IRISH & AMERICAN PAEDIATRIC SOCIETY MEETING

45th Annual Meeting

SEPTEMBER 25 – 29, 2013
CHARLESTON MARRIOTT HOTEL,
CHARLESTON, SOUTH CAROLINA, USA
**Irish and American Paediatric Society Meeting**  
**September 25 – 29, 2013  
Charleston, South Carolina  USA**  
*Chairs Rita M. Ryan and Jimmy McElligott*

### Scientific Sessions

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<td>Thursday, October 1, 2009</td>
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<td>Charleston Marriott Hotel</td>
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<td>Friday, October 2, 2009</td>
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<td>Saturday, October 3, 2009</td>
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**Thursday, September 26, 2013 – at the Marriott**

7:45 Welcome and Opening of Meeting – President Dean Wilcox

**Session I – Moderator: Tom Clarke, MD, General Pediatrics**

8:00 Alan Macken *Travel award winner*  
Ireland  
Endothelial Dysfunction Studies in Children Whose Fathers had Premature Cardiovascular Disease

8:15 Tony Ryan  
Cork Univ Maternity Hosp  
A Longitudinal Study of the Duration of Breastfeeding in Term and Preterm Infants

8:30 Katie King *Travel award winner*  
MUSC  
Academics and Physical Activity in Title One Elementary Schools: Changing What We Can

8:45 Michael A. Mandarano  
U Buffalo  
Morganella Morganii an unusual case of a sore throat in a 2 year old girl

9:00 Orla Walsh *Travel award winner*  
Univ Hospital, Galway, Ireland  
Perceptions of Diabetes Patients Their Parents, And Healthcare Professionals On Diabetes Transition In The West Of Ireland: The Mater Study

9:15 Parnell Donahue  
Brentwood, Tennessee  
Parenting, Pediatricians, and Our Changing Culture

9:30 **Thomas Cone Founders Lecture:**  
*David Bundy, MD, MUSC, Vice Chair for Pediatric Quality and Safety*  
**“Adviser him we don’t have fuel’ Learning from Avianca Flight 52”**

10:15 break

**Session II – Moderator: Gene Dempsey, MD, Neonatology, University College, Cork, Ireland**

10:45 Jim Kiger  
MUSC  
When are preemies “out of the woods”? Changing patterns of infant mortality in a tertiary care NICU

11:00 Jack Widness  
U Iowa  
Post-Transfusion Red Cell Survival of Neonatal Autologous and Adult Donor Biotin Labeled RBCs, Measured Concurrently, Are Not Different
11:15 Tony Ryan  
Univ College, Cork, Ireland  
Helping Babies Breathe (HBB) Sudan: Training of Trainers: Workshop Evaluation

11:30 John Romond  
U Kentucky  
Variant Tricuspid Atresia: A Circulatory Dilemma

11:45 Grace Dukes  
Travel award winner  
U Kentucky  
Pediatric Inflammatory Myofibroblastic Tumor of the Heart: A Case Report with Sudden Cardiac Death and Review of the Literature

Friday, September 27, 2013 — at Medical University of South Carolina (MUSC)

7:00 Leave for Medical University of South Carolina

8:00  
**William Kidney Founders Lecture:**  
*Eleanor Molloy, MD, National Maternity Hospital, Dublin, Ireland*  
"Inflammation in Neonatal Encephalopathy: a Possible Therapeutic Target?" (Pediatric Grand Rounds)

Session III – Moderator: Bill Basco, MD, MUSC, General Pediatrics

9:00 Evan Allie  
(resident)  
MUSC  
Outcome Analysis For Two Treatment Options in Acute Pediatric Coin Ingestion: Surgical Management Versus Bougieange

9:15 Brendan Gerard Loftus  
School of Medicine, NUI Galway, Ireland  
Acute Childhood Asthma in Galway City, West Of Ireland From 1985-2005: Relationship To Air Pollution And Climate

