a right upper extremity ecchymosis. A complete blood count revealed pancytopenia with a hemoglobin of 11.6 g/dL, platelet count of 36 x 10^9/L, and a white blood cell count of 1.8 x 10^9/L with circulating promyelocytes.

### MOLECULAR REMISSION IN A JEHOVAH’S WITNESS WITH ACUTE PROMYELOCYTIC LEUKEMIA USING ALL-TRANS-RETINOIC ACID AND ARSENIC TRIOXIDE WITHOUT BLOOD PRODUCT SUPPORT

Eubanks J, Bigelow C. University of Mississippi Medical Center, Jackson, MS.

**Case Report:** Treatment of acute promyelocytic leukemia in patients whose religious beliefs prohibit transfusion of blood products is an exceptional therapeutic challenge as chemotherapy-induced myelosuppression can result in life-threatening cytopenias. The use of all-trans-retinoic acid (ATRA) and arsenic trioxide has been associated with less hematologic toxicity than standard anthracycline-based regimens, thus providing a treatment option for these patients. We report a case of a Jehovah’s Witness who was successfully treated with ATRA and arsenic trioxide induction and consolidation without blood product support.

A 57 year old male Jehovah’s Witness presented with fatigue and poor exercise tolerance. Examination showed conjunctival pallor and a right upper extremity ecchymosis. A complete blood count revealed pancytopenia with a hemoglobin of 11.6 g/dL, platelet count of 36 x 10^9/L, and a white blood cell count of 1.8 x 10^9/L with circulating promyelocytes. Disseminated intravascular coagulopathy was present as evidenced by a prothrombin time of 18 sec, D-dimer of 22,640 ng/mL and a fibrinogen of
Case Report: The tyrosine-kinase inhibitor imatinib mesylate has dramatically improved the prognosis for patients with metastatic gastrointestinal stromal tumors (GIST). This rare tumor of mesenchymal origin commonly expresses c-KIT (CD117 antigen), one of the three tyrosine-kinases targeted by imatinib.

Here we describe a case of a 58 year old female with recurrent, metastatic GIST who has been progression-free for over twelve years on high-dose imatinib. The patient was first diagnosed with an epithelioid leiomyosarcoma of the stomach in 1983, with subsequent gastrectomy in the same year. In February of 2002, an enlarged liver was palpated during routine examination, and computed tomography (CT) scan identified multiple metastatic liver lesions. Liver biopsy identified the masses as a recurrence of the original gastric tumor, with positive staining for c-KIT. The patient was initiated on 400 mg of oral imatinib daily in March of 2002, with a subsequent dose increase to 800 mg daily. While current guidelines recommend dose titration to 800 mg only in the face of tumor progression, at the initiation of her therapy, dosing recommendations for imatinib in this disease were not well established. The patient expressed a desire to maintain a high-dose regimen, and has tolerated adverse effects including periorbital edema, nausea, multiple loose bowel movements daily as well as cytophenias.

Her performance status remains high, she walks several miles per week on a treadmill and continues to work full time. Serial Positron Emission Tomography - computed tomography (PET-CT) scans every 6 months have shown relative stability of multiple liver lesions which measure between 3.5 cm and 8.0 cm in size with SUV ranging from 7.7 to 16.8. Given the significant degree of her tumor burden to start with, the radioisotopic and clinical stability in her case is truly remarkable. Our case demonstrates that prolonged high-dose imatinib therapy can prevent progression of metastatic GIST for over twelve years with modest toxicity and a well maintained quality of life.

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TARGETED THERAPY TRANSLATES TO TWELVE YEAR PROGRESSION FREE SURVIVAL IN METASTATIC GASTRO INTESTINAL STROMAL TUMOR

Wann D, Chakraborty K, Jaishankar D. Quillen College of Medicine, ETSU, Jonesborough, TN.

45 mg/dL. Fluorescence in situ hybridization showed the presence of the PML-RARα gene fusion and reverse transcription polymerase chain reaction studies identified PML-RARα mRNA transcripts, thus establishing the diagnosis of acute promyelocytic leukemia. The patient received induction with ATRA 45 mg/m²/day and arsenic trioxide 0.15 mg/kg five days weekly for five weeks. Epoetin alfa was given throughout therapy to aid in maximizing erythropoiesis. Hematologic toxicity with a nadir hemoglobin of 5.7 g/dL and platelet count of 27 x 10^9/L resulted in a delay in induction weeks four and five. Induction therapy was followed by ATRA and arsenic trioxide consolidation.

At the completion of consolidation, patient had achieved a molecular remission and currently continues on maintenance therapy.

While treatment of Jehovah’s Witnesses with acute promyelocytic leukemia is challenging, this case demonstrates that a molecular remission can be obtained without blood product support with combination ATRA and arsenic trioxide induction and consolidation with epoetin alfa support.

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DIFFUSE IDIOPATHIC PULMONARY NEUROENDOCRINE CELL HYPERPLASIA (DIPNECH) AND ROLE OF SOMATOSTATIN ANALOGS

Chauhan A, Ramirez RA, LSU Health Sciences Center, New Orleans, LA.

Purpose of Study: To analyze the contributing factors of thromboembolic events in the pediatric population at Ochsner Medical Center, including subsequent management and the use of prophylactic treatment.

Methods Used: ICD-9th Revision codes were used to identify patients 0-21 years of age admitted to Ochsner Medical Center between July 1, 2009 and September 31, 2013 who were diagnosed with a thromboembolic event during their treatment stay. Individuals were excluded if their event occurred or was diagnosed at an outside facility. Charts were reviewed to verify the validity of the event, investigate possible risk factors and identify pre and post event treatment regimes.

Summary of Results: 117 of the 340 identified patients have been reviewed to date, with 49 meeting study parameters. 13 patients had a thrombus as a complication of a general medical illness or due to an unknown cause. 11 cases were complications of central venous catheters or peripheral IV lines. 11 cases were attributed to surgical procedures. 7 cases were related to unrepaired congenital heart defects. 4 cases were related to hormonal changes (OCP, pregnancy). 3 cases were attributed to infection.

Risks for thromboembolism in this population included: Age: 44.9% of patients were found to be < 1 year old; Gender: Male=Female; Hypercoagulable State: 2 cases had an identified primary hypercoagulability.

Other contributing factors: immobility (17 cases); 2nd hand smoke exposure (4 cases); maternal diabetes (3 cases); perinatal asphyxia (2 cases).

17 patients were on prophylactic therapy (heparin, LMWH, warfarin or aspirin) at the time of their event. 8 patients received a thrombophilia workup. For 6 patients, this was their second or more thromboembolic event.

Conclusions: The risk of thromboembolism in the pediatric population is low when compared to adults, but has been increasing over the last decade due to medical advancement. Few treatment guidelines currently exist, and possible prophylactic intervention needs further investigation to determine its benefit in reducing or preventing these events in children.

Upon completion of the chart review and literature search, we intend to propose prevention and treatment strategies for the pediatric age group at Ochsner Medical Center.
FATALLY ASPARAGINASE-INDUCED HEPATOTOXICITY: RADIO-PATHOLOGICAL FINDINGS

Chalhoub C, Chalhoub E, Hammod D, Lewin E, Safah H, Tulane University, New Orleans, LA; Estafan University, New Orleans, LA and Lebanese University, Beirut, Lebanon.

Case Report: Although hepatotoxicity due to Asparaginase is a well-known adverse event, none of the previous reports show the acute changes in radiologic findings.

A 76 year old female was diagnosed with acute lymphocytic leukemia. She received chemotherapy (CALGB 9111 regimen) including L-Asparaginase on D6 and Pegaspargase on D9 (delayed/switched due to availability). On D20, total bilirubin (max 14.3 mg/dL), alkaline phosphatase (1430 U/L), AST (136 U/L) and ALT (159 U/L) started to increase. CT abdomen showed diffuse severe fatty infiltration of the liver, which was absent on CT 12 days prior (Fig1). Liver biopsy showed severe macrovesicular and microvesicular steatosis. The patient subsequently transitioned to hospice.

Liver injury from Asparaginase occurs in less than 5% of cases. The exact mechanism is not clear. Severity varies from asymptomatic elevation of liver enzymes to fatal liver failure. Treatment is mainly supportive (isolated reports of liver transplantation, or treatment with vitamin B complex and L-carnitine).

CT scan findings of L-asparaginase induced fatty liver can develop very rapidly (days). Early imaging may help prevent further hepatotoxicity.

FIGURE 1.

Infectious Diseases 1
Concurrent Session
2:00 PM
Friday, February 27, 2015

IMPACT OF MULTIPLE CENTRAL LINES ON CENTRAL LINE-ASSOCIATED BLOODSTREAM INFECTION RATES
Cock J, Chernetsky Tejedor S, Steinberg J, Robichaux C, Jacob J, Emory University School of Medicine, Atlanta, GA and Emory University School of Medicine, Atlanta, GA.

Purpose of Study: Central line-associated bloodstream infections (CLABSIs) are leading causes of healthcare associated infections and inter-institutional comparison of CLABSI rates are used for pay-for-performance measures. The CDC definition of CLABSI has a denominator of one line-day if one or more central lines are present on a given day. This method may be unfavorable to institutions with high patient acuity. We compared the effect of using the standard denominator to one that uses the actual number of lines.

Methods Used: CLABSI rates were calculated for all adults in two 500-bed hospitals from 12/1/2009 to 6/30/2011 using standard CDC definitions. Two denominators for CLABSI rates were used, 1) the standard CDC method and 2) a modified method counting n central lines in one patient in one day as n central line-days. CLABSI rates were determined for ICUs and non-ICU (wards).

Summary of Results: Among 15,843 hospital admissions there were 205 CLABSIs (80 in ICUs; 125 in wards). Using the CDC method, there were 142,840 central line-days (52,776 in ICUs; 90,064 in wards). The modified method identified 166,562 central line-days (70,122 in ICUs; 96,440 in wards). In ICUs, 37% of admissions contributing line-days had more than one central line compared to 11% of admissions to wards. Use of the modified denominator compared to the standard denominator reduced the CLABSI rate per 1000 central line-days by 25% in ICUs (1.52 vs. 1.14) but only 7% (1.39 vs. 1.30) for wards.

Conclusions: By counting more than one central line per patient day, CLABSI rates in ICUs decreased by 25% falling below the rates in the wards. These results suggest that institutions with high acuity of patient care may be at a disadvantage for inter-institutional CLABSI comparisons when using the current methodology.

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<th>All Units</th>
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<td>205</td>
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<td>125</td>
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<tr>
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<td>Modified</td>
<td>166,562</td>
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<td>CLABSI Rate (per 1,000 line-days)</td>
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A UNIQUE CASE OF PNEUMOCOCCAL EOSINOPHILIC MENINGITIS IN A LUPUS PATIENT
Khuon D, Arnold SR, Patel A, Bagga B. 1University of Tennessee Health Sciences Center, Memphis, TN; 2St. Jude Children’s Research Hospital, Memphis, TN and 3Le Bonheur Children's Hospital, Memphis, TN.

Case Report: A 15 year old female with history of systemic lupus erythematosus (SLE) on mycophenylate mofetil and abatacept presented with fever, headache, neck stiffness, and hearing loss. On physical exam, she appeared ill and anxious with meningismus and difficulty hearing. She had no known exposure to tuberculosis. White blood cell count was 54,700 cells/mm3 (2% bands, 71% neutrophils, 24% lymphocytes, 3% monocytes). C-reactive protein was 368.2 mg/L and erythrocyte sedimentation rate was 98 mm/hr. Ceftriaxone and vancomycin were initiated for presumed bacterial meningitis before blood and cerebral spinal fluid (CSF) cultures due to difficulty obtaining samples. CSF studies seven hours after antibiotics demonstrated protein 293 mg/dL, glucose 44 mg/dL, 0 RBC/mm³, and 5624 WBC/mm³ (12% lymphocytes, 20% monocytes, 68% eosinophils). Bacterial CSF and blood cultures were negative for growth. Tests for histoplasmosis, tick-borne infections, and enteroviruses were negative. Tuberculin skin test, and CSF acid-fast stain and culture were negative; interferon gamma release assay was indeterminate. CT and MRI brain were negative. Day 3 CSF demonstrated
protein 144 mg/dL, glucose 43 mg/dL, 0 RBC/mm³, and 558 WBC/mm³ (7% lymphocytes, 22% monocytes, 71% eosinophils). S. pneumoniae CSF PCR was positive on day 1 (cycle threshold (CT) values 30, 33.47, 35.71 from three runs) and day 3 (CT value 33.44). The patient completed 14 days of ceftriaxone and vancomycin. Microbiological diagnosis of pneumococcal meningitis can be challenging as cultures obtained after antibiotics may be negative. SLE predisposed our patient to invasive pneumococcal disease; however, CSF eosinophilia and negative cultures led to a diagnostic dilemma. S. pneumoniae identification by PCR of CSF confirmed the clinical diagnosis, preventing additional testing and ensuring appropriate therapy. This report highlights eosinophilic meningitis as an unusual presentation of pneumococcal meningitis in an SLE patient and raises awareness of CSF pneumococcal PCR as a viable diagnostic test in pretreated meningitis.

COFACTORS FOR MORTALITY IN HIV-POSITIVE DIALYSIS PATIENTS

Williams E¹, Nahman S¹, Colombo R¹, Kintzheimer K¹, Kheda M¹, Huber L¹, Baer S², ¹Georgia Regents Univ, Augusta, GA and ²Augusta VAMC, Augusta, GA.

Purpose of Study: The use of highly active antiretroviral therapies for Human Immunodeficiency Virus (HIV) has changed it from a fatal to a chronic illness. End-stage-renal disease with HIV increases the risk for mortality and the need for specialized health care. The United States Renal Data System (USRDS) is one of the largest cohorts of HIV positive hemodialysis (HD) patients, which provides a unique opportunity to define risk factors for morbidity and mortality in HIV-positive HD patients.

Methods Used: All incident adult HD cases from the USRDS for calendar years 2005-2008 were queried for a diagnosis of HIV, demographics, and potential clinical covariates, using ICD-9 diagnosis codes submitted for Medicare billing. Form 2728 was used to define the form of access at initiation of HD. Survival analyses were performed to correlate these with mortality. The proportional hazards (PH) assumption were tested for all variables of interest, and Cox PH or extended Cox models were used to assess risk factors for mortality in the HIV-positive cohort, with hazards ratios reported.

Summary of Results: Clinical covariates correlated with an increased risk for mortality in HIV positive HD patients include Histoplasmosis (relative risk (RR) 1.78, 95% confidence interval [CI] 1.11-2.85), Mycobacterium tuberculosis (RR 1.56, 95% CI 1.06-2.29), liver cirrhosis (RR 1.51, 95% CI 1.24-1.84), cerebrovascular disease (RR 1.45, 95% CI 1.24-1.70), any AIDS defining illness (RR 1.41, 95% CI 1.23-1.61), Mycobacterium avium complex (disseminated/extravascular) (RR 1.40, 95% CI 1.06-1.85), bacteremia (RR 1.32, 95% CI 1.18-1.47), wasting syndrome (RR 1.29, 95% CI 1.13-1.46), Hepatitis C (RR 1.22, 95% CI 1.09-1.37), and Candida colonization (RR 1.17, 95% CI 1.03-1.33). Demographic covariates associated with an increased risk for mortality include being ≥65 years (RR 1.48, 95% CI 1.16-1.89), unemployed (RR 1.61, 95% CI 1.29-2.02), or disabled (RR 1.60, 95% CI 1.27-2.02). History of influenza vaccination (RR 0.39, 95% CI 0.35-0.44) and combined pneumococcal and influenza vaccination (RR 0.30, CI 0.17-0.51) were protective factors.

Conclusions: Histoplasmosis was the clinical factor with the greatest risk for mortality among HIV-positive dialysis patients. History of an influenza vaccine and the combined influenza and pneumococcal vaccine were protective.

FATAL CASE OF ESCHERICHIA COLI SEPTICEMIA WITH ECTHYMA GANGRENOsum

Fenié M¹, Mohamed A², Ruddle D¹, Patel P¹, ¹East Tennessee state university, Johnson City, TN and ²Texas Tech University Health Science Center, Lubbock, TX.

Purpose of Study: 1-Report a rare case of ecthyma gangrenosum(EG).

Methods Used: Case report and review of literature

Summary of Results: A 70 year old male with history of multiple myeloma(MM) was awakened at 4 AM by severe right upper extremity pain. MM therapy included lenalidomide and dexamethasone. Temperature on admission was 102.7 F. Right elbow exam revealed edema and severe tenderness to light palpation. Vancomycin was started empirically for cellulitis. Within 6 hours, he became hypotensive and was started on stress dose steroids and pressors. Lab results revealed absolute neutrophil count of 2100/ul and lactate of 3.4. Elbow x-ray did not reveal fracture or subcutaneous gas. Surgery and infectious diseases were consulted; empiric piperacillin-tazobactam and clindamycin therapy was added. Swelling around the elbow continued to get worse and the patient started to develop bullae. The patient continued to deteriorate despite aggressive therapy and death was pronounced within 18 hours from admission. Blood and elbow wound cultures revealed E. coli post-mortem.

Discussion:

EG is a “necrotic blister”. The term is used to describe lesions that begin with papules surrounded by erythema and edema with evolution into hemorrhagic, necrotic ulcers. The lesions typically appear between the umbilicus and the knees that progress rapidly within 12 to 24 hours. The condition is often considered pathognomonic for P. aeruginosa bacteremia, however, EG has been observed in patients with blood cultures growing Klebsiella, Serratia, and Aeromonas hydrophila. Seven adult cases related to E. coli have been reported (Patel et al), with the above case being the eighth. A skin biopsy was not obtained, however, the diagnosis is supported with E. coli wound and blood cultures in the setting of severe rapid disease progression and immunosuppression (almost always associated with EG). Pathologically, EG is described as bacterial invasion of the media and adventitia of deep veins in the dermis, sparing the intima and lumen. The bacterial invasion results in marked fibrin exudation and frank hemorrhage, followed by bulla formation leading to necrosis of the dermis.

Conclusions: Fatal septicemia may have skin manifestations every clinician should be familiar with.
CATHETER-INDEPENDENT RISK FACTORS FOR BACTEREMIA IN HEMODIALYSIS PATIENT

Simmons P1, Colombo R1-2, Kintziger K1, Nahman S1, Kheda M1, Baer S2-1, Huber L1, 1Georgia Regents University, Augusta, GA and 2Charlie Norwood VA Med Center, Augusta, GA.

Purpose of Study: Hemodialysis (HD) for end stage renal disease (ESRD) is performed through catheters (CATH), arteriovenous grafts (AVGs), or arteriovenous fistulas (AVFs). AVF/AVGs are associated with lower infection rates compared to CATHs. Bacteremia (BAC) occurs in nearly 25% of HD patients, CATH vascular access at the time of HD initiation has been shown to have an almost twofold increased risk for BAC. Excluding CATH related infection may provide a unique perspective of baseline risk factors for BAC in HD. This study assessed CATH-independent risk factors for BAC by focusing on the AVF/AVG HD population.

Methods Used: All incident HD cases from the United States Renal Data System (USRDS) for calendar years 2005-2008 were queried for a diagnosis of BAC and several clinical covariates using ICD-9 diagnosis codes submitted for Medicare billing. Data was limited to adults who began dialysis with an AVG or AVF and had no evidence of CATH placement prior to BAC. Descriptive statistics were performed for demographic data and select comorbidities. Factors associated with BAC risk were assessed using log-binomial regression. CATH-independent risk factors for death were also evaluated.

Summary of Results: 45,341 patients initiated HD with either an AVG or AVF during the period of study, among whom 8,802 (19.4%) were diagnosed with BAC. 3,477 cases met criteria for CATH-independent BAC (60% had evidence of CATH placement at or prior to BAC). Demographics for the CATH-independent BAC cohort included: 60% male, 66% White, 59% age 65 years or above, and 69% with AVF. Diabetes mellitus was the most common comorbidity (60%). AVGs were associated with a higher risk of BAC than AVFs [adjusted relative risk (aRR) 1.35, 95% confidence interval (CI) 1.27-1.45]. Multiple CATH independent risk factors for BAC were identified, including HIV infection (aRR 2.22, 95% CI 1.81-2.71). BAC was associated with a reduced median survival time in ESRD patients dialyzing via AVF/AVG.

Conclusions: BAC is an important clinical concern in HD as it negatively impacts survival. Identifying CATH-independent factors associated with the development of BAC in HD patients may promote heightened clinical suspicion among physicians caring for patients with these comorbidities and potentially allow for earlier targeted intervention.

LOPINAVIR USE AND HUMAN PAPILLOMAVIRUS INFECTION IN HUMAN IMMUNODEFICIENCY VIRUS POSITIVE WOMEN

Dugan KB1, Delille CA1, Burk RD2, Palefsky J3, Massad L2, Reimers L2, Minkoff H1, Xie X2, D’Souza G5, Xue X2, Cole C1, Anastos K2, Wingood G1, Stricker H1, Ofotokun F1, 1Emory University School of Medicine, Atlanta, GA; 2Albert Einstein College of Medicine, Bronx, NY; 3University of California San Francisco, San Francisco, CA; 4Washington University School of Medicine, St. Louis, MO; 5Maimonides Medical Center, Brooklyn, NY; 6Johns Hopkins School of Public Health, Baltimore, MD and 7Georgetown University Medical Center, Washington, DC.

Purpose of Study: Lopinavir (LPV), an HIV protease inhibitor (PI), inhibits human papillomavirus (HPV)-mediated p53 degradation in vitro. Cervical application of LPV was associated with clearance of cervical dysplasia in a small clinical trial. It is unknown whether oral LPV use reduces HPV risk in HIV positive women.

Methods Used: Specimens and data were obtained from 2793 HIV positive women enrolled in the Women’s Interagency HIV Study in 1994/95 or 2002/03 at semiannual follow-up. During each visit, questionnaires were administered and cervicovaginal lavage (CVL) specimens for HPV DNA testing by MY09/MY11 PCR were obtained.

Summary of Results: 1291 HIV positive women receiving highly-active antiretroviral therapy (HAART) provided CVLs and were included in the analysis. LPV use was reported by 233 (18%). Compared to non-users, LPV users had a lower median CD4 count (298 vs 428 T-cells/mL, P=0.001) and were more likely to have plasma HIV viral load >4000 copies/mL (43% vs 26%, P=0.001). There was no difference between LPV-users and non-users in HPV DNA positivity (52% vs 45%, P=0.13). Multivariate GEE models included women on effective HAART (undetectable HIV viral load) and adjusted for age, CD4, race/ethnicity, smoking, and number of recent sex partners to model HPV prevalence and HPV clearance in LPV users, other PI users, and non-PI users. While not statistically significant, LPV users had a higher HPV prevalence (OR=1.41, P=0.13) and lower HPV clearance (OR=0.47, P=0.06) compared to non-users.

Conclusions: These data reveal no reduction in HPV prevalence or increase in HPV clearance with oral LPV, suggesting oral and topical LPV use might have different impacts. Future studies examining associations between HPV infection and cervical LPV drug concentrations are warranted. It is unlikely that LPV-based HAART will reduce the burden of HPV infection in HIV positive women.
A LONGBITUDINAL STUDY OF IMMUNE RESPONSES TO GROUP A STREPTOCOCCAL ANTIGENS FOLLOWING PHARYNGEAL INFECTIONS IN PEDIATRIC SUBJECTS

Hysmith ND1,3,4, Kaplan E2, Clary P2, Johnson D2, Penfound T1,4, Dale J1,4,
1University of Tennessee Health Science Center, Memphis, TN; 2University of Minnesota, Minneapolis, MN; 3St. Jude Children’s Research Hospital, Memphis, TN and 4The Department of Veteran Affairs Medical Center, Memphis, TN.

Purpose of Study: Multiple antigens of group A streptococci (GAS) are being considered as vaccine components, yet little is known about the human immune responses to these antigens following natural infection. We evaluated immune responses following pharyngeal infections in pediatric subjects using a panel of 28 GAS antigens.

Methods Used: Fifty-six pediatric subjects (ages 6-15 years) were evaluated during a 24-month longitudinal study. A total of 235 serum samples and 58 positive throat cultures (13 different emm types) representing new acquisitions of GAS were obtained. ELISA was performed with streptolysin O (SLO), DNaseI, C5a peptidase (SCPA), GAS40, streptococcal serine esterase (SSE), serum opacity factor (SOF), fibronectin binding protein (FBP54), three M-related protein peptides (Mrlp-III), and 18 M peptides.

Summary of Results: Increases in SLO and/or DNaseB antibodies were observed following 32/58 (55%) new GAS acquisitions. In 34/58 (58%) new acquisitions there were increases in type-specific M antibodies corresponding to the infecting emm type. No new GAS acquisitions of the same emm type were observed when type-specific antibodies were present. Of the remaining 8 common antigens, there was a response to an average of 1.6 antigens (range 0-4). Antibody responses to GAS40 and SCPA after GAS acquisition were seen in 45% and 33% of subjects, respectively. Twelve cases of immunologically significant GAS acquisition were only detected by antibody increases to GAS40 or SCPA.

Conclusions: Immunologically significant GAS infections in children were associated with humoral immune responses to common and type-specific antigens. Persistence of newly acquired GAS in the posterior oropharynx was not influenced by significant and sustained immune responses to type-specific or shared antigens. Sensitivity of commonly used clinical markers of GAS infection (SLO and DNaseB) can be improved from 55% to 75% by the addition of common antigens. The human immune responses to GAS antigens provide important information regarding potential vaccine formulations designed to prevent GAS infections.

IMPACT OF AN ANTIBIOTIC STEWARDSHIP PROGRAM AT A CHILDREN’S HOSPITAL: AN ANALYSIS OF ANTIBIOTIC USE AND ACQUISITION COSTS

Ferguson-Paul K1,2,3, Arnold SR1,2, Bagga R1,2, Lee K1,4, 1Le Bonheur Children’s Hospital, Memphis, TN; 2St. Jude Children’s Research Hospital, Memphis, TN; 3University of Tennessee Health Sciences Center, Memphis, TN and 4University of Tennessee Health Science Center, Memphis, TN.

Purpose of Study: To determine the impact of the Le Bonheur Children’s Hospital’s Antibiotic Stewardship Program (ASP) on acquisition costs and antibiotic days and to compare these to reported outcomes from other institutions.

Methods Used: Antibiotic purchasing data was obtained from our wholesaler’s database for August 2011-July 2014. Antibiotic usage data was extracted from the Pediatric Health Information Systems (PHIS) database for August 2011- March 2014. Antibiotic usage data was normalized to 1000 patient days to allow comparison over time. Changes in antibiotic use and purchasing associated with implementation of ASPs at other children’s hospitals were determined by literature review.

Summary of Results: Average monthly antibiotic days/1000 patient days declined by 21.8% over 3 years of ASP. Total antibiotic acquisition costs decreased from $902,996 to $730,015 over 3 years (savings of $173,000; 20% decrease). Average monthly targeted antibiotic days/1000 patient days declined by 30%. Targeted antibiotic acquisition costs declined from $285,689 to $108,336 over 3 years (savings of $177,000; 55% decrease). Other pediatric ASPs have, on average, shown a 22% reduction in restricted antibiotic costs and 14% decrease in restricted antibiotic use. Overall antibiotic use reductions of 3 - 6% and cost reductions of $50,000-$100,000 have been reported.

Conclusions: Both usage and acquisition costs of targeted and overall antibiotics have decreased since the inception of ASP at our institution 3 years ago. Our reductions in antibiotic use and costs compare favorably with reports from other ASPs. Continued efforts are needed to ensure antibiotic use and acquisition continue to decline without compromising patient outcomes.

PREDICTORS OF MORTALITY IN END STAGE RENAL DISEASE PATIENTS WITH INFECTIVE ENDOCARDITIS

Powell R1, Steinberg J2, Jacob J2. Emory University School of Medicine, Decatur, GA and 3Emory University School of Medicine, Decatur, GA.

Purpose of Study: Patients undergoing dialysis for end stage renal disease are at increased risk for infective endocarditis (IE), but the predictors of mortality from IE in this population are unclear.

Methods Used: Chronic dialysis patients with IE from 1992 to 2012 in two academic medical centers were retrospectively identified using ICD-9 codes and validated using the modified Duke criteria and chart review. Outcome was tracked until death or loss to follow up. Categorical variables were compared using chi-square and continuous variables with the t-test or Fisher’s exact test. Cox proportional hazard regression analysis was performed to identify independent predictors of mortality.

Summary of Results: Over 21 years, there were 258 patients with IE; 87% were black and 58% used a catheter for vascular access. Mortality rates were 24.2% in the hospital, 27.9% at 30 days, 57.0% at 1 year, and 75.8% at 3 years. Altered mental status at admission (55.1% vs 23.5%, p=0.001), embolic stroke (36.1% vs 27.4%, p=0.001), vegetation ≥ 1.5 cm (70.0% vs 46.8%, p=0.01), thrombocytopenia (34.6% vs 15.8%, p=0.004), and age ≥ 65 (40.3% vs 24.7%, p=0.01) were more common in patients who died at 30 days compared to those who survived, while valve replacement surgery (15.3% vs 30.7%, p=0.01) and infection with enterococci (6.9% vs 19.9%, p=0.01) were less common. Age ≥ 65 (HR 4.6, 95% CI 2.7 - 7.9), embolic stroke (HR 2.9, 95% CI 1.7 - 5.0), thrombocytopenia (HR 2.9, 95% CI 1.2 - 3.4), and prosthetic valve endocarditis (HR 2.3, 95% CI 1 - 4.9) were associated with increased mortality by Cox regression analysis. No factors were found to be protective on multivariate analysis.

Conclusions: Mortality is high in dialysis patients with endocarditis, particularly in older patients and those with embolic stroke. While valve replacement surgery is not an independent predictor of survival, it may improve survival in an appropriately selected population of patients. Cox proportional hazards model for mortality in dialysis patients with infective endocarditis.

Variable | Hazard Ratio (95% CI) | P-value
---|---|---
Age > 65 | 4.6 (2.7 - 7.9) | <0.0001
Embolic stroke | 2.9 (1.7 - 5.0) | <0.0001
Planter < 150 | 2.0 (1.2 - 3.4) | 0.006
Prosthetic valve | 2.3 (1.1 - 4.9) | 0.03

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EVALUATION OF A SIMULATED PATIENT ENCOUNTER TO IMPROVE PEDIATRIC INTERNS’ MEDICAL KNOWLEDGE AND COMMUNICATION SKILLS

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Purpose of Study: To evaluate the impact of using a simulated patient encounter (SPE) as part of an educational module to improve trainees’ medical knowledge and ability to communicate with parents expecting delivery of a late-preterm infant

Methods Used: All first year pediatric (PGY1) residents completed a videotaped SPE at the start and end of their first year of training. Residents in the original group (N=30) completed the standard first-year curriculum. Residents in the intervention group (N=30) participated in an educational module designed to teach morbidity common to the late-preterm infant as well as communication skills in the setting of a prenatal consultation. The module consisted of review of the individual’s initial SPE, a didactic component, and a group role-playing activity. The residents’ knowledge regarding the late-preterm infant and communication skills at the end of the first year were assessed by graduate medical educators, who reviewed and scored the end-of-year SPEs completed by both groups.

Summary of Results: Residents in the intervention group demonstrated higher overall scores (p<.0004) and higher scores in the specific areas of content (p<.0001) and objective interpersonal skills (p<.0001) than residents in the control group. There was no significant difference detected in subjective communication skills scores between groups.

Conclusions: The addition of SPEs in addition to traditional didactic instruction is an effective method for teaching first-year pediatric residents content knowledge and objective interpersonal skills in the setting of a prenatal consultation for impending delivery of a late-preterm infant.

A DETERMINATION OF THE OPTIMAL TIME FOR A NEONATAL INTUBATION SKILLS REFRESHER FOR CLINICIANS WHO DO NOT UTILIZE THE SKILL ROUTINELY

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Purpose of Study: The Neonatal Resuscitation Program recommends skill refresher courses every two years. We believe this interval is too long. We set out to delineate the skill decay of novice trainees six, twelve, and eighteen months after learning a new skill when it is not used routinely.

Methods Used: We previously randomized 110 third-year medical students into three different neonatal manikin endotracheal intubation training sessions (weekly practice for four straight weeks, consecutive practice for four days over one week, or no additional practice). We rated students on their ability to pick the proper equipment in addition to a term newborn manikin (preparation), the steps of intubation (performance), the length of time it took to successfully intubate the manikin, and the number of attempts before a successful intubation. In this follow-up study, we similarly re-evaluated 33 of the original students. These students were randomized to be retested six, twelve, or eighteen months after their original evaluation.

Summary of Results: ANOVA of the students’ performances did not show any significant decay in the skill acquisition as measured by changes in preparation scores, performance scores, time to successful intubation, or number of attempts before achieving a successful intubation at any of the follow-up evaluations. When follow-up performances were evaluated based on the original practice group assignment, ANOVA demonstrated a significant decrease in preparation score changes and an increase in intubation times in the weekly practice group compared to the other two groups.

Conclusions: No statistically discernible decay was noted six, twelve, or eighteen months after the initial evaluations. Sub-group analysis showed a decrease in preparation scores and an increase in intubation times among the weekly practice group, with no other effects noted resulting from the original group assignment.

NEONATAL SIMULATION TRAINING IMPROVES PEDIATRIC TRAINEES’ COMPETENCE IN DELIVERY ROOM RESUSCITATION

Dang S, Shook L. University of Kentucky, Lexington, KY.

