and all protocol/indication liver biopsies were reviewed and graded for steatosis (0–3), inflammatory (NASH) activity (0–3), and fibrosis (0–4) by a single pathologist blinded to clinical data. **Results:** *Clinical Data:* LT for CC was performed for 90 of 832 (11%) adult LT pts, mean age 53 ± 12 yrs (range 21–69 yrs), 54% men, and follow-up of 81 ± 62 mo (range 0–170 mo). At LT, the prevalence of hepatoma was 4% and mean calculated MELD score was 16.4 ± 6.2 (range 6–33). *Survival:* One and five-yr pt survival were 88% and 85% respectively. Repeat LT was required for 4 of 90 (4%). Cardiovascular disease (CVD) deaths occurred in 11 of 27 (41%) pts, 5 (55%) of which were within one yr of LT. *Metabolic Parameters:* IR was noted in 52% of pts pre-LT. During follow-up, no change was noted in weight, BMI, or glycemic control when compared to pre-LT values (p = ns). During the follow-up period, triglycerides, total and LDL cholesterol increased ($p < 0.01$) and HDL cholesterol declined ($p < 0.01$). *Histology:* At LT, grade 1 and 2 steatosis were noted in 39% and 6% respectively. At 1, 3, and 5 yrs post-LT, steatosis grade ≥ 1 was noted in 39%, 76%, and 89%; steatosis ≥ grade 2 was noted in 11%, 69% and 71% of pts respectively. Mean steatosis grade was 0.91 ± 0.9, 0.96 ± 0.87, and 1.46 ± 0.96 ($p < 0.01$). Mild NASH was observed in 1/3 of pts and did not increase over time. Comparable NASH grade was 1.06 ± 0.59, 0.68 ± 0.71, and 0.66 ± 0.89 ($p < 0.05$), and fibrosis from 1.09 ± 1.15, 1.0 ± 0.97, and 1.5 ± 1.3 ($p <$ ns). One pt developed cirrhosis by 5 yr post-LT. **Conclusions:** NAFLD is a common sequela for pts undergoing LT for CC. Progression to advanced fibrosis was uncommon in this cohort and survival is excellent. IR is present in a majority of patients with CC with CVD is the leading cause of mortality post LT. Hyperlipidemia primarily correlates with recurrent NAFLD post-LT.

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ASSOCIATION OF CD4 COUNTS WITH TETANUS ANTIBODY TITERS AMONG HIV-INFECTED PATIENTS. K. Alagappan, B. Donohue, C. Hoey, S. Geboff, M.H. Kaplan, J. Cervia, R. Silverman, Long Island Jewish Medical Center, North Shore-LIJ Health System, New Hyde Park, NY.

Background: Approximately 90% of the US population has protective levels of anti-tetanus-antibody (ATA). Elderly individuals demonstrate lower protective levels of ATA compared with the general population; decreased immune system function has been postulated. It is unknown if human immunodeficiency virus (HIV) infection is also associated with non-protective ATA. **Objective:** Determine if there is an association between CD4 counts and ATA in HIV-infected patients. **Methods:** A convenience sample of outpatients from 2 HIV specialty clinics was recruited from May 2000 until July 2004. ATA was measured by ELISA, and a level greater than 0.15 IU is considered protective. CD4 counts were measured by flow cytometry within 60 days of ATA testing. The lowest CD4 count documented in the clinic record was also documented (nadir CD4). For analysis, a CD4 count of < 100 cells/μL was considered severe illness. Data were analyzed with standard parametric and non-parametric testing. Logistic regression models were developed to test association with tetanus protection. **Results:** 262 subjects were included with a mean age of 43 years (range 20 to 67 years). A total of 227/262 (87%) had protective ATA. The CD4 count for patients with protective ATA was 459 cells/μL compared to 353 cells/μL for nonprotective ATA ($p = 0.055$). When the nadir CD4 count was analyzed, patients with protective ATA had 208 cells/μL and nonprotective had 128 cells/μL ($p = 0.006$). Those with nadir CD4 counts < 100 cells/μL were less likely to have protective ATA (86/106, 81%) than those with nadir counts ≥ 100 cells/μL (141/156, 90%, $p = 0.03$). After adjusting for factors that may influence ATA, including country of birth, logistic regression models found an association between lower CD4 and less tetanus protection trending toward significance for contemporaneous CD4 counts ($p = 0.06$). The adjusted association between ATA and CD4 was significant when nadir CD4 counts were analyzed ($p = 0.01$). **Discussion:** HIV-infected individuals have generally similar protection to tetanus as the general US population. However, among HIV patients, a history of lower CD4 counts is more often associated with non-protective tetanus titers. This suggests that in some patients the memory for this antigen is lost, particularly when CD4 counts drop below 100 cells/μL. **Conclusion:** Clinicians must be aware that HIV-infected patients with low CD4 counts may be at greater risk for non-protective ATA. Support received from North Shore-LIJ Research Institute, GCRC, Grant # M01 RR018535.