9:30 Nuala Quinn  
Travel award winner  
Children’s Univ Hosp, Dublin  
A Historical Perspective Of Pediatric Publications – A Bibliometric Analysis

9:45 Brielle Weinstein  
(medical student)  
MUSC  
Pediatric Metabolic Syndrome Study: Correlations Between Simple Diagnostics And Symptoms Of Early Diabetes

10:00 Colby Day  
(resident)  
MUSC  
Resident Comfort and Adherence to Best Practice Guidelines for Pediatric Well Visits

10:15 Break and **Poster Session:**

Sarah Adams  
MUSC  
The Initiation of Inhaled Corticosteroids in the Emergency Department

Chike Onwuneme  
Cumulative Dietary and Supplemental Vitamin D Intake in Preterm Infants: how much Vitamin D is enough?

Nuala Quinn  
Travel award winner  
Our Lady’s Children’s Hospital Crumlin, Dublin  
Childhood SLE And Sweet’s Neutrophilic Dermatosis – A Rare Association
Meaghan Flessa MUSC Conserved Regulation of the Tinman Gene Nkx2.5 During Cardiac Outflow Tract Development: A Window into Novel Congenital Heart Disease Entities.

Colby Day MUSC Implementation and Revision of Communication Skills Curriculum in a Pediatric Residency Program: PediComm

K Brokus St, Munchin’s Regional Maternity Hospital, Limerick, Ireland Translocation of gene 9p,18q in a Neonate with Central Diabetes Insipidus

Tom Hulsey MUSC Regionalized Perinatal Care In The South Carolina Lowcountry Perinatal Region: 1998 – 2011

Session IV – Moderator:  Ward Rice, MD, Neonatology, Cincinnati Children’s Medical Center

10:45 Daryl Peter Butler Travel award winner Univ College Cork, Ireland SKIPI-SKIN PUNCTURES IN PREMATURE INFANTS

11:00 Aisling Smith Travel award winner Univ College Cork, Ireland Echocardiography Parameters Predicting Ductus Arteriosus Patency at One Month in Pre-Term Infants

11:15 Gene Dempsey Univ College Cork, Ireland Neonatal Outcomes in Elective Caesarean Sections at Term

11:30 Talia Glasberg MUSC Incidence of Necrotizing Enterocolitis (NEC) with Recurrent Antibiotic Use in Very Low Birth Weight (VLBW) Infants

11:45 Heather Smith Travel award winner Cinti Children’s Intestinal Microbial Colonization Differs Prior to Diagnosis of Bronchopulmonary Dysplasia

Saturday, September 28, 2013 – at the Marriott

7:45 Business meeting

Session V – Moderator:  Jimmy McElligott, MD, General Pediatrics

8:15 Donald Ian Macdonald Natalie K. Wallis Travel award winner U Kentucky Progeria - Old Disease, New Insights

8:45 Carrisann Woods Geisinger Medical Center, Danville, Pennsylvania, USA Familial Translocation T(11;13)(P15.4;Q32.1) Resulting In Partial Trisomy 11p And Monosomy 13q : Report of a New Case and Review of the Literature

9:00 Elaina Ann Pirruccello Travel award winner U Kentucky Rare Cause of Pediatric Acute Abdomen, Cured by Surgical Resection: A Case Report of Bleeding Into a Retroperitoneal Lymphangioma

9:15 Jan van Eys Vanderbilt U A Brief History Of Christmas Disease
Fred Burke Founders Lecture:
Carol Wagner, MD, Professor of Pediatrics, MUSC
“Vitamin D in Pregnancy and the Neonate”

10:00 Break

Session VI – Moderator: Eleanor Molloy, MD, Neonatology, University College, Dublin