Purpose of Study: To improve the competency and comfort level of graduating Pediatric residents in neonatal resuscitation and code management through use of high fidelity simulation labs.

Methods Used: The Lean 8-step problem solving model was used for this project to identify key points of occurrence where house officers in training begin to lag behind in resuscitation skills. Resuscitation protocols were developed by a multidisciplinary resuscitation committee and incorporated staged inclusion of house officers as competency goals are achieved. Success was determined by house officer survey and direct observation of delivery room skills. Skill acquisition was systematically checked through a “Clinical ladder” system.

Summary of Results:
1. Residents are more comfortable providing CPAP via bag and mask as compared to T-piece and mask (11% vs. 22%).
2. Majority of residents don’t feel comfortable in being at the head of the bed, intubating or running a code for < 28 weeks gestational age infant.
3. Ideal situation is 100% of trainees be competent in NRPI by the time they graduate, our survey shows gap of almost 90% which is excessive.
4. Most residents felt that there are less clinical opportunities available for improving their competency specially in terms of intubating and running codes.
405 EYE-TRACKING IN THE ASSESSMENT OF DIGITAL PATHOLOGY APPLICATIONS
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Purpose of Study: Digital pathology has been demonstrated as a useful tool for applications requiring remote access to diagnostic material. The use of digital imaging in pathology will undoubtedly become a viable option for pathology departments to meet the increasing demands of cost reduction, access to specialist expertise, and educational resources. In order to achieve optimal diagnostic capability through digital means, it is important to understand the visual process by which pathologists arrive at image-based diagnoses. In the digital workflow, little is known about the effect of display device and image presentation on pathologist gaze and interpretation.

Methods Used: Pathologists and trainees were recruited to evaluate sets of diagnostic images in a natural viewing state using Tobii remote eye-tracking systems integrated with a variety of digital pathology platforms. We used a Philips UFS whole slide scanner and hosted the scans on https://slide-atlas.org, a high-performance web-based digital pathology system. Telepathology platforms utilized an Olympus BX43 microscope equipped with a digital camera. Through this method, the diagnostic material and characteristics of the viewing screen could be controlled across digital modalities. Eye-tracking data including fixation count and duration, as well as patterns of eye gaze and slide movement were analyzed and compared across sample categories.

Summary of Results: We present an analysis of the use of eye-tracking methodology under the different viewing modes on digital presentation devices. We have also examined the visual scanning patterns among pathologists with a range of experience, including assessment of the consistency of gaze patterns by individuals, and features of digital images affecting the educational process.

Conclusions: In this first quantitative comparison of gaze utilizing a variety of digital pathology platforms, we observed similarities in localized visual gaze patterns by individuals, and features of digital images affecting the educational process.

406 IMPLEMENTATION OF AN INFANT SKELETAL TRAUMA PATHWAY
Murray L, Doshi P, Farst K. University of Arkansas, Little Rock, AR.

Purpose of Study: The study reviews the items included for evaluation in an infant skeletal trauma pathway for the 3 years following pathway implementation.

Methods Used: A retrospective review of infants evaluated as part of an institution wide infant skeletal trauma pathway during a 3 year period following implementation of the pathway was undertaken. The cases were divided into 4 age groups (0 to <3 months, 3 to <6 months, 6 to <9 months, 9 to <12 months). Data was collected regarding the location of the index fracture, whether abuse was suspected at the time the index fracture was identified, results of the history and testing items obtained during pathway evaluation, and whether the suspicion for abuse changed following the evaluation. Testing for significant differences between the categories was performed with Fisher’s exact test.

Summary of Results: There were 250 cases identified for review. 65 infants were in the 0 to <3 month category (26%), 74 were 3 months to <6 months (30 %), 65 were 6 months to <9 months (26 %), and 46 were 9 months to <12 months (18 %). Skull fractures were the most common index fractures (59 %) followed by fractures of the femur (17 %) and humerus (13 %). Additional fractures were found in 34 infants (14 %). Abuse was suspected from the index fracture in 129 infants (52 %). Abuse was suspected after the pathway work up was complete in 96 infants (38 %). There was no statistical difference in the distribution of cases between age groups comparing suspicion of abuse pre and post pathway evaluation (p = 0.2177). Bruising was documented at the fracture site in only 18 % of infants. An alkaline phosphatase level was tested in 112 of infants (45 %) and was abnormal in 9 (4 %). Rickets was not diagnosed in any of the infants.

Conclusions: Implementation of a pathway to assess for co-occurring injuries and medical conditions that could contribute to fracture risk offers an opportunity to collect information objectively which can decrease bias involved in deciding which patients may be at risk for future injury from abuse.

407 EVALUATION OF A PATIENT HANDOFF CURRICULUM FOR PEDIATRIC RESIDENTS
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Purpose of Study: Due to recently mandated duty hours, the number of patient handoffs between resident physicians has increased substantially. Resident discontinuity is linked with longer length of hospital stay for patients, increased laboratory testing and increased medication errors. The ACGME now requires that residency programs formally train residents in patient handoff. However, there is not an approved curriculum for teaching handoff etiquette. We aim to improve patient handoff amongst pediatric residents using a standardized handoff curriculum.

Methods Used: Through our quality improvement project, we plan to implement a standardized “SIGN-OUT” mnemonic and teach the approach in a small group, interactive setting to our post junior residents. We will evaluate the extent to which resident participation in the formal handoff curriculum results in improvement of their handoff abilities. This will be measured by a validated verbal handoff evaluation tool both before and after the session is taught. In addition, qualitative feedback regarding the benefits of the handoff session will be obtained using an anonymous pre- and post-curriculum survey.

Summary of Results: Inter-Rater Reliability (IRR) amongst our group was assessed using two-way mixed, absolute Intra-Class Correlations to assess the degree that raters provided consistency in their ratings of handovers across subjects. The resulting ICC for content was extremely high (ICC = .95), indicating that we had a very high degree of agreement and suggesting that the handoffs were scored similarly across raters. Our data will be collected September, 2014 through January, 2015. Data will be analyzed and statistics resulted in January, 2015. We plan to present the complete findings in February, 2015.

Conclusions: We hypothesize that the session will result in a more standardized and complete checkout approach amongst our residents and, therefore, an improvement in their handoff evaluations. The improved patient handoff will benefit patient safety in the hospital. In addition, the innovative information gathered with this study has high potential for generalization to other residency programs, as there are few studies of formal patient handoff curriculum.

408 REDUCING UNNECESSARY CONTINUOUS PULSE OXIMETRY
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Purpose of Study: In 2013, the “Choosing Wisely” campaign was instituted to encourage physicians to be conscious of ordering unnecessary medical tests.
One of the top five recommendations was to limit the use of continuous pulse oximetry in children with acute respiratory illnesses to only patients requiring supplemental oxygen. This mandate prompted our group to examine our own use of continuous pulse oximetry, beginning with non-respiratory patients. We also examined the frequency with which nursing appropriately followed the oximetry order.

Methods Used: Data were manually collected for a 6 week period in March and April of 2014. Patients with a non-respiratory diagnosis admitted to all general pediatrics resident teams and an attending-only hospitalist team were included. We recorded the admitting diagnosis, age, admitting oximetry order, and method of pulse oximetry monitoring the patient was actually receiving.

Summary of Results: Data from 227 patients were reviewed with a variety of diseases represented, including gastroenteritis, seizures, sepsis evaluations, and failure to thrive; although specific care of each patient was not investigated, the assumption was made that intermittent pulse oximetry would be appropriate based off the admitting diagnosis. Of those 227 patients, 151 (66.5%) had continuous pulse oximetry ordered, 27 (11.8%) had intermittent orders, and 49 (21.5%) had no order referencing oximetry monitoring. Of those 151 patients with continuous pulse ox, 16 were actually on intermittent monitoring and 22 had no monitoring. Of those 27 with intermittent orders, 17 (63%) were receiving continuous monitoring and 2 had none. Of the 49 with no pulse ox ordered, 32 (65.3%) were on continuous oximetry and 3 (6%) were on intermittent. These results suggest that physicians are often reflexively ordering continuous pulse oximetry monitoring unnecessarily, and the physician order was followed by nursing only 60% percent of the time.

Conclusions: At our institution, there is evidence of overutilization of continuous pulse oximetry. Physician ordering tendency and nursing practice were identified as two key drivers. A pediatric department and nursing education campaign is ongoing with plans for follow up data collection in October 2014; results of our education intervention should be available prior to poster presentation.

CONCLUSIONS

We present 2 cases where bypass grafts were thought to be patent.

Case 1: A 74-year-old male with CABG presented with inferior ST elevation MI. On a previous angiogram, his saphenous vein graft (SVG) to posterior descending artery (PDA) was thought to be totally occluded. This time coronary angiography revealed his SVG-PDA was partially (and not totally) patent. An aortogram can also be done to increase the sensitivity.

Case 2: A 70-year-old female with CABG presented with chest pain, and acute NSTEMI. Since SVG to obtuse marginal (OM) artery was considered occluded on a previous angiogram, a DES was implanted in the native bypassed OM. Three months later, angiogram revealed SVG-OM was widely patent, making the prior intervention probably unnecessary.

missings for the potential for inappropriate revascularization

Bolorunduro OB, Cheema Y, Kouzoum RN. University of Tennessee Health Science Center, Memphis, TN.

Case Report: Purpose: Vascular intervention in coronary artery bypass graft (CABG) patients requires accurate knowledge of prior native coronary and grafted anatomy. Selective angiogram of bypass grafts can be challenging, especially in emergent cases and yet is extremely important in decision making. This becomes especially relevant when there is the need to intervene on a native coronary artery that was previously bypassed. It is important to confirm the absence or occlusion of that graft prior to intervention on the corresponding bypassed native vessel.

Case Report: We present 2 cases where bypass grafts were thought to be occluded on prior angiography, but follow-up angiography revealed them to be patent.

1) A 74-year-old male with CABG presented with inferior ST elevation MI. On a previous angiogram, his saphenous vein graft (SVG) to posterior descending artery (PDA) was thought to be totally occluded. This time coronary angiography revealed his SVG-PDA was partially (and not totally) occluded proximally with fresh thrombus. Aspiration thrombectomy was then performed followed by drug eluting stent (DES) placement.

2) A 70-year-old female with CABG presented with chest pain, and acute NSTEMI. Since SVG to obtuse marginal (OM) artery was considered occluded on a previous angiogram, a DES was implanted in the native bypassed OM. Three months later, angiogram revealed SVG-OM was widely patent, making the prior intervention probably unnecessary.

Conclusions: These cases highlight an important yet under-reported conundrum. Clear and selective angiogram of bypass grafts can be challenging, but remains extremely important in decision making. It is important to confirm the occlusion of bypass grafts prior to intervention on the corresponding bypassed native vessel, to avoid unnecessary interventions. Non-selective aortic root angiogram is usually performed for this purpose; cardiac computed tomographic angiogram can also be done to increase the sensitivity.

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Voxel-based morphometry of patients with schizophrenia and schizoaffective disorders

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Purpose of Study: Neuroimaging has shown that schizophrenia (SZ) and schizoaffective disorders (SD) are associated with neuroanatomical abnormalities, which are suggestive of diffuse brain pathology. Identifying the morphometry of SZ and SD is critical to understanding the etiopathophysiology in order to prescribe treatment. This study utilized voxel-based morphometry to investigate regional gray matter (GM) volume differences in SZ, SD, and healthy controls.

Methods Used: 50 patients (22 - SZ and 28 - SD) and 47 controls were randomly selected. Ethical approval and consent were obtained. A Siemens Sonata 1.5T scanner was used for imaging. T1-weighted MRI data was acquired using MP-RAGE sequences. VBMM was used for comparing GM volume differences between groups. Multiple comparison error was corrected using family-wise error (FWE) based correction (p < 0.05).

Summary of Results: Regional GM decreases were identified in schizophrenics at the ventromedial prefrontal cortex, the right and left temporal areas, and the somatosensory region in the left parietal lobe versus controls. SD patients demonstrated significant reductions in the mediadorsal frontal lobes and the left lateral temporal and parietal lobes, as well as thalamic and cingulate regions. Comparing SZ and SD demonstrated greater decreases with FWE. However, uncorrected (p < 0.001) results indicated that SD patients had reductions in thalamic and dorsal parietal regions compared to SZ, while SZ showed decreases in the ventral prefrontal cortex relative to SD.

Conclusions: Different regions of reduced GM volume are identified in SZ and SD patients compared to controls. These regions are related to components of the pathoetiology underlying symptoms and neuropsychological manifestations of schizophrenia.

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Segmentation test-retest of perfusion computed tomography-obtained blood-brain barrier in prediction of infarct expansion following acute ischemic stroke

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Purpose of Study: Deterioration of the blood-brain barrier is believed to contribute to infarct extension following acute ischemic stroke. Patient outcomes have been shown to be inversely proportional to final infarct volume; thus, prediction of infarct growth is one of the most important prognostic criteria in stroke care. The goal of this study was to evaluate the use of perfusion CT obtained blood-brain barrier permeability (BBBP) to predict infarct expansion.

Methods Used: 30 patients with acute ischemic stroke, who underwent perfusion CT on admission, were enrolled in this study. Ethical approval and consent were obtained. BBBP was measured using the Gjedde-Patlak method, and follow-up imaging was subsequently performed at least 5 days after stroke onset. Infarct expansion ratio was defined as the percentage of initial penumbra turned to infarct. Linear regression was used to test the relationship between increased microvascular permeability and infarct expansion ratio.

Summary of Results: A proportional association was found between increased BBBP (measured in penumbra and entire ischemic volume) and infarct expansion ratio (R = 0.40, p < 0.05). Predictability of infarct expansion was substantially improved with subsequent imaging compared to untreated patients compared to patients who underwent recanalization therapy (R=0.53 and 0.43, p < 0.05 for permeability value measured in penumbra and the entire ischemic volume, respectively.)
Conclusions: This study established that increased microvascular permeability is a potential early predictor of infant expansion and radiological outcome in patients following acute ischemic stroke, particularly in those treated conservatively.

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REVERSIBLE POSTERIOR LEUKOENCEPHALOPATHY SYNDROME INDUCED BY ATIXINIB

Klomjit S 1, Hosiriluck N 1, Laengvejkal P 2, Panikkath D 1, Nugent K 1, 2, "Texas Tech Health Sciences Center, Lubbock, TX; 2Texas Tech Health Sciences Center, Lubbock, TX and 3Texas Tech Health Sciences Center, Lubbock, TX.

Case Report:

BACKGROUND: Reversible posterior leukoencephalopathy syndrome (RPLS) is a clinical constellation which includes headache, confusion, decreased level of consciousness, and seizures in association with reversible focal vasogenic edema of posterior cerebral white matter on magnetic resonance imaging (MRI). Atixinib is a novel oral anti-vascular endothelial growth factor (VEGF) multikinase inhibitor with antiangiogenic activity. This is the second case report that demonstrates that atixinib can cause RPLS.

CASE: A 71-year-old white man with metastatic renal cell carcinoma was previously treated with everolimus. His tumor progressed despite an initial response. He then received bevacizumab for four cycles, but his tumor still progressed. He was, therefore, started on atixinib. One month after atixinib treatment, he was then brought to the hospital after episodes of seizure activity-jerking movements in both upper and lower extremities. Vital signs showed a blood pressure of 140/70 mmHg and body temperature of 100.8°F. The patient was admitted and developed another seizure with elevated systolic blood pressures of 170-190 mmHg. Atixinib was discontinued. Patient was treated with levetiracetam (1,000 mg daily) and lonazepam. He was initially given broad spectrum antibiotics and acyclovir for suspected intra-cerebral infection, but this treatment was stopped after cerebrospinal fluid tests were normal. CT scan of the head showed no abnormalities. MRI of the brain showed T2 hyperintensity of cerebellum and occipital lobes. The patient slowly improved, and he was discharged home with levetiracetam. He had not had any seizures since discharge. The follow-up MRI of the brain showed resolution of the abnormalities.

DISCUSSION: Atixinib is a well-known cause of elevated blood pressure via its VEGF-2 inhibitory effects. RPLS is a rare complication of atixinib which should be recognized since stopping the medication is critical. Bevacizumab is also known to cause RPLS which we should take into consideration when the patients receive VEGF inhibitors.

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NOVEL MUTATIONS IN THE RNA BINDING PROTEIN HETEROGENEOUS NUCLEAR RIBONUCLEAR PROTEIN A1 IN MULTIPLE SCLEROSIS PATIENTS: IMPLICATIONS FOR NEURODEGENERATION

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Purpose of Study: Multiple sclerosis (MS) is the most common autoimmune disease of the central nervous system (CNS) in humans. Identification of somatic mutations associated with the pathogenesis of MS could lead to new methods for diagnosing and treating MS. Considering heterogeneous nuclear ribonuclear protein A1 (hnRNP A1) is an autoantigen associated with MS, we hypothesized that single nucleotide variants (SNVs) might be present in the transportin binding site of hnRNP A1, which would alter hnRNP A1's function.

Methods Used: DNA was isolated from brain and peripheral blood mononuclear cell (PBMC) DNA from 25 patients. PCR was performed that included exons 8 and 9 of hnRNP A1 DNA. PCR products were DNA sequenced and analyzed for mutations.

Summary of Results: Results show that healthy control patients lacked SNVs in the transportin binding site of hnRNP A1. In contrast, all primary progressive MS patients had at least one SNV that resulted in an amino acid substitution in the transportin binding site. SKNSh cells transfected with WT hnRNP A1 DNA showed hnRNP A1 localized within the nucleus and no signs of stress or apoptosis. In contrast, mutant hnRNP A1 showed mis-localization of hnRNP A1 to the cytoplasm in a granular fashion, suggesting the presence of stress granules. Stress granule formation was confirmed by staining for TDP-43. Further, cells transfected with mutant hnRNP A1 DNA stained for caspase 3 and showed additional signs of apoptosis, such as molecular blebbing and fragmented nuclei.

Conclusions: We discovered novel somatic genomic SNVs that have the potential to contribute to the pathogenesis of MS. We were able to determine that transfection of mutant hnRNP A1 results in hnRNP A1 mis-localization to the cytoplasm, formation of stress granules, and apoptosis. Future studies will focus on the in vivo effects of mutant hnRNP A1 isolated from the brains of MS patients and the functional significance of mutations in PBMCs by examining the effect of the SNVs on the immune response in MS patients.

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MISS CIS...MISS MS! (CLINICALLY ISOLATED SYNDROME...MULTIPLE SCLEROSIS)

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Case Report: A 32yo African American presented with one month of intermittent tingling in his toes that progressed to weakness of the right leg and blurry vision. Eventually he could not walk without support and came to the hospital. 5 months before he had right sided facial numbness and tingling, was diagnosed with Bell's palsy, treated and released. PMH and FH were neg. PE: A&O, vitals NL, lungs, heart and abd normal, neuro: right ptosis, decreased visual acuity, horizontal nystagmus, left lateral gaze palsy. Strength 3/5 in right UE and LE (5/5 on left) and patchy loss of fine touch in right LE; gait was ataxic. Labs were normal and tox screen neg. Brain and spine MRI showed scattered T2 weighted foci in the periventricular white matter, mid-brain, frontal lobe, optic nerves, and anterior 2/3 of spinal cord at C4. CSF showed high protein, no WBC's, 2 oligoclonal bands. increased IgG synthesis. The patient was dx with relapsing remitting MS and treated with high dose IV methylprednisolone and beta interferon. He showed slow but noticeable recovery. 3) Secondary Progressive and 4) Primary Progressive. Dx is primarily clinical but supported by MRI, CSF oligoclonal bands and increased IgG index. High dose pulse steroids and interferon are commonly used though multiple disease modifying agents are used and under study. Dx at the CIS stage is a major goal of current research since prompt dx at this point may delay the occurrence of a 2nd attack up to 5 years. This case reminds internists to consider MS in any young person with unusual motor or sensory deficits.

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ACUTE KIDNEY INJURY IN SUBARACHNOID HEMORRHAGE IS ASSOCIATED WITH HIGHER DEATH AND DISABILITY

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Purpose of Study: Acute kidney injury (AKI) in setting of subarachnoid hemorrhage (SAH) maybe associated with higher mortality. We conducted this study to determine the effect of AKI on outcomes of SAH patients treated in a large national cohort.

Methods Used: We analyzed data from all patients admitted to US hospitals between 2005 and 2011 with a primary discharge diagnosis of SAH and secondary diagnosis of AKI. The effect of AKI on discharge outcomes was...
analyzed after adjusting for potential confounders using logistic regression analysis.

Summary of Results: Of 173,053 patients with SAH, 8,302 (4.8%) had AKI. In AKI group, 290 (3.5%) patients were on dialysis. SAH patients with AKI had higher rates of moderate to severe disability (40.7% versus 36.6%; p<0.0001) and in-hospital mortality (39.4% versus 20.1%; p<0.0001) compared to those without AKI. After adjusting for potential confounders SAH patients with AKI had higher odds of moderate to severe disability [odds ratio (OR) 1.3, 95% confidence interval (CI) (1.1-1.5, p=0.02)] and death [OR(2.0, 95% CI) (1.8-2.3, p=0.0001)].

Conclusions: AKI in patients with SAH is associated with significantly higher rates of death and disability. It remains unclear whether aggressive fluid replacement and bicarbonate administration can reduce the high rates of adverse outcomes.

416 DIFFUSE MUSCLE WEAKNESS: A CASE OF INFANTILE BOTULISM

Paccione R, Remedios P, Gautreaux J, English R. Children’s Hospital of New Orleans, New Orleans, LA.

Case Report: A seven month old male with no prior past medical history was brought to the Emergency Department due to concerns of floppiness by his family. He was reportedly acting differently and was unable to hold his head up. On physical exam, pertinent findings included profound hypotonia, proximal greater than distal muscle weakness and areflexia. Evaluation included a lumbar puncture, CT head, EEG and EMG, all of which were negative. Nerve conduction studies did not show any evidence of demyelination, ruling out Guillain-Barre syndrome. Repetitive stimulation showed facilitation of compound muscle action potentials, indicative of pre-synaptic neuromuscular junction disorder. Lambert-Eaton antibodies were negative. Further history revealed the patient had been given home-containing herbal supplements for his cough. Based on these findings, the diagnosis of botulism was made. Treatment was initiated with Botulism Immune Globulin with immediate but gradual improvement over the next several weeks.

Discussion: Infant botulism, though rare, is the most common form of human botulism in the United States. It is caused by swallowed spores of Clostridium botulinum. Inadequately prepared food, canned foods and honey are common vectors. The spores germinate, temporarily colonize the lumen of the large intestine and produce botulinum neurotoxin. The toxin is devastating, as it can cause irreversible binding to the neuromuscular junction. Due to the varying degrees of severity, it is necessary to watch these patients closely with frequent monitoring. Treatment with Botulism Immune Globulin has been effective at neutralizing the toxin and improving clinical outcomes.

Conclusions: Infant botulism is a condition that may be avoided if education regarding the potentially harmful effects of homeopathic remedies that include honey is explained to families. Though honey is often thought to be associated with spore ingestion, commonly the cause of spore ingestion is unknown. A judicious diagnosis is imperative for achieving optimal treatment results and reducing mortality from rapid disease progression.

417 ACUTE RESPIRATORY FAILURE REQUIRING MECHANICAL VENTILATION AS INITIAL PRESENTATION OF MYASTHENIA GRAVIS

Fitzgerald LM, Fitzgerald S, Roy T. East Tennessee State University, Johnson City, TN.

Case Report: A 42 year old female patient was transferred to the medical ward following a three day admission to the intensive care unit for unexplained respiratory failure requiring endotracheal intubation. She had one prior admission for respiratory failure requiring intubation several months prior but an etiology was never determined. Two weeks prior she had an upper respiratory tract infection treated with antibiotics but there was no history of chronic obstructive pulmonary disease, asthma, smoking, drug abuse or other medical history. On initial review of systems, the patient presented with fluctuating muscle weakness, dysphagia, aspiration, and intermittent diplopia. Physical exam showed an obese female with normal vital signs, mild bilateral ptosis, diplopia with repeated eye movements, and 4/5 muscle strength in large muscle groups. An ice pack was placed over the left eye for two minutes with resultant improvement in the ptosis compared to the right. Blood was drawn for anti-acetylcholine receptor antibodies and a neurology consult was placed. A diagnosis of myasthenia gravis was made and she was discharged home with neurology follow up and appropriate treatment. This is a case demonstrating the novel presentation of myasthenia gravis with severe bulbar symptoms as myasthenic crisis precipitated by an upper respiratory tract infection.

418 FLACCID PARALYSIS IN A FIVE YEAR OLD WITH ENTEROVIRUS

Murphy M, Rajendra R, Chang E, Deputty S. LSUHSC, New Orleans, LA.

Case Report: A five year old female presented with fever and vomiting for four days and two days of neck stiffness with left proximal arm weakness. Initial CSF studies showed 54 WBCs with lymphocytic predominance and remainder of CSF studies were normal.

On examination, patient was in acute distress with neck movement. She was able to shrug her right shoulder but unable to move her left shoulder. Left upper extremity strength was 0/5 proximally and 4/5 distally. Right upper extremity and bilateral lower extremity strength were 5/5. Reflexes were absent in all extremities. The remainder of her neurological exam including cranial nerves and sensation was normal.

MRI of the brain was normal and spine showed swelling and edema from C2-C6-C7 with increased signal on T2 centrally located in the grey matter. She also had a normal viral panel for rhino/enterovirus and RSV. One day after admission she developed respiratory distress secondary to left hemidiaphragm paralysis. She was intubated and started on high dose steroids and plasmapheresis. A repeat LP was performed and CSF was negative for West Nile, Parechovirus and Enterovirus. Her weakness progressed and by day four of admission she had no proximal or distal movement of left upper extremity and minimal distal movement of her right upper extremity and bilateral lower extremities. She also developed paresthesias of the 6th and 7th cranial nerves and remained areflexic.

Due to worsening paresis, plasmapheresis was continued for 11 doses, followed by four days of IVIG. Over the course of two weeks, cranial nerve palsies resolved and proximal and distal strength of right upper extremity returned, but no improvement of left upper extremity and minimal distal improvement of bilateral lower extremities. EMG showed pure motor axonopathy. She also continued to require ventilator support and now is trach dependent.

This disease process is polio-like in that it is a pure motor syndrome with no loss of sensation and only affecting grey matter on imaging. With recent reports from the CDC about enterovirus D68 and flaccid paralysis, we suspect that our patient has a viral induced pure motor syndrome involving the spinal cord. Her sputum and CSF is currently being tested further by the CDC.

419 INFANTILE PSEUDOTUMOR CEREBRI

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Case Report: A previously healthy 7-month old female presented with a two-day history of bulging fontanel and morning-time vomiting. She was otherwise well-appearing, and no other symptoms were present. There was no recent trauma or insect bites, nor any sick contacts. Five days prior to presentation, she was started on oral fluconazole for treatment of a clinically diagnosed fungal diaper rash that failed to resolve with topical therapy. The only pertinent physical exam finding was a bulging fontanel. Vital signs, CBC and CMP were all within normal limits, as was a Head CT. Thyroid, cortisol and vitamin-A levels were measured and returned normal values. MRI, MRA and MRV studies were conducted and were normal. A lumbar puncture was performed yielding an opening pressure of over 350mmH2O of clear fluid. CSF analysis and cell counts were normal. 8mL of CSF were drained, lowering the pressure to 180mm and resolving the bulge. The patient tolerated the procedure without any complications, and was discharged the next day with a diagnosis of pseudotumor cerebri.

Pseudotumor Cerebri is a syndrome of intracranial hypertension in the absence of increased ventricular size, a mass lesion, infection or malignancy,
and with normal brain parenchyma. Diagnosis is defined by the revised modified Dandy criteria: if present, signs and symptoms are of generalized increased and are without localization, except for 6th nerve palsy, there is increased CSF opening pressure in properly performed lumbar puncture, CSF composition is normal, there is no cause for intracranial hypertension is identified. While there have not been many reliable studies investigating the normal opening pressure in children, normal values a currently held to be <180mmH2O in those younger than 8 years of age. The condition also seems to affect boys and girls equally. Treatment is aimed at symptoms and preservation of visual function, with acetazolamide and other diuretics being first-line options, and if medical management fails, surgical treatment to relieve the pressure should be considered. Serial lumbar punctures have been discouraged in children. While many drugs such as vitamin-A, tetra-cyclines and oral contraception have a well-documented association, there have been no documented cases to date of an association with fluconazole.

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REVERSIBLE MOVEMENT DISORDER DUE TO ANTIPHOSPHOLID SYNDROME

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Case Report: Antiphospholipid syndrome (APS) is a hypercoagulable state associated with antiphospholipid and anticardiolipin antibodies. Most patients have ischemic complications secondary to arterial or venous thrombosis. APS can also cause a variety of non-thrombotic neurological clinical findings. We present clinical and neuroimaging findings of a teenager who presented with a transient ataxia and ballistic chorea associated with transient neuroimaging changes in association with persistently elevated antiphospholipid and anticardiolipin antibodies.

A previously healthy 14-year-old female presented with rapidly progressing dysarthria, difficulty walking, talking and writing associated with uncontrollable involuntary movements of all extremities and worsening of headache without alteration of consciousness. She received intravenous immunoglobulin (IVIG) 1g/kg/dose on the third day of illness and rapidly improved. Within one week, all involuntary movements had stopped with almost complete return to baseline.

MRI of brain on presentation showed acute infarction of bilateral cerebellar superior hemisphere with right more than left. CT angiography of the brain and neck were normal. Antiphospholipid IgM was 38 MPL on day 2 and 51 MPL after 3 weeks. Anticardiolipin IgM was 18 MPL at 3 weeks. Lupus anticoagulant, ANA and ds DNA antibodies were negative. Initial Coagulation profile and 5 days later respectively showed PTT of 39 then 34 seconds, INR 0.9 then1, PT 13 then15 seconds. Fibrinogen was within normal limits. Platelets were normal. The remaining Thrombophilia work up was negative. Follow-up MRI after 1 week showed transient white matter diffusion defect involving bilateral cerebellar hemispheres which has largely improved from initial scan. MRV was negative.

MRI changes and neurological findings can be transient in primary antiphospholipid syndrome (PAPS). We present a patient with most of her neurological manifestation can’t be explained by cerebellar infarction found on MRI. We hypothesize that these manifestations were caused by direct effect of antiphospholipid and anticardiolipin antibodies.

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ISOLATED NEUROSARCOIDOSIS IN A FIFTEEN-YEAR-OLD MALE

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Case Report: Neurosarcoidosis has been noted in approximately 5 to 10% of all cases of adult sarcoid. However, only 41 cases of neurosarcoidosis have been reported in the pediatric population, with eight of those cases being an isolated neurosarcoidosis. We present the ninth case of a child with the initial presentation of isolated neurosarcoidosis. A fifteen-year-old male from Honduras presented with a one year history of intermittent headache, vomiting, ataxia, and a resolved Bell’s Palsy. An extensive work-up was completed in Honduras, including serial brain MRIs, lumbar puncture, brain biopsy. However, a diagnosis was unable to be confirmed. On physical exam, focal neurologic deficits included brisk reflexes at bilateral patellae, clonus at bilateral ankles, and ataxia on tandem gait. Brain MRI done at the time of presentation was compared to previous studies and showed continued progression of disease, including enhancement of cerebellar folia and leptomeninges with progression to cerebral and central white matter. Seven months after his initial presentation in Honduras, a chest CT revealed pulmonary nodules bilaterally. Angiotensin converting enzyme was elevated in the CSF but not in the serum. Serum testing for tuberculosis, coccidiosis, HIV, syphilis, and HSV were negative. Brain biopsy was reviewed and found to have multiple perivascular noncaseating granulomas with scattered Langhans type giant cells. Based on our patient’s imaging, laboratory studies, and biopsy findings we concluded that our patient had neurosarcoidosis.

Our literature search revealed 41 reported cases of neurosarcoidosis in children. The most common manifestations included cranial neuropathies (25%), seizures (20%), and hypothalamic deficits (25%) with the latter two being more likely in younger children. Diagnosis is made by biopsy; however, other studies such as imaging and laboratory tests can aid in diagnosis. Treatment typically includes corticosteroids and/or immunosuppressants, as was initiated in our patient.

Although neurosarcoidosis is rare in children, patients with focal neurological findings and abnormal imaging who have had a negative infectious work-up should be considered for autoimmune diseases such as neurosarcoidosis.
**COMPARISON OF OUTCOMES OF VERY LOW BIRTH WEIGHT INFANTS FED HUMAN MILK FORTIFIED WITH LIQUID ACIDIFIED HUMAN MILK FORTIFIER VERSUS POWDERED FORTIFIER**

Thoni N1, Shah S1, Sweeney A1, Talati A1, 1UTHSC, Memphis, TN and 2Regional One Health, Memphis, TN.

**Purpose of Study:** Very low birth weight (VLBW) infants have higher nutritional needs and human milk is not enough to provide adequate nutrition. The use of human milk fortifiers (HMF) has provided additional nutrients that help to meet the needs of VLBW infants and have been associated with better growth outcomes. Different fortifiers are available with differences in their content. We compared the growth and feeding tolerance of VLBW infants fed human milk fortified with acidified liquid HMF (lHMF) vs powdered HMF (pHMF).

**Methods Used:** A retrospective chart review was done from April 2012 to May 2014 for VLBW infants admitted to our NICU. 25 VLBW infants fed pHMF and 24 fed pHMF were evaluated. A 4-week period was reviewed starting the day after the infant was off TPN and receiving fortified human milk. Daily weight, weekly length and head circumference (HC) were recorded. Relevant serum chemistries were recorded weekly. Daily protein, calorie, and lipid intake were calculated.