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NONSURGICAL TREATMENT OF DUPUYTREN'S DISEASE. M.A. Badalamente, L.C. Hurst, Department of Orthopaedics, State University of NY at Stony Brook, NY.

Dupuytren's disease is a fixed flexion contracture deformity of the hand currently treated by surgical fasciectomy of the collagenous cords responsible for the finger joint contractures. Our prior Phase 2 controlled, clinical trials have clearly indicated safety and efficacy of collagenase injection therapy as an alternative to surgery. The purpose of this Phase 3 study was to further test collagenase injection as a nonsurgical treatment for Dupuytren's disease. **Methods:** 35 patients were enrolled in the random, placebo, double-blind control protocol allowing for an assignment to 0.58 mg collagenase or saline placebo, maximum three injections. At this time, the control treatment code may not be unmasked. However, patients had the option to enroll in an additional protocol either for treatment failure or because they had additional contracted joints of the same or opposite hand. The second protocol allowed for a maximum five open label, 0.58 mg, collagenase injections, given at one month intervals. Seventeen patients were treated in the open label protocol, 12 male and 5 female, mean age 62 years, 12 metacarpophalangeal (MP) joints and 16 proximal interphalangeal (PIP) joints. The initial mean MP and PIP joint contractures were 45° and 43°, respectively. Collagenase injections, 0.58 mg, were to the collagenous cords, using an insulin syringe, 0.25 mL for MP joints and 0.20 mL for PIP joints. Patients were seen in serial follow up at 1, 7, 14 and 30 days, and 2, 3, 6, 9 and 12 months. **Results:** 92% of the MP joints and 75% of the PIP joints treated achieved full, normal finger extension (0°), most within one week of one collagenase injection. Results were sustained in the longer term, mean follow-up, 125 days for MP joints and 107 days for PIP joints. There was no loss of normal finger flexion, or normal grip strength. Adverse events included pain associated with "cord rupture," hand edema and palmar ecchymosis. These resolved without problems in 7–14 days. **Conclusions:** Collagenase injection therapy for Dupuytren's disease continues to show clear merit in Phase 3 clinical trials as a safe and effective alternative to surgical fasciectomy. National, multicenter, Phase 3 study of this promising method will begin in the next several months. This study was supported by the FDA (Grant 001437), the NIH (GCRC-M01RR10710) and the Biospecifics Technologies Corp.

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MEDICATION USE AND CLINICAL OUTCOMES FROM PREADMISSION TREATMENT THROUGH ONE-YEAR FOLLOW-UP FOR CHILD PSYCHIATRIC INPATIENTS. J.C. Blader, Stony Brook State University of New York, School of Medicine, NY.