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<td>David Healy</td>
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<td>Structured Promotion of Breast Milk Expression is associated with a reduction in the length of hospitalisation for the very preterm infant</td>
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<td>11:00</td>
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<td>11:15</td>
<td>Elena Rivers</td>
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<td>11:30</td>
<td>Chris Clark</td>
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<td>The Tinman gene Nkx2.5 regulates metabolic stress genes during cardiac outflow tract development</td>
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<tr>
<td>11:45</td>
<td>Tamika Katherine Rozema</td>
<td>U Kentucky</td>
<td>“Spells” in an otherwise healthy infant</td>
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THE INITIATION OF INHALED CORTICOSTEROIDS IN THE EMERGENCY DEPARTMENT
Sarah Nicole Adams, MD Christina Cochran, MD Carolyn Word, MD Olivia Titus, MD James R Roberts MD, MPH Annie L Andrews, MD MSCR
-Department of Pediatrics, Medical University of South Carolina, Charleston SC

Background: It has been demonstrated that patients with asthma seen in the emergency department (ED) are unlikely to attend outpatient follow-up. Emergency Department physicians can participate in preventive care delivery by initiating inhaled corticosteroid (ICS) therapy.

Objectives: Determine the rate of prescribing ICS at discharge and identify factors associated with higher prescribing rates.

Methods: A retrospective chart review was done of patients, ages 2-18 years, seen in and discharged home from the ED with a primary diagnosis of acute asthma exacerbation from June 2011 to July 2012. It was then determined whether an ICS was prescribed for patients not already on a home ICS. Demographic and clinical variables recorded included race, age, insurance, asthma severity score, and number of albuterol treatments administered. Chi-square tests were used to determine the association between demographic and clinical variables and ICS initiation. Logistic regression models were then used to determine factors significantly associated with initiating ICS at discharge.

Results: A total of 117 patients with the diagnosis of acute asthma exacerbation were eligible for ICS initiation. Only 17.09% of eligible patients were started on inhaled corticosteroid therapy. Race was found to be significantly associated with the initiation of ICS in the ED. In addition, non-white patients were significantly less likely to have therapy initiated at time of discharge (OR 0.227, 95% CI 0.066-0.780)

Conclusions: There is significant room for improvement in delivering preventative care in the acute care setting of the emergency department for children with asthma. Improving the rate of ICS initiation will likely prevent future asthma exacerbations.
OUTCOME ANALYSIS FOR TWO TREATMENT OPTIONS IN ACUTE PEDIATRIC COIN INGESTION: SURGICAL MANAGEMENT VERSUS BOUGIEANG

Allie, E., Blackshaw, A., Losek, J., Tuuri, R.

Background:
Coin ingestion with esophageal entrapment is a common problem managed in the Pediatric Emergency Department (PED). At most pediatric institutions, children undergo sedated retrieval of the coin under general anesthesia using endoscopy, forceps or foley dislodgement. Esophageal bougienage is another technique for coin advancement that has demonstrated excellent safety, efficacy, and costs, but as the body of supporting literature is small, bougienage is not used in most PED’s. In our PED, medically stable children without significant prior gastro-esophageal surgery or disease, who have witnessed coin ingestions of less than 24 hours, undergo bougienage. Our objective was to report our PED’s experience with esophageal bougienage versus surgical retrieval with regard to clinical outcomes and costs.

Methods:
We conducted a retrospective review of all children with lodged esophageal coins presenting to The Children’s Hospital of South Carolina PED from January 2004 to October 2012. The following data were abstracted: presenting symptoms, location and type of coin, age, sex, exclusion criteria for bougienage, efficacy and complications of chosen procedure, return visits, length of stay (LOS), and hospital charges. The senior reviewer re-abstracted 10% of charts for a total of 672 data points re-reviewed, with 98% agreement. T-tests and 95% confidence intervals were used for comparisons. Outcome measures were compared between the group eligible for bougienage that initially underwent bougienage with the group eligible for bougienage but undergoing surgical retrieval and those who did not meet bougienage criteria.