**Summary of Results:** Gestational age and gender were similar, while pHMF had a higher weight (1170g vs 1057g, p=0.03) and length (38cm vs 36.2cm p=0.03) prior to initiation. After 4 wks there was no significant difference between the 2 grps for length, weight or HC. Protein and lipid intakes were higher in lHMF group, but there was no significant caloric difference during weeks 2-4. Mean serum chemistry values prior to initiation showed no significant differences. After 4 weeks there was a significantly higher protein (p=0.02), albumin (p=0.003), sodium (p=0.02), and potassium (p=0.002) in the lHMF grp. The increase in growth velocity in lHMF grp was significantly higher compared to pHMF grp. Both grps showed a decrease in serum potassium. In 2 grps with no difference between the 2 grps during any week. There was no difference in the number of times feedings were held for more than 24 hrs, pHMF grp showed significantly more episodes of feeding intolerance in wk 3.

**Conclusions:** Infants fed lHMF received higher protein and lipid leading to higher total protein and albumin levels after four weeks. Better improvement in growth velocities between wks 1-4 was seen in lHMF grp. Protein intake need to be optimized VLBW infants to prevent postnatal growth restriction.

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**EFFECT OF SURFACTANT PROTEIN A ON GENE PROFILE IN HUMAN RETINAL ENDOTHELIAL CELLS**

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**Purpose of Study:** We have previously shown that Surfactant Protein A (SP-A) modulates human retinal endothelial cell function in vitro and showed a decrease in neovascularization in SP-A-/- mice in the oxygen induced retinopathy (OIR) model. We hypothesize that SP-A decreases inflammatory signals and alters the expression of angiogenic factors in retinal reduced retinopathy (OIR) model. We hypothesize that SP-A decreases inflammation and alters the expression of angiogenic factors in retinal endothelial cells (HREC’s) in culture newborns (G<29 weeks gestation) with umbilical vein catheter (UVC) and/or artery (UAC) placements. Mortality is significantly higher in premature infants associated with birth weight, gestational age, and UVC placement. We hypothesize that UVC placement directly causes increased mortality via thrombotic or embolic events.

**Methods Used:** Utilizing a nested case-control study design, we expanded data collection to include all electronic or written medical record data regarding the management, morbidities, and mortalities in premature (<29 weeks gestation) with umbilical vein catheterization. We expanded the database to include all patients with UVC placements who survived and compared those with newborns who survived UVC placements.

**Conclusions:** We have previously reported that in extremely premature newborns (<29 weeks gestation) with umbilical vein catheterization (UVC) and/or artery (UAC) placements, mortality is significantly higher in newborns with UVC placements compared to those of newborns who survived UVC placements.
T10 (low-lying). Of survivors, only 15% had low-lying lines following the initial placement film. Of non-survivors, 43% had low-lying lines (p=0.027). Though significant differences in high-lying lines, platelet counts, WBC counts, metabolic profiles, transfusions, or medication usage were not demonstrated, cranial ultrasonographic scan (CSS) results differed. Of survivors with CSS (n=67), only 3% had grade 4 intraventricular hemorrhage (IVH) - occurring only on the right - and 69% had normal CSS. Of non-survivors with CSS (n=8), 50% (p<0.001) had grade IVH - only on the left if unilateral or greater on the left if bilateral - and only 25% (p=0.022) had normal CSS. Conclusions: Low-lying UVCs are significantly more common in non-survivors and may contribute to mortality by promoting thrombosis and/or embolism in the slow-flowing portal circulation. Moreover, the significantly increased occurrence of left grade 4 IVH in non-survivors suggests possible right-to-left shunting of thrombotic or air emboli across the patent ductus arteriosus into the left carotid circulation contributing to grade 4 IVH occurrence.

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OMEPRAZOLE POTENTIATES HYPEROXIA-INDUCED DEVELOPMENTAL LUNG INJURY IN NEWBORN MICE

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Purpose of Study: Hyperoxia contributes to bronchopulmonary dysplasia (BPD) in preterm infants. We showed that omeprazole (OM) protects adult mice against hyperoxic lung injury via an hydrogen receptor (AhR) dependent mechanism. Whether OM protects newborn mice against hyperoxia-induced developmental lung injury is unknown. Therefore, we tested the hypothesis that omeprazole-treated newborn mice will have decreased inflammation and alveolar simplification upon exposure to hyperoxia.

Methods Used: Within 12 h of birth, wild type C57BL/6J (WT) pups were immediately exposed to either air (normoxia) or 85 % O2 (hyperoxia) for 14 d. WT newborn mice were either exposed to either air or hyperoxia, were administered 10 or 25 mg/kg of OM or an equivalent volume of the vehicle, polyethylene glycol (PEG), i.p. once daily from birth for 14 d. The dosing regimen was based on our finding that at least 25 mg/kg/d of i.p. OM is required to activate the AhR. Additionally, we used OM at a dose of 10 mg/kg/d to determine its AhR independent effects. Following exposure, the lungs and bronchoalveolar lavage fluid (BALF) of the mice were harvested to determine inflammation, angiogenesis, and alveolarization. Lung inflammation was determined by immunohistochemistry (neutrophils and macrophages) and BALF protein concentration, alveolarization was determined by lung morphometry (alveolar size), and angiogenesis was determined by vonWillebrand factor immunostaining.

Summary of Results: OM treatment (10 to 25 mg/kg) increased oxygen toxicity in a dose-dependent manner. OM-treated mice were significantly growth restricted upon exposure to hyperoxia. Furthermore, OM increased hyperoxia-induced lung macrophage influx. Interestingly, mice treated with high-dose OM had persistent acute lung inflammation as evident by the presence of hyaline membranes, neutrophil infiltrates, and proteinaceous debris in the alveolar space. Additionally, high-dose OM increased alveolar simplification (alveolar size) and BALF protein concentration. However, OM at either dose did not decrease lung angiogenesis.

Conclusions: Contrary to our hypothesis, OM potentiates alveolar simplification and inflammation in a model of hyperoxia-induced lung injury in newborn mice. These findings suggest the need to be cautious before instituting OM therapy in neonates.

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IMPROVING COMPLIANCE WITH EVIDENCE BASED GUIDELINES REGARDING SEDATION FOR NON-EMERGENT NEONATAL INTUBATIONS: A QUALITY IMPROVEMENT PROJECT

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Purpose of Study: In older children and adults, tracheal intubation is performed after adequate anesthesia to decrease pain and discomfort, facilitate intubation, and decrease adverse reactions. In 2010 the American Academy of Pediatrics (AAP) published guidance that remedication should be used for all newborn intubations except emergent events. Despite these recommendations, multiple studies indicate heterogeneous implementation of these guidelines. We aimed to determine if a combined approach of regular multidisciplinary educational sessions and a new electronic medical record (EMR) documentation tool would improve Walter Reed National Military Medical Center Neonatal Intensive Care Unit (WRNNMC NICU) compliance to greater than 80%.

Methods Used: A quality improvement project was conducted and subsequent IRB approval obtained to study the impact retrospectively. Pre-intervention records from a 12 month period were reviewed to determine existing compliance rates for all infants admitted to WRNNMC NICU who underwent endotracheal intubation. After implementing a new educational curriculum, multidisciplinary educational sessions and a new endotracheal intubation note template in the EMR, records were reviewed over an 11-month post-intervention period.

Summary of Results: During 2012 to 2013, 62 non-emergent intubations were performed in the WRNNMC NICU. Only 55% of infants (34/62) received some form of sedation. Overall compliance rate with guideline-recommended medications was 59%. Over the 11-month period of this intervention, compliance rates increased to 82% (69/82) in 2014. In 2015 compliance rates increased to 78% (53/68) with sedation. Compliance with guideline-recommended medications improved to 87% (46/53).

Conclusions: Our combined approach of educational sessions and a new EMR tool led to marked improvement in compliance with AAP guidelines regarding sedation use for neonatal intubations. The new documentation reinforced the importance of sedation use and appropriate medication selection. Further study regarding the impact these methods have on trainee success at intubation is indicated. Utilizing a combined method to facilitate quality improvement is a successful approach when new practice guidelines are published.
Purpose of Study: Therapeutic approaches to treat pulmonary hypertension (PH) pathogenesis involve pulmonary artery smooth muscle cell (PASMC) proliferation. Loss of phosphatase and tensin homolog deleted on chromosome 10 (PTEN) contributes to PH pathogenesis. We previously identified PTEN as a PASMC-specific hypoxia inducible factor 1alpha (HIF-1alpha) target. In this study, we evaluated the role of PTEN in PASMC proliferation in PH settings.

Methods: We compared PASMC proliferation in normoxic (21%O2) and hypoxic (1%O2) conditions with or without RGD (10μM) treatment. We measured the expression of HIF-1alpha, PTEN, phospho-PTEN, and PTEN mRNA levels by qPCR.

Results: PASMC proliferation was significantly higher in hypoxic compared to normoxic conditions. RGD treatment attenuated PASMC proliferation in hypoxic conditions. HIF-1alpha and PTEN mRNA levels were significantly elevated in hypoxic conditions compared to normoxic conditions. RGD treatment attenuated HIF-1alpha and PTEN mRNA expression.

Conclusions: PTEN is a target for therapeutic intervention in PH. RGD may be a potential therapeutic agent for PH treatment.
of PTEN depletion in PH pathogenesis remains to be defined. MicroRNA (miRNA), small RNA molecules that regulate gene expression through post-transcriptional gene silencing, modulates cellular proliferation. This study examines the role of miRNA-21 in hypoxia-induced PASMC proliferation and PTEN depletion and explores if the antiproliferative effects of the peroxisome proliferator-activated receptor gamma (PPARγ) activator, rosiglitazone (RSG), occurs through modulation of miRNA-21 expression.

**Methods Used:** HPASMC monolayers were exposed to normoxia or hypoxia (1% O2) for 72 hours ± treatment with RSG (10 μM) during the last 24 hours of exposure. Male C57Bl/6 mice were exposed to normoxia or hypoxia (10% O2) for 3 weeks ± RSG (10 mg/kg/day) via oral gavage during the last 10 days. MiRNA and mRNA levels in cell lysates or lung homogenates were quantified by qRT-PCR. Mature miRNA-21 mimic or antisense oligonucleotides were transfected to overexpress or inhibit miRNA-21. HPASMC proliferation was subsequently measured using MTT assay or cell counting.

Finally, PTEN expression was silenced using siRNA to examine its role in HPASMC proliferation.

**Summary of Results:** MiRNA-21 overexpression, hypoxia exposure, or siRNA-mediated depletion of PTEN enhanced proliferation of HPASMC. MiRNA-21 knockdown tended to increase PTEN levels and attenuated hypoxia-induced HPASMC proliferation. RSG attenuated hypoxia-induced reductions in HPASMC proliferation, but did not inhibit HPASMC proliferation induced by PTEN depletion. RSG attenuated hypoxia increases in lung miRNA-21 levels in vivo.

**Conclusions:** MiRNA-21-mediated reductions in PTEN play a central role in HPASMC proliferative responses to hypoxia. RSG confers antiproliferative effects by preventing hypoxic increases in miRNA-21 and reductions in PTEN. Ongoing studies are designed to define how RSG attenuates miRNA-21 and additional targets regulated by miRNA-21 that modulate hypoxic PASMC proliferation.

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**TNF ALPHA REGULATES AQUAPORIN 1 AND AQUAPORIN 3 IN PULMONARY MICROVASCULAR ENDOTHELIAL CELLS**

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**Purpose of Study:** Inflammation plays a key role in endothelial hyperpermeability, TNF alpha is a major proinflammatory cytokine that been implicated in the development of pathological conditions including lung edema. Water channels aquaporins AQP1 and AQP3 play a role in fluid retention. In the present study, we investigated whether TNF alpha affects fluid internalization by regulating AQP1 and AQP3.

**Methods Used:** Human primary pulmonary microvascular endothelial cells (PMVEC) were cultured according to supplier’s recommendations. PMVEC were treated with TNF alpha (10ng/ml) for 24 hours and fluid internalization was assessed by FITC-dextran internalization using fluorescent microscopy. AQP1 and AQP3 mRNA level was analyzed using real-time PCR. 18S gene was used as endogenous control gene. Data was analyzed using ANOVA.

**Summary of Results:** TNF alpha increased fluid internalization. Treatment of PMVEC with TNF alpha significantly increased the number of internal FITC-dextran vesicle compared to control cells. AQP1 and AQP3 mRNA level were significantly increased following TNF alpha treatment. TNF alpha increased AQP1 mRNA by 2.9-folds and AQP3 mRNA by 2.6-folds compared to control (p < 0.05).

**Conclusions:** These data suggest that TNF alpha increases endothelial hyperpermeability and regulates water channel proteins AQP1 and AQP3, which seem to play a role in fluid internalization in PMVEC. These data also indicate that inflammation mediates endothelial hyperpermeability and AQP1 and AQP3 may be novel therapeutic targets for pulmonary edema treatment.

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**PEER EDUCATION TO IMPROVE IDENTIFICATION AND TREATMENT OF PATIENTS WITH SEPSIS**

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**Purpose of Study:** The Surviving Sepsis Guidelines (SSG) were developed to enhance identification, treatment, and outcomes of patients with sepsis, severe sepsis, or septic shock. We hypothesized educating residents monthly about SSG would improve recognition, treatment and mortality.

**Methods Used:** A Peer Education Program (PEP) was instituted and data for the twelve months before and the nine months after PEP were compared. The PEP is a teaching program to review the criteria for diagnosing and treating sepsis. PEP residents gave mini-lectures to the medicine resident teams each month. The goals of the PEP were: review diagnostic criteria of sepsis, Early Goal Directed Therapy (EGDT), case studies, providing SSG cards and placing posters in-call rooms and break areas, and OUMC goals for sepsis identification and treatment. Patients with sepsis were identified by a nurse specialist who manually reviews all admissions to OU Medical Center. Physician documentation, adherence to early EGDT, and mortality were analyzed.

**Summary of Results:** The incidence of sepsis, severe sepsis and septic shock in the period before and after the intervention was similar. The PEP significantly improved physician recognition. Despite increased recognition, adherence to treatment guidelines did not improve. Interestingly, sepsis mortality rates showed a trend toward improvement (p = 0.06) regardless of continued poor implementation of EGDT.

**Conclusions:** PEP significantly increased recognition and improved outcomes for septic patients even in the absence of detectable treatment change.
BUILDING PATIENT CONFIDENCE IN THE RIGHT TREATMENT MODALITY: USE OF ALBUTEROL HFA INHALERS DURING INPATIENT ASTHMA EXACERBATIONS

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Purpose of Study: A mainstay of the acute inpatient management of asthma is the short acting beta agonist. The two main delivery systems are the metered dose inhaler (MDI) and the nebulizer machine. Patients and families often have perceptions of increased efficacy of nebulized treatment, due in part to its predominance in the hospital setting. However, multiple studies indicate that MDI delivery is at least as effective as nebulizer treatments. At our institution, nebulized treatments are more expensive and used more often than MDI. Our goal was to educate providers regarding the benefits of MDI delivery.

Methods Used: We administered surveys to providers at our institution to assess attitudes regarding Albuterol delivery systems. These demonstrated a preference for nebulized delivery. Reported barriers to MDI use included unfamiliarity with dosing and parental preference for nebulized treatments. We then presented research in favor of MDI use to providers during a one-hour conference. We also provided weight-based MDI dosing cards. Chart reviews before and after our interventions were done to assess (1) whether MDIs were used during acute inpatient management of asthma, (2) if the proper dose was ordered, and (3) MDI prescription at time of discharge.

Summary of Results: After interventions were made, physician inpatient use of HFAIs increased from 45% to 66% (p=0.001). Correct inpatient dosing of HFAIs improved dramatically from 4% to 51% (p=0.0001). Discharge to home with HFA improved slightly, from 74% to 79% (p=0.4415).

Conclusions: We have demonstrated a clear change in provider practices after peer-to-peer education on the efficacy of MDIs in the inpatient setting. Future research from our group will focus on parental perceptions of the relative efficacy of MDI versus nebulized albuterol treatment, how these perceptions may be influenced by inpatient hospitalizations, and how these parental perceptions impact medical adherence.

DECREASED LUNG FUNCTION IN PATIENTS WITH CYSTIC FIBROSIS RELATED DIABETES

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Purpose of Study: Pulmonary insufficiency is a major cause of death in patients with cystic fibrosis (CF). Several factors such as pancreatic status, chronic pseudomonas infection and low body mass index have been found to be associated with declining lung function. Herein, we studied the correlation of CF related diabetes (CFRD) to bronchiectasis and average forced expiratory volume in 1 second (FEV1).

Methods Used: A retrospective multicenter study was conducted including patients with CF who were followed at Nemours Delaware and Pensacola from March 2004-2014. Patients with CFRD were sought and investigated for the presence or absence of bronchiectasis. Their average FEV1 was also studied. The data was then analyzed using Chi-square and Mann-Whitney U test.

Summary of Results: A total of 194 patients were analyzed of whom 28 (14.4%) were diagnosed with diabetes. In patients with CFRD, 20 of 28 (71.4%) had bronchiectasis whereas 54 of 166 (32%) of CF patients without diabetes had bronchiectasis (Chi-square p<0.001). Mean FEV1 in patients with CFRD was 74.9% as opposed to 96.5% in CF patients without diabetes (Mann-Whitney U test, p<0.001).

Conclusions: Patients with CF related diabetes have higher risk of bronchiectasis and lower lung function compared to those without diabetes.

COMMUNITY ASSOCIATED STAPHYLOCOCCUS AUREUS SEPSIS IN PREVIOUSLY HEALTHY CHILDREN

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Purpose of Study: We previously reported an increasing incidence of severe staph aureus (SA) sepsis in previously healthy patients with high mortality. Objective of this follow up study is to describe our current practice and outcomes

Methods Used: Retrospective chart review from 2009-2013 of all previously healthy children admitted to the pediatric intensive care unit (PICU) with microbiologic confirmation of SA. Data shown in median and IQR.

Summary of Results: 48 children were included: 34 with Methicillin Resistant SA (MRSA) and with 14 Methicillin Sensitive SA (MSSA). Age at admission was 99(8.145) months. Hospital length of stay (HLOS) was 22(13.35) days and PICU LOS 6.3(4.16) days. There was a non-significant trend in MRSA cases to be younger [54(9.140) v/s 114(90,156) months], have longer PICU LOS [58.16 v/s 5.12 days] and longer HLOS [22(17.35) v/s 149.45 days]. Symptoms preceding admission were respiratory tract infection (33%), joint and bone (JB) infection (29%) and skin infection (20%). Skin infections were more frequent in MSSA group (38% v/s 11%, p=0.02). 88% of patients presented as septic shock, 67% requiring vasocautive drugs (10% >2 agents). Ventilatory support was required in 83%, 36% developed multi-organ failure (MOF). Bacteremia duration was 4(1-7) days (55% >3 days, 36% >5 days). Duration of bacteremia was correlated with PICU LOS (r=0.4) and MOF (r=0.85). MRSA was isolated in 2 sites 38% and ≥ 3 sites in 36%. MSSA was isolated in 2 sites 50% and no isolation in ≥ 3 sites (p<0.01). All patients were empirically treated with Vancomycin. No patient was treated with monotherapy, 3 antibiotics (AB) were used in 35% of cases and 4 AB in 33%. One patient with MRSA sepsis died. 30% had significant morbidity at discharge: 4 patients with tracheostomy, 1 patient on home mechanical ventilation, and 1 patient on dialysis.

Conclusions: Community acquired SA is an important cause of sepsis in previously healthy children. MRSA was usually multifocal and MSSA was more frequently isolated from skin infections. Empiric AB frequency included double coverage. Mortality decreased with respect to our previous report, but morbidity remained high. Future studies are needed to confirm our findings in a larger population to support recommendations for empiric antibiotic treatment.
Conclusions: NaDC1 transports α-KG and is the predominant, but not exclusive, mechanism of control of renal α-KG excretion. Also α-KG is transported via the Ca-sensitive transporter in OK cells. Furthermore based on the FEs > 1.0, substantial amounts of α-KG are secreted into the proximal tubule simultaneously with reabsorption.

Thus NaDC1 in the proximal tubule regulates acid-base transport in the distal tubule via the regulation of α-KG transport. NaDC1 represents a novel mechanism of proximal tubule-distal tubule communication in the kidney.

Summary of Results: The Npr1 gene-disrupted mice displayed the significant reduction of Foxp3+ expression in 0-copy (77.5%) and 1-copy (71.5%) mice compared with 2-copy wild-type mice. Similarly, CD25+ expression was reduced in 0-copy (75%) and in 1-copy (60%) mice compared with wild-type controls. In contrast, the total CD4+ count was comparatively up-regulated by 40% in 0-copy and 31% in 1-copy mice compared with 2-copy control mice. Treatment with rapamycin showed a substantial increase of Foxp3+ cells by 17.38% (P < 0.001) in 0-copy and 8.23% (P < 0.01) in 1-copy mice and CD25+ T cells by 62.2% (P < 0.001) in 0-copy and 38.1% (P < 0.001) in 1-copy mice.

Conclusions: These results demonstrate that phenotyping levels of T regulatory cells in Npr1 gene-disrupted 0-copy and 1-copy mice correspond to wild-type mice. The treatment of 0-copy and 1-copy mice with rapamycin renders elevation of Tregs, suggesting the potential roles of Npr1 in down-regulation of pro-inflammatory immune conditions.

Conclusions: NaDC1 transports α-KG and is the predominant, but not exclusive, mechanism of control of renal α-KG excretion. Also α-KG is transported via the Ca-sensitive transporter in OK cells. Furthermore based on the FEs > 1.0, substantial amounts of α-KG are secreted into the proximal tubule simultaneously with reabsorption.

Thus NaDC1 in the proximal tubule regulates acid-base transport in the distal tubule via the regulation of α-KG transport. NaDC1 represents a novel mechanism of proximal tubule-distal tubule communication in the kidney.

Summary of Results: The Npr1 gene-disrupted mice displayed the significant reduction of Foxp3+ expression in 0-copy (77.5%) and 1-copy (71.5%) mice compared with 2-copy wild-type mice. Similarly, CD25+ expression was reduced in 0-copy (75%) and in 1-copy (60%) mice compared with wild-type controls. In contrast, the total CD4+ count was comparatively up-regulated by 40% in 0-copy and 31% in 1-copy mice compared with 2-copy control mice. Treatment with rapamycin showed a substantial increase of Foxp3+ cells by 17.38% (P < 0.001) in 0-copy and 8.23% (P < 0.01) in 1-copy mice and CD25+ T cells by 62.2% (P < 0.001) in 0-copy and 38.1% (P < 0.001) in 1-copy mice.

Conclusions: These results demonstrate that phenotyping levels of T regulatory cells in Npr1 gene-disrupted 0-copy and 1-copy mice correspond to wild-type mice. The treatment of 0-copy and 1-copy mice with rapamycin renders elevation of Tregs, suggesting the potential roles of Npr1 in down-regulation of pro-inflammatory immune conditions.

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Summary of Results: CaSRs activate PKC through Gq, but also inhibit adenylate cyclase (AC) through Gi. To mimic Gi, cells were pre-incubated with an AC inhibitor, MDL-12,330A (50µM for 30min) followed by Suc uptake. The addition of MDL inhibited uptake in both Ni (0.09±0.01 to 0.05±0.01, p=0.01) and low Ca (0.13±0.01 to 0.08±0.01, p=0.01). In CUBS, Suc uptake was also inhibited in Ni (0.6±0.02 to 0.3±0.03, p=0.01) and low Ca (0.5±0.04 to 0.2±0.03, p=0.01). These results are very similar to our previous studies examining CaSR activation by spermine. If MDL inhibits transport via AC inhibition, 8-BrcAMP (8-Br) should inhibit MDL activity. In OK cells, MDL eliminated the Ca-sensitivity; i.e. Ni (0.05±0.004) and low Ca (0.05±0.002). In low Ca, treatment with 8-Br reversed the inhibitory effects of MDL. Compared to MDL controls, in low Ca, 8-Br significantly increased uptake (0.049±0.002 to 0.063±0.002, p<0.05).

Conclusions: Thus stimulation of the CaSR, PKC and AC pathways through both Gq and Gi leads to inhibition of the Ca-sensitive dicarboxylate transport process in OK cells and indicates CaSR involvement in the regulation of Ca-sensitive citrate transport.

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INTRADIALYTIC HYPERTENSION: ROLE OF SYMPATHETIC NERVOUS SYSTEM OVERACTIVATION

Park S, Lin AM, DaCosta D, Park J. Emory University, Atlanta, GA.

Purpose of Study: End-stage renal disease (ESRD) patients with a paradoxical increase in blood pressure (BP) during hemodialysis, defined as intradialytic hypertension (IDH) are at significantly higher risk of cardiovascular events and mortality. The mechanisms underlying IDH are unknown. We hypothesized that ESRD patients prone to IDH have overactivation of SNS activity during orthostatic stress induced by volume removal during hemodialysis. We further hypothesized that SNS overactivation is mediated by oversensitization of the volume-sensitive cardiopulmonary baroreceptors, resulting in an exaggerated static stress induced by volume removal during hemodialysis. We further hypothesized that SNS overactivation is mediated by oversensitization of the volume-sensitive cardiopulmonary baroreceptors, resulting in an exaggerated static stress induced by volume removal during hemodialysis. We further hypothesized that SNS overactivation is mediated by oversensitization of the volume-sensitive cardiopulmonary baroreceptors, resulting in an exaggerated static stress induced by volume removal during hemodialysis.

Methods Used: 14/C Suc uptake were performed in OK cells and human retinal pigmented cells stably transfected with hNaDC1 (CUBS) in Ni and low Ca buffer.

Summary of Results: CaSRs activate PKC through Gq, but also inhibit adenylate cyclase (AC) through Gi. To mimic Gi, cells were pre-incubated with an AC inhibitor, MDL-12,330A (50µM for 30min) followed by Suc uptake. The addition of MDL inhibited uptake in both Ni (0.09±0.01 to 0.05±0.01, p=0.01) and low Ca (0.13±0.01 to 0.08±0.01, p=0.01). In CUBS, Suc uptake was also inhibited in Ni (0.6±0.02 to 0.3±0.03, p=0.01) and low Ca (0.5±0.04 to 0.2±0.03, p=0.01). These results are very similar to our previous studies examining CaSR activation by spermine. If MDL inhibits transport via AC inhibition, 8-BrcAMP (8-Br) should inhibit MDL activity. In OK cells, MDL eliminated the Ca-sensitivity; i.e. Ni (0.05±0.004) and low Ca (0.05±0.002). In low Ca, treatment with 8-Br reversed the inhibitory effects of MDL. Compared to MDL controls, in low Ca, 8-Br significantly increased uptake (0.049±0.002 to 0.063±0.002, p<0.05).

Conclusions: Thus stimulation of the CaSR, PKC and AC pathways through both Gq and Gi leads to inhibition of the Ca-sensitive dicarboxylate transport process in OK cells and indicates CaSR involvement in the regulation of Ca-sensitive citrate transport.

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ADHERENCE TO ANTI-HYPERTENSIVE DRUGS AND 1-YEAR HOSPITALIZATIONS IN A COHORT OF PREVALENT DIALYSIS PATIENTS

OKECHUKWU O1, Trivedi D1, Wang L1, Wan J1, Kovesdy C1,2, Gosmanova E1,1UNIVERSITY OF TENNESSEE, MEMPHIS, TN and 2VAMC, MEMPHIS, TN.

Purpose of Study: Inadequate adherence to anti-hypertensive drugs (AHD) was linked to increased risk of hospitalizations in the general hypertensive population. However, little is known about effects of adherence to AHD in hemodialysis (HD) patients. We investigated the association between adherence to AHD and 1-year hospitalizations in a cohort of prevalent HD patients.

Methods Used: This was a prospective cohort study involving 58 patients with ESRD and hypertension (mean age 55 years, 96.5% African-Americans, 59.3% males) undergoing HD for at least 12 months. Adherence to AHD was determined at baseline using pharmacy dispensation records and calculating proportion of days covered (PDC) in the previous 2 months. Adherence to AHD was considered as good (PDC>80%) or poor (PDC<80%). The association between adherence to AHD and subsequent 1-year hospitalizations was assessed using univariate and multivariate logistic regression models after adjusting for age, race, gender, and the number of AHD.

Summary of Results: At baseline, only 17.2% of patients (N=10) had good adherence to AHD. The average pre-dialysis mean(SD) SBP and DBP were 157.9(19.4) and 85.3(13.2) mmHg and 154.6(16.6) and 85.7(9.7) mmHg in patients with good and poor adherence, respectively. No correlation was observed between levels of SBP and DBP and the levels of adherence (p=NS). During 1-year follow up period, 41 patients (71%) had at least one hospitalization for any cause, including 23 patients (40%) who were hospitalized for cardiovascular (CV) causes. In univariate analysis, good adherence to AHD was associated with 89% lower risk of overall hospitalizations (OR 0.11, 95%CI 0.03-0.32, p=0.05); however, no difference was observed in CV hospitalizations (OR 0.13, 95% CI 0.02-1.12) between patients with good and poor adherence. Similarly, in the adjusted analysis, patients with good adherence to AHD remained at lower risk for overall hospitalizations (OR 0.16, 95% CI 0.03-0.80, p=0.02) but not for CV hospitalizations (OR 0.17, 95%CI 0.02-1.37) as compared with patients with poor adherence.

Conclusions: Good adherence to AHD was associated with reduced risk of hospitalizations in prevalent HD patients. Therefore, it is important to monitor and promote adherence to AHD in order to improve outcomes in this population.
HYPERTENSIVE CRISIS: THE CAUSATIVE EFFECTS OF NON-STERoidal ANTI-INFLAMMATory DRUGS (NSAIDS)

Gonzales H, Sander GE. Tulane University Medical Center, New Orleans, LA.

Case Report: A 62 year-old woman with no past medical history presented with headache and was found to have hypertensive emergency with a blood pressure (BP) of 225/110 mmHg. She had been taking ibuprofen 800 mg four times daily for the past several weeks for radicular pain. Physical exam showed flushing and mild non-pitting edema of the digits. Ophthalmologic and cardiac exams were normal. Initial work-up showed normal renal function with proteinuria on urinalysis, mild hypokalemia, and a normal EKG and CT head. The patient was started on a diuretic, angiotensin receptor blocker, and calcium channel blocker and advised to discontinue ibuprofen. Over the next few weeks, home BPs decreased to <120/80 mmHg, her symptoms and proteinuria resolved, and BP medications were tapered to single-drug therapy with good control.

Discussion: Often thought of as benign medications, NSAIDs are among the most frequently used. However, they have been shown to have serious side effects including hypertension (HTN), renal failure, gastrointestinal bleeding, bronchospasm, and severe cardiovascular complications such as myocardial infarction, stroke, and congestive heart failure. This case illustrates the effect of NSAIDs on BP, an often overlooked etiology of secondary HTN. The mechanism of HTN from NSAIDs involves alterations in prostaglandin metabolism, with the net effect of sodium and water retention and relative vasoconstriction. Patients who are particularly vulnerable to these effects include the elderly and those with chronic kidney disease and diabetes. Further, it has been established that NSAIDs may blunt the effects of many anti-HTN drugs leading to resistant HTN. Even modest increases in BP within the normal range, known as the “iceberg effect,” may still increase a patient’s cardiovascular risk and mortality.

Conclusions: It is critically important to recognize the ability of NSAIDs to cause severe and resistant HTN. When patients are taking NSAIDs, BPs should be monitored and patients informed of the cardiovascular risks when elevated BPs arise. If elevated BPs occur, discontinuation of NSAIDs is a reasonable first-line approach.

QUALITY IMPROVEMENT PROCESS AND FELLOW EDUCATION IMPROVES REFERRAL RATES FOR TRANSPLANT AND DIALYSIS ACCESS

regmi A1,2, Nahman S1,2, Desai J1,2, Saith S1,2, Afzal U1,2, Iwuagwu N1,2, Ugwu E1,2, White J1,2, GRU, Augusta, GA and ’Charlie Norwood VA, Augusta, GA.

Purpose of Study: Important function of a CKD clinic are timely referrals for renal transplantation/HD access surgery.

Methods Used: As part of quality assurance initiative in our CKD Clinic at the Charlie Norwood VA we evaluated all patients seen by nephrology fellows from Jan -Apr 2013- Phase 1 and assessed referral patterns for renal transplantation and dialysis on CKD 4/5. At conclusion of Phase I results were reviewed with fellows/faculty: need for early referral emphasized; plans for second audit defined. Then referral patterns for patients seen from Aug- Nov 2013 - Phase 2. Lastly, referral rates for eGFR < 20 ml/min was reassessed & reasons for non-referral noted.

Summary of Results: After intervention, increase in referral access rate was seen, but limited effect on transplant referral. Reasons for non-referral in 24 patients:advanced age-5,acute malignancy-4,patient refusal-3,unstable mental health-1,unknown-1.

Conclusions: Rates of referral for transplantation and dialysis access may improve by chart audits. The reasons for non-referral may be due to complex medical problems but patient refusal is also an issue. Addressing non referral issues may help increase transplant and vascular access referrals in advanced CKD.

BACTEREMIA AND MORTALITY IN KIDNEY TRANSPLANT PATIENTS RETURNING TO DIALYSIS

Huber L1,2, Petit J1, Kheda M1, Colombo R1, Baer S1,2, Kintziger K1, Nahman S1,2,2 Georgia Regents University, Augusta, GA and 'Charlie Norwood VAMC, Augusta, GA.