Background: Recent clinical research has established an evidentiary basis for the pharmacotherapy of several psychiatric disorders of childhood. However, children hospitalized for psychiatric disorders seldom experience robust response to monotherapy with the agents studied, and consequently receive polytherapy involving medication combinations that lack data to support their efficacy. Longitudinal, observational data on medication use and outcomes for this patient group may furnish information useful to the development of controlled research on the potential effects of medication combinations. **Objectives:** 1) To assess the utilization of specific pharmacotherapy strategies for child psychiatric inpatients from pre-admission care through 12 months after discharge. 2) To examine associations between these strategies and postdischarge outcomes. **Method:** Prospective follow-up of 83 5–13 year-old children admitted to acute inpatient care for aggressive behavior in the context of a disruptive behavior disorder. Treatment and symptom severity data were obtained via standardized rating scales at admission, discharge, and 3, 6 and 12 months after discharge. **Results:** *Utilization:* Number of concurrent medications increased over assessment times. Changes in children's pharmacotherapy occurred most frequently during hospitalization and 3 months after discharge, but even between postdischarge assessments over half experienced treatment changes, most often initiation of new agents. Treatment with antipsychotics and mood stabilizers increased over assessment times while SSRI treatment decreased. *Outcomes:* Children treated with stimulants and risperidone 3 months after discharge had significantly improved ratings, adjusted for admission scores and concurrent medications; improvements were not apparent at 6- and 12-month follow-ups. Children treated with SSRIs at 6 months postdischarge had higher problem severity ratings. Those who maintained lithium and SSRI treatment between 6 and 12 months displayed improvements. Medication interactions suggested better outcomes with combined risperidone and lithium at 12 months and less advantageous outcomes for children treated with alpha-agonists unaccompanied by stimulants. **Conclusions:** Complexity of pharmacotherapy for child inpatients ratchets upwards from admission through one year after discharge. Hospital-initiated treatment is commonly altered soon after discharge and throughout follow-up. Within the limitations of observational methodology, postdischarge outcomes seem related to specific pharmacotherapy regimens, some of which may improve children's functioning while others may worsen it.

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UNIVERSITY OF FLORIDA GENERAL CLINICAL RESEARCH CENTER DNA BANK. M. Brantly, T. Mathews, W. Hyde, D.W. Theriaque, C. Wang, University of Florida, Gainesville, FL.

Introduction: Genetic studies depend on well-characterized phenotypes to establish relationships between disease and genetic mutations. Researchers typically study and collect DNA from a single group of subjects and may not have ready access to other study populations that may be excellent controls for their study population. The University of Florida General Clinical Research Center (UFGCRC) evaluated approximately 1000 new study subjects in 2003–2004. To allow GCRC researchers to expand their gene-based studies beyond their own study groups the GCRC established a DNA bank that enrolled subjects admitted to the GCRC inpatient and outpatient facilities. **Methods:** Following consent, up to 20 mL of whole blood was obtained and the attending physician designated up to 10 ICD-9 codes that best characterized the study subject. A secure database, reporting and tracking system was developed using MySQL/PHP and Freezerworks 1.0.4 (DataWorks Development, Mountlake Terrace, WA). Demographic information was collected from all study subjects. DNA was extracted from whole blood using a Gentra Puregene DNA extraction kit (Minneapolis, MN). DNA concentration and purity were established by spectrometry at UV wavelengths of 260 and 280 nm. The quality of extracted DNA was further determined by demonstrating that the alpha-1-antitrypsin gene could be amplified using Taq polymerase (Promega, Madison, WI). **Results:** To date 324 subjects have consented to participate in the GCRC DNA bank. Twenty-two percent of the subjects are male. Ethnicity-race percentages are 71.3% Caucasian, 21% African-American, 6.5% Hispanic and 1.6% are Asian. Age distribution is 22.8% are > 50 years old, 23.5% age 36–50 years, 45.7% age 20–35 years and 8% are less that 20 years old. Disease categories included 36% normal, 18.2% endocrine, 13.3% cardiovascular, 10.5% gastrointestinal, 9.3% genital-urinary, 4.9% respiratory, 7.7% infectious disease, 33.3% rheumatology and 14.2% other diseases. The mean DNA concentration is 306 ± 208 μg/mL (n = 212), DNA purity 1.98 ± 0.07 (A260/A280), mean amount of DNA/subject, 839 μg. The alpha-1-antitrypsin gene was successfully amplified in 99% to the subjects evaluated to date. **Conclusion:** The GCRC has established a new resource for GCRC investigators to utilize for the purpose extending their gene-based research. Access to the sample collection is through a GCRC Advisory subgroup panel, which upon approval of a brief application provides the investigator with normal and disease specific DNA samples.