Results:
Two-hundred-forty-five patients presented to our PED with lodged esophageal coins. One-hundred-sixty-five patients were eligible for bougienage, 80 were ineligible. Of the bougienage-eligible, 129 had successful bougienage, 7 failed, and 29 underwent endoscopy. Of the 80 ineligible patients, 71 had surgical retrieval, 9 had inappropriate bougienage; of these inappropriate bougienage there were 3 failed attempts with no serious complications.

The average LOS for all 135 eligible cases that initially underwent bougienage was 137.3 (SD 54 minutes) and 769 (SD 535 minutes) for the surgical cases and 9 bougienaged, but ineligible cases. The difference in the means was 632.4 minutes, 95% CI for the difference was 723.7 to 541.1, (p-value = 0.000).
The average hospital charges for 135 eligible cases that initially underwent bougienage were $952.80 (SD 417) and $7022 (SD 3132) for the surgical cases and 9 bougienaged but ineligible cases, (p=0.000).
The difference in the means was $6,069, 95% CI for the difference was $6,606 to $5,533, (p-value = 0.000). The charge for the eligible children undergoing bougienage was significantly less.

Children undergoing bougienage were significantly older, mean age 4 (SD 2 years), than those who went to surgery, mean age 2 (SD 3 years), (p=0.000).

For the successful bougienage group, 22 minor complications were observed and five return visits to clinics or PED were reported. The surgical group had ten minor complications reported and two significant complications including one traumatic esophageal diverticulum and one endotracheal tube dislodgement requiring reintubation.

Conclusions:
For eligible children, esophageal bougienage is a safer and more cost-effective management strategy for lodged esophageal coins.
Case Report:

Baby boy X was born to a thirty-eight-year-old G3P1021 female at 39\textsuperscript{4/7} weeks gestational age via C-section, indicated for breech presentation. The mother has a history of two previous miscarriages at 14 and 7 weeks gestational age. Ultrasounds performed at 25, 31, 35, and 37 weeks gestation showed polyhydramnios.

Upon delivery, baby requiring four minutes of bag mask ventilation and IV fluid resuscitation. APGAR scores were 5 at one minute, 8 at five minutes, and 10 at ten minutes.

Pertinent physical exam findings include a birth weight of 2.92 kg, head circumference of 34cm, low set ears with abnormal cartilage formation of antihelices, a large sagital suture with communicating fontanelles (anterior fontanelle measured approximately 4x5cm and posterior fontanelle measured approximately 3x4cm), mild scleral icterus, micrognathia, short neck, nuchal fat pad, rectus abdominus muscles palpated laterally, but rectus sheath not appreciated medially, palpable spleen, undescended testes bilaterally, contractures at proximal interphalangeal joints bilaterally, clinodactyly, short femurs, and small feet.

On day three, serum sodium increased to 155 (repeated measurement of 152) mmol/L. The patient had been passing more urine than usual. Due to this persisting hypernatremia, we ordered for a serum osmolality that came back at 320mOsm/kg [282-300]. Urinalysis showed a specific gravity of 1.005 and repeat measurements showed 1.010 x 2 occasions.

We determined that the cause of the patient’s hypernatremia was likely secondary to diabetes insipidus (DI). A desmopressin challenge test was ordered to determine if the patient’s DI was central or nephrogenic. One dose of desmopressin (DDAVP 60 nanogm) corrected his urine specific gravity to within normal rage. Thus, we concluded that his ADH receptors at the level of the nephron were functioning properly, and his DI was central in etiology. Baby boy X will require synthetic ADH for the rest of his life.

Results of karyotyping showed a translocation of chromosome 9p and 18q. This chromosomal abnormality has never before been documented.

As this is a rare finding, we cannot conclude that this is the etiology of the patient’s physical appearance or DI. We may, however, learn more from the case of baby boy X with close follow-up care. This case supports the indication for karyotyping future neonates with similar findings.
A LONGITUDINAL STUDY OF THE DURATION OF BREASTFEEDING IN TERM AND PRETERM INFANTS
Bussmann N. (MD), Ryan A. (MD), O’Shea C-A (Clinical Research Nurse)
Cork University Maternity Hospital, Wilton, Cork, Ireland

Background: As part of a longitudinal study of proteomics and proteolysis in human breast milk we enrolled breast-feeding mothers of term and preterm babies. The purpose of this study was to improve these mothers in terms of duration of breastfeeding in the context of a breastfeeding study.