Purpose of Study: Kidney transplantation is the preferred mode of therapy for end stage renal disease (ESRD) and compared to dialysis is associated with improved survival. However, transplant failure necessitates a return to dialysis after graft loss (DAGL). DAGL patients may be immunocompromised, suggesting a greater risk for infection, including bacteremia. To investigate this question, we assessed the effect of bacteremia on mortality in DAGL patients from the United States Renal Data System (USRDS).

Methods Used: All incident ESRD and transplants from the USRDS for calendar years 2005-2008 were queried and DAGL patients identified. From the DAGL cohort mortality and associated risk factors were assessed. ICD-9 diagnosis codes were used to identify diagnoses. Cox regression models were used to determine the relative hazard of death associated with DAGL. We used bivariate and multivariable models to assess these associations. Using backwards elimination of non-significant variables, a final model was obtained, and the hazard ratios (HR) and 95% confidence intervals (CI) determined.

Summary of Results: There were 443,890 incident dialysis patients and 24,839 transplants. After patients with incomplete datasets and death with a functioning transplant were excluded, 1049 DAGL patients remained for analysis. Demographics were: median age 47 years, 64% white, and 57% male. By the end of the study period (12/31/08) 109 patients had died. Significant mortality occurred in those with (vs those without) stroke (31% vs 10%), bacteremia (29% vs 7%), age > 47 (14% vs 7%), MI (24% vs 10%), CHF (21% vs 8%), diabetes (15% vs 8%) and hypertension (13% vs 3%). In an adjusted model, the HR and CI for death were significant for bacteremia (HR 3.12 CI 1.2, 4.61), CHF (HR 2.24 CI 1.50, 3.34) and age > 47 (HR 1.76 CI 1.15, 2.68).

Conclusions: Bacteremia carries an over 3 fold risk of death in DAGL patients. We would speculate that chronic immunosuppression may play a role in this observation. If so, these data suggest that intensified efforts at prevention of bacteremia may be of benefit to DAGL patients.
Summary of Results:

A retrospective analysis was done from 2009-2011 in patients (P) who had angioplasty or another intervention at the same target lesion. Five Hundred records were reviewed, 93 P. developed stent restenosis.

Conclusions: The incidence of stent restenosis was 1.8% of the total population analyzed. No difference was seen in the type of stent used.

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(25.7%) participants had elevated Berlin scores indicating high-risk for OSA. Mean (SD) of cIMT was 0.66 (0.15) mm, and 87 (9.5%) and 161 (17.6%) participants had eccentric and concentric LV hypertrophy, respectively. In log-linear regression models adjusted for age, race, sex, education, current smoking, regular alcohol consumption, total cholesterol, high-density lipoprotein cholesterol, and type-2 diabetes, participants at high-risk of OSA were 1.35 (95% CI: 1.07-1.71) times more likely to be in the highest quartile of cIMT (≥0.74 mm), were 1.68 (1.09-2.58) times as likely to have eccentric hypertrophy, and 1.64 (0.95-2.82) times as likely to have eccentric hypertrophy, compared to those at low-risk of OSA. Similar positive associations were observed for persistent snoring but not for persistent daytime sleepiness. After further adjusting for current obesity status in the models, the association of OSA risk with cIMT remained consistent while the associations with L V hypertrophy was attenuated. There was no significant effect modification by race or sex.

Conclusions: Being at high risk OSA was associated with substantially higher risk of subclinical cardiovascular disease in this biracial, semi-rural, community-based population of young-to-middle age adults.

Allergy, Immunology, and Rheumatology II

Concurrent Session

1:00 PM
Saturday, February 28, 2015

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THE TRANSITIONAL B CELL CHECKPOINT INCOMPLETELY SELECTS CELLS THAT LACKED BONE MARROW PRE BCR EXPRESSION

Khatt M1, Blackburn Y1, Watkins L2, Zhuang Y1, Burrows B1,2, Schroeder H1,2,1 UAB, Birmingham, AL and 2University of Alabama at Birmingham, Birmingham, AL.

Purpose of Study: Expression of the pre-B cell receptor (preBCR), which is created by the association between a nascent Mu heavy chain (HC) and surrogate light chain proteins (SLC), marks an early bone marrow check point during B cell development. Loss of any of the pre BCR components causes a block in B cell development, allowing only a few B cells expressing an abnormal repertoire to survive and enter the peripheral compartments, a state mimicking many autoimmune diseases. In unpublished studies, we have found that preBCR selection adjusts the composition of third complementary region of the H chain (CDR-H3), thereby limiting autoreactivity and optimizing H chain amino acid content. The extent to which subsequent selection steps can be used to adjust the CDR-H3 repertoire in mice lacking the preBCR is unknown.

Methods Used: To test whether CDR-H3 content in mature B cells could be properly regulated in the absence of the preBCR, we sorted mature B cell subsets in the spleen and the peritoneal cavity from SLC-deficient (Ak KO) BALB/c mice and then amplified and cloned their CDR-H3s sequences.

Summary of Results: We found that the splenic T1 transitional checkpoint was able to limit the passage of B cells expressing Mu HCs with CDR-H3s using reading frame (RF)2 that would otherwise have been removed at the pre-B cell stage. However, these Ak KO mice produced IgM dsDNA binding antibodies, suggesting that this T1 selection step was incomplete and failed to control the repertoire.

Conclusions: Our findings suggest that the preBCR selection step plays an essential role in controlling autoimmunity by controlling repertoire content.

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IMPACT OF LONG ACTING ANTIHISTAMINE ON HISTAMINE SKIN PRICK TEST RESPONSIVENESS IN NORMAL HUMANS

Tharpe C, Montgomery D, Sunesara I, Marshall G. University of Mississippi Medical Center, Brandon, MS.

Purpose of Study: In skin prick testing (SPT), a positive histamine control (HC) determines if a patient is able to produce an appropriate wheal/flare response which validates the results of allergy testing. A positive response is defined as a wheal that is 3mm greater than that produced by a negative saline control. HC concentrations of 1mg/ml and 6mg/ml are widely used. There is concern that allergen SPT may have reduced sensitivity when patients have been taking oral antihistamines if the 6mg/ml HC is used which may remain positive despite anti-histamine induced suppression of allergen SPT responses. Thus, the purpose of this project is to determine if a positive response to a 1mg versus a 6mg HC predicts restored skin test reactivity to SPT after administration of fexofenadine.

Methods Used: SPT was done on 21 subjects using 1mg and 6mg HCs, a negative glycercin-saline control, and dust mix, weed, tree and grass mix allergen solutions. All had a history of allergic symptoms and were not on medications that could impair the skin response (e.g. antihistamines). Wheal and flare reactions were measured at 8 minutes after testing controls and at 15 minutes after testing allergens. After initial testing (day 0), subjects were given fexofenadine 180mg and repeat testing was done on day 2, 4, and 6. Linear mixed models were used to test main effects and interactions. A p-value of <0.05 was deemed statistically significant.

Summary of Results: Of the 21 subjects tested, the average wheal size for the 1 mg HC was 4.4mm (95% CI: 4.23 to 4.60) and for the 6 mg HC was 5.7 (95% CI: 5.40 to 6.01). The difference in wheal size between 1 mg and 6 mg was 1.3mm (95% CI: 0.92 to 1.65, p<0.001). The interaction of time and dose were not statistically significant on wheal size (p=0.514), and over time there were no statistically significant changes in wheal size (p=0.336) (Fig. 1).

Conclusions: Despite larger wheal size observed in 6mg versus 1mg HCs, fexofenadine 180mg did not suppress HC positivity or allergen SPT results as soon as 2 days after stopping antihistamine. Furthermore, these results imply that fexofenadine may not need to be held more than two days prior to SPT in the presence of either the 6mg or 1mg HC.

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CONNECTIVE TISSUE DISEASE MAY BE A SIGNIFICANT RISK FACTOR FOR DEVELOPMENT OF NON-UREMIC CALCIPHYLAXIS

Fowler AH1, Maher L1,2, Majithia V1,2, 1University of Mississippi Medical Center, Jackson, MS and 2“G.E.Sonny Montgomery VA Medical Center, Jackson, MS.

Purpose of Study: Calciphalaxis is well reported in the setting of end stage kidney disease. Non-uremic calciphylaxis (NUC) is a rare and poorly understood condition seen in association with multiple underlying diseases. We report 2 new cases and review the previously reported cases of NUC in the setting of CTD.

Methods Used: Two cases of NUC in association with CTD were identified recently at our institution. Both patients were female, had severe rheumatoid arthritis (RA), normal renal function and were treated with corticosteroids and TNF-inhibitors. Development of cutaneous nodules and ulcers lead to biopsy where in a histopathologic diagnosis of calciphylaxis was made. A systematic review of literature for NUC was performed using PUBMED with the MeSH headings of “non-uremic calciphylaxis, rheumatic disease, connective tissue disease and calciphylaxis”. Cases of calciphylaxis in the setting of chronic kidney disease or ESRD were excluded. The remaining case reports were compiled and reviewed.

Summary of Results: A total of 76 cases of NUC were identified, 16 (21%) of which were associated with underlying CTD, including our two patients. The majority of patients were female with RA and were commonly treated with corticosteroids and other immunosuppressants. Mortality rate was high in this population (56.3%).

Conclusions: CTD was seen in 21% of patients with NUC suggesting a significant association in its pathogenesis. In this setting, NUC is reported most often in women with RA. Other possible associations seem to be the use of corticosteroids and immunosuppressants. The exact role and contribution

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Number of cases (%)</th>
<th>Sex</th>
<th>Incidence of death (%)</th>
<th>Immunosuppressant</th>
</tr>
</thead>
<tbody>
<tr>
<td>RA</td>
<td>8 (50%)</td>
<td>f</td>
<td>1 (50%)</td>
<td>6 DMARD</td>
</tr>
<tr>
<td>SLE</td>
<td>3 (18.7%)</td>
<td>f</td>
<td>2 (66.7%)</td>
<td>3 CS</td>
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<tr>
<td>PMR/GCA</td>
<td>3 (18.7%)</td>
<td>f</td>
<td>2 (66.7%)</td>
<td>3 CS</td>
</tr>
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<td>Crohn’s Disease</td>
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<td>f</td>
<td>1 (100%)</td>
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<tr>
<td>Sarcoidosis</td>
<td>1 (6.3%)</td>
<td>f</td>
<td>1 (unknown outcome)</td>
<td>1 CS</td>
</tr>
<tr>
<td>Totals</td>
<td>16</td>
<td></td>
<td>9 (56.3%)</td>
<td>16 DMARD (33.8%)</td>
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</table>

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Connective tissue disease may be a significant risk factor for development of non-uremic calciphylaxis.
of these factors remains unclear and further studies are needed to fully characterize these findings.

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ATOPY AND ANAPHYLAXIS: AN UNRECOGNIZED RISK
Paccione R, Amin AN, Avant RA, Engel LS, Jain N. LSU Health Sciences Center, New Orleans, LA.
Case Report: INTRODUCTION: Radio-contrast media has been implicated as an uncommon cause of anaphylaxis.
CASE: A 49-year-old woman with a past medical history of peanut allergy described as shortness of breath, coronary artery disease, asthma, hypertension, and cocaine abuse presented with one day of chest pain. Vital signs on admission: Temperature 99.6 °F (37.6 °C), heart rate 100 beats per minute, respiratory rate of 18, BP: 160/100 mmHg, SpO2 98% on room air. Pertinent physical exam findings included an obese body habitus with distinct heart sounds, difficult to assess JVP due to body habitus, expiratory wheezes throughout and, 2+ pitting edema in bilateral lower extremities. EKG showed sinus tachycardia with non-specific ST/T wave changes. Pertinent labs included troponin’ s 0.03, 0.47, and 3.54 ng/ml. She was diagnosed as a NSTE MI and subsequently a diagnostic coronary angiography was performed. Months later she became acutely short of breath and hypoxic. She was noted to have injected sclera at this time. She went into pulse-less electrical activity and despite resuscitation efforts; she died.
Post mortem autopsy examination revealed laryngeal edema and tracheal narrowing secondary to laryngeal spasm. Tryptase was 166 μg/L. Due to the acuity of her presentation and elevated tryptase level her cause of death was deemed anaphylaxis due to IV radio-contrast media.

DISCUSSION: Severe systemic reactions occur in 1:1000 exposures with death in 1:10,000-40,000 exposures. The cause of the anaphylactoid reaction is not the iodine in the IV contrast but the hypertonicity. Newer contrast media with low osmolality are much safer. In this case, Ultravist 370 was used which is considered a low osmolality media. Though the rate of adverse events in low osmolality media are reduced, they are not absent. There are no diagnostic tests to predict an adverse reaction to IV contrast. Patients with a previous reaction have a 17-35% chance of recurrence on re-exposure. Although our patient had a peanut allergy, no clear linkage exists between previous reaction have a 17-35% chance of recurrence on re-exposure. Al- though our patient had a peanut allergy, no clear linkage exists between previous reaction have a 17-35% chance of recurrence on re-exposure. Although our patient had a peanut allergy, no clear linkage exists between previous reaction have a 17-35% chance of recurrence on re-exposure. Although our patient had a peanut allergy, no clear linkage exists between previous reaction have a 17-35% chance of recurrence on re-exposure. Although our patient had a peanut allergy, no clear linkage exists between previous reaction have a 17-35% chance of recurrence on re-exposure.

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A DRUG INDUCED REVERSIBLE ENCEPHALOPATHY
DAS D, Naik H, Jaishankar D, CHAKRABORTY K. East Tennessee State University, Johnson City, TN.
Case Report: Headache is a common clinical problem. Its generality can lead to missed opportunity to find an acute etiology promptly. A correlation of clinical history and radiological imaging can help to establish a right diagnosis. Here we present a case of recurrent headache as a presenting symptom of an uncommon clinical syndrome. A 71 year old male presented with headache, nausea, vomiting and seizure two days after receiving cyclophosphamide for vasculitis. Past medical history was significant for recent diagnosis of microscopic polyangiitis, renal insufficiency, anemia and remote lung resection for bronchiectasis. Labs were remarkable for BUN of 34, creatinine of 2.16, HGB 11.0 and normal electrolytes. Examination revealed elevated blood pressure, tachycardia, fine bi-basal crackles but no focal neuro-deficit. A magnetic resonance imaging (MRI) showed increased signal intensity in the subcortical white matter and cortex of parietal lobes and frontal lobes. A follow-up MRI in five days showed complete resolution of this abnormality. MRI changes were felt to be due to vasculitis. Approximately five weeks later patient re-developed similar event after receiving next dose of cyclophosphamide. A MRI brain re- demonstrated increased signal within parietal lobes. A repeat MRI brain obtained in four weeks was unremarkable supporting a diagnosis of posterior reversible encephalopathy syndrome (PRES). Taking clinical presentation into account. With change of treatment to Rituximab patient did well without recurrence of similar symptoms. PRES, first described in 1996 can be of from use of immunosuppressive therapy, vasculitis, uncontrolled hypertension, eclampsia, hemolytic uremic syndrome and connective tissue diseases. PRES also may mimic other disorders such as infarction, demyelinating disorders, encephalitis/ meningitis and cerebral vasculitis. Pathogenesis of PRES is linked to reversible subcortical vasogenic edema in the parietal and occipital lobes. PRES is both a clinical and radiographic diagnosis. Clinical presentations include headaches, nausea/vomiting, seizures and visual disturbances. MRI shows edema in the parietal and occipital lobes. It is important for internist to be aware of PRES in the context of above detailed etiologies for causing recurring headache.

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LOW DOSE METHOTREXATE INDUCED TOXICITY IN A PATIENT WITH GRANULOMATOSIS WITH POLYANGIITIS
Chiang E, Majithia V. University of Mississippi Medical Center, Jackson, MS.
Case Report: Methotrexate (MTX) is an anti-folate cytotoxic medication commonly used in cancer and rheumatic disease conditions. The side effects of MTX is often proportional to the cumulative dose and timing of administration. Potential adverse events are pulmonary toxicity, myelosuppression, gastrointestinal toxicity, and mucositis.

A 64-year-old woman with history of granulomatosis with polyangiitis, treated with MTX, presented with facial swelling, oral mucositis, hemoptysis, and respiratory failure. Laboratory data showed leukopenia, anemia, negative ANA, negative C/P-ANCA, and serum MTX < 0.02 μmol/L, RF 23 IU/mL, ESR 124 mm/hr, CRP 31.6 mg/dl. Chest radiograph showed bilateral infiltrates and CT chest revealed possible diffuse alveolar hemorrhage (DAH) and superimposed infection. Subsequent bronchoscopy with biopsy was negative for DAH and infectious work-up. A presumptive diagnosis of MTX induced toxicity was made. IV antibiotics, high dose solumedrol, and leucovorin rescue were started. Conditions improved 24 hours after therapy; bilateral infiltrates, dyspnea, and mucositis quickly resolved. Patient was discharged in stable condition with continuation of high dose of prednisone. MTX related toxicity is significant, life threatening and commonly presents with pulmonary or liver toxicity, and hematologic suppression. The practitioners for toxicity are diagnosis and consequences. The choices for MTX administration and decrease creatinine clearance (ARF=CKD). Obtaining serum MTX level is often not helpful due to low doses used for rheumatic

ACQUIRED ACRODERMATITIS ENTEROPATHICA AND HYPOTHYROIDISM IN A SEVERELY ATOPIC INFANT
Shah NN, Paris K. LSU New Orleans Health Sciences Center, New Orleans, LA.
Case Report: It is important to evaluate the nutritional deficiencies in severely atopic infants in order to treat possible non-atopic causes of persistent dermatitis. We describe a case of acquired acrodermatitis enteropathica and endocrinopathy in an atopic child.
A full term 11 month old male with history of failure to thrive, GERD, and atopic dermatitis presented to our hospital with fever, dehydration, edema, and dermatitis. At 6 months of age, the patient’s mother elected to feed with rice milk, exclusively due to positive serum specific IgE testing to various foods. Our initial laboratory evaluation revealed anemia, lymphopenia, hypoalbuminemia, and transaminisits. His growth parameters were <3rd percentile, and he had anasarca. Skin exam revealed denuded, scaly, well demarcated erythematous plaques over the diaper area and erythematous crust lesions around the mouth and eyes. The rash appeared 1 month prior to admission, and was not responsive to topical steroids. Skin biopsy was consistent with acrodermatitis enteropathica. Inpatient evaluation of his malnutrition further revealed markedly elevated thyroid stimulating hormone and low serum zinc. An elemental diet and levothyroxine were started, and the patient’s symptoms improved. Appropriate re-evaluation of his serum MTX level is often not helpful due to low- doses used for rheumatic
diseases and a short serum half-life. The formation of MTX-PG is a critical step and polyglutamate concentrations are of clinical relevance in the treatment of RA. However, the association of MTX-PG concentration to toxicity has not been established due to large variation with the median times to reach steady state in red blood cells.

MTX is a commonly used maintenance therapy in small vessel vasculitis and is relatively safe. However, in rare cases may be associated significant toxicity and pulmonary toxicity that can mimic DAH. Patients should be closely followed with history and physical and laboratory monitoring following accepted guidelines. Quick recognition and treatment can lead to rapid and complete response.

Patients on leflunomide can experience weight loss and diarrhea, but colitis is rare. We present a case of a rheumatoid arthritis patient on eight year leflunomide therapy who developed all three. A 65 year old white male with rheumatoid arthritis (RA), chronic kidney disease, and a 10-month history of intermittent diarrhea and 35 lb weight loss was admitted to the hospital with worsening diarrhea. Labwork revealed anemia, acidosis, and worsening renal function. Leflunomide had been held three weeks before due to gradually increasing creatinine and he was placed on prednisone for RA. At admission, he had a 2-week history of several watery stools daily, without blood or mucus. On physical exam, he had weakness and chronic synovitis, but otherwise was stable. The initial differential diagnosis included medication-induced diarrhea, inflammatory bowel disease, infection, malignancy, and endocarditis. Stool studies showed no infectious source. A transthoracic echocardiogram without vegetations, and is relatively safe. However, in rare cases may be associated significant toxicity and pulmonary toxicity that can mimic DAH. Patients should be closely followed with history and physical and laboratory monitoring following accepted guidelines. Quick recognition and treatment can lead to rapid and complete response.

A third of patients on leflunomide can develop diarrhea and weight loss, most often in the first two weeks of treatment. His inflammatory colitis could be due to a reaction to leflunomide or a co-existing colitis. Case reports have shown ulcerative, hemorrhagic, and lymphocytic colitis thought to be induced by leflunomide. Also complicating matters was a three week history of diffuse myalgia and weakness, lack of fever, and normal white blood count did not support endocarditis. A computerized tomography scan showed gastric wall thickening and no colonic abnormalities. Colonoscopy revealed no strictures, ulcerations, masses, or bleeding, but showed patchy erythema in the ascending, transverse, and descending colon. Biopsies throughout the colon showed chronic inflammation with focal acute inflammation and crypt abscesses. Leflunomide had been held indefinitely, and the patient was discharged on prednisone monotherapy for his RA with improvement of his symptoms and weight gain.

A NEW AUTOANTIBODY FOR THE EXPLANATION OF PERSISTENT MYOPATHY FOLLOWING DISCONTINUATION OF IMG-COA REDUCTASE

Hudson CT, Andrew RB, Enged LS. LSU Health Sciences Center, New Orleans, LA.

Case Report: An 81-year-old African American woman with a past medical history of chronic kidney disease stage V and hyperlipidemia presented with two weeks of nausea and fatigue and one week of diffuse myalgia and weakness which progressed until she was bed bound. On exam, she was found to have diffuse decreased muscle tone and proximal muscle weakness with normal reflexes and sensation. Initial lab work showed Cr elevated above baseline, AST of 1484 units/l, ALT of 593 units/l, and a CK level that was unreportable by the lab with the maximum value being 40,000 units/ml. Having suspected statin induced rhabdomyolysis at the time of admission, the patient’s rosuvastatin was discontinued and her AKI was treated with fluids and hemodialysis. When the patient failed to improve as expected and given the rapid progression of her weakness, there was concern for an autoantibody mediated myopathy. Additional lab work showed an aldolase of 225 u/l, a myoglobin of greater than 5000 ng/ml, a CRP of 158, and an ESR of 75. Negative rheumatologic workup included anti-SSA, anti-SSB, anti-smith, anti-RNP, anti-histone, anti-jo-1, C3, C4, and ANA. In addition, a serum sample was sent for the anti-200/100 autoantibody and she was started on high dose prednisone. Within a day of initiating treatment, the patient’s weakness improved on exam, and her CK began to trend down. She was discharged soon after her AKI resolved and was noted to be markedly improved at her clinic follow-up appointment. She was found to be untreated/100 Bible.

DISCUSSION: The family of autoimmune myopathies is characterized by a lack of disease specific findings on muscle biopsy as well as the clinical features of proximal muscle weakness following exposure to statin therapy that does not resolve with removal of the statin. Additionally, patients may have autoantibodies against myositis-specific antibodies, for example, anti-signal recognition particle. Proper therapy requires differentiation of immune-mediated myopathies from other etiologies because only the former will likely respond to immunosuppression.

Clinical Epidemiology and Preventive Medicine
Concurrent Session
1:00 PM
Saturday, February 28, 2015

ASSOCIATIONS BETWEEN RESISTANT HYPERTENSION AND MENTAL AND PHYSICAL QUALITY OF LIFE IN OLDER ADULTS

Ahia C1,2, Joyce C1, Holt EW2, Frohlich ED3, Re RN2, Irving M4, Munner P1, Krouth-Wood M2,3,1 University of Queensland, Brisbane, QLD Australia; 2Ochsner Clinic Foundation, New Orleans, LA; 3Tulane University, New Orleans, LA and 4University of Alabama at Birmingham, Birmingham, AL.
**Purpose of Study:** Few studies have characterized the association between resistant hypertension, a known cause of higher cardiovascular event rates, and mental and physical quality of life in older adults.

**Methods Used:** We included participants from the Cohort Study of Medication Adherence (CSMA) with established hypertension, an antihypertensive medication fill in the 90 days prior to blood pressure (BP) taken before enrollment and a medication possession ratio >0.8. Resistant hypertension was defined as systolic BP $\geq 140$ mmHg or diastolic BP $\geq 90$ mmHg with use of three classes of antihypertensive drugs including a diuretic, or treatment with $\geq 4$ antihypertensive drugs including a diuretic, irrespective of BP values. Participants without resistant hypertension were defined as having controlled BP (systolic BP $<140$ mmHg and diastolic BP $<90$ mmHg) and filling less than 3 classes antihypertensive drugs. Low quality of life was defined by the RAND Medical Outcomes Study 36-item tool using the lowest tertile for physical and mental component scales assessed at baseline.

**Summary of Results:** In this analysis (n=1108), 49.4% of participants were 75 years or older and 27.3% had resistant hypertension. The prevalence of low quality of life in participants with versus without resistant hypertension was 30.9% and 30.6% (p=0.91), respectively, for mental quality of life, and 43.2% and 29.3% (p<0.0001) for physical quality of life. After adjustment for age, race, education, body mass index, hypertension duration, and comorbidity, those with resistant hypertension had a higher odds of low physical quality of life (OR = 1.59, 95% CI 1.19-2.13, p<0.01) compared to those without resistant hypertension. There was no association between resistant hypertension and low mental quality of life (OR = 0.94, 95% CI 0.70-1.27).

**Conclusions:** Older adults with resistant hypertension had worse physical quality of life compared to those without resistant hypertension. Further research is needed to explore if interventions can improve physical quality of life in older adults with resistant hypertension.

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**AUTOMATIC TEXT MESSAGE BASED VACCINATION REMINDERS TO IMPROVE COMPLIANCE**

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1University of Kentucky College of Medicine, Lexington, KY; 2Immunization Chhattisgarh, Trust, Bangalore, India and 3The Johns Hopkins University, Baltimore, MD.

**Purpose of Study:** National Immunization Survey 2013 (MMWR 08/2014) revealed that only 70% children in the United States completed the recommended primary vaccination series by 35 months of age. This series should ideally be completed by 15 months to protect children from several vaccine preventable diseases. Routine immunizations are often delayed or missed, and are influenced by caregivers being unaware of the vaccine dosing times or their due dates. Immunization reminders have been shown to improve compliance, but studies suggest that traditional reminders have a low impact.

Currently about 92% Americans own a cell phone. This makes text messaging based reminder system an effective tool to inform caregivers about upcoming immunizations.

**Methods Used:** We have developed a web based platform where patients in pilot clinics are enrolled. This platform automatically identifies their upcoming vaccines based on the CDC guidelines and sends reminders via a text message to their caregivers. We started recruiting caregivers of newborn children born at one study site for this prospective randomized control study starting August 1, 2014. Our main outcome measure is compliance and timeliness of receipt of immunizations among the intervention group (who receive reminders) compared to the control group (who don’t receive reminders).

**Summary of Results:** The study is ongoing and outcome data is pending collection at the time of submission of this abstract. In the first 2 months of recruitment, of the 236 children born at the site, 65% met the inclusion criteria (caregivers understood English and have a cell phone with a text messaging). 76% of these were interested in the study and consented for randomization. We plan to recruit at least 1000 patients over the next few months and compare the on-time immunization rates between the intervention and the control group. We will be calculating relative risk and odds ratio and will report their 95% confidence intervals.

**Conclusions:** We predict that this method will improve compliance and timeliness of receipt of vaccines among children. We hope that in long term, clinics across the state and eventually the country will employ this system to improve immunization coverage.

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**DECREASED MORTALITY RISK ASSOCIATED WITH FRACTURE IN THE ESRD POPULATION**

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1Medical College of Georgia, Georgia Regents University, Augusta, GA and 2Charlie Norwood Veterans Administration Medical Center, Augusta, GA.

**Purpose of Study:** There is increased mortality risk associated with hip fractures compared to not fracturing among end-stage renal disease (ESRD) patients. This study compared the five-year mortality risk for hip fractures to non-hip fractures, recurrent hip fractures, and multiple skeletal site fractures. We hypothesized that recurrent hip fractures, recurrent non-hip fractures, and multiple skeletal site fractures would have a greater mortality risk than those who had a non-recurrent hip or other type of fracture.

**Methods Used:** The sample included 26,775 ESRD patients who experienced any type of fracture from 1997 to 2008 in the United States Renal Data System (USRDS). The International Classification of Diseases, 9th revision (ICD-9) codes 805-808 and 811-829 were used to identify types of fractures. A recurrent fracture was defined as having the same ICD-9 code after six months. Multiple skeletal site fractures were defined as having a different ICD-9 code at any time. The Cox Proportional Hazards model included ESRD patients who have had any type of fracture, while controlling for demographic (age, sex, race), comorbidities (cardiovascular diseases and multiple skeletal site fractures). A sub-analysis was conducted for those who have had at least one hip fracture.

**Summary of Results:** Among the ESRD patients in the USRDS who experienced any type of fracture, 39% (n=10,475) had at least one hip fracture. The hazard ratio (HR) for recurrent hip fractures was 0.47 (95% Confidence Interval (CI) [0.40, 0.56]) compared to those who had one hip fracture. Those who had multiple skeletal site fractures had a lower mortality risk (HR=0.57, 95% CI [0.50, 0.64]) than those who had one hip fracture. Patients who had any type of recurrent fracture (HR=0.41, 95% CI [0.36, 0.46]) or multiple skeletal site fractures (HR=0.72, 95% CI [0.69, 0.75]) had a lower risk for mortality compared to those who had a non-recurrent fracture.

**Conclusions:** Our data suggest a protective effect against five-year mortality risk for having had recurrent and multiple skeletal site fractures among ESRD patients, compared to having had a non-recurrent fracture. This effect is interpreted to be due to close monitoring and treatment of ESRD patients who have fractured.

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**A MEDIATION MODEL LINKING ADIPOSITY AND CHILDHOOD FOREARM BONE STRENGTH VIA INSULIN RESISTANCE**

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**Purpose of Study:** Given that type 2 diabetes is increasingly being recognized as a risk factor for skeletal fracture combined with recent evidence that 1 in 4 adolescents is on the fast track to type 2 diabetes, it is vital to understand the effects of insulin resistance (a key pathology in type 2 diabetes) on bone strength at the forearm, the most common fracture site in children.

**Methods Used:** Objective: This study investigated the relationship between adiposity and forearm bone strength in 320 children aged 9-13 years, and whether insulin resistance is a mediator of this relationship.

Design: Homeostasis assessment of insulin resistance (HOMA-IR) was calculated from fasting blood samples of glucose and insulin. Bone strength at the radial diaphysis was determined from volumetric bone mineral density and bone geometry parameters, as measured by peripheral quantitative computed tomography, to calculate strength-strain index (SSI), an estimate of torsional bone strength. Fat mass and fat-free soft tissue (FFST) mass were measured by dual-energy X-ray absorptiometry. Associations between fat mass and SSI and the mediating role of HOMA-IR were analyzed with multiple linear regression models and Sobel’s test.
Summary of Results: Results: Controlling for sex, race, pubertal stage and FFST mass, fat mass was inversely associated with radial bone SSI (P<0.01) and positively associated with HOMA-IR (P<0.01). The effect of fat mass on radial bone SSI was no longer significant (P=0.16) after adjusting for the effect of HOMA-IR, which remained significant (P<0.01). Mediation analyses demonstrated that 32% of the association between fat mass and radial bone SSI is mediated by HOMA-IR.

Conclusions: Our data suggest that elevated adiposity may induce insulin resistance with the resulting hyperinsulinemia leading to suboptimal bone strength at the radius, the most common fracture site in children.

MEASURING HEALTH CARE UTILIZATION IN LOW INCOME COMMUNITIES

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Purpose of Study: Adequate measures of health care utilization must assess both the quantity and the quality of health care services. In this study we use data from the Labor Market Health Care Survey (LMHCS) - a longitudinal study of labor history, health status, and health care utilization, conducted between 2006 and 2014- to assess these two dimensions of health care utilization among low income adults.

Methods Used: The LMHCS enrolls participants from households randomly selected from high poverty census tracts. Households are selected using a two-stage cluster probability sample. Data was collected using a questionnaire administered in person. Survey participants were asked if they had a regular doctor or health care provider and if they knew their doctor's name. They were also asked if they had received test for high blood pressure, high blood sugar, and high cholesterol in the current year or the previous year.

Summary of Results: Participants in the LMHCS (n=148) consist of 57% females and 43% males. Approximately 61% of respondents reported that they had a regular doctor or health care provider, but only 43% reported they knew their doctor's name. When asked about medical tests in the current or previous year, 56% reported having their blood pressure measured, 47% reported an examination of blood sugar levels, and 43% reported a test for high cholesterol. Only 34% reported having all three examined. Our data indicate that having a regular health care provider correlates with having blood pressure, blood sugar, and cholesterol examined, where at least one of the three tests were performed in the past year. In addition, for participants who have a regular health care provider and know the names of their physicians, our data shows that they were more likely to have blood sugar and cholesterol tests than those who did not know the names of their physicians.

Conclusions: In summary, we report that having a regular health care provider and knowing the names of individual physicians significantly effect the likelihood of having blood pressure, blood sugar, and cholesterol examined. The role of having a regular provider and knowing the names of individual physicians is increasingly important, particularly in low-income communities where hypertension, diabetes, and coronary artery disease are prevalent.