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ASSOCIATION IN BREAST AND COLON CANCER SCREENING BEHAVIOR IN WOMEN. R. Carlos, Department of Radiology, University of Michigan Medical Center, Ann Arbor, MI.

Purpose: Gender-based psychosocial factors appear to influence colorectal cancer (CRC) screening adherence. Given its near universal acceptance by the public, screening mammography represents a potential "teachable moment" for educating patients about the risk of CRC. Accordingly, to better understand screening behaviors among women, data from the Behavioral Risk Factors Surveillance Survey (BRFSS) were analyzed to identify potential relationships that would allow interventions to enhance CRC screening. **Methods:** Women 50 years and older who participated in the BRFSS 2001 survey were included in the analysis. CRC, breast and cervical cancer screening adherence with American Cancer Society guidelines was determined. Breast and cervical cancer screening adherence and general health and demographic characteristics were used as predictors of CRC screening adherence. **Results:** After adjusting for sociodemographic factors in a multivariate analysis, women 60–69 years old (adjusted OR 1.50, $p < 0.01$) and 70–79 years old (adjusted OR 1.39, $p < 0.01$), having achieved at least some high school (adjusted OR 1.62, $p < 0.01$) or college (adjusted OR 2.11, $p < 0.01$) education, having health coverage (adjusted OR 1.67, $p < 0.01$) or a personal physician (adjusted OR 1.60, $p < 0.01$), and adherence to screening mammography (adjusted OR 2.42, $p < 0.01$) and Pap smear (adjusted OR 1.70, $p < 0.01$) independently increased the likelihood of CRC screening adherence. Women in self-reported good general health were less likely to adhere to CRC screening guidelines (adjusted OR

0.79, p < 0.01). Current smokers were also less likely to adhere to CRC screening guidelines compared to women who never smoked or formerly smoked (adjusted OR 0.76, p < 0.01). Participants who adhered to both mammography and Pap smear guidelines were significantly more likely to adhere to CRC screening (51.5% CRC screening adherence) when compared to women who adhered to neither screening test (8.2% CRC screening adherence), with an adjusted OR 5.67 (p < 0.001). Participants who adhered to both mammography and Pap smear guidelines were significantly more likely to adhere to CRC screening compared to women who adhered to either screening test (38.0% CRC screening adherence) with an adjusted OR 1.94 (p < 0.001). **Conclusions:** Women with up-to-date mammography and cervical cancer screening were more likely to be up-to-date with CRC screening. Regardless of the increased association between non-CRC-related cancer screening and CRC screening, rates of CRC screening utilization remained low in these otherwise compliant populations.

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CRITICAL INJURIES ARE HIGHER IN UNRESTRAINED THAN RESTRAINED PEDIATRIC VICTIMS OF MOTOR VEHICLE COLLISION. L. Chan, K. Reilly, J. Telfer, University of Arizona, Tucson, AZ.