Methods: The initial study was designed and carried out at Cork University Maternity Hospital. Mothers of pre-term and term infants were approached in the post natal ward and the neonatal unit by a clinical research nurse. They were provided with an information leaflet on the above study and then consented for participation. Mothers provided breast milk weekly from the start of the study (i.e. from when the infant was born) and then at one week, three week, six week and ten week intervals once full term corrected gestational age was reached. A total of 56 infants were enrolled in the study and followed during this time frame. During this period breast-feeding support was also offered with the help of the breast-feeding consultant.

Results: Of 54 mothers recruited, half (27) had term (mean gestational age 39/40 +/-) and half preterm (mean gestational age 30/40 +/-) babies. At 10 weeks post birth 82% of mothers of term babies were still breastfeeding compared to 42% of mothers of preterm babies. Of those not breastfeeding, 69% of preterm babies were formula fed prior to reaching full term corrected gestational age.

Conclusions: Breastfeeding mothers of term and preterm babies are highly altruistic and readily consented to longitudinal studies on human breast milk and more are still breastfeeding (82% and 52%) compared to national averages 10 weeks post-delivery. Enrolled in research studies along with enhanced breastfeeding support may be a motivating factor for these mothers.
**SKIPI-SKIN PUNCTURES IN PREMATURE INFANTS**
Authors: Dr. Daryl Peter Butler MD BCh, Dr. Daragh Finn MD BCh, Orla Sheehan Dr. Eugene Dempsey MD BCh
Cork University Maternity Hospital, University College Cork, Wilton, Cork, Ireland.

**Aim:**
To quantify the number of skin punctures in premature infants in the first 2 weeks of life in a tertiary neonatal centre. To assess whether there is an association between skin punctures and sepsis in premature neonates.

**Background:**
Premature infants’ skin integrity is crucial to preventing infection yet the skin is punctured for a number of reasons in the first weeks of life. This includes intravenous cannulation, blood sampling for electrolyte and glucose monitoring. There is little information about how often the skin integrity is broken.

**Methods:**
A prospective observational study of all premature infants (< 35 weeks) born over a 6 month period from December 2012 to May 2013 in Cork University Maternity Hospital. Each skin puncture a premature neonate received in their first 2 weeks of life was documented. This included skin punctures for intravenous cannulation, blood sampling, capillary gases, glucose measurements and long line insertion. Relevant demographics such as gestation at birth, birth weight, episodes of sepsis and other co-morbidities were also documented. Sepsis was defined as either blood culture positive sepsis or clinically, where antibiotics were given for a minimum of 5 days. Total numbers of skin punctures were then correlated with incidences of sepsis and other relevant demographics.

**Results:**
104 infants were recruited, 3 infants were excluded due to inadequate data collection and 2 excluded as they were transferred to another center, therefore, 99 infants were included in the analysis. Overall the median (range) number of skin punctures per infant was 21 (2-51) the majority (median 15) occurring in the first week of life. For those infants born < 28 weeks gestation there was a higher number of skin punctures in the second week of life (median 8 in 1st week versus mean 14 in second week). For gestational ages >28weeks there was a higher number of skin punctures in the first week of life, median 16 (2-40). One infant had 29 skin punctures in the first 72 hrs. Capillary blood gases accounted for the majority of skin punctures in all gestational ages. 15 infants had sepsis during the study period, 5 of which were blood culture positive. Overall mean skin punctures in infants with sepsis over the 2 week period was 23.7. Of those who had positive blood cultures mean skin punctures in the first 2 weeks of life was 29.2. Median day of life for positive blood culture was day 8 (range 8-15). Patients with clinical sepsis were more immature and lower birth weight. We found no correlation between number of skin punctures and sepsis.