MAXIMIZING HEALTHY FOOD PURCHASES AMONG FOOD ASSISTANCE PROGRAM BENEFICIARIES

Cohen A 1, Hanks RS 3, Fruh S 2, Arrieta MI 1. 1University of South Alabama, Mobile, AL and 2University of South Alabama, Mobile, AL.

Purpose of Study: Programs such as the Supplemental Nutrition Assistance Program (SNAP) increase low-income families’ access to food. The ways beneficiaries use these programs, however, have been linked to costly health problems such as obesity and metabolic syndrome. Researchers have identified education in budgeting and shopping as a means to enhance low-income families’ abilities to make healthy food purchases. We proposed to design an educational tool promoting healthy and frugal food purchases.

Methods Used: We: 1) searched for education or training programs available to beneficiaries in Mobile, AL, 2) conducted focus groups with Community Health Advocates - community members seeking to improve the health status of local residents- to tailor our curriculum to local practices, and 3) interviewed 15 beneficiaries in the community. During the interviews, we assessed local beneficiaries’ diets, shopping behaviors, perceptions of health, barriers to making healthy food purchases, and willingness to participate in an educational program. We noted the amount of money beneficiaries spend on food in addition to government subsidies.

Summary of Results: Based on the information gathered we selected Cooking Matters at the Store - an initiative designed to optimize low-income families’ food shopping behaviors - as a foundational model, and designed a curriculum to help beneficiaries make healthy food purchases and minimize additional spending. The curriculum includes a short piece of literature and a four- to six-week course that includes grocery store tours and engaging activities such as cooking demonstrations, taste tests, and presentations. During the next phase of this study, we will recruit 10-15 beneficiaries to participate in the course and evaluate its impact on food security, perceptions of health, and budget allocation, resulting in more efficient utilization of food assistance programs like SNAP.

Conclusions: Government food education programs are scarce, and nonprofit organizations fail to serve adequately the 90,000 beneficiaries in Mobile County. A program demonstrating actual savings derived from frugal shopping practices can enhance beneficiaries’ abilities to maintain healthy diets.

GAPS IN MEDICAID COVERAGE AND PROVIDER ASSIGNMENT IN PEDIATRIC PATIENTS ASSIGNED TO A RESIDENT CLINIC

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Purpose of Study: Studies have increasingly shown the benefits of provider care continuity, which is often a byproduct of continuous medical insurance coverage. This study evaluated the gaps in insurance coverage for patients <18 years of age who qualify for Oklahoma Medicaid and were assigned at least once during the calendar year to our academic pediatric clinics.

Methods Used: Data was compiled using patient eligibility and provider assignment files from the Oklahoma Healthcare Authority. Data was examined for patients <18 years of age at the start of eligibility coverage in 2012, who were not born during 2012 and were not new enrollees. Patients also had to be assigned at least once to The University of Oklahoma pediatric academic clinics from January through December of 2012. We then analyzed the continuity of coverage in terms of number of days covered for the year and the continuity in clinic assignment when the patients were eligible. Analysis was performed using SAS v9.3.

Summary of Results: A total of 11,169 eligible patients were identified. Of those 6,179 patients (55.3%) had enrollment eligibility covering the entire calendar year. In fact, 5,955 (53.3%) of all patients had a single enrollment period covering the entire year. These leaves 4,990 patients (44.7%) with one or more eligibility periods with gaps in coverage. Of those patients with full year coverage, only 1,050 (17%) had a single provider assignment period covering the entire year.

Conclusions: This study demonstrates that approximately 53% of patients on Medicaid assigned to a resident clinic have consistent medical insurance coverage for an entire calendar year. Despite having consistent coverage, only 17% had a single consistent clinic assignment during that time period. Future plans include evaluating duration of and reasons for coverage gaps and changes in provider assignment.

NEXT-GENERATION SEQUENCING REVEALS NOVEL GENETIC VARIANTS IMPLICATED IN MULTIPLE EPIPHYSEAL DYSPLASIA AND PRIMARY OSTEOARTHRITIS

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Purpose of Study: Multiple epiphyseal dysplasia (MED) is a genetically inherited chondrodysplasia that is characterized by delayed and irregular ossification of the epiphyses leading to early-onset osteoarthritis (OA). To date, single mutations within a total of 6 genes have been found responsible for 50% of MED cases; however, the genetic defects that are
responsible for the remaining 50% of MED cases are unknown. Furthermore, very little is known about the genetic basis for primary osteoarthritis.

Methods Used: To identify novel candidate genes implicated in MED/ OA, we performed whole-exome sequencing via the Ion Proton next-generation sequencer (Ion Torrent/Life Technologies) among direct relatives within families that displayed autosomal dominant forms of MED/ OA, excluding families that carried mutations in one of the six previously identified genes. Analysis of exome-sequencing was performed with VariantCaller (Ion Torrent Suite Software) and Ion Reporter ver. 4.2 software, which utilizes algorithms including PolyPhen, SIFT, and Grantham scales to predict effects of the sequence changes. Additionally, in a separate group of patients undergoing elective total knee replacement surgery for primary OA, we analyzed candidate genes identified by the exome analysis.

Summary of Results: Exome-sequencing and analysis revealed novel genetic variants that may potentially play a causative role in MED and primary OA. We analyzed co-segregation of these specific gene changes with the MED/OA phenotype in the MED afflicted families as well as in a separate patient group undergoing elective knee replacement surgery for primary OA.

Conclusions: Overall, our studies identify novel genetic variants associated with the MED/OA phenotype and provide new insight towards understanding the genetic basis of primary osteoarthritis.

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SEAT BELTS FOR TEEN DRIVERS: WHY THEY DON'T USE THEM
Monroe K1, Nichols E2, Nichols M3, King WD1. 1University of Alabama, Birmingham, AL and 2Auburn University, Auburn, AL.

Purpose of Study: Teen driving is a high priority public health concern, especially for Alabama which ranks fourth in teen driving deaths. Alabama has had a primary seat belt law since 1999. The purpose of our study was to look at seat belt usage among teens, to understand what factors contribute to their use, and to determine reasons for nonuse.

Methods Used: A teen driving survey was distributed to Jefferson County High School students between the ages of 15 and 18 years old. The survey included questions related to seatbelt use from the National Youth Risk Behavior Survey and supplemental detail questions using the same format. This survey was voluntary and anonymous and approved by the Institutional Review Board.

Summary of Results: 1,014 students returned the survey (540 females, 464 males, 10 unknown). 48% of students (486 of 1014) reported they always wear their seatbelt while the other 52% of students reported not always wearing their seatbelt (35 never, 58 rarely, 119 sometimes, and 314 most of the time). No significant difference was found in seat belt use regarding gender, signing of driving contracts, or crash history. Students whose parents wore seat belts had 3.2 times the odds of wearing a seat belt compared to students whose parents did not use seat belts (OR= 3.2, 95% CI (2.4, 4.2)). White students had 1.6 times the odds of wearing their seat belt compared to non-whites (OR=1.6, 95%CI (1.2, 2.2)). Students who had Driver's Education had 1.7 times the odds of wearing their seat belt compared to those who did not have Driver's Education (OR=1.7, 95%CI (1.3, 2.2)). Top reasons for not wearing seatbelts included: forgetting (401 students), no desire (178 students), and having signed a contract with parents (31% of students who answered yes) reporting having done so > 4 times. 23% students admitted to riding in a car with a driver who had been drinking and 32% of those had done so > 4 times. Having had parents talk about drinking and driving (O.R. 0.7 (CI 0.5,0.9)), and having taken a drivers education class (O.R. 0.6 (CI 0.5,0.9)) had a significant effect on teenage drinking while driving. Gender, race, awareness of the Graduated Driver License Law, and talking to a doctor about drinking and driving had no significant effect. Interestingly, having had a car crash (O.R.1.8 (CI 1.3,2.6)) and having signed a driver's contract (O.R. 2.1 (CI 1.4,3.4)) were both associated with drinking and driving. High rates of non-response to the car crash history and parental contract questions were also documented (31.6% and 51.2%, respectively).

Conclusions: These results suggest that a multi-level approach, which targets the family, may be effective in increasing parental self-efficacy and therefore help to reduce tobacco initiation in the homes of African Americans.

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TEENAGE DRINKING AND DRIVING BEHAVIORS
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Purpose of Study: In Alabama, motor vehicle crashes (MVC) are the leading cause of teenage death. This study was conducted to learn more about the trends in teen drinking behavior as it relates to driver and passenger behavior.

Methods Used: A blinded, voluntary survey based on the Youth Risk Behavior Survey was given to high school students in Jefferson County, Alabama between the ages of 15 and 18 years old. Basic descriptive statistics, odds ratios and their 95% Confidence Intervals were provided using True Epistat version 5.0, Gustafson TL, Epistat Services.

Summary of Results: A total of 1,023 students completed the survey. Of those, 584 (56.4%) students reported having drank alcohol in the last 30 days with 31% reporting they had drunk > 4 times. Driving and drinking was a prevalent risk behavior for teens. In a car with someone who has been drinking was more prevalent for the subjects than driving and drinking. Discussion with parents and drivers education classes may have a preventive influence on teen drinking and driving. Having a history of a car crash and having signed a contract with parents was associated with teen drinking and driving, the survey did not specify which came first (the high risk behavior or the crash/contract). Also, both survey questions had very high no response rates, so we cannot offer valid conclusions about these findings.

Conclusions: The study shows drinking and driving is a prevalent risk behavior for teens. Riding in a car with someone who has been drinking was more prevalent for the subjects than driving and drinking. Discussion with parents and drivers education classes may have a preventive influence on teen drinking and driving. Having a history of a car crash and having signed a contract with parents was associated with teen drinking and driving, the survey did not specify which came first (the high risk behavior or the crash/contract). Also, both survey questions had very high no response rates, so we cannot offer valid conclusions about these findings.

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EXAMINING THE EFFECTIVENESS OF A MULTI-LEVEL INTERVENTION ON PARENTAL SELF-EFFICACY FOR DELIVERING AN ANTI-TOBACCO SOCIALIZATION MESSAGE IN A PREDOMINANTLY AFRICAN AMERICAN POPULATION
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Purpose of Study: Tobacco use kills 480,000 people every year. Recent studies have shown that higher parental self-efficacy in delivering an anti-tobacco socialization message can reduce teen smoking by 28%. However, self-efficacy in a minority population has not been thoroughly investigated and intervention programs tailored to minorities are lacking. The purpose of this study was to investigate the effect of a new tobacco cessation intervention on parental self-efficacy in a largely African American population.

Methods Used: The study participants included 4th graders (n=329) and at least one parent or guardian (n=344) from both rural and urban settings. The participants were randomly assigned to control or intervention groups. The control group received a traditional health class for children and general health information for parents, including self-help information and contact information for the state toll-free Quit-Line for parent smokers. The intervention group received school and home-based interventions aimed at promoting the development of healthy behaviors, specifically tobacco prevention, as well as parental self-efficacy. Smokers in the intervention group also received nicotine replacement therapy and tailored motivational interviewing via telephone or face-to-face for 8 weeks.

Parental self-efficacy was measured at pre and post intervention, using a 7-point Likert scale.

Summary of Results: It was found that parental self-efficacy scores in the intervention group increased significantly (pre=63.1, post=65.1, p=0.0037), while there was no increase in the control group (pre=65.7, post=65.0, p=0.4399).

Conclusions: These results suggest that a multi-level approach, which targets the family, may be effective in increasing parental self-efficacy and therefore help to reduce tobacco initiation in the homes of African Americans.
THE RATIO OF WEIGHT/HDL-CHOLESTEROL AS AN INDICATOR OF INSULIN RESISTANCE IN THE NATIONAL HEALTH AND NUTRITION EXAMINATION SURVEY 2005-2012

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Purpose of Study: Studies have shown that insulin resistance precedes the onset of type 2 diabetes mellitus (DM). A simple measure to detect insulin resistance may lead to a more widespread recognition of hyperinsulinemia in non-diabetic disorders such as Alzheimer’s disease. We have previously shown that the ratio of weight to HDL-cholesterol (WT/HDL) correlates positively with insulin requirement in men following cardiac pulmonary bypass surgery, suggesting that an increase in WT/HDL may indicate patients with insulin resistance. In this study we examine the utility of WT/HDL as simple indicator of insulin resistance in a large ambulatory population.

Methods Used: Data from NHANES 2005-2012 were analyzed for the risk factors of the metabolic syndrome defined by the 2005 AHA/NHLBI updated ATP III criteria. We designated metabolic syndrome as surrogate of insulin resistance and the outcome variable for this study. We performed the multivariable logistic regression analysis after excluding participants with age below 20, pregnancy, and prediabetes or DM (fasting serum glucose <100 mg/dL, A1c <5.7% and on no treatment) (n=11,696). We also compared WT/HDL to HOMEostat model assessment (HOMA-IR) and A1c with ROC curves.

Summary of Results: WT/HDL mean values are shown in the table. WT/HDL is strongly associated with metabolic syndrome: adjusting for age, gender, race, and education, OR 5.46, 95% CI 4.98-5.98, p<0.001. Comparing the Area Under the ROC Curve of WT/HDL vs HOMA-IR and A1c shows that the ratio is the best indicator of metabolic syndrome, 0.85 vs 0.73 and 0.59, respectively, all p value <0.001.

Conclusions: The simple ratio WT/HDL is a better indicator of metabolic syndrome, indicative of insulin resistance, than HOMA-IR and A1c and can identify individuals at risk of cardiovascular disease or other disorders with insulin resistance before they demonstrate prediabetes or DM.

IDEAL BODY WEIGHT IS MORE ACCURATE THAN TOTAL BODY WEIGHT FOR ADEQUATE LEVOthyroxine REPLACEMENT IN OBSESE PATIENTS

Glynph K, Gosmanov A. UT Health Science Center, Memphis, TN

Purpose of Study: Recommendations for levothyroxine (LT4) replacement in patients with hypothyroidism are based on evidence from studies in non-obese subjects. The aim of our study was to compare LT4 regimens required to achieve euthyroidism between obese and non-obese hypothyroid patients.

Methods Used: We retrospectively identified female patients in endocrinology clinic who had total thyroidectomy for benign goiter or stage I thyroid cancer and who achieved euthyroid state on LT4 therapy after surgery. Obesity was defined as body mass index (BMI) of ≥30 kg/m2. Clinical characteristics and thyroid-stimulating hormone (TSH) values were analyzed using Student’s t-tests and Chi-square tests.

Summary of Results: We identified 29 hypothyroid females (14 non-obese/15 obese, 16 whites/13 blacks, average age 48±17 years and final TSH 1.1±0.8mIU/L). There was no difference between the groups in age and TSH, but obese patients weighed more (104.5±21.5 vs 68.6±10.6kg, P<0.001) and had higher BMI (38.2±8.5 vs 25.8±2.8, P<0.001). Compared with non-obese, obese women required higher LT4 dose (149±38.5 vs 102±12.8mcg, P<0.001). There was no difference between the groups in LT4 per total body weight (TGW) (1.45±0.37 vs 1.52±0.26 mcg/kg, P=0.59), however, LT4 per ideal body weight (IBW) was higher in obese than in non-obese females (2.63±0.71 vs 1.89±0.29 mcg/kg, P<0.001). The LT4 dosing patterns were similar in obese and non-obese patients independent of menopausal status. We found marked variability in LT4 dose in obese patients. When we categorized obese patients who became euthyroid on LT4 dose of 100-150mcg (n=10) vs 150-200mcg (n=5), age, weight, and BMI were not different between the subgroups. The LT4 dose per TBW (1.27±0.21 vs 1.80±0.4, P=0.004) and IBW (2.23±0.48 vs 3.44±0.18, P<0.001) differed significantly in obese females who achieved euthyroidism on LT4 dose ≤150mcg vs obese taking >150mcg, respectively.

Conclusions: Compared with non-obese, obese females after total thyroidectomy require higher LT4 dose per IBW but not per TBW. Among obese women, there was significant heterogeneity in final LT4 dosage not explained by known clinical features. Using IBW as opposed to TBW could be a more accurate initial approach for successful therapy of hypothyroidism in a majority of obese females.

ASSOCIATIONS OF SOCIOECONOMIC STATUS WITH MARKERS OF MINERAL METABOLISM IN COMMUNITY-DWELLING ADULTS

Lysa CN, Panwar B, Judd S, Jenny N, McClellan W, Rizk D, Gutierrez O. 1University of Alabama, Birmingham, Birmingham, AL; 2Emory University, Atlanta, GA and 3University of Vermont, Burlington, VT

Purpose of Study: To determine in a multi-center study, if implementation of a multidisciplinary intensive diabetes program (MIDP) at one site will reduce the proportion of poorly controlled DM when compared to two other comparison sites. The variables measured included age, sex, insurance and A1c levels. Patients with A1c over 10% were classified as poorly compliant and were enrolled in an intensive program including DM education, nutrition consultation, social service and psychological interventions, weekly phone calls to monitor glucose levels and insulin doses and monthly clinic visits. Longitudinal comparisons of the percentage of poorly controlled patients were made using Chi-square analysis.

Summary of Results: In 2004 at baseline, there was statistically higher percentage of poorly controlled DM at the MIDP site compared to the other two comparison sites. (MIDP site: 27.82%, n=74; site two: 19.68%, n=195, site three:14.23%, n=101; p<0.001). There was no difference between the three centers for 2012 at end of study period after starting MIDP.

Medicaid population analysis: In 2004 at baseline, there was statistically higher percentage of poorly controlled DM in MIDP site compared to the other two comparison sites. (MIDP site:45.12%, n=37; site two: (25.4%, n=72); site three: 18.2%, n=37; p<0.001). In 2012, at the end of the study, there were no differences among the sites.

MIDP site analysis (Medicaid): Comparing baseline 2004, with 2012 after starting MIDP, there was a statistically significant decrease in the percentage of diabetes who were poorly controlled. (2004: 45.1%, n=37; 2012: 29.08%, n=57) (Chi-square p < 0.001).

Conclusions: This study provides evidence that MIDP resulted in a reversal of the higher percentage of poorly controlled DM patients found at baseline for the MIDP site compared to two other sites over 9 years. Additionally, a greater impact was noted for Medicaid patients who had dramatic reductions in proportion of patients in poor control.
Purpose of Study: Disorders of mineral metabolism are associated with increased risk of cardiovascular and kidney disease. Prior studies showed that lower socioeconomic status (SES) was associated with disturbances in phosphorus homeostasis, but the association of SES with other markers of mineral metabolism has been studied in less detail.

Methods Used: Serum phosphorus, calcium, intact parathyroid hormone (PTH), 25-hydroxyvitamin D (25(OH)D) and fibroblast growth factor 23 (FGF23) were measured in a random sample of 1,040 participants of the Reasons for Geographic and Racial Differences in Stroke (REGARDS) study, a national cohort of 30,239 community-dwelling adults. Associations of SES markers (annual family income, county level poverty, Diez-Roux neighborhood SES score) with markers of mineral metabolism were examined in multivariable regression models.

Summary of Results: There were no associations of individual income or county level poverty with markers of mineral metabolism in analyses adjusted for age, race, and sex. Greater neighborhood SES scores (indicating lower neighborhood disadvantage) were associated with lower concentrations of PTH (P<0.001), FGF23 (P<0.001), and higher concentrations of 25(OH)D (P<0.001) in unadjusted models. After adjustment for age, race, and sex, greater neighborhood SES score remained associated with lower FGF23 (P<0.001), but not PTH or 25(OH)D. The inverse association between neighborhood SES score and FGF23 remained statistically significant (P=0.03) after further adjustment for indexes of kidney function, body mass index, diabetes, smoking, annual income and educational achievement.

Conclusions: Greater neighborhood SES score was independently associated with lower FGF23 concentrations among black and white community-dwelling adults. Future studies should determine environmental factors that may explain these findings.

SEX DIFFERENCE IN ENDOTHELIAL FUNCTION AMONG ADOLESCENTS
Verma A, Singh R, Sheehan C, Vasilyeva T. Texas Tech University Health Sciences Center, Amarillo, TX.

Purpose of Study: Endothelial function is a marker of cardiovascular health. Endothelial dysfunction sets in childhood, long before clinical manifestations of atherosclerosis. Reactive hyperemia index (RHI), a non invasive measure of endothelial function has linear correlation with the Framingham index, diabetes, smoking, annual income and educational achievement.

Methods Used: Two major markers of endothelial function- RHI and ET-1 were measured in a random sample of 1,040 participants of the Reasons for Geographic and Racial Differences in Stroke (REGARDS) study, a national cohort of 30,239 community-dwelling adults. Associations of SES markers (annual family income, county level poverty, Diez-Roux neighborhood SES score) with markers of mineral metabolism were examined in multivariable regression models.

Summary of Results: Greater neighborhood SES score was independently associated with lower FGF23 concentrations among black and white community-dwelling adults. Future studies should determine environmental factors that may explain these findings.

CALCIUM PERTURBATIONS AND BONE TURNOVER IN PATIENTS WITH ACROMEGALY: A PROSPECTIVE CONTROLLED STUDY
Constantin T 1, Tangpricha V 1,2, Shah R 1, Ioachimescu O 2, Oyesiku N 1,2, Emory University, Atlanta, GA.

Purpose of Study: Some patients with acromegaly (ACM) have hypercalcemia, usually due to primary hyperparathyroidism or less likely, 1,25(OH)2D excess. ACM patients are at risk of vertebral fractures. We present a prospective study on calcium homeostasis and bone markers in ACM.

Methods Used: ACM group included 22 patients who underwent surgical or medical therapy. Control group included 22 nonfunctioning pituitary adenomas (NFPA). Among ACM, 17 had improved/normalized IGF-1 at end of follow-up; this subgroup was used for analyses. Gender, serum prolactin and eGFR were similar between groups, but ACM group was younger (43±13 vs 54±9). Serum IGF-1, GH, phosphorus, CTX, and PINP were higher and PTH was lower in ACM vs NFPA group (p<0.01). Serum calcium, 25(OH)D and 1,25(OH)2D were similar between groups. In ACM, serum calcium correlated with IGF-1 (R2=0.41, p=0.01) and PTH inversely correlated with IGF-1 (R2=0.49, p<0.05). Bone markers did not correlate with age. After ACM treatment, serum and urine calcium, phosphorus and CTX decreased, PTH increased (p<0.01) and 25-OH D, 1,25(OH)2D or PINP did not change.

THE EFFICACY OF HIGH PROTEIN VS HIGH CARBOHYDRATE DIET IN REVERSAL OF IMPAIRED GLUCOSE TOLERANCE TO NORMAL GLUCOSE TOLERANCE IN OBSESE ADULTS
Garber KC, Kitabchi AE, Stentz F. University of Tennessee Health Science Center College of Medicine, Memphis, TN.

Purpose of Study: The purpose of this study is to identify diet modifications that facilitate weight loss and conversion of patients with impaired glucose tolerance (IGT) to normal glucose tolerance (NGT). Specific aims are to determine if a High Protein (HP) or High Carbohydrate (HC) diet is more effective in conversion of IGT obese adults to NGT; and to identify if an HP or HC diet contributes to DNA methylation changes that can be linked to conversion to IGT from NGT.

Methods Used: 16 obese, pre-diabetic adults were randomized to a HP or HC diet for 6 months with all food provided. The HP diet consisted of 30% protein, 30% fat, 40% carbohydrates while the HC diet consisted of 15% protein, 30% fat, 55% carbohydrates. The diets were distributed by percentage of daily kcal derived for each subject. An Oral Glucose Tolerance Test (OGTT) was performed at 0, 3, & 6 months to determine IGT/NGT status. DNA was extracted from OGTT 0 minute blood samples of the 0 and 6 month visits and DNA methylation was determined. A DEXA Scan was taken at 0 and 6 months to measure body composition. Weight was measured weekly.

Summary of Results: The HP diet had a 100% conversion rate to NGT while the HC diet had a 37.5% conversion rate. A HP diet was more effective in conversion of IGT obese adults to NGT; and to identify if an HP or HC diet contributes to DNA methylation changes that can be linked to conversion to IGT from NGT.
**Conclusions:** Uncontrolled ACM is associated with calcium alterations that are PTH-independent and correlate with disease activity. Both bone resorption and formation increase in uncontrolled ACM. Six months post-treatment, only bone resorption decreases. Further research is needed to identify factors involved in increased bone resorption and calcium perturbations in ACM.

<table>
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<th>Serum calcium (mg/dL)</th>
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<th>Post-treatment</th>
<th>p value</th>
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<td>9.4±0.4</td>
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**Fractional excretion of calcium**

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<th>Serum Phos (mg/dL)</th>
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<th>p value</th>
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<td>4.3±0.7</td>
<td>3.9±0.8</td>
<td>0.002</td>
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</tbody>
</table>

**PHT** (mmHg)

| 36.3±19            | 48.9±16.7 | 0.01 |

(p= 0.01) with two comparisons; control vs. AAb+ and control vs. DM1. Comparison between the 500 most significant genes of each analysis showed that only 62 genes (12.5%) were the same. Data mining of those lists of genes showed the following enrichments in both: 1) canonical pathways and KEGG pathways of mitochondrial dysfunction and oxidative phosphorylation; 2) gene ontologies associated with oxidative/reductive and mitochondrial functions. Top candidates for upstream regulators were also somewhat similar for AAb+: RICTOR, IFN-β-1a; for DM1: RICTOR, PDX1. Promoter and de novo pathway analyses of the two lists also gave similar results. In the AAb+ group: cellular growth/apoptosis (Elk-1), Autoimmunity (E2F-1), Insulin regulation (Sp-1). In the DM1 group: cellular growth/apoptosis (Evi-1/ETF) and Autoimmunity (E2F).

**Conclusions:** Expression profiles of islets from AAb+ and DM1 donors both differed significantly from those of normal controls. However, the genes that separated those two groups from controls were dissimilar. When we analyzed what pathways were represented in those gene lists we found larger degree of overlap. Both non-diabetic AAb+ and DM1 donor islets had clear indications of changes in genes associated with oxidative phosphorylation and mitochondrial function. As beta cells are highly dependent on ATP production as a mechanism of insulin secretion, the loss of genes essential for mitochondrial function may explain the dysfunction of these cells during development of DM1.

**IMPACT OF GENETIC VARIANTS IN THE ENDOCANNABINOID SYSTEM IN AFRICAN-AMERICANS**

Katalenich B1, Thethi TK1, Sigel A2, Japa S1, Tasker J1, Liu S1, Nguyen T1, Larrazolo J1, Carpio G1, Lovre D1, Hwn W1, Stell CA1, Conroy K2, McDuffie R1, Fonseca V1, Tulane University, New Orleans, LA; Nevada State College, Henderson, NV and Tulane University, New Orleans, LA.

**Purpose of Study:** Genetic variants of endocannabinoid receptor Type 1 gene (CNR1) and fatty acid amide hydrolase (FAAH) polymorphisms play a prominent role in clinical expression of obesity and the metabolic syndrome (MS).

**Methods Used:** 314 African-American (AA) participants from a cross-sectional study were analyzed to assess association of mutations in CNR1-3813, CNR1-4895 and FAAH-385 genes with clinical and biochemical measurements of obesity.

**Summary of Results:** Odds ratios (OR) and modeling analysis was conducted. Participants with mutation CNR1-3813 were more likely (OR = 1.7, p= 0.0387) to have elevated fasting plasma glucose than those without. Participants with CNR1-4895 mutation were more likely to have hyperlipidemia (OR=2.0, p=0.0152) and diabetes mellitus (DM) (OR=2.1 p=0.0249) than those without. Participants with FAAH-385 mutation were more likely to have elevated triglycerides (OR=1.7 p=0.0363), be obese (OR=1.9 p=0.0186) and defined as having MS (OR=1.6 p=0.0382) than those without. CNR1-3813 mutation was associated with abdominal obesity, hypertension (HTN), triglycerides, MS, and chronic kidney disease (CKD). CNR1-4895 mutation was associated with triglycerides, estimated glomerular filtration rate (eGFR), vitamin D deficiency, HDL, hypertension, glucose and CKD. FAAH-385 mutation was associated with hyperlipidemia, HTN, DM, glucose, triglycerides, eGFR, systolic BP, cardiovascular disease (CVD), MS, and VLDL.

**Conclusions:** Initial results show significant associations between the CNR1-3813, CNR1-4895 and FAAH-385 gene mutations and risk factors for obesity, DM and CVD. Individuals with these mutations may be able to change their risk factors such as obesity to reduce outcomes such as DM.

**IMPROVED DETECTION OF ELEVATED BLOOD PRESSURE IN CHILDREN WITH TYPE 2 DIABETES MELLITUS AFTER IMPLEMENTATION OF ELECTRONIC MEDICAL RECORD**

Prestel C1, Paulo R2, Shatat I3, Bowly D2, Lewis K2, Medical University of South Carolina, Charleston, SC; MUSC, Charleston, SC and MUSC, Charleston, SC.

**Purpose of Study:** Hypertension (HT) during adolescence tracks into adulthood. As cardiovascular disease is the leading cause of death in adults, it is important to diagnose and treat at early stages. The diagnosis of elevated blood pressure (BP) in children is weight, height and sex specific which requires provider to check both growth charts and BP tables to identify elevated measurements. Frequently, in a busy clinical setting elevated BP goes unrecognized. In this study, we examine the impact of electronic medical record (EMR) smartphrase that calculates height, age and sex specific BP percentile. BP% smartphrase was implemented.

**Methods Used:** BP of children with T2DM followed at our outpatient Pediatric Diabetes clinic for 6 months before and 6 months after implementing the BP % smartphrase were analyzed. Elevated BP in the hypertensive range was defined as ≥95% systolic/diastolic. Chi square was used to analyze difference in detection rate of elevated BP before vs after implementation.

**Summary of Results:** 118 patients with T2DM were analyzed. Average age was 14.3yo, with 65% females and 56% African-Americans. All were obese (BMI ≥95%). 54% have Medicaid. 7 (6%) have been diagnosed with HT and on antihypertensive medications prior to the start of the study.

Of the 69 patients seen before implementation of the EMR smartphrase, 30 (44%) had BP ≥95% but only 6 (20%) were recognized. After implementation, 68 patients were seen, 24 (35%) had BP ≥95% and significantly more patients [15 (62%)] were recognized (P=0.001). This led to higher frequency of counseling, instruction to monitor BP at PCP’s office, or referral to the hypertension clinic.

**Conclusions:** There is a high prevalence of elevated BP among children with T2DM. Implementing an EMR smartphrase increased the rate of elevated BP detection, and thus improved care. Utilizing features in EMRs that facilitate detection of children at higher risk of cardiovascular morbidities should be encouraged and systematized.

**COMPARISON OF THREE INSULIN TREATMENT REGIMENS ON HEMOGLOBIN A1C IN PEDIATRIC TYPE 1 DIABETES**

Nelson D1, Henpe P2, Vargas A3, Scribner R2, Velasco-Gonzalez C2, Gomez R1, Louisiana State University Health Sciences Center School of Medicine - New Orleans, New Orleans, LA; Louisiana State University Health Sciences Center

**Purpose of Study:** The purpose of our study was to determine differences in islet gene expression profiles between organ donor groups relevant to DM1.

**Methods Used:** Cryo-sections were obtained from pancreata of organ donors from the following 3 groups: DM1 with insulin+ islets (n=21), AAb+ without DM1 (n=12), and age, sex, ethnicity matched controls (n=18). Laser-capture was used to isolate islets (30-40 per donor). RNA was immediately isolated and stored at -80°C. The quantity and integrity of the RNA was determined. The RNA was amplified and transcriptome analysis performed with Affymetrix Human 2.0ST arrays.

**Summary of Results:** The data was filtered by expression value leaving 20,757 probe sets. Statistical analysis was completed using Student’s t-Test (p< 0.01) with two comparisons; control vs. AAb+ and control vs. DM1. Comparison between the 500 most significant genes of each analysis showed that only 62 genes (12.5%) were the same. Data mining of those lists of genes showed the following enrichments in both: 1) canonical pathways and KEGG pathways of mitochondrial dysfunction and oxidative phosphorylation; 2) gene ontologies associated with oxidative/reductive and mitochondrial functions. Top candidates for upstream regulators were also somewhat similar for AAb+: RICTOR, IFN-β-1a; for DM1: RICTOR, PDX1. Promoter and de novo pathway analyses of the two lists also gave similar results. In the AAb+ group: cellular growth/apoptosis (Elk-1), Autoimmunity (E2F-1), Insulin regulation (Sp-1). In the DM1 group: cellular growth/apoptosis (Evi-1/ETF) and Autoimmunity (E2F).

**Conclusions:** Expression profiles of islets from AAb+ and DM1 donors both differed significantly from those of normal controls. However, the genes that separated those two groups from controls were dissimilar. When we analyzed what pathways were represented in those gene lists we found larger degree of overlap. Both non-diabetic AAb+ and DM1 donor islets had clear indications of changes in genes associated with oxidative phosphorylation and mitochondrial function. As beta cells are highly dependent on ATP production as a mechanism of insulin secretion, the loss of genes essential for mitochondrial function may explain the dysfunction of these cells during development of DM1.
Purpose of Study: Compare hemoglobin A1c (HbA1c) levels in pediatric type 1 diabetes patients receiving either two daily injections (2DI), multiple daily injections (MDI), or insulin pump therapy when controlling for demographic, race and social determinant variables.

Methods Used: Patients (n=88) at Children's Hospital were recruited with informed consent and demographic information was self-reported. Home addresses were used with US Census Data to assign a concentrated disadvantage index (CDI) to each patient. Patients were stratified into three groups based on their prescribed insulin treatment regimens. Hemoglobin A1c (HbA1c) was used as a metric of metabolic control.