Purpose: Despite public efforts encouraging use of child safety restraints, our emergency department (ED) continues to evaluate children involved in motor vehicle collisions (MVC) who are unrestrained. To emphasize the need for further educational and legislative measures to convince guardians to use proper safety restraints, we conducted a study to compare injuries, death, and financial cost between unrestrained and restrained pediatric victims of MVC and also to determine the compliance of safety restraint use in this population. **Methods:** This retrospective chart review compared restrained children (RC) to unrestrained children (URC) (< 14 years old) of MVC who presented to our ED from 1/1/3–12/31/3. Our ED is in an urban, teaching hospital with an annual 68K census, which is racially, ethnically, and socioeconomically diverse. Our level 1 trauma center encompasses a catchment area > 40K square miles with a 1.3 million population. Charts were excluded if use or nonuse of safety restraint was not documented. The two groups were compared for age, gender, medical charges, hospital duration, injuries (intraabdominal, intrathoracic, intracranial and fractures), admission, surgery, transfusion of blood products, and intubation. Use of restraint was reported as a percentage. Chi-square was used for analysis of gender and t-test with 95% CIs for age, medical charges, and duration of admission. Odds ratios and 95% CIs were calculated for URC with respect to RC for dichotomous data. All statistical tests were two tailed using a significance level of 0.05. **Results:** 353 patients were identified. 17 (4.8%) lacked documentation on car restraint use. Of the remaining 336, 255 (76%) were RC and 81 (24%) were URC. The URC group was statistically older (8.86 years, CI: 7.94, 9.78) than the RC (6.95 years, CI: 6.42, 7.48). Length of hospital stay of 1.94 days (CI: 0.75, 3.12) for URC was significantly longer than RC, 0.10 days (CI: 0.02, 0.21). Cost of care for URC ($14,754, CI: $7676, $21,831) was significantly higher than RC ($1996, CI: $1207, $2786). Odds of admission was 14.48 times (CI: 5.91, 38.63) higher in URC than RC. Odds of serious injuries were higher in URC: intraabdominal [OR = 20.16 (CI: 2.36, 930.68)], intrathoracic [OR = 13.09 (CI: 1.26, 647.05)], and fractures [OR = 5.85, CI: 2.13, 16.89]. Nine (11.11%) URC had an intracranial bleed versus no RC. The URC group had higher need for surgery [OR = 13.09 (CI: 3.30, 74.33)] and transfusion [OR = 27.61 (CI: 3.56, 1229.85)]. Ten URC (12.35%) required intubation versus no RC. The only two mortalities were URC. **Conclusion:** Serious injuries and cost of care were significantly higher in unrestrained than restrained victims of MVC. In this population, there was room for improvement in terms of compliance with child safety restraint.

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CAUSES OF DEATH IN ADULT SICKLE CELL DISEASE PATIENTS AT HOWARD UNIVERSITY. D.S. Darbari, J. Kwagyan, S. Rana, V.R. Gordeuk, O. Castro, Center for Sickle Cell Disease and Department of Pediatrics, Howard University, Washington, DC.

Purpose of Study: To analyze causes of death among adults with sickle cell disease (SCD) at a single institution over a 25 year period. **Methods:** A single physician (OC) recorded causes of death among 141 adult SCD patients he treated and knew well from 1976 to 2001. Causes of death were determined by autopsy report and/or clinical assessment. **Results:** One hundred fourteen (80.9%) of the patients had SS phenotype and 66 (46.8%) were female. The mean ± SD age at death was 36 ± 11 years with a range of 18 to 80 years. The leading causes of death were pulmonary hypertension (PHT) (n = 37; 26.2%), sudden death (n = 33; 23.4%), renal failure (n = 32; 22.7%), sepsis (n = 26; 18.4%), thrombo-embolism (n = 21; 14.9%) and cirrhosis (n = 16; 11.4%). Most patients had more than one diagnosis contributing to death. When causes of death that occurred after 1990 (n = 80) were compared to those that occurred in 1990 or earlier (n = 61), fat embolism was significantly lower (2.5% vs 13.1%; p = 0.02) while the categories of sepsis (25.0% vs 9.8%; p = 0.028) and PHT (38.8% vs 9.8%; p < 0.001) were significantly higher after 1990. When associations among various causes of death were explored, significant relations were found between PHT and thromboembolism and between cirrhosis and iron overload. Thrombo-embolism contributed to death in 8 (7.7%) of non PHT patients versus 13 (35.1%) of the PHT group (p < 0.001). Iron overload contributed to death in 3 (2.4%) of the non-cirrhosis patients versus 7 (43.8%) of the cirrhosis group (p < 0.001). Renal failure as a cause of death increased significantly with age (13.5% in patients 30 years and lower vs. 47.4% in patients over 40 years of age; p < 0.001) **Conclusions:** Life expectancy in SCD remains low. The increased diagnosis of pulmonary hypertension as a cause of death since 1990 may be due to increased awareness of this condition and also to SCD individuals with severe disease living long enough to develop this complication. The decreased diagnosis of fat embolism as a cause of death in recent years may be explained by early and aggressive management of acute chest syndrome (ACS) with transfusions, for fat embolism often complicates ACS. The associations of renal disease as a cause of death with increasing age and the finding of a high convergence of iron overload and cirrhosis as causes of death are consistent with current clinical understanding of these conditions. Whether some of the sudden unexplained deaths are due to unrecognized pulmonary hypertension should be investigated in future studies.