**Conclusion:**
Our study demonstrates that premature infants’ (< 35 weeks gestation) skin is punctured frequently across all gestational ages within the first two weeks of life. The most common reason for skin punctures is for capillary gas measurements. For infants born < 28 weeks gestation the majority of skin punctures occurred in their second week of life. A multidisciplinary team effort is required to reduce skin punctures in preterm infants.
THE TINMAN GENE NKK2.5 REGULATES METABOLIC STRESS GENES DURING CARDIAC OUTFLOW TRACT DEVELOPMENT

Christopher D Clark, BS¹, Anthony J Horton, BS¹, and Kyu-Ho Lee, MD-PhD¹. ¹Pediatrics, The Children’s Hospital of South Carolina, Charleston, SC, United States.

Background: Congenital heart disease is the most common form of congenital anomaly and the leading cause of death from birth defects. Insight into the underlying causes has been provided by studies of key genes regulating signaling and transcriptional events during heart development. The Nkx2.5 transcription factor is one such gene: Its expression is required for the proper differentiation and growth of heart cells. Nkx2.5 null mutation in mice results in severe right heart and outflow tract (OFT) hypoplasia, and Nkx2.5 point mutations in humans are associated with heart defects involving the cardiac OFT, likely due to dysregulation of downstream target genes in cardiac precursors.

Objective: To identify direct downstream target genes of Nkx2.5 in OFT precursors and assess their function in normal heart development

Design/Methods: Analysis of expression microarray data from wildtype and Nkx2.5 null mouse embryos and in silico promoter sequence analysis were used to identify candidate Nkx2.5 target genes. Quantitative and qualitative mRNA expression analysis, ChIP analysis and promoter assay were used to further evaluate potential target genes.

Results: We identified two genes closely linked and highly conserved in mammals that appear directly regulated by Nkx2.5 in cardiac OFT precursors, Ccdc117 and Xbp1. Nkx2.5 is required for the normal expression of Ccdc117 and Xbp1 in OFT precursors. Xbp1 is a key regulator of autophagic stress responses to cellular damage and the expression of Xbp1 downstream target genes is decreased in developing hearts of Nkx2.5 mutant embryos.

Conclusions: Morphologic defects due to Nkx2.5 pathway mutation may result in part from compromised response of cardiac precursors to metabolic stress during development.
RESIDENT COMFORT AND ADHERENCE TO BEST PRACTICE GUIDELINES FOR PEDIATRIC WELL VISITS
Colby L. Day MD, Kimberly L. Hays MD, Anna C. Collins MD, Chang L. Wu MD, Daniel C. Williams MD, Pediatrics, Medical University of South Carolina, Charleston, South Carolina

BACKGROUND: The American Academy of Pediatrics published the Bright Futures guidelines to promote best practice for pediatric well visits. Current understanding of resident comfort and adherence to these guidelines is lacking. Our objective is to assess resident comfort and adherence to best practice guidelines for pediatric well visits as an ongoing quality improvement project.

METHODS: This study is a prospective observational study. Baseline resident comfort and adherence to best practice guidelines was first assessed through an anonymous survey utilizing visual analog scales. Following this baseline assessment, resident adherence to best practice guidelines was assessed through chart reviews of 2 year old well visits randomly selected from July 2012 through February 2013. The chart reviews specifically assessed documentation of screens and procedures recommended by best practice guidelines. These included height, weight, body mass index, head circumference (HC), oral health and dental home assessments, lead and hemoglobin screens, developmental and autism assessments, as well as tuberculosis, lipid, hearing, vision, and blood pressure (BP) risk assessments. Interventions implemented included a comprehensive reference document and resident education in several venues. Chart reviews were performed at 2 month increments over the period of interventions. Adherence was based on an all-or-none assessment of resident documentation. Comparison