Statistical analyses were performed using SAS 9.3. Population demographics were generated using descriptive statistics, and means between groups were compared using ANCOVA followed by post hoc analysis with Tukey’s procedure. Results were considered statistically significant at p<0.05.

Summary of Results: See Table 1

Conclusions: 1) Pump therapy was associated with significantly lower HbA1c levels compared to 2DI therapy when controlling for gender, age, or BMI; however, this effect was lost when controlled for differences in race or CDI.
2) There was no significant difference in HbA1c between MDI and pump therapy.
3) Race and environmental stress (CDI) may contribute differences in HbA1c observed between insulin treatment groups.

Adjusted Mean HbA1c

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<th>MDI</th>
<th>PUMP</th>
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Health Care Research and Quality Improvement
Concurrent Session
1:00 PM
Saturday, February 28, 2015

RECENT TRENDS IN COMPUTED TOMOGRAPHY (CT) IMAGING IN PEDIATRIC PATIENTS IN EMERGENCY DEPARTMENTS (EDs): WHAT IS THE ROLE OF ALTERNATIVE IMAGING MODALITIES?

Kakarakia K, Johnson LW. Texas Tech University HSC, Lubbock, TX.

Purpose of Study: The role of imaging in pediatric radiation exposure has brought increased awareness to diagnostic testing for patients, and especially to use of CT (computed tomography). There are national efforts, such as Image Gently, encouraging physicians to be more cognizant of radiation dosage, areas scanned, and the necessity of testing. Some children’s hospitals have demonstrated declining CT utilization, with a concomitant increase in alternative imaging modalities such as magnetic resonance imaging (MRI) and ultrasound (US). We sought to determine whether this trend translates to the general pediatric population seeking emergency care in a nationally representative sample of EDs.

Methods Used: Retrospective analysis of data from the 2005-2010 National Hospital Ambulatory Medical Care Survey (NHAMCS), a nationally-representative four-stage probability sample of ED visits. Primary variables of interest were different forms of imaging utilization: CT, Magnetic Resonance Imaging (MRI), and ultrasound (US). We generated descriptive statistics and determined bivariate associations of imaging use with hospital characteristics with chi-square tests. All analyses were completed using SUDAAN.

Summary of Results: There were 48,857 pediatric visits during the study period with a national estimate of 172,998,933 visits. For these visits, annual frequency of CT utilization ranged from 4.7% in 2005 to 6.2% in 2010, (p=0.003). Rates of ultrasound imaging ranged from 0.8%-1.3% of visits and MRI imaging rates ranged from 0.1% to 0.9% of visits with no differences among the years. Ultrasound was performed more often in teaching hospitals than in non-teaching hospitals (1.5% versus 0.9%, p<0.003). CT imaging occurred less often in child-focused emergency department compared with general EDs (3.9% vs 5.7%) and there were no differences in MRI or US imaging utilization.

Conclusions: CT imaging in children does not appear to be declining in this nationally representative sample of ED visits. Furthermore, our analysis did not demonstrate an increase in use of alternative imaging modalities. This suggests an ongoing need for education and implementation of protocols for best imaging practices in imaging in the emergency department.

IS LUMBAR PUNCTURE NECESSARY IN THE EVALUATION OF FEBRILE LOW RISK INFANTS 28 TO 90 DAYS OF AGE?

Hines L1, Muhlinghause M2, Anderson M2, Naifeh M1. 1University of Oklahoma Health Sciences Center, Oklahoma City, OK and 2University of Oklahoma, Oklahoma City, OK.

Purpose of Study: Febrile infant is a common chief complaint in the pediatric population and the assessment of febrile infants 28-90 days of age is controversial. Guidelines for evaluation of febrile infants 28 to 90 days of age vary and include evaluation and treatment based on risk-stratification. The primary aim of this study was to evaluate the impact of lumbar puncture (LP) performance on clinical outcomes in low risk, febrile infants 28 to 90 days of age.

Methods Used: Medical records were reviewed for all children 28-90 days of age presenting to the emergency department between January 1, 2006 and December 31, 2011 with a diagnosis of fever or sepsis. Infants were considered low risk if WBC count was between 5,000 and 15,000, without underlying condition, ear, soft tissue or bone infection and had not received antibiotics prior to presentation.

Summary of Results: 624 infants met inclusion criteria for the study; of those, 126 were high risk and 498 were low risk. We compared low risk infants who received LP (n=277) to those who did not receive LP (n=214) on a number of variables including hospital admission, length of stay and antibiotic use. 56% of low-risk patients received a LP and 53% were admitted to the hospital. None of the 278 low-risk infants who received a LP had a positive CSF culture. Significantly more patients in the high risk group had a positive CSF culture (5 of 86 who received LP). Low risk infants who received LP were more likely to receive antibiotics (LP 94.2% vs no LP 16.59% p<0.0001) and more likely to be admitted to the hospital (LP 70.4% vs no LP 16.82% p<0.0001).

Conclusions: Our data suggest that lumbar puncture among low risk, febrile infants 28 to 90 days of age may lead to unwarranted higher rates of antibiotic use and hospitalization. Our data also suggest that risk stratification appropriately identifies the patients at risk for severe illness and in need of further evaluation and treatment including LP and antibiotics.

HYPONATREMIA CONSEQUENCES FOR HOSPITALIZED PEDIATRIC PATIENTS

Thakore PS, Sartor Z, Letvuchik T, Vasylyeva T. Texas Tech Health Sciences Center, Amarillo, TX.

Purpose of Study: The aim of this study is to characterize the prevalence of hyponatremia and its sequelae, namely length of hospital stay (LOS), in pediatric patients.

Methods Used: Retrospective chart review of pediatric patients who were admitted to Northwest Texas Healthcare System Hospital in Amarillo from October to December 2012 was done after TTUHSC IRB approval. Patients were divided into three diagnosis groups: bronchiolitis, asthma exacerbation, and gastroenteritis. Other diagnoses and patients without serum chemistries were excluded. Age, sex, type and rate of intravenous fluid (IVF) replacement and duration of hospital stay were recorded. Prevalence and odds ratio for hyponatremia (Na < 135) in patients on traditional IVF were calculated. LOS was compared for patients with and without hyponatremia using an unpaired T test.

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**Conclusions:** Documentation of asthma severity was significantly increased. Retrospective chart review was performed on inpatient asthma exacerbations are a leading cause of pediatric hospitalizations. Guidelines from the National Heart, Lung and Blood Institute (NHLBI) on asthma management recommend assessment of disease severity and prescribing of inhaled corticosteroids (ICS) for patients with persistent asthma based upon assessment of five symptoms. This study assessed adherence to these guidelines as reflected in physician documentation admitted to the Asthma Clinical Pathway (ACP) at Children’s of Alabama.

**Methods Used:** Asthma exacerbations are a leading cause of pediatric hospitalizations. The study assessed adherence to asthma severity classification and ICS prescription on admission and discharge for patients admitted to the ACP. ACP eligibility criteria included: patients being ≥2 years old with a primary diagnosis of asthma and absence of co-morbid respiratory disease. Chi square tests and two sided independent t-tests were performed.

**Summary of Results:** Patients (n=230) were 67% male, with an average age of 6.7 years, with no significant differences between groups. The documentation of symptom assessment and the percentages of patients on ICS at admission and prescribed at discharge was not significantly different between the two groups. Asthma severity classification was significantly greater in 2014 despite no change in symptom assessment documentation.

**Conclusions:** Documentation of asthma severity was significantly increased in the 2014 group despite no change in symptom assessment documentation between groups. Symptom assessment documentation was less than half of the five symptoms per the NHLBI guidelines. However, a consistent increase in the percentage of patients on ICS at discharge in comparison to admission is noted in both groups, which reflects awareness of disease severity despite lacking documentation. We postulate that developing a standardized approach to obtaining all of the historical components to address disease severity will improve adherence to guidelines and improve asthma management.

**CAN A BRIEF EDUCATIONAL INTERVENTION IMPROVE THE USE OF SPIROMETRY IN CHILDREN?**

Singh N, Dasari P, Mehdi N, Anderson M, Naifeh M. Oklahoma University School of Community Medicine, Tulsa, OK and Oklahoma University of Health Sciences, OKC, OK.

**Purpose of Study:** Determine if an educational intervention on the appropriate use and interpretation of spirometry increased use of spirometry in the diagnosis and management of asthma in children followed in our resident clinics.

**Methods Used:** An anonymous online survey adapted from Janson et al of faculty and residents was conducted. This was followed by an educational intervention consisting of one formal lecture for the general pediatrics faculty and residents (available online for those unable to attend) and a small group session prior to the start of resident Continuity Clinic. A post-intervention survey was administered. Pre- and post-surveys were not matched. Clinic billing data from our 3 general pediatrics clinics were reviewed nine months before and after the study period. Appropriate statistical methods were used for analyses.

**Summary of Results:** Sixty-seven faculty and residents were consented. A total of 56 responded to the pre-survey (34%) with 29 responding to the post-survey (43%). A majority of respondents (85%) to both surveys correctly identified spirometry as the preferred lung function test but only 16% of pre-survey respondents reported using it. This improved to 59% of post-survey respondents (p<0.0001). Respondents to both surveys had difficulty interpreting spirometry results correctly (38% pre 28% post p=0.33) but correctly identified appropriate treatment (73%, 76%). Respondents were more likely to report feeling comfortable interpreting spirometry on the post-survey (35%, 74%, p<0.001). There was increased use of spirometry in our resident clinics after the intervention. In fact, 95% of all spirometry billed occurred after the intervention (p < 0.0001).

**Conclusions:** A short educational intervention improved the use of spirometry in our pediatric resident clinics. Unfortunately, knowledge was not improved. More sustained training is needed to improve knowledge of spirometry interpretation.

**DOCUMENTATION OF ASThma SEVERITY CLASSIFICATION AND INHALED CORTICOSTEROID PRESCRIBING AT PATIENT DISCHARGE FROM A TERTIARY CHILDREN’S HOSPITAL**

Molina AL, Roitsch C, Magruder T, Narayanan S, Wall T, Walley S. University of Alabama at Birmingham, Birmingham, AL.

**Purpose of Study:** Asthma exacerbations are a leading cause of pediatric hospitalizations. Guidelines from the National Heart, Lung and Blood Institute (NHLBI) on asthma management recommend assessment of disease severity and prescribing of inhaled corticosteroids (ICS) for patients with persistent asthma based upon assessment of five symptoms. This study assessed adherence to these guidelines as reflected in physician documentation admitted to the Asthma Clinical Pathway (ACP) at Children’s of Alabama.

**Methods Used:** Retrospective chart review was performed on inpatient physician documentation during 2 time intervals regarding asthma symptom assessment, severity classification and ICS prescription on admission and discharge for patients admitted to the ACP. ACP eligibility criteria included: patients being ≥2 years old with a primary diagnosis of asthma and absence of co-morbid respiratory disease. Chi square tests and two sided independent t-tests were performed.

**Summary of Results:** Patients (n=230) were 67% male, with an average age of 6.7 years, with no significant differences between groups. The documentation of symptom assessment and the percentages of patients on ICS at admission and prescribed at discharge was not significantly different between the two groups. Asthma severity classification was significantly greater in 2014 despite no change in symptom assessment documentation between groups. Symptom assessment documentation was less than half of the five symptoms per the NHLBI guidelines. However, a consistent increase in the percentage of patients on ICS at discharge in comparison to admission is noted in both groups, which reflects awareness of disease severity despite lacking documentation. We postulate that developing a standardized approach to obtaining all of the historical components to address disease severity will improve adherence to guidelines and improve asthma management.

**IMPROVING THE ELECTRONIC DISCHARGE SUMMARY: A PEDIATRIC QUALITY IMPROVEMENT PROJECT**

Lala S, Nayyar G, Andrews T, Howard-Tran A, Salgado B, Broussard M, Cooper P. Louisiana State University, Shreveport, LA.

**Purpose of Study:** Electronic discharge summaries (EDS) are a form of handoff communication between the inpatient and outpatient settings. To be effective in describing inpatient care, discharge summaries must include critical information, including accurate discharge medications, an updated problem list, and follow-up recommendations. These items help prevent medication errors, and may decrease re-admission rates. There has been scarce data that focuses on creating and implementing a clinically relevant EDS in concordance with JCAHO standards. EDS are becoming more prevalent with time and health care systems continue to increase their focus on transitions of care, making the analysis of the discharge summary an integral part of quality improvement. The objective of this study was to improve our hospital EDS by creating a JCAHO-compliant discharge summary template and assess its impact on quality of care and compliance of EDS on the pediatric ward of our hospital for further improvements, including hospital-wide implementation, in the future.

**Methods Used:** A comprehensive evidence-based discharge summary template was created at an academic medical center-based pediatric training program. A retrospective chart review was conducted of a sample of discharge summaries from before and after implementation to monitor JCAHO compliance, adoption, and outcomes of new discharge summary templates.

**Summary of Results:** Twenty-one charts from before implementation and twenty-five charts from after implementation of a JCAHO-compliant discharge summary template were analyzed evaluating ten variables, including...
usability of the medication list and primary care handoff information, and were scored based on inclusion and accuracy of information. Compared with charts prior to implementation of the improved template, the new EDS had statistically significant scores (P<0.05) on seven of ten tested variables using a two-tailed student's t-test and chi-square analysis.

Conclusions: The implementation of a standardized, JCAHO-compliant discharge template can improve the quality and context of EDS in an academic medical setting and can be applied to other situations to assist with compliance and quality improvement.

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CLINICIAN SATISFACTION BEFORE AND AFTER TRANSITION FROM A BASIC TO A COMPLETE ELECTRONIC HEALTH RECORD
McCoy AB1,2, Milani R 1, Holt EW1, Krousel-Wood M1,2, 1Ochsner Health System, New Orleans, LA and 2 Tulane University, New Orleans, LA.

Purpose of Study: Healthcare organizations are transitioning from basic to comprehensive electronic health records (EHRs) to meet Meaningful Use requirements and improve patient safety. Yet, full adoption of EHRs is lagging and may be linked to clinician dissatisfaction. In depth assessment of satisfaction before, during, and after EHR transition is rarely done.

Methods Used: Using an adapted published tool to assess clinician adoption, satisfaction, and perceptions with EHRs, we surveyed clinicians at a large, non-profit academic medical center before (baseline), and 6-12 months (short term follow up) and 12-24 months (long term follow up) after transition from a basic, locally-developed to a comprehensive, commercial EHR. Satisfaction with the EHR (overall and by component) was captured at each interval. We used McNemar's chi-squared test to compare satisfaction between baseline and follow up assessments.

Summary of Results: We received all three surveys from 47 eligible clinicians (recapture rate=57%). Overall EHR satisfaction was highest at baseline (85%), lowest post short term follow up and declining over time: 71% (short term follow up) and 62% (long term follow up) were statistically significant (p<0.02, 0.03 respectively), but the difference between baseline and long term follow up was not (p=0.41). This trend was similar for satisfaction with EHR components designed to improve patient safety including clinical decision support, patient communication, health information exchange, and system reliability. Conversely, at baseline, short term and long term follow up, perceptions of productivity (45%, 26%, 28%, respectively), ability to provide better care with the EHR (81%, 43%, 49%, respectively), and satisfaction with available resources (62%, 34%, 40%, respectively), were lower at both short and long term follow up compared to baseline (p<0.05).

Conclusions: Overall clinician satisfaction with EHR decreased after the initial transition to a comprehensive EHR but increased over time. Persistent dissatisfaction at short and long term follow up may be driven by lagging determinants of dissatisfaction may increase full adoption of EHRs. Further investigation in larger populations is warranted.

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PUBLIC HEALTH INTERVENTIONS FOR NEONATAL ABstinence SYndrome
Reynolds EW. Norton Healthcare/Kosair Children's Hospital, Louisville, KY.

Purpose of Study: Neonatal Abstinence Syndrome (NAS) is a rapidly growing problem in the US. Recent data shows that nearly $750,000,000 is spent on NAS, 70-80% coming from public money, indicating a true public health problem. The purpose of this study is to identify current state public health responses to NAS.

Methods Used: This project is a 10-question survey completed from Nov 2013-Feb 2014. Contact was made with the Department of Public Health in each state to participate. Respondents were asked if their state had a plan to deal with NAS. Programs were described, funding sources were identified and policies regarding post-hospital care were discussed. Respondents were asked about abusive head trauma and SIDS in former NAS infants.

Summary of Results: 49 states completed the survey (1 abstention). 31 states have a program to deal with NAS. 19 have existed >1 year, 7-1 year, 5 are organizing programs. 3 states with no program plan to start one within a year. Programs include taskforces associated with a Governor's Office, Attorney General's Office, Public Health Department, Legislative Advisory Committee, or State Perinatal Quality Collaborative. 15 programs have an identified funding source which include: Title V Grant money, Budget item in Governor's Office, State AG or Health Department, Medicaid, University or Private Support, Legislative Appropriation or Other Federal money. 49 states have Prescription Drug Monitoring Programs. 5 respondents felt that the treatment needs of substance abuse during pregnancy are met in their states. 21 states have policies regarding hospital discharge of NAS infants (automatic referral to Child Protective Services, Eligibility for Early Intervention Services, etc). 17 states have programs to aid families after discharge, but most of these are not specific to NAS. Most respondents suspect an increase in abusive head trauma and suicide in former NAS infants, but could not provide statistical evidence to support the idea because data systems are not designed to link birth and death certificates in this way.

Conclusions: States are responding to the growing NAS epidemic as a true public health emergency. Most do not have dedicated funding for these initiatives and depend on volunteer stakeholders. Most do not have programs specifically dedicated to NAS. Better data systems are needed to measure mortality statistics of infants affected by NAS.

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PHENOTYPE CORRELATION OF DIFFERENT GENOTYPES IN PATIENTS WITH CYSTIC FIBROSIS
Neemuchwala F1, Vala S1, Chidekel A2, Burns J1, Efidemir O2, 1Florida State University, Pensacola; FL, 2Nemours Children’s Clinic, Pensacola, FL and 3 Nemours Children’s Clinic, Wilmington, DE.

Purpose of Study: Cystic fibrosis (CF) is the most common autosomal recessive disease of Caucasians. During the last several years, newborn screening has facilitated early detection of disease and many new mutations have been identified. One of the most common questions of parents is what the prognosis will be for their child. Hence, the aim of our study was to investigate correlation between phenotypic manifestations of the different genotypes in patients with cystic fibrosis.

Methods Used: A retrospective multicenter study was conducted including patients with CF who were followed at Nemours Delaware and Pensacola from March 2004-2014. Different genotypes were investigated for presence or absence of certain phenotypic features. The genotypes were categorized into three different groups based on the status of delta F508 mutation: no delta F508 mutation, heterozygous for the delta F508 mutation and homozygous for the delta F508 mutation. Clinical manifestations of CF were recorded including bronchiectasis, diabetes, pancreatic insufficiency, sinusitis requiring surgery, meconium ileus, age at first pneumoniason, average body mass index and forced expiratory volume in one second (FEV1). The data was analyzed using Chi-square and ANOVA.

Summary of Results: A total of 198 patients were studied with 91 homozygous for delta F508 mutation, 79 heterozygous for delta F508 and 28 with no delta F508 mutation. Pancreatic insufficiency was found in 100% of patients with the homozygous delta F508 genotype, 75.9% in heterozygous patients and 50% in patients with no delta F508 mutation (Chi-square p<0.001). There was no statistical relationship between genotype status and presence of bronchiectasis, diabetes, sinusitis requiring surgery, meconium ileus, age at first pneumoniason, average body mass index and FEV1.

Conclusions: Our study concludes that pancreatic insufficiency correlates significantly with delta F508 mutation in CF patients whereas other phenotypic manifestations do not. Hence knowledge of genotype will help predict pancreatic status but not lung function for patients with cystic fibrosis.

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GUT MICROBIOTA, PERMEABILITY AND SYSTEMIC INFLAMMATION IN CHRONIC KIDNEY DISEASE (STAGE 4 AND 5) WITH T2DM AND HEALTHY SUBJECTS
Singh R, Weinheimer G, Vasylyeva T. TTUHSC, Amarillo, TX.

Purpose of Study: The aim of the study is the assessment of gut microbiota, plasma zonulin, and inflammatory cytokines (TNF-alpha, IL-6) in conjunction with FGF-23, ET and levels of LPS in CKD (stage 4 and 5) patients with Diabetic Nephropathy (DN).

Gut Microbiota, Permeability and Systemic Inflammation in Chronic Kidney Disease (Stage 4 and 5) with T2DM and Healthy Subjects

Neemuchwala et al.

Summary of Results:

- A total of 198 patients were studied with 91 homozygous for delta F508 mutation, 79 heterozygous for delta F508 and 28 with no delta F508 mutation.
- Pancreatic insufficiency was found in 100% of patients with the homozygous delta F508 genotype, 75.9% in heterozygous patients and 50% in patients with no delta F508 mutation.
- There was no statistical relationship between genotype status and presence of bronchiectasis, diabetes, sinusitis requiring surgery, meconium ileus, age at first pneumoniason, average body mass index and FEV1.

Conclusions:

- Our study concludes that pancreatic insufficiency correlates significantly with delta F508 mutation in CF patients whereas other phenotypic manifestations do not. Hence knowledge of genotype will help predict pancreatic status but not lung function for patients with cystic fibrosis.
Conclusions: Gut microbiota and increased gut permeability in patients with advanced CKD secondary to DN results in high level of circulating LPS that mediates chronic inflammation which was implicated in deterioration of cardiovascular health.

A RETROSPECTIVE REVIEW OFAMYOTROPHIC LATERAL SCLEROSIS PATIENTS AND ADHERENCE TOAMERICAN ACADEMY OF NEUROLOGY’S PRACTICE PARAMETER

Rajendra R 1, Gutierrez A 2, LSUHSC, New Orleans, LA and 2LSUHSC, New Orleans, LA.

Purpose of Study: ALS is a rapidly progressive and fatal motor neuron disease. Although the only disease modifying therapy is Riluzole, there are other measures that should be implemented to improve the quality of life of ALS patients. There are four categories of recommendations for ALS patients including therapeutic options, evaluation and monitoring of disease status and associated symptoms, patient safety, and end of life care. The goal of our study was to perform a retrospective chart review of deceased Amyotrophic Lateral Sclerosis (ALS) patients and our adherence to American Academy of Neurology (AAN) practice measures.

Methods Used: A retrospective chart review was performed on deceased patients from our ALS multidisciplinary clinic from 1999 to 2012. Forty-eight patients were identified and the sample excluded those with incomplete medical records (n=10). Data recorded included: date of diagnosis; use, dosing, and initiation of Riluzole; cognitive/behavioral screening; ALS-symptom screening; respiratory symptoms and the use of noninvasive ventilation; nutrition screening and PEG initiation; hospice intervention; and history of falls.

Summary of Results: Average time between onset of symptoms to diagnosis was approximately 15 months. 92% of patients were prescribed Riluzole. Cognitive/behavioral screening was performed on 13% of patients. 95% of patients were asked about ALS associated symptoms and 89% required pharmacotherapy. All patients were asked about respiratory symptoms and 87% were screened dysphagia symptoms, but none received nutritional screening. 39% of patients had documented falls and 32% were referred for hospice care.

Conclusions: Overall, we adhere to most measures of the AAN practice parameter. Our least documented measures included cognitive/behavioral screening, fall query, nutritional screening, and end of life care. In order to implement these measures a comprehensive template should be available on our electronic health records system so all measures are discussed throughout the disease process in order to improve the quality of life of patients and their caregivers.

Hematology and Oncology II
Concurrent Session

1:00 PM
Saturday, February 28, 2015

PRELIMINARY CLINICAL TRIAL INVESTIGATING THE ABILITY OF PLANT EXOSOMES TO ABRAGE ORAL MUCOSITIS INDUCED BY COMBINED CHEMOTHERAPY AND RADIATION IN HEAD AND NECK CANCER PATIENTS

Redman R 1, Perez C 1, Bumpous J 1, Potts K 1, Dunlap N 1, Silverman C 1, Wu X 2, Rai S 2, Mu J 1, Zhang H 1, Miller DM 1, 2University of Louisville, James Graham Brown Cancer Center, Louisville, KY; 2University of Louisville, Louisville, KY and 3University of Louisville, Louisville, KY.

Purpose of Study: Oral mucositis is one of the most common and severe side effects of radiation and chemotherapy for squamous cell carcinoma of the head and neck, leading to pain, weight loss, and interruptions in treatment. New approaches to lessen the impact of oral mucositis are urgently needed. Plant exosomes have been shown to prevent both acute and chronic inflammation in animal models. This study is designed to test the hypothesis that grape exosomes will reduce the severity of oral mucositis in patients undergoing chemotherapy and radiation for head and neck squamous cell carcinoma.

Methods Used: Subjects receiving concurrent cisplatin and radiation for squamous cell carcinoma of the head and neck were randomized 1:1 to the “grape exosomes” arm or “standard treatment” arm of the study. Exosomes were prepared using Good Laboratory Practice. Subjects in the exosome arm swish and swallow one aliquot of exosomes daily prior to radiation. Severity of mucositis was scored weekly by subjects in both arms using the WHO Oral Mucositis Scale. Primary endpoint is to determine whether grape exosomes reduce the severity of oral mucositis. Secondary endpoints include safety, tolerability, survival, and biomarker analysis.

Summary of Results: A total of 33 subjects are included in this analysis. Arms were well-balanced for disease site (oropharynx vs. non-oropharynx), AJCC stage, and radiation dose. The proportion of subjects in the exosome arm able to maintain a solid diet throughout treatment (≤WHO grade 2) was significantly higher than in the standard treatment arm (29% vs. 0. p=0.0445). There was no significant difference in grade 3 or higher other toxicity between the two arms (p = 0.5536).

Conclusions: Oral administration of grape exosomes may decrease the severity of oral mucositis without increasing other toxicities in patients receiving concurrent chemotherapy and radiation for squamous cell carcinoma of the head and neck.
oropharyngeal cancer from May 2010 - May 2014. Patients who had undergone robotic surgery for an oropharyngeal cancer from May 2010 - May 2014 were included in this study if they received robotic surgery for an oropharyngeal cancer.

**Summary of Results:** The average hospital stay after TORS surgery was 3.8 days with a range of 1-8 days. Most resections (64%) were complete without any tumor left in patient, R0. There was one intraoperative complication (7.1%), one postoperative complication (7.1%), three tracheotomies (21.4%), zero undergoing prolonged intubation, three PEG tube placements (21.4%), and one nasogastric tube placement (7.1%). 92% of patients were classified as ‘alive without disease’ at last follow-up. The time period for follow-ups ranged from 1.7-49.7 months. This is due to the 4 year range of the study (4/2010-4/2014). Disease Free Survival (DFS) rate was calculated taking the date of last follow-up from the date that the patient was classified as ‘disease free’. The overall DFS is 15.4 months with a range of 0.5-42.7 months.

**Conclusions:** TORS offers a sophisticated surgical intervention that allows oncologically sound resections while limiting functional damage and morbidity and hence permitting short hospital stays and faster recovery than conventional surgery. It permits pathological analysis of disease that can help stratify tumors into low and high risk based on true pathologic data rather than clinical and radiological tumor staging, permitting further intensification or deintensification of therapy with additional chemo radiation based on tumor pathology, hence offering the opportunity of individualized or personalized cancer treatment vs a blanket therapy with CRT for all OP Cancer patients. TORS can also be a good alternative for surgical salvage after failed CRT and its feasibility must be considered prior to conventional open surgery.

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**EFFECT OF ANGIOTENSIN CONVERTING ENZYME INHIBITION ON OUTCOMES IN PATIENTS WITH HEAD AND NECK SQUMOUS CELL CARCINOMA**

Linden C, Redman R, Rai S, Rai A. *University of Louisville, Louisville, KY.*

**Purpose of Study:** Studies suggest that inhibition of the renin-angiotensin system (RAS), with angiotensin-converting enzyme inhibitors (ACE-I) or angiotensin receptor blockers (ARB), may be associated with both improved outcomes as well as mitigation of radiation-induced toxicities in cancer patients. This study aims to determine whether treatment with an ACE inhibitor in head and neck cancer patients undergoing radiation with or without concurrent chemotherapy is associated with improved survival or a decrease of treatment-related toxicity, specifically oral mucositis and renal insufficiency.

**Methods Used:** A retrospective chart review of consecutive patients with biopsy-proven squamous cell carcinoma of the head and neck treated with radiation with or without chemotherapy was associated with improved survival or a decrease of treatment-related toxicity, specifically oral mucositis and renal insufficiency.

**Methods Used:** A retrospective chart review of consecutive patients with biopsy-proven squamous cell carcinoma of the head and neck treated with radiation with or without chemotherapy was associated with improved survival or a decrease of treatment-related toxicity, specifically oral mucositis and renal insufficiency.

**Summary of Results:** 19.6% of patients were taking ACE-I during the course of their treatment. Characteristics of the cohorts (+)ACE-I versus (-)ACE-I were well balanced. Improved overall survival was noted in the (+)ACE-I cohort as only 10 patients had expired at time of analysis whereas 18/41 in the (-)ACE-I cohort had expired (logrank p 0.094). The majority of deaths (73.7%) were cancer-related. Three patients died secondary to treatment-related toxicity, all of whom were in the (+) ACE-I cohort.

There was no difference in the overall incidence of oral mucositis between the cohorts. Although only three patients developed grade ≥3 mucositis, none of who were taking ACE-I. There was no difference in the incidence of renal insufficiency or any grade ≥3 toxicity (logrank p 0.49).

**Conclusions:** The overall survival curve suggests that ACE-I may improve cancer-specific survival in patients with head and neck squamous cell carcinoma undergoing radiation with or without concurrent chemotherapy, despite a lack of significant effect on treatment-induced toxicity.
A CT Brain to evaluate a potential stroke reveals a lytic bony lesion over the anterior C1 vertebra and occipital condyle of the calvarium. An infiltrating tumor of the clivus and base of the occipital skull is seen on a MRI cervical spine. Barium swallow indicates severe oropharyngeal dysphagia secondary to lingual dysfunction and cranial nerve IX and X impairment. Prior serum electrophoresis is positive for a 1.7 gram IgG monoclonal spike with 10% plasmacytosis on bone marrow biopsy. A plasmacytoma is suspected, and a biopsy is performed, only to reveal a poorly differentiated adenocarcinoma prostate. Serum PSA is high at 18. Treatment with high dose steroids, radiation and androgen deprivation therapy is initiated.

Bilateral XII nerve palsy is an extremely rare condition. Tumors such as metastatic carcinomas, gliomas and acoustic neuromas account for half the reported cases. Common sites of metastases in prostate cancer are the vertebrae, pelvis and ribs. Unusual areas of metastases include the orbital, intracranial and cervical region. Cranial nerve palsy results from neural/foraminal compression or infiltration and rarely can be the only presenting sign of metastatic disease as in our patient. It is important to be aware of uncommon presentations of common diseases such as prostate cancer to expedite diagnosis and treatment.

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THE EFFECT OF BMI AND GROWTH ON CHILDREN WITH SICKLE CELL DISEASE
Van Buren JW1, Kaulfers A1,3, Wilson F1,2, University of South Alabama, Mobile, AL; 1University of South Alabama, Mobile, AL and 3University of South Alabama, Mobile, AL.

Purpose of Study: The purpose of our study is to look at the children in our area with sickle cell anemia to compare them to standard growth charts, as well as to see if there is a protective effect of low height, weight, and/or BMI in regards to vaso-occlusive crisis in sickle cell disease.

Methods Used: We did a chart review of 147 children with Sickle Cell Disease in our area, aged 2 to 20 years. We reviewed both hospital records and clinic records from 2003 to present. We compared the height, weight, and BMI to CDC growth charts and counted the number of admissions to our hospital that were related to vaso-occlusive crisis only. Children were categorized into three groups: Hemoglobin SS (n=87), Sickle Beta Thalassemia (n=19), and Hemoglobin SC (n=41), then into Male and Female and Children (ages 2-11) and Adolescents (age 12-20). Associations were examined between BMI and hospital admissions within each group and subgroup by means of a Pearson Correlation Coefficient, or r.

Summary of Results: The HB SS and SC group had patients more than 2 SD below the mean height for age, 14% and 5% respectively. Our analysis indicated a mean correlation between BMI and hospital admission in children with sickle beta thalassemia (r=0.3), adolescent females with HB SC (r=0.3) and children with HB SS(r=0.4). Only one group had a high correlation of BMI to hospitalizations; adolescent males with HB SC (r=0.6) had a high negative correlation.

Conclusions: We conclude that a higher percentage of sickle cell patients fall more than 2 SD below their mean height, confirming the perception that children with sickle cell disease have difficulties meeting their growth potential. We found that there is a positive correlation between 3 of the groups: Children with sickle beta thalassemia, adolescent females with HB SC, and children with HB SS, suggesting a possible protective factor of low BMI. In our group of adolescent males with HB SC, we found a negative correlation, suggesting a possible protective factor of higher BMI. Our study looked strictly at a correlation between BMI and hospital admissions and not confounding variables. We believe that the benefits of maintaining a healthy BMI outweigh the possibility of less vaso-occlusive crises in lower weight individuals.

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THE MIGHTY MICROPARTICLE: A NOVEL MECHANISM FOR CANCER RELATED THROMBOSIS
desai P, Rigby P, LSU Health Sciences Center, New Orleans, LA.

Case Report: Patients with cancer have an increased incidence of thrombosis and emboli; however, the mechanism behind this phenomenon is not well understood. Studies have shown that patients with increased levels of microparticles harboring tissue factor had a higher incidence of thrombi. Also, patients with solid tumor malignancies have a higher level of multiple types of microparticles; some harboring tissue factor. Microparticles form from cell membranes of each of the different blood cell lines, including cancer cells. Microparticles circulate in the blood and interact with other cells in the circulatory system. Tissue factor is an important molecule in the clotting cascade leading to thrombus formation, and the increase in microparticles harboring tissue factor could be a cause of the increased incidence of thrombosis in patients with cancer. Our hypothesis is that tissue factor microparticles are upregulated in patients with solid tumor malignancies, and increase the risk for patients developing a deep venous thrombosis.

Our goal is to examine if patients with cancer, who develop deep venous thrombosis, have a higher level of tissue factor microparticles. We will measure tissue factor levels in a small subset of patients with solid malignancies and known deep venous thrombosis to assess a correlation. If an association is found, then this could propose a possible mechanism for the hypercoagulable state seen in cancer, and suggest a possible target for future therapies aimed to decrease the morbidity and mortality related to clot formation in cancer patients.

Tissue Factor-Initiated Coagulation

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HIGH ALTITUDE BONE MARROW CHANGES ON MRI
Master S, Devarakonda S, Burton G, Mills G, Feist Weiller cancer center, Shreveport, LA.

Case Report: Hyperactive bone marrow can mimic marrow involvement by cancer that can be seen occasionally as an incidental finding on MR spine.

Case Report: A 70 y/o Caucasian male with past medical history of degenerative joint disease had complaint of severe low back pain. His physical exam including neurological exam was normal except for antalgic gait due to back pain. MR spine done at outside hospital was read as malignancy involving marrow. Of note, patient was in Colorado (High Altitude) for past 3 months. He returned to Shreveport (Sea level) and was seen at our Cancer Center for further work up. Patient had a detailed work up for malignancy involving marrow and it was unrevealing. A repeat MR spine done at our facility showed resolving changes as below.

A diagnosis of marrow hyperplasia secondary to high altitude was made and confirmed by criteria outlined below.

Discussion: When there is increasing demand for hematopoiesis and it exceeds existing red marrow capacities, then there is repopulation of yellow marrow by hematopoietic cells, a phenomenon referred to as marrow hyperplasia / conversion. Causes of hyperplasia include chronic severe anemia, marrow infiltration by tumor, effects of granulocyte-macrophage colony stimulating factor and increasing oxygen demand of as in marathon runner and high altitude. Criteria helpful to distinguish between red marrow from infiltrating marrow are as follows. Unlike infiltrative process, conversion follows pattern of normal marrow and is more symmetrical. Secondly, signal intensity...
of red marrow is slightly greater that skeletal muscle and infiltration has much higher intensity. Thirdly, reconversion does not cause cortical break down or adjacent soft tissue mass.

### Infectious Diseases II

**Concurrent Session**

1:00 PM Saturday, February 28, 2015

**507 RISK FACTORS FOR AN ABNORMAL ANAL PAP SMEAR IN HIV+ ADULTS IN NEW ORLEANS**

Nanfro J, Frontini M, Hagensee M. LSU, New Orleans, LA.

**Purpose of Study:** The frequency of anal cancer is 5-10 times higher in HIV+ individuals as compared to the general population. Anal cancer prevention in HIV+ individuals starts with anal Pap smear screening. Similarly to cervical pap smears, this screening can help identify high grade lesions that may develop into cancer. New Orleans ranks high among US cities in rates of new HIV and AIDS diagnosis. The HIV Outpatient Program clinic in New Orleans started an anal cancer screening program in 2012. The goal of this study is to identify risk factors for abnormal pap smears in our HIV+ patients.

**Methods Used:** Data was gathered through the EMR of all results of the anal paps taken from 7/2012-5/2014. These were compared to demographic and clinical characteristics including age, race, gender, CD4 cell count, HIV viral load (VL), year of collection, and clinician performing the test. Pap smears were read using the Bethesda criteria developed for cervical Pap smear and categorized as unsatisfactory, normal, ASCUS, LSIL, HSIL, or cancer.

**Summary of Results:** A total of 403 anal pap smears from 164 patients were obtained over the study period. Patients were 54.3% female and 78% African American (AA) with a mean age of 47.2, mean CD4 count of 480.2 cells/ml and median HIV viral load of 171 copies/ml. Of the 403 samples collected, 9.9% were unsatisfactory. Unsatisfactory Paps were more often in females, AA race, and those with a higher CD4 count (all p<0.05). Abnormal pap was more likely in patients who were male, Caucasian and younger. No difference was found in patients based on HIV VL or CD4 count. Dysplastic pap was more common among patients who were male, Caucasian and had CD4 counts less than 500. HIV VL load had no significant correlation.

**Conclusions:** Most pap smears were adequately collected and all providers collected the samples with equal adequacy. Fortunately, those less likely to have an adequately collected pap smear were those at lower risk for having an abnormal pap. Abnormal pap and dysplasia were seen more often in younger, Caucasian men with lower CD4 counts. We are continuing to expand this cancer screening by follow up of the patients with abnormal anal paps with anoscopy, directed biopsy, and treatment of dysplastic lesions.
1.22-2.04) and for rCDI was 2.13 (95% CI 1.60-2.80). Among NAAT-switch labs, although numerically higher, the median ratio of rCDI compared to the median ratio rCDI was not significant different (p=0.55). Rejection of formed stool was adopted in 11 of 12 NAAT-switch labs and in 0 of 3 non-switch labs.

Conclusions: After switching to NAAT, median increases of 71% for rCDI and 113% for rICD were noted, while CDI cases remained stable in labs continuing to use EIA. Implementation of stool rejection policies was common with the switch to NAAT and may have limited additional false positive test results. CDI diagnosed by NAAT must be correlated with clinical symptoms.

510 COMPARISON OF DIRECTLY OBSERVED ISOPIAZID-RIFAPENTINE WITH SELF ADMINISTERED ISOPIAZID AND RIFAMPIN REGIMENS FOR LATENT TUBERCULOSIS INFECTION—A PUBLIC HEALTH PERSPECTIVE

Yamin A1,2, Kempker R3,1, Borinste E2, Hensen R2. 1 Fulton County Health Department, Atlanta, GA and 2 Emory School of Medicine, Atlanta, GA.

Purpose of Study: Tuberculosis (TB) rates in the United States have been on the decline since. It is estimated that more than 80% of TB cases in the U.S. are the result of re-activated latent infection. A strategy focused on screening and treatment of LTBI is imperative to continue to decrease prevalence and case rate of TB in the U.S.

Methods Used: A retrospective observational cohort study design was utilized for this study. All patients evaluated for latent tuberculosis infection (LTBI) and agreeing to treatment at Fulton County Health Department TB clinic from January 2012 to December 2013 were included. Patients were tested for LTBI with either a tuberculin skin test (TST) or interferon-gamma release assay (IGRA). LTBI regimens included either 9 months of daily self-administered isoniazid (9I), 4 months of daily self administered rifampin (4R) or 3 months of weekly directly observed INH rifampine (3HRP).

Summary of Results: From January 1 to November 30, 2013, 321 patients were screened during the study period and 635 (39%) were offered treatment. Among these 635 patients, 496 (78%) accepted LTBI treatment and 356 (56%) of those offered treatment and 72% of those accepting treatment finished their recommended regimen. The majority of patients (n=294, 55%) received a 4R regimen, 33% (n=158) a 9I regimen, and 13% (n=62) received 3HRP. Patients with HIV were significantly more likely to receive 9I (79%) versus 4R (19%) or 3HRP (2%) while homeless patients were more likely to receive 4R (81%) as compared to either 9I (16%) or 3HRP (3%). Significantly more patient patients receiving 9I (12%) had their treatment regimen changed due to side effects as compared to 4R (4%) or 3HRP (5%), p<0.01.

Conclusions: In comparison to the treatment regimens of 9I and 4R, the newly approved regimen of weekly isoniazid and rifapentine had low rates of side effects and high completion rates. Patients receiving 3HRP did have lower rates of side effects of the HIV, so it will be important to next evaluate their treatment completion rates remain high among this high-risk population.

511 IN VITRO SYNERGISTIC ACTIVITY OF CASPOFUNGIN AND POLYMIXIN B AGAINST FLUCONAZOLE-RESISTANT CANDIDA GLABRATA

Kelly EA, Ashenafi DS, Pankey GA. Ochsner Clinic Foundation, New Orleans, LA.

Purpose of Study: Candida spp. account for most invasive fungal infections and the emergence of fluconazole (FL) and caspofungin (CS) resistance is problematic. Overcoming resistance with synergism of two drugs may be useful. In a recent in vitro study, colistin (polymyxin E) and CS were shown in vitro synergy against additional C. glabrata isolates. The purpose of our study was to extend this finding by evaluating polymyxin B (PO) and CS for in vitro synergy against additional C. glabrata isolates.

Methods Used: Eight FL-resistant C. glabrata isolates from the Florida State University, Pensacola, FL and 2 UT Southwestern Medical Center, Dallas, TX.

Purpose of Study: Respiratory syncytial virus (RSV) is the most important respiratory pathogen of early life. Previously, we have demonstrated that infant baboons infected with human RSV develop tachypnea, inflammatory responses and pathologic changes similar to those of human infants. In order to develop a better understanding of the pathogenesis of RSV infection, we carried out an analysis of pulmonary function tests (PFT) in RSV-infected infant baboons.

Methods Used: 4 week old baboons (n=6) were sedated, intubated and infected with RSV by instilling 5x10^8 PFU of human RSV intratracheally. Oxygen saturation (SpO2), arterial blood gases (ABG) and PFT were evaluated just prior to infection, and on days 5, 7 and 10 following infection. ABG were measured in intubated spontaneously breathing animals. PFT were measured by an Anea ventilator during positive pressure ventilation in animals with apnea induced by IV propofol.

Summary of Results: PFT did not change from baseline over the course of the study. Average O2 alveolar-arterial gradient (A-aO2) increased from 25.0 to 32.9 on day 5 (31.6% increase, p=0.023) with a subsequent trend back toward normal values. Average CO2 alveolar-arterial gradient (A-aCO2) did not change significantly. Changes in PFT were most pronounced on day 5: static and dynamic compliance decreased from Day zero values by 3.2% (p<0.06) and 10.5% (p=0.1), respectively, ratio of compliance in last 20% of inspiration time to total dynamic compliance increased by 18.2% (p=0.09), peak expiratory flow rate decreased by 9.3% (p=0.07), and work of breathing increased by 21% (p=0.067).

Conclusions: This is the first study, which showed that experimental RSV infection in baboons induces a statistically significant decrease in oxygenation as measured by A-aO2, and trends toward a decline in pulmonary compliance, a reduced peak expiratory flow rate, and an increase in work of breathing. Although PFT parameters did not achieve statistical difference, we expect that significant changes would be observed in a larger group of animals. PFT might be used to determine the effectiveness of therapeutic or prophylactic interventions in RSV infection.

513 THE LIKELIHOOD OF SERIOUS BACTERIAL INFECTIONS IN NEONATES WITH INCIDENTAL HYPERTHERMIA

Seddik TB1, Lee AE2,3, 1 Florida State University, Pensacola, FL and 2 UT Southwestern Medical Center, Dallas, TX.

Purpose of Study: Temperature symptoms including fever, temperature instability and hypothermia are among the most important indicators of serious bacterial infection (SBI) in neonates. Newborns with incidental hypothermia present an interesting diagnostic challenge. Newborns with hypothermia identified on well child visits are admitted for inpatient evaluation. Although literature is sparse regarding the management of
such neonates, many undergo a full evaluation for SBI. The purpose of this study is to identify and characterize the association between hypothermia and SBI in otherwise healthy neonates.

**Methods Used:** A retrospective chart review was utilized to analyze neonates who presented to the outpatient setting for a well newborn visit, found to have hypothermia and subsequently admitted to the inpatient unit. The association of certain factors -such as weight and feeding patterns- with hypothermia was also observed. We only included neonates with hypothermia who were evaluated for serious bacterial infection. We then compared the incidence of SBI in the hypothermia group with a cohort of well febrile neonates who were also evaluated for SBI.

**Summary of Results:** Over a period of five years, 59 hypothermic neonates and 104 febrile neonates were analyzed. All patients were evaluated with at least a blood culture. Within the hypothermia group, 85% of patients underwent urinary catheterization and 64% underwent a lumbar puncture. The incidence of SBI in well appearing hypothermic neonates is significantly lower than the incidence of SBI in febrile infants (3.4% Vs 25.7% p <0.05). All blood cultures and cerebrospinal fluid cultures were negative in the hypothermia group. Two patients had significant positive urine cultures for urinary pathogens, one of whom had a urinary tract infection with associated pyuria.

**Conclusions:** It is crucial to perform a comprehensive physical exam looking for signs of bacterial infection in neonates presenting with hypothermia, however, a full evaluation for SBI may not be necessary. Urine studies seemed to have a higher yield if a sepsis work up is considered. A larger, multi-institutional study is recommended to further evaluate the need for complete SBI evaluation in this particular population.

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**ERADICATION OF MULTIDRUG RESISTANT AND EXTREMELY DRUG RESISTANT PSEUDOMONAS INFECTIONS IN THE BURN ICU IN AN URBAN PUBLIC HOSPITAL**

Kandiah S2,3, Wong J2, Wang W2,3, Robinson Z2, Cole M2, Nair N2, Drake T2, Ray S2,3. Emory University School of Medicine, Atlanta, GA & Grady Memorial Hospital, Atlanta, GA.

**Purpose of Study:** To describe a successful intervention by antimicrobial stewardship and hospital epidemiology in an outbreak of multi-drug resistant and extremely drug resistant Pseudomonas infections (MDRPa and XDRPa) in the burn ICU (BICU).

**Methods Used:** Surveillance of pharmacy and microbiology data identified an increasing number of resistant Pseudomonas infections in our 8-bed BICU at Grady Memorial Hospital between October 2013 and May 2014. A real time BICU-specific antibioticogram was created and shared with the BICU attending. Carbapenem (CPM) restriction and new empiric antibiotic therapy recommendations for BICU patients were implemented in May 2014. Universal contact precautions and careful environmental cleaning were employed in the BICU before and during the intervention. Incidence rate of MDR/XDR Pa infections was compared before and after the intervention.

**Summary of Results:** During the 8-month outbreak period, 13 patients had Pa infections; 8 (62%) were drug resistant (3 MDRPa, 8 XDR). Prior CPM use was identified in 10 (78%) of Pa infection patients. In the 4 months since instituting CPM restriction, 2 patients have had Pa infections and 0 (0%) were drug resistant. Pa infection incidence rates before and after the intervention were 8.6/1000 pds and 4.2/1000 pds respectively (p=NS) and the elimination of MDR/XDR Pa infections has been sustained for 4 months to date.

**Conclusions:** Antimicrobial stewardship programs can employ unit-specific antibiotics in conjunction with real time surveillance by hospital epidemiology and infection prevention to intervene successfully during an outbreak of MDR/XDR Pseudomonas

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**POSSIBLY BEYOND USUAL CELLULITIS**

Fenire M, Rudd D, Patel P. East Tennessee State University, Johnson City, TN.

**Purpose of Study:** 1-Report a case of Edwardsiella tarda (E. tarda) bacteremia and cellulitis including images. 2-Raise awareness regarding infections caused by marine exposure

**Methods Used:** Case report and review of literature

**Summary of Results:** A 71 year old male with known history of atrial fibrillation and mechanical valve replacements (mitral and aortic) presented with right upper extremity redness. While fishing in a local lake the day prior to admission, he received a small puncture wound to the distal right forearm by a fin of a bass. Within hours, he noted worsening erythema around the puncture site that became tender and spread to involve his entire forearm (Panel 1). Upon presentation, peripheral blood cultures were obtained and empiric antibiotic therapy with vancomycin was initiated; however, the erythema worsened over the next 48 hours. Infectious disease was consulted on hospital day 3, and ceftriaxone was added. Blood cultures revealed E. tarda (Panel 2). Vancomycin was discontinued and ceftriaxone dose was increased to 2 grams. Transthoracic echocardiography was obtained and revealed no evidence of vegetation. The patient responded well to ceftriaxone therapy and was discharged home to complete a 2 week course of ceftriaxone and to follow up in clinic. Discussion: In 1965, Ewing et al introduced a new family of Enterobacteriaceae - E. tarda. The "tarda" refers to the inability of this species to ferment most carbohydrates. E. Tarda has been recovered from a large variety of freshwater and marine life, including turtles, water tortoises, fish, pelicans, alligators, seals, toads, snakes, and lizards. This species is an oxidase-negative, catalase-positive, gram-negative bacilli that is motile by means of peritrichous flagellae. Most strains of E.tarda are sensitive to ampicillin, chloramphenicol, tetracycline, and aminoglycosides; but not to colistin. While E. tarda most commonly causes gastroenteritis, its range of pathology can be serious in patients with underlying liver disease, diabetes mellitus, immunosuppression, or malignancy. In patients with extraintestinal infections, mortality rate was reported up to 44%.

**Conclusions:** Clinicians should maintain a high level of suspicion for gram negative organisms caused by marine exposure particularly in immuno-compromised host. Failure to do that may lead to higher morbidity and mortality.

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**Conclusions:** Antimicrobial stewardship programs can employ unit-specific antibiotics in conjunction with real time surveillance by hospital epidemiology and infection prevention to intervene successfully during an outbreak of MDR/XDR Pseudomonas
DIFFERENTIAL INDUCTION OF HOST GENES BY ACHROMOBACTER SPECIES

Swenson C1, Sadikot R2,3, University of Florida, Gainesville, FL; 2Emory University, Atlanta, GA and 3Department of Veterans Affairs, Atlanta, GA.

Purpose of Study: Achromobacter are environmentally ubiquitous gram-negative bacteria that can be opportunistic pathogens affecting multiple organ systems, including the respiratory tract. Although the organism was first isolated and described in the early 1970s, little is known about the molecular pathways involved in its pathogenesis, specifically pertaining to host responses. We and others have previously shown that lipid mediators generated by the cyclo-oxygenase pathway play a critical role in host immune response to infections. In these studies we sought to investigate the signaling mechanisms involved in the host response to Achromobacter.

Methods Used: Two Achromobacter species, xylosoxidans and denitrificans, were isolated from clinical specimens of a patient with recurrent Achromobacter infection. Human macrophage and epithelial cells were infected with Achromobacter (MOI 1-10) for 6 and 24 hours. Expression of inflammatory mediators, including cyclooxygenase 2 (COX2), prostaglandin E2 (PGE2), inducible nitric oxide species (iNOS), and cytokines (TNF-α and IL-1β) were determined by RT-PCR or ELISA. Summary of Results: Both Achromobacter species induced COX2 with production of PGE2 in human macrophages and epithelial cells, with higher mRNA concentrations in the A. xylosoxidans-infected specimens. The induction of COX2 by Achromobacter was dependent on NF-κB activation. A. xylosoxidans induced iNOS in macrophages, however, surprisingly, A. denitrificans inhibited the induction of iNOS.

Conclusions: Like other gram-negative pathogens, Achromobacter isolates are capable of inducing COX2 followed by production of PGE2 in host macrophages and epithelial cells in vitro. PGE2 production by gram-negative organisms has been shown to impair bacterial clearance, and thereby help establish infection. Macrophage iNOS production was inhibited by A. denitrificans, but not A. xylosoxidans. This differential induction of host genes by the two species may be responsible for the differences in the pathogenicity of these two clinical isolates. Further research into these complex signaling mechanisms is warranted.

ASSOCIATION BETWEEN VITAMIN D LEVEL AND OTHER BIOMARKERS IN OBESITY OF CHILDREN AND ADOLESCENTS

Radulescu A1, Wallace C1, Li Y1, Killian M2, Bada H1, University of Kentucky, Lexington, KY; 1University of Kentucky, Lexington, KY and 2University of Kentucky, Lexington, KY

Purpose of Study: To determine the association between vitamin D level and other biomedical abnormalities in pediatric patients with obesity.

Methods Used: We reviewed medical records of obese children seen in the Pediatric High Risk and Bayley's III scores in this small cohort. Significant weight gain and adiposity have been associated with duration of breastfeeding, we evaluated infant feeding history provided by the mother. We found no significant differences in the duration of breastfeeding in the DHA supplemented and placebo groups (6.6 ± 2.0 vs 5.0 ± 1.5 months, p=0.51).

Conclusions: These data suggest that maternal dietary DHA supplementation in pregnancy may play a protective role in early life against the development of excess adiposity.

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Conclusions: These data suggest that maternal dietary DHA supplementation in pregnancy may play a protective role in early life against the development of excess adiposity.
Purpose of Study: To compare outcomes of infants pre and post initiation of a feeding protocol providing an exclusive human milk (HM)-based diet.

Methods Used: In a multicenter cohort study, infants who received a bovine-based diet (BOV) of mother’s milk fortified with bovine fortifier and/or formula were compared to infants who received a newly introduced exclusive HM feeding protocol. Infants <1250 g birth weight were included. Excluded infants had major congenital anomalies or died in the first 12 hours of life. Study data were collected for 2-3 years prior to and after introduction of an exclusive HM-based diet. Primary outcome was necrotizing enterocolitis (NEC) and/or death.

Summary of Results: 1386 infants were studied from 3 centers in Texas, Illinois and California. There were no differences in baseline demographics or growth of infants.

Conclusions: Premature infants who received an exclusive HM-based diet had a significantly lower incidence of NEC and/or death. HM group also had a reduction in late onset sepsis and threshold Retinopathy of Prematurity. This multicenter study further emphasizes the multiple benefits of an exclusive HM-based diet and demonstrates improved outcomes after implementation of such a feeding protocol.

Outcomes

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<th>BOV (%)</th>
<th>HM (%)</th>
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<tr>
<td>NEC (%)</td>
<td>14.8</td>
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<td>&lt;0.0001</td>
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<td>Medic NEC (%)</td>
<td>5.5</td>
<td>2.2</td>
<td>&lt;0.01</td>
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<td>Surgical NEC (%)</td>
<td>9.2</td>
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<tr>
<td>NEC and/or death (%)</td>
<td>27.8</td>
<td>19.7</td>
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<td>Late-onset sepsis (%)</td>
<td>27.4</td>
<td>18.3</td>
<td>&lt;0.001</td>
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<tr>
<td>Threshold Retinopathy of Prematurity (%)</td>
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</table>

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PROVIDERS DO NOT DIAGNOSE OVERWEIGHT/OBESITY IN CHILDREN

Burgess K. University of Alabama, College of Community Health Sciences, Tuscaloosa, AL.

Purpose of Study: Research finds parental discernment of child over-weight/obesity is poor. Further, studies suggest patients have better awareness of weight status if health providers discuss it with them. As obesity is plaguing our healthcare system, providers should inform patients of weight status routinely. If providers do not diagnose obesity, its incidence could be underestimated.

University Medical Center, in Tuscaloosa, AL, provides care to a large number of children in both the pediatrics and the family medicine clinics. This study utilized electronic health data to determine whether providers document overweight/obesity and to estimate with what frequency.

Methods Used: EHR report pulled BMIâ€¼ile for age, diagnoses, and other demographic information for children aged 2-18 years for a 6 month period. Children were categorized as being overweight or obese using the CDC definition.

Summary of Results: 3818 patients were seen during a 6 month period by 57 providers. 16% (602) were overweight and 22% (832) were obese. A diagnosis of “overweight” was only given to 1% of overweight patients, and a diagnosis of “obese” was only given to 10% of obese patients. 48% of these diagnoses were made concurrently with a patient’s annual check up. 315 overweight children presented for check ups, and 1% were diagnosed as overweight at that time. 396 obese children presented for check ups, and 12% were diagnosed as obese at that time. Additionally, 5% of obese children were misdiagnosed as overweight, 37% of those diagnoses occurring at time of check up.

Conclusions: Overweight and obesity are under diagnosed, and diagnosis does not occur until children are very obese. As unhealthy weight is such an important health problem, providers should take any opportunity to discuss unhealthy weight status with patients, especially at the more comprehensive annual check up visits. Providers did not consistently capitalize on these opportunities. More investigation is necessary to determine why providers do not consistently diagnose unhealthy weight status. Because research indicates that patients and families do not recognize overweight and obesity, it is important for providers to make this diagnosis and discuss with patients for improved awareness and subsequent behavior changes.

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DURATION OF CHILDHOOD OBESITY AND RELATION TO MIDDLE-AGE OBSTRUCTIVE SLEEP APNEA RISK: THE BOGALUSA HEART STUDY

Hu T1, Bertisch S2, Chen W1, Harville E1, Redline S1, Bazzano L1, 1 Tulane University, New Orleans, LA; 2Beth Israel Deaconess Medical Center, Boston, MA and 3Brigham and Women’s Hospital, Boston, MA.

Purpose of Study: We prospectively examined the association between duration of overweight and obesity (OW) in childhood and subsequent risk of OSA in 844 adults with ≥ 2 measures of body mass index (BMI) between 4 and 18 yrs of age.

Methods Used: Childhood OW was defined using age and gender specific BMI ≥85th percentile based on 2000 CDC Growth Charts. Duration of OW was calculated using the presence or absence of OW at each follow-up examination: For participants normal weight at baseline who then became and remained OW through the last examination, or participants who were OW throughout, duration was calculated as the cumulative number of consecutive OW yrs. If participants were OW then ever became normal weight during childhood (N=84), duration was not calculated. After an mean follow-up period of 35 years (in 2010; IQR 1yr), high-risk for OSA was determined using Berlin Questionnaire score as the primary outcome. Secondary outcomes included persistent snoring and daytime sleepiness.

Summary of Results: Of those included in the analysis, 42.3% were male and 33.6% were Black. At baseline mean (SD) age was 9.9 (2.9) yrs and proportion of OW individuals was x. At follow-up, individuals were mean age of 23.4 (8.5) yrs and had a mean BMI of 31 kg/m2. In total, 217 (25.7%) had elevated Berlin scores indicating high-risk for OSA. Mean (SD) of OW duration was 5.2 (2.5) yrs. In multivariable log-linear regression models adjusted
for baseline age, race, sex, follow-up time, education, current smoking status, alcohol consumption, physical activity and current OW status, participants with an OW duration of 1-4 yrs, 4-8 yrs, and 8+ yrs were 1.19 (95% CI 0.90-1.57), 1.23 (0.92-1.63), and 2.29 (1.67-3.15) times more likely to be high-risk for OA as compared to those who were never OW. Significant linear trends were present across categories of OW duration (P for trend 0.006). Similar positive trend was observed for persistent snoring but not for daytime sleepiness. There was no significant effect modification by race or sex.

**Conclusions:** This community-based cohort study suggests that longer OW duration childhood was associated with high-risk for OA in middle-age.

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**ADHERENCE TO LOW CARB AND LOW FAT DIETS IN RELATION TO WEIGHT LOSS AND CARDIOVASCULAR RISK FACTOR REDUCTION**

Hu T1, Stuchlik P1, Yao L2, Reynolds K1, Wheulton P1, He J1, Bazzano L1.

**Tulane University, New Orleans, LA; 2University of Minnesota, Minneapolis, MN**

**Purpose of Study:** We compared indicators of dietary adherence between two dietary interventions and examined their relationship with efficacy using data from 148 obese adults (Mean age 47 y; Mean BMI, 35 kg/m2; 11.5% men; 51% Black) who participated in a clinic trial comparing the effect of a LCD (net carb ≤30 g/d) with the LFD (<30% fat, <10% saturated fat) on changes in weight and CVD risk factors.

**Methods Used:** Indicators of dietary adherence included attendance at dietary counseling sessions which provided the same behavioral curriculum for weight loss to each group, deviation from the macronutrient goal of the diet and participants’ actual intake, and the cumulative percentage of urinary ketones detected at each of the 26 behavioral sessions. A composite adherence score was created based on these 3 indicators. Outcomes included changes in body weight, body composition and CVD risk factors at 12 mo.

**Summary of Results:** There was no significant difference in the attendance at dietary counseling sessions between the groups (57% in LCD group vs 52% in LFD group). In the LCD group, 45% of individuals met the carb goal while 55% were above, with a mean of 198 deviation representing a carb intake of about 119 g. In the LFD group, 56% and 28% of individuals met total and saturated fat goals, and those who were above had mean deviations of 23% and 46%, representing intakes of about 37% total fat and 10% saturated fat. At 12 mo, compared to the LFD group, the LCD group had a higher cumulative percentage of ketones detected in urine (25% vs 8%, P<0.001). There was no significant difference in composite scores for adherence between the groups [Mean (SD): 64 (19) in LCD group vs 61 (20) in LFD group]. In the LCD group, a higher composite score reflecting adherence the LCD was associated with more weight loss, loss of fat mass and preservation of lean mass. Indicators of adherence to a LCD diet were not associated with blood pressure, lipids, glucose or C-reactive protein. None of adherence indicators was associated with any study outcome in the LFD group.

**Conclusions:** Despite similar adherence between the LCD and LFD, adherence to the LCD resulted in greater weight loss and better improvement in body composition at 12 mo.
However, the combined treatment led to significantly worse scores in neurodevelopmental performance, compared to individual treatment alone. LPS immunostaining showed that both LPS and hyperoxia treatments increased the number of activated microglia, which was further enhanced by the combined treatment.

**Conclusions:** Our data suggest that intraperitoneal LPS-induced neuroinflammation may interact with hyperoxia, resulting in enhanced neurodevelopmental disturbances, possibly involving microglia activation.

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**527 ACCURATE ANALYSIS OF DILUTED HUMAN MILK**

Smith BW,Radmacher PG,Adamkin D. University of Louisville, Louisville, KY.

**Purpose of Study:** Human milk (HM) analyzers, adapted from the dairy industry, are a new tool for improving HM nutrition for the preterm infant. The amount of native milk needed for the analysis (~16 mL) may present an obstacle to routine use. The purpose of this study was to determine the maximum dilution (minimum volume) of HM that yields reliable results for protein, fat and lactose.

**Methods Used:** This study was determined to be exempt by the University of Louisville IRB. Samples of frozen, native HM (remaining post-discharge, scheduled for disposal) were obtained with names removed. Milk was thawed and warmed in a water bath at 40°C for 10 minutes. A control HM sample was included to ensure device performance. Undiluted HM was analyzed (Calais Human Milk Analyzer, North American Instruments, Solon, OH) for macronutrient composition: total protein (P), fat (F), lactose (L), and Energy (E) was calculated from the macronutrient results. Subsequent analyses were done with 1:2, 1:3, 1:5 and 1:10 dilutions of each sample with distilled water.

**Summary of Results:** There were no statistically significant differences in concentration (g/dL) between native and diluted HM samples for P, L, or F at the 1:2 or 1:3 dilutions. However, P concentration did show a statistically significant difference with 1:5 and 1:10 dilutions (1.26 ± 0.99 and 1.46 ± 0.25 vs. 1.15 ± 0.16, p=0.004). F concentration was significantly different at the 1:10 dilution (2.74 ± 1.29 vs. 3.31 ± 1.34, p=0.029). L concentration was significantly different at the 1:5 and 1:10 dilutions (6.74 ± 0.08 and 6.81 ± 0.18 vs. 6.62 ± 0.13, p=0.001). When comparing the agreement between diluted and undiluted samples, statistically significant differences existed for P and F for all dilutions (p=0.015 and p=0.005). L was significant at the 1:2 and 1:10 dilutions only (100.8 ± 1.7% and 102.9 ± 2.5%, p<0.001).

**Conclusions:** The Calais Human Milk Analyzer can be used with diluted HM samples and return results within 5% of values from undiluted HM at a 1:2 or 1:3 dilution. At a 1:5 or 1:10 dilution, however, results vary as much as 10%. The most variability was with P (slightly higher values) and F (slightly lower values). At the 1:2 and 1:3 dilutions these differences appear to not be significant in the context of nutritional management. However, the accuracy and reliability of the 1:5 and 1:10 dilutions are questionable.

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**528 FETAL ENVIRONMENT ALTERS PANCREATIC DEVELOPMENT IN NON-HUMAN PRIMATES**

Palaczuk JI, Blanco C, Nathanielsz P, Li C, Quinn A. University of Texas HSC, San Antonio, TX.

**Purpose of Study:** The fetal pancreas shows developmental plasticity to its environment. The objective of this study is to examine histological changes in the fetal endocrine pancreas after exposure to antenatal corticosteroids (ANS), maternal obesity (MO) and maternal nutrient restriction (MNR) and correlate to pancreatic markers for differentiation, function, and proliferation in non-human primates.

**Methods Used:** Twenty-three fetal baboons were delivered via C-section near term as follows: 6 Control (CTR), 6 MNR (70% global diet), 5 MO (45% energy fat, 4% glucose, and unlimited fructose), and 6 ANS (betamethasone). Pancreatic tissue was obtained and immunohistochemistry performed. Relative percent areas of the microscopic structures were calculated utilizing the Computer Assisted Stereology Toolbox 2.0 system. Expression of IGFI, Ki67, HNF4α, and PDX1 was quantified. Statistical analysis was performed with SPSS 17.0.

**Summary of Results:** Birth weight was lower in MNR and MO fetuses compared to CTR and ANS (p<0.01). The total pancreatic weights as % of body weight was similar. Maternal serum insulin was higher in MO group compared to all groups (p<0.05). No differences were found in fetal serum insulin. Fetal α-β-δ cell percent area comprised 10.8±4.3, 13.3±3.3 and 10.9±3.5 (mean, ±S.E.) of endocrine pancreas in CTR animals, respectively. Alpha cell percent area was increased in ANS fetuses by 2.7, 2.0, and 1.8 fold when compared to MO, MNR and CTR fetuses (p=0.03, 0.1, 0.1, respectively). Beta cell area decreased by 66% in the MNR fetuses as compared to CTR (p=0.07) and was paired with a decrease in IGFI, HNF4α, Ki67, and PDX1 expression (p=0.05 vs. all groups). Beta cell area positively correlated with the expression of HNF4α and Ki67 (p=0.05, R2=0.9). IGFI and Ki67 were increased in the MO fetuses as compared to CTR (p=0.01). Delta cell percent area was not altered.