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RANDOMIZED CONTROLLED TRIAL OF ABLATION FOR BARRETT'S ESOPHAGUS. G. Dulai, D. Jensen, G. Cortina, A. Ippoliti, David Geffen School of Medicine at UCLA, Los Angeles, CA.

Background: Endoscopic ablation of Barrett's esophagus has been described using a combination of high dose proton pump inhibitors and a variety of modalities for thermocoagulation. Randomized comparisons of ablation strategies have not been published. **Methods:** Referred subjects were screened to identify those with 2–7 cm of Barrett's esophagus without high grade dysplasia or cancer. Included subjects received pantoprazole 40 mg twice daily, followed by randomization to treatment with argon plasma coagulation (APC) versus multipolar electrocoagulation (MPEC). The primary outcome measure was number of treatment sessions to endoscopic ablation. **Results:** 235 subjects were screened, and 52 randomized. The mean length of Barrett's esophagus was 3.1 cm in MPEC versus 4.0 cm in APC (p = 0.03), but treatment groups were otherwise similar with regard to baseline characteristics. The mean number of treatment sessions to endoscopic ablation was 2.9 for MPEC versus 3.8 for APC (p = 0.04) in an intention to treat analysis, though p = 0.249 after adjustment for difference in Barrett's esophagus length. The proportions achieving ablation above the gastroesophageal junction endoscopically were 88% MPEC versus 81% APC (p = 0.68) and histologically 81% MPEC versus 65% APC (p = 0.21). The mean time per 1st treatment session was 6 minutes with MPEC versus 10 minutes with APC (p = 0.01) in per protocol analysis. There were no serious adverse events, but transient moderate to severe upper gastrointestinal symptoms occurred post-MPEC in 8% versus 13% post-APC (p = 0.64). **Conclusions:** Although there were no statistically significant differences, ablation of Barrett's esophagus with pantoprazole and MPEC resulted in numerically fewer treatment sessions and a greater proportion of subjects achieving endoscopic and histologic ablation than treatment with pantoprazole and APC.

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THE ARG16/GLY β_2-ADRENERGIC RECEPTOR POLYMORPHISM ALTERS THE CARDIAC OUTPUT RESPONSE TO ISOMETRIC EXERCISE. J.H. Eisenach, T.L. Pike, R. Chettiar, A.M. Oviedo, L.A. Sokolnicki, S. Masuki, K.H. Rehfeldt, M.J. Joyner, Mayo Clinic College of Medicine, Rochester, MN.

Our laboratory has demonstrated that healthy, normotensive adults homozygous for glycine (Gly) of the Arg16/Gly β_2-adrenergic receptor polymorphism produce 1) greater forearm β_2-receptor mediated vasodilation and 2) a higher heart rate (HR) response to isometric handgrip than arginine (Arg) homozygotes. To test the hypothesis that the higher HR response in Gly16 subjects serves to maintain the pressor response (increased cardiac output, CO) in the setting of augmented peripheral vasodilation to endogenous catecholamines, we measured continuous HR (ECG), arterial pressure (AP, Finapres), and CO (transthoracic echocardiography) during isometric, 40% submaximal handgrip to fatigue in healthy subjects homozygous for Gly (n = 25; mean age ± SE: 30 ± 1, 9 women) and Arg (n = 14, age 29 ± 1, 8 women). Resting data were similar between groups. Handgrip produced similar increases in AP, venous norepinephrine and epinephrine concentrations; however, HR increased greater in the Gly group (62 ± 5% increase from baseline vs. 44 ± 4%, $p < 0.05$) which correlated with a higher CO (Gly: 7.8 ± 0.4 L/m vs. Arg: 6.6 ± 0.3, $p < 0.05$) and a tendency toward a lower systemic vascular resistance. We conclude that Gly16 homozygotes generate a higher CO to maintain the pressor response to handgrip, in part consistent with augmented peripheral vasodilation. Supported by HL-63328, GCRC RR-00585, NCRR K23–17520.