**Conclusions:** The adjusted fetal pancreatic weights is not affected by maternal environment while alpha cell area increases with exposure to ANS and beta cell area decreases with MNR. The expression of fetal pancreatic markers for endocrine cell function, proliferation, and differentiation is altered depending on maternal nutrient intake. Disruption of pancreatic development at these critical periods may have long lasting consequences.

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**529 HYPERGlyCEMIA INDUCES EXAGGERATED PLACENTAL INFLAMMATORY RESPONSE AFTER INFECTION**

Yee A 1,2, Aneja E 2, Lin X 2, Wan-Huen P 2, Hanna N 2. University of Alabama at Birmingham, Birmingham, AL and 2Winthrop University Hospital, Mineola, NY.

**Purpose of Study:** Preterm delivery (PTD) remains an important cause of neonatal morbidity and mortality. Although its pathogenesis is multifactorial, infection-induced inflammation is the most common identifiable cause. Recent studies suggest that type 2 diabetes is an inflammatory disease. However, there are no studies investigating the effect of hyperglycemia on placental inflammatory response specially when associated with infection. It is the objective of this study to investigate the effects of hyperglycemia on human placental production of pro- and anti-inflammatory cytokines with or without infection.

**Methods Used:** Term human placentas (n=10) were obtained from elective C-sections. Placental explant culture model was utilized. Placental cultures were pretreated with either high glucose (25mMol/L) or normal glucose (5.5mMol/L) in 5% O2 and CO2 for 24 hours. Cultures were treated with either: LPS (Lipopolysaccharide; established model for infection) or media control. The media were harvested at 3, 6, 18, and 24 hours. ELISA was used to measure IL-1β and IL-10 levels in supernatants. Data was analyzed using paired t-test and Wilcoxon rank sum test.

**Summary of Results:** As expected placental explants without LPS stimulation had low levels of IL-1β and IL-10 in supernatants of both high and normal glucose groups. LPS stimulation induced significant increase in IL-1β and IL-10 secretion. However compared to normal glucose treatment, high glucose treatment significantly exaggerated the LPS-induced IL-1β production at the 18 and 24 hour time points (p<0.05). Interestingly, high glucose treatment significantly decreased the placental pro-pregnancy IL-10 production in the presence of LPS compared to normal glucose treatment at the 18 and 24 hour time points.

**Conclusions:** In the presence of LPS, high glucose causes a significant increase in the production of pro-inflammatory cytokine IL-1β and decrease in the anti-inflammatory cytokine IL-10, indicating an exaggerated placental inflammatory response to infection. This suggests that patients with hyperglycemia and intrauterine infection may have increased risk of PTD.

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**530 STAT3 INHIBITORS ATTENUATE THE SEVERITY OF EXPERIMENTAL NEC**

Scott B, Chaaban H, Lawrence S, Eckert J. OUHSC, Oklahoma City, OK.

**Purpose of Study:** Necrotizing enterocolitis (NEC), the leading cause of death from gastrointestinal disease in premature infants, is characterized by acute inflammation of the intestinal mucosa and can quickly progress to bowel necrosis and death. Despite advances in neonatal care, survival has remained unchanged due to lack of therapies for this devastating disease. The pathogenesis of NEC involves the induction of toll-like receptors (TLR), resulting in activation of pathways that induce an intense pro-inflammatory response in neonates. Down-regulation of TLR-mediated inflammatory response has been shown by inhibition of STAT3 pathways. The use of STAT3

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inhibitors in NEC models has not been explored. We hypothesize that DLL3, a novel inhibitor of STAT3 derived from curcumin, is well tolerated in mice pups and can decrease the inflammatory response in a NEC model.

Methods Used: To determine safety and tolerability, mouse pups were exposed to DLL3 (50mg/kg bw) from P9-14. To determine the effect of DLL3 on NEC, we used a model by Zhang et al in which P15 mice are injected with dithiozone to induce Paneth cell loss, followed by *K. pneumoniae* infection. Pups were euthanized when in distress or 10 hrs post NEC challenge. Gross examination of small bowel were compared between groups. Sections of bowel were harvested for histopathology analysis and protein quantification. NEC severity will be compared using the standardized NEC scoring by a blinded pathologist. Small bowel protein quantification and group comparison of inflammatory cytokines/chemokines will be processed via Procarta Plex Mouse Cytokine & Chemokine Panel 1A.

Summary of Results: DLL3 given orally is well tolerated in mice pups without adverse effects. Using the Paneth cell ablation NEC model, DLL3 treated group showed a gross decrease in intestinal inflammation. Small bowel histopathologic evaluation, inflammation scoring, and quantification of inflammatory cytokines/chemokines are currently in progress.

Conclusions: We show that a novel STAT3 inhibitor, DLL3, can be safely administered to neonatal mice. Furthermore, our appreciation of a reduction in gross bowel inflammation is very promising and suggests a possible role for reducing inflammation in NEC. We are hopeful that completion of histopathologic evaluation of bowel tissue and quantification of inflammatory cytokines/chemokines will further support our hypothesis.

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OUTCOME OF INTRAUTERINE GROWTH RESTRICTED PRETERM INFANTS

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Purpose of Study: To evaluate growth outcomes and major morbidities in preterm intrauterine growth restricted (IUGR) extremely low birth weight infants.

Methods Used: Retrospective case-control study evaluating IUGR preterm infants born between a five year period in a level III NICU. IUGR neonates were matched to controls (1:1) by extremely low birth weight. Outcome measures included growth velocities, TPN days, and total enteral calories at 36 weeks CGA.

Summary of Results: Forty-eight infants (24 IUGR, 24 controls) were identified in the NICU and divided into aliquots. Samples analyzed fresh and after freezer storage for 3 months. After 6 months of freezer storage the NO concentration was 6.5 (a reduction of 62%, p<0.05) after freezer storage for both 3 and 6 month periods. Premature infants at risk for necrotizing enterocolitis (NEC). Mothers of preterm infants often express concern about the pH of the various feeds including parenteral are needed. Incidence of NEC below 17

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DOES THE ADDITION OF A LIQUID HUMAN MILK FORTIFIER RESULT IN SUSTAINED METABOLIC ACIDOSIS IN VERY LOW BIRTH WEIGHT INFANTS?

Sternad L, Gates A, Bhatia J. Georgia Regents University, Augusta, GA.

Purpose of Study: The purpose of this study was to investigate if an acidified liquid human milk fortifier [LHMF] given to premature infants leads to sustained metabolic acidosis.

Methods Used: This was a retrospective study of very low birth weight infants [Birth weight<1500g] admitted to the NICU and identified as receiving human milk, mother's own or donor, as part of their enteral diet. Data collected included birth weight, gestational age, day of addition of liquid human milk fortifier, CO2, Blood urea nitrogen and weight gain. All laboratory values obtained were recorded. From the data, time to regain birth weight, and weight gain from RTBW to 2000g were calculated. Incidences of CO2 values below 17 mEq/L were considered acidic.

Summary of Results: Forty-six infants met the criteria and were identified. Of these, 27 were fed mother's own milk [MOM] and 19 received donor breast milk [donor] obtained from a milk bank. Both feedings were fortified when enteral intake approximated 80 mL/kg/d. Infants were weaned from parenteral nutrition [PN] as enteral feedings were advanced. Fortification was started at a mean age of 24.17 days and full fortification was achieved between 35 and 40 days of life. Incidence of CO2 below 17 is depicted in the Table. Mean age at RTBW was 12.11 [SD 5.89] ALL days with MOM 10.37 [5.18] and Donor 14.53 [6.38] days. Growth velocities were 15.64 [2.98] ALL, 15.98 [2.65] MOM and 15.15 [3.4] donor, g/kg/d. Growth velocity was comparable to data in the literature. BUN [data not shown] values were similar in the two groups and there was no correlation between BUN and weight gain in this cohort.

Conclusions: These data demonstrate a lack of evidence for sustained metabolic acidosis in VLBW infants provided acidified LHMF. More studies about the pH of the various feeds including parenteral are needed. Incidence of CO2 below 17

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COMPARISON OF LACTOFERRIN ACTIVITY IN FRESH AND STORED HUMAN MILK

Raafq NA, Radmacher PG, Adamkin D, Telang S. University of Louisville, Louisville, KY.

Purpose of Study: Lactoferrin (Lf), a protein found in human milk (HM), has been shown to have both anti-inflammatory and antioxidant activity in the neonatal gut. HM has been shown to be protective for preterm infants at risk for necrotizing enterocolitis (NEC). Mothers of preterm infants often express concern about the pH of the various feeds including parenteral are needed. Incidence of NEC below 17

<table>
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<tr>
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<th>Before LHMF [PN]</th>
<th>During LHMF +PN</th>
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<tr>
<td>ALL</td>
<td>14/276</td>
<td>16/336</td>
<td>3/166</td>
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<tr>
<td>MOM</td>
<td>12/160</td>
<td>2/139</td>
<td>1/64</td>
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<tr>
<td>DONOR</td>
<td>3/117</td>
<td>13/195</td>
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BREASTFEEDING INTENTION IN MOTHERS WITH HIGH RISK FOR DRUG USE

Halloran DR1,2, Ryne E1,2, Borawski A1, Hinyard L1. 1Saint Louis University, St Louis, MO and 2Saint Louis University, St Louis, MO.

Purpose of Study: Although breastfeeding has been shown to reduce the severity of the neonatal abstinence syndrome symptoms in infants, it is
unclear what percent of women who are high risk for drug abuse choose to breastfeed. This study will examine the rate of breastfeeding intention in women who are high risk for drug abuse.

Methods Used: This is a retrospective cohort of all viable infants (~24 weeks gestation) born at a tertiary care center in 2013. High risk women were defined as those with a known substance use history, poor prenatal care, or HIV+ based on the mother’s chart. Breastfeeding intention is obtained at admission and was abstracted along with factors associated with breastfeeding rates. Descriptive and Cochran-Mantel-Haenszel statistics were calculated.

Summary of Results: There were a total of 3082 infants born during the study period. 1626 (53.3%) women were noted to be high risk including 48 (1.6%) women who were known drug users. 1553 (53.2%) non-high risk women planned to exclusively breastfeed compared to 48 (29.6%) high risk women. Among the 48 known drug users, 8 (16.3%) women intended to breastfeed. Overall, women who were high risk were less likely to intend to breastfeed (OR 0.37 (95% CI 0.26, 0.52); p<0.001). Among high risk women, multiparous women and known drug abusers were 79% (OR 0.21 (95% CI 0.09, 0.51); p<0.001) and 64% (OR 0.36, 95% CI (0.15, 0.83); p=0.02) less likely to intend to breastfeed, respectively. Maternal age and gestational age were not associated with intention in high risk women.

Conclusions: Women who are high risk for drug abuse are less likely to intend to breastfeed. Further data will be presented based on infant chart abstraction on actual breastfeeding rates and additional characteristics associated with breastfeeding.

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COMPARISON OF CONVENTIONAL AND NOVEL HEARING SCREENING TESTS IN NICU

Gulati R1, Yellamsetty A2, Bhagat S2, Talati AJ1, 2UTHSC, Memphis, TN and 1University of Memphis, Memphis, TN.

Purpose of Study: Hearing loss is 10-20 times more likely in NICU infants who are at risk for speech delay. Screening programs are designed to utilize Otoacoustic emission (OAE) tests and Auditory Brainstem Response (ABR) after birth. NICU infants are vulnerable to middle ear diseases which can interfere with the predictive power of OAE/ABR. It is important to resolve the location of problem within the ear, as treatment of middle ear disorders differ from that of sensorineural deafness. Our aim was to determine the feasibility of a novel hearing test to detect middle ear diseases. We compared tympanometry (TM) with wide band reflectance (WBR) measurements and their relationship to conventional ABR and OAE.

Methods Used: Neonates who failed ABR were recruited after getting informed consent; Infants with congenital malformations of ears or with preexisting tympanic membrane perforation were excluded. Demographic data, ABR results and other pertinent data were recorded. TM, WBR and OAE were performed by an audiologist using standard techniques. Phi coefficient was calculated for comparisons using SPSS.

Summary of Results: 29 infants were included in the study with 58 ears tested. 72% (21/29) were males, median BW was 2380gms and median GA 34.5 weeks. Median age at the time of hearing tests was 23 days. Of the 58 ears 5 % passed both TM and ABR, 12 % passed only ABR, 36 % passed only TM, while 46% failed both. 58% of ears failed TM and all patients failed WBR at one or all of the frequency. When ABR was compared with WBR and TM, they were not correlated indicating different pathology. TM and DPOAE test results are related (p<0.05) with TM and WBR test results were unrelated and independent of each other. WBR test results had stronger numerical associations with DPOAE test results compared with the associations between TM and DPOAE.

Conclusions: WBR as a middle ear screening test is feasible in neonates, and a significant number of neonates fail hearing screens probably of middle ear problems. If identified early, this can be treated rather than waiting several months for a repeat test as is current practice.

Renal, Electrolyte and Hypertension II
Concurrent Session
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CITRATE TRANSPORT IN NaDC1 KNOCKOUT MICE

Hering-Smith K1,2, Huang W1, Coleman-Barnett J1, Hamm LL1, 1Tulane University, New Orleans, LA and 2Tulane University, New Orleans, LA.

Purpose of Study: Urinary citrate is the major endogenous inhibitor of calcium nephrolithiasis. Regulation of urinary citrate has been presumed to occur solely by the sodium dependent dicarboxylate cotransporter (NaDC1) located on the apical membrane of the proximal tubule.

Methods Used: Using NaDC1 knockout (KO) and heterozygous (Het) mice (originally generated by Ho et al, Kidney International 2007) along with their wild type litter (WT) mates, we studied citrate and other Krebs cycle intermediates in urine with mice on either a normal diet (NI) or on a 72 hour acid loading protocol (AL).

Summary of Results: Plasma citrate concentrations did not differ substantially in any of the conditions. On NI diets, fractional excretion (FE) of citrate was higher in KO animals (0.24±0.02) than in WT (0.13±0.05); Hets were intermediate (0.18±0.04). Therefore, although KO of NaDC1 increases citrate excretion, substantial reabsorption of citrate still occurs in these animals (fractional reabsorption = 1-FE).

With acidosis FE in WT was nearly abolished (0.003±0.001) but did not change in the KO animals (0.24±0.02 NI vs 0.25±0.02 AL diet). Surprisingly Het animals responded somewhat to acidosis but not completely, FE of 0.10±0.06 with AL and 0.18±0.04 with NI diet.

Urineary excretion of other dicarboxylates paralleled that of citrate except that urine excretion of each decreased significantly in KO animals with acidosis.

Conclusions: Thus, in examining FE in NaDC1 KO mice we found that most of renal reabsorption of citrate is not via NaDC1; although NaDC1 appears to account for the response to acidosis for citrate. Interestingly, the reabsorption of other Krebs cycle intermediates did respond significantly to acidosis. Also and surprisingly, heterozygous animals do not have normal citrate reabsorption indicating that two normal alleles are important for normal citrate transport.

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RENOPROTECTIVE EFFECTS OF PARICALCITOL IN DIABETIC KIDNEY DISEASE

Parekh KK, Yego C, Prabhakar S. Texas Tech University Health Sciences Center, Lubbock, TX.

Purpose of Study: The role of vitamin D in health and disease has been constantly expanding and the therapeutic significance of vitamin D and its analogues in CKD and ESRD is well established. Recent experimental studies alluded to the anti-proteinuric effects of vitamin D analogues in CKD. The mechanisms underlying such effects remain unclear, although vitamin D could suppress renin angiotensin system (RAS). The aims of the present study are to examine renal effects of paricalcitol (PAR) in a rat model of diabetic nephropathy and to explore potential mechanisms underlying such effects.

Methods Used: We used obese ZSF rats, a murine model of nephropathy in type II diabetes, in four different groups- control, losartan (LOS) - a known angiotensin receptor blocker , paricalcitol (PAR) and LOS + PAR. Losartan was administered 25 mg/L in the drinking water. Paricalcitol was injected subcutaneously 1 μg/kg BW twice weekly. Rats were studied from 18 to 30 weeks of age with weekly monitoring of body weights, and water intake. At 30 weeks, rats were sacrificed and kidneys were harvested, homogenates examined for expression of eNOS and TGF-β. Blood and urine samples were obtained at the start and end of the study to evaluate renal function.

Summary of Results: At 30 weeks ZSF rats exhibited hyperglycemia, hypertension, nephrosis, renal failure and obesity. Compared to control rats (1805 ± 374 mg/dL) both LOS (796 ± 108 mg/dL, p<0.035) and PAR (964 ± 612 mg/dL, p=0.038) reduced urinary protein excretion but LOS+PAR (529 ± 62 mg/dL, p=0.02) reduced more than either agent alone. The creatinine clearance (expressed in L/kgBW/d) in control rats at 30 weeks was

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RET IS A DIRECT POSITIVELY REGULATED TARGET OF ANGIOTENSINII(ANGII) IN THE URETERIC BUD(UB)

Song R, Preston G, Yosypiv IV. Tulane University; New Orleans, LA.

Purpose of Study: Our recent studies identified Ret signaling as a key novel mediator of Ang II-induced UB branching morphogenesis (Song, MÖD, 2010). Here, we tested the hypothesis that Ang II induces transcriptional activation of Ret.

Methods Used: To determine whether Ang II regulates Ret gene transcription in UB tip cells, E12.5 kidneys of RetGFP+ knock-in mice, which express GFP under control of Ret promoter, were grown ex vivo and treated or not with Ang II (10-5 M) for 48 hours. GFP fluorescence was analysed with SlideBook4.1 software. The effect of Ang II on luciferase reporter activity from the human Ret promoter construct (-1545 to -5270, AF032124, Genebank) was examined in UB cells in vitro. Protein lysates were analyzed for Luciferase activity using Beckman LD400 plate reader. The data were factored per β-galactosidase activity and protein concentration.

Summary of Results: GFP fluorescence intensity was higher in Ang II-treated compared to control kidneys (2313±102 vs. 1992±53 pixels, p<0.05). Luciferase activity was increased in Ang II-treated compared to non-treated UB cells (2010±400 vs. 620±40 units, p<0.01).

Conclusions: We conclude that Ang II is a direct activator of Ret gene transcription in the UB epithelium. Future studies will identify transcription factors that mediate effects of Ang II on Ret gene expression.

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ANGIOTENSIN CONVERTING ENZYME TYPE 2 AND A DISINTEGRIN AND METALLOPROTEINASE 17 ACTIVITY IN HYPERTENSION AFTER TREATMENT WITH COMMON ANTIHYPERTENSIVE MEDICATIONS

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Purpose of Study: We previously reported that hypertension leads to a decrease in Angiotensin Converting Enzyme 2 (ACE2) expression and activity while A Disintegrin and Metalloproteinase 17 (ADAM17) is up-regulated in the central nervous system. The goal of the present study was to investigate the effect of anti-hypertensive medications on the activity of ACE2 and ADAM17 in the heart.

Methods Used: Hypertension was induced in mice, by uninephrectomy and proteinuria and slowed the renal failure while these effects were amplified in the PAR+LOS group. We conclude that PAR has renoprotective effects that are additive to those of LOS. Besides inhibiting RAS, PAR may affect other pathways including vasoactive and growth factors that are incriminated in CKD.

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A JAK/STAT PATHWAY MEDIATES ACTIVATED MACROPHAGE-INDUCED ANGIOTENSINII AUGMENTATION IN RENAL PROXIMAL TUBULAR CELLS

O’Leary R, Miyata K, Sato R. Tulane University School of Medicine, New Orleans, LA.

Purpose of Study: The development of angiotensin II (Ang II)-dependent hypertension presents with increased infiltration of macrophages (MΦ) into the kidney, leading to elevation of pro-inflammatory cytokines. Interleukin-6 (IL-6) seems to play an important role as IL-6 production is enhanced in activated MΦ and elevated IL-6 is found in Ang II-dependent hypertension. In vitro studies have shown that IL-6 contributes to the elevated expression of angiotensinogen (AGT) via activation of the JAK/STAT pathway in renal proximal tubule cells (PTC), which can activate the intrarenal renin-angiotensin system (RAS). These findings suggest that IL-6 produced by infiltrating MΦ leads to stimulation of intrarenal AGT and acceleration of the development of hypertension and renal injury. However, direct evidence for the contribution of JAK/STAT signaling to this process has not been demonstrated. This study was performed to demonstrate the importance of the activated JAK/STAT pathway in MΦ-IL-6 mediated AGT augmentation in PTC.

Methods Used: Cultured rat MΦ were treated with 0-10-5 M Ang II for up to 48 hr. Subsequently, PTC were incubated with the collected medium for periods of 30 min and 24 hr. STAT3 activity in the PTC was evaluated by measuring STAT3 phosphorylation levels using western blot analysis. AGT mRNA levels in PTC after 24 hr of treatment were evaluated by real-time RT-PCR.

Summary of Results: IL-6 expression levels were increased in Ang II-treated MΦ. Elevated levels of phosphorylated STAT3 (P-STAT3) protein (1.78 ± .09 ratio to control) and STAT3 nuclear trafficking were shown in PTC at 30 min. Addition of a neutralizing IL-6 antibody to the collected culture medium of Ang II-stimulated MΦ attenuated the increase in levels of P-STAT3. Additionally, AG490, a JAK2 inhibitor, also suppressed the elevation of P-STAT3 in PTC. AGT mRNA levels in PTC after 24 hr of treatment with MΦ medium were increased. The elevation of AGT mRNA was countered by addition of either an IL-6 neutralizing antibody or AG490.

Conclusions: These results establish an important role of JAK/STAT signaling in immune cell-IL-6 mediated AGT induction in the kidneys. AGT induction in PTC can stimulate intrarenal RAS and the development of associated disease.

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RISK FACTORS FOR DEATH FROM PERITONITIS IN HOSPITALIZED PERITONEAL DIALYSIS PATIENTS

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Purpose of Study: Peritonitis (PTN) in peritoneal dialysis (PD) patients is usually treated in an outpatient setting, but PTN patients with septic symptoms or severe abdominal pain may be admitted for management. We have shown that PTN in PD in-patients is associated with DM, bacteremia (BAC) and MRSA infection (Falk, J Invest Med 62:554, 2014). In the present work we queried the United States Renal Data System (USRDS) to assess risk factors for mortality in PD patients hospitalized for PTN.

Methods Used: All incident PD cases from the years 2005-2008 from the USRDS were included and defined according to dialysis modality on the most recent form. PTN and all comorbidities were defined by ICD9 diagnosis codes. Only PD at PTN diagnosis was included in this study. Survival analysis was performed using Cox regression.
Summary of Results: The analysis included 24,522 PD patients. PTN occurred in 3575 (14.6%). Demographics showed: 54% male, 72% Caucasian and 64% ≥ age 65 years. 27.9% of patients on PD died (39.4% of PTN, 25.9% of non-PTN). Clinical co-variates conferring the greatest risk of death (hazard ratio (HR), 95% confidence intervals (CI)) included decubitus ulcer (HR 3.74, CI 3.41, 4.09) and age ≥ 65 years (HR 3.36, CI 3.20, 3.52). Conditions with HR > 2.0 included cirrhosis, BAC, PVD, endocarditis, C. difficile colitis, BAC with SIRS, candidemia, and pancytopenia. DM and MRSA infection showed HR of 1.76 and 1.79, respectively. The most common covariates with significant HR for death included age ≥ 65 years (N=8,679, 35.4%), DM (N=7,550, 30.8%), PTN, BAC (N=2,197, 9.0%), PVD (N=1,993, 8.1%), and BAC w/SIRS (N=1,788, 7.2%). All other significant co-morbidities were present in < 3% of the patients.

Conclusions: PTN leading to a hospital admission occurred in nearly 15% of incident PD patients. Older age, DM and infectious complications are common morbidities with an increased risk of death. Less common conditions also confer an increased risk of death, but we would speculate they may also act as surrogate markers for disease severity. Aggressive treatment of PTN in older diabetics, with any suspicion of systemic infectious complications, is supported by this study.

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RISK FACTORS FOR CLOSTRIDIUM DIFFICILE INFECTION IN DIALYSIS PATIENTS

Tirath A 1, Colombo R 1, Baxer S 1,2, Huber L 1,2, Kintziger K 1, Kheda M 1, Nahman S 1,2, 1Georgia Regents University, Augusta, GA and 2Norwood VAMC, Augusta, GA.

Purpose of Study: Clostridium difficile (C diff) infection is a serious complication of broad-spectrum antibiotic use; however, other risk factors may also be present. Hemodialysis patients (HD) are often treated with large doses of broad-spectrum antibiotics due to the frequent occurrence of bacterial infections. Clostridium difficile infection is a common cause of nosocomial infection and infection in HD patients, and as such, is not an easily modifiable risk factor. On the other hand, there may be conditions that predispose to C diff that are independent of bacteraemia. To address this question, we assessed the occurrence of C diff infection in HD patients from the USRDS.

Methods Used: All incident HD cases from the USRDS for calendar years 2005-2008 were queried for a diagnosis of C diff infection. Survival and risk factors associated with C diff infection and mortality were also assessed. ICD-9 diagnosis codes were used to identify diagnoses. Cox regression models were used to determine the relative hazard of death associated with a C diff diagnosis compared to other known risk factors for death in ESRD patients. We used bivariate and multivariable models to assess these associations. Using backwards elimination of non-significant variables, a final model was obtained, and relative risks (RR), hazards ratios (HR) and 95% confidence intervals (CI) determined.

Summary of Results: For the 4-year period of study, C diff was identified in 17,853/419,875 (4.25%) patients. The incidence was >9% in patients with HIV, UTI, diverticulitis, GI bleeding, and osteomyelitis. In the adjusted model, the greatest relative risk (RR) of C diff was seen in patients with HIV (RR 2.68, 95% CI 2.39, 2.99), age 65 years or older (RR 1.76, CI 1.70, 1.81), bacteraemia (RR 1.74, CI 1.68, 1.80), and diabetes (RR 1.54, CI 1.49, 1.59). Risk factors associated with increased hazard of death included older age (HR 2.27, cirrhosis (HR 1.76), and HIV (HR 1.37), while GI hemorrhage, urosepsis, UTI, and Hepatitis C (HR 1.31-1.30) also significantly increased the rate of death (all p-values <0.05). Median survival time in C diff patients was 694.2 days vs. 488.60 in C diff negative patients.

Conclusions: C diff is a common comorbidity in ESRD patients, and identification of relevant risk factors for the infection and mortality may allow for management modifications for patients at risk.

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ALLOGENEIC KIDNEY TRANSPLANTATION IN YUCATAN MINIATURE SWINE AND YORKSHIRE PIGLETS: GENOTYPING AND OUTCOMES

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Purpose of Study: Yucatan miniature swine (YMS) are commonly used for experimental kidney transplantation due to extensive characterization of their histocompatibility antigens (swine leukocyte antigens (SLA)) that predict rejection. However, YMS are 5-10 fold more expensive vs outbred Yorkshire piglets (Yorks). Advances in SLA genotyping may allow for predicting rejection in Yorks. To test this question, we performed dual exchange allogeneic kidney transplantation (DEAK) in 4 pigs (2 YMS and 2 Yorks) and assessed rejection and its association with SLA genotypes.

Methods Used: Two YMs (Pig #9, SLA haplotype Lr-4/5.6, blood type A; and Pig #10: SLA Lr-5.4, A, and 2 Yorks (Pig #7: SLA Lr-6/12 22.15b, non-A; and Pig #8: SLA Lr-4/40.12, A) underwent DEAK. Species couples were operated simultaneously using isoflurane. For YMS Pig #9, the left kidney (LK9) was resected, ex vivo perfused (heparin/bicarb/manitol in LR), and stored in an ice bath. LK10 was then removed, perfused and stored on ice while LK9 was orthotopically transplanted into #10. Then LK10 was transplanted into #9. Both R kidneys were removed. The same procedure was performed with the Yorks. No immunosuppression was used, and animals were sacrificed on POD 2-10.

Summary of Results: Pigs were 30 kg females with mean pre-op creatinine levels of 1.05 mg/dL. For YMS, Pig #10 rejected LK9 and died on POD-6 (cret 19.6). Pig #9 accepted LK10 with creat 2.4 at sacrifice on day 9. For Yorks, Pig #7 showed hyperacute rejection of LK8 (POD-2, creat 12.1) and Pig #8 demonstrated vascular rejection of LK7 (POD-2, creat 4.4). For all pigs the kidneys appeared well perfused at sacrifice. For YMS, one SLA haplotype mismatch (Lr-6.7) led to rejection of LK9 by Pig #10, whereas a complete SLA haplotype match in the other direction of LK10 allowed graft acceptance by Pig #9. Both animals were blood type A. For Yorks, hyperacute rejection occurred in pig #7 due to blood type incompatibility. Vascular rejection in pig #8 resulted from a 2 class I haplotype-one class II haplotype mismatch.

Conclusions: SLA genotyping and blood typing predict rejection patterns in both YMS and Yorks following DEAK. Yorks may be a reasonable alternative to YMS for the study of experimental kidney transplantation.
A COMPARISON OF THREE INDUCTION THERAPIES ON PATIENTS WITH DELAYED GRAFT FUNCTION AFTER KIDNEY TRANSPLANTATION

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Purpose of Study: Delayed graft function (DGF) after kidney transplant is associated with high risk of rejection and inferior graft survival. The optimal induction therapy remains unknown. We compare the outcome of different induction with either methyl prednisolone, basiliximab or alemtuzumab in patients with DGF.

Methods Used: From 2006 to June 2013, 725 patients received a primary kidney transplant in our institute, and 212 (29%) had DGF that required dialysis. Among them, 58 received methyl prednisolone (group 1), 56 received basiliximab (group 2) and 98 received alemtuzumab (group 3).

Summary of Results: Protocol biopsies were performed and our maintenance consisted of tacrolimus and mycophenolate. Low-dose of steroids was also given to patients in group 1 and 2. The 1-year cumulative incidences of biopsy-confirmed and clinically-treated acute rejection were 27.4%, 19.8% and 10.1% in group 1, 2 and 3 respectively (p=0.007). The rejection rate was significantly lower in group 3 compared with group 1 (p=0.002) and group 2 (p=0.03%) than in group 1. It also trended lower in group 3 compared with group 2 (p=0.05). About 1/3 rejection episodes were diagnosed by protocol biopsy before DGF recovered. The 1-year graft survival were 90%, 96% and 100% in group 1, 2 and 3 (log rank p=0.006). Group 1 had an inferior graft survival than group 2 (p=0.03) and group 3 (p=0.002). The 1-year patients survival were not significantly different (96.6%, 98.2% and 100%, log rank p=0.81).

Conclusions: Our study supports the usage of antibody induction in patients with (the risk of) DGF, as it decreases the incidence of acute rejection and improves the graft survival than steroid induction alone.

PATHOGENESIS OF CONTRAST-INDUCED NEPHROPATHY IN DIABETICS AND EXPLORING NEW THERAPEUTICS

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Purpose of Study: Contrast media (CM)-induced nephropathy (CIN) is associated with significant clinical and economic consequences. However, an effective and reliable strategy to prevent CIN remains elusive. We studied aged diabetic (db/db) mice, as a novel model of CIN and explored renoprotective strategies using N-acetyl-L-cysteine (NAC) and other agents.

Methods Used: We first evaluated protective effects of various doses of NAC (1 mM - 6 mM) against contrast-toxicity in HK-2 cells exposed to iodoxanol (50 mg iodine/ml) for 24 hr. To evaluate the role of diabetes on the vulnerability to CIN, 24-wk-old db/db and non-diabetic (db/m) male mice (N = 5 - 10) were used. To create CIN, 24-wk-old db/db male mice (N = 3 - 5) were dehydrated for 24 hr and nonionic low-osmolar (iohexol) or isoosmolar (iodixanol) was injected via a jugular vein catheter at a dose of 3 g of iodine/kg bw. Treatment group received NAC (100 mg/kg bw) 1 hr before and 6 hr after iohexol injection. All mice were sacrificed 24 hr after CM injection.

Summary of Results: NAC at 2 mM dose was significantly more effective than other doses against iodixanol toxicity in HK-2 cells. 24-wk-old db/db mice became obese, polyuric, developed glucosuria, ketonuria and had increased GFR, systolic blood pressure, urine NGAL, and renal tubular damage compared to db/m mice. 24 hr after iohexol injection, db/db mice showed significant decrease in GFR and significant increases in serum creatinine, urine and kidney KIM-1 levels, and kidney damage histologically compared to control db/db mice. After iohexol administration, the mRNA levels of immune system markers CD11b, CD68 and TLR2 were significantly upregulated in kidney. Iohexol also caused metabolic acidosis in db/db mice as evidenced by decreased levels of blood HCO3 and total CO2. Treatment with NAC showed an improvement in reducing kidney pathology.

Conclusions: Elderly db/db mice develop spontaneous nephropathy, hypertension, and are prone to CIN. Innate immunity mediated by TLR2 play an important role in CIN. NAC with other antioxidant and anti-inflammatory agents could be a reliable preventive strategy in elderly diabetics.