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ASCERTAINING THE PARENTAL PERSPECTIVE OF CARING FOR A CHILD WITH BILIARY ATRESIA. J. Erlichman,* J. Arvay-Nezu,* J. Shea,+ B.A. Haber,* *The Children's Hospital of Philadelphia, Philadelphia, PA; +The University of Pennsylvania School of Medicine, Philadelphia, PA.

Purpose of Study: Ascertaining the parental perspective of caring for a chronically ill child provides valuable insights for clinicians striving to improve patient/family satisfaction, coping, and compliance. The purpose of the present study was to understand the parental impact of caring for a child with biliary atresia (BA). **Methods:** Families were recruited from a registry of BA patients seen at a single center. Three focus groups were held in summer and fall of 2003 with the parents of children with BA. A total of 18 parents of 12 children with BA participated in the three group sessions. Within 48 hours of each session the investigators met as a team to aggregate the data. **Study Design:** Through content analysis, statements generated during the focus group sessions were indexed and categorized. Four themes emerged from the three focus group sessions: emotional stress, need for social support, lack of education, and financial burden. Emotional stress was exemplified through feelings of "guilt, anger, helplessness, fear of the future, worry." The ideal social support structure was comprised of four elements, though not all received support from all four sources: support from the hospital staff, friends, family, and co-workers. Every parent wanted to know the cause and course of BA and many felt frustrated by this lack of information. The financial impact of BA was evidenced in many ways. Some parents lost employment, stopped working, or switched jobs as a result of their child's condition. These consistently reiterated themes illustrate the parental impact of caring for a child with BA. **Conclusions:** Focus group sessions such as those discussed in this abstract delineate disease-specific issues that need to be incorporated into the comprehensive medical model when dealing with the child with BA. As has been shown for other disease models, meeting these parental needs will optimize patient health care delivery; improve compliance, coping, and overall family satisfaction with the patient care experience.

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VARIATIONS IN *MSX1* AND *PAX9* ARE ASSOCIATED WITH HUMAN TOOTH AGENESIS. S.A. Frazier-Bowers, M.A. Torain, J.T. Wright, University of North Carolina, Chapel Hill, NC.

Congenitally missing teeth (tooth agenesis) represents the most common craniofacial anomaly in man, affecting up to 20% of the population worldwide. Tooth agenesis is a clinically heterogeneous disorder affecting specific tooth types at different rates. Recent evidence also confirms the role of genetic heterogeneity in this common dental anomaly, with two genes, *MSX1* and *PAX9*, contributing to the majority of cases published. Previous studies have associated mutations in *PAX9* with molar oligodontia, while others have associated mutations in *MSX1* primarily with premolar hypodontia. Most of these studies have focused on individuals who are otherwise normal and affected with this disorder. **Objective:** Our objective was therefore to determine if alterations in *MSX1* and *PAX9* might also be responsible for tooth agenesis in families also affected with dental syndromes including tricho dento osseous syndrome (TDO) or amelogenesis imperfecta (AI). **Methods:** To test whether mutations in *MSX1* and *PAX9* are responsible for cases of tooth agenesis occurring with specific dental anomalies, we used direct sequencing of PCR products. Clinical examination, pedigree analysis, followed by PCR-based mutational analysis of *MSX1* was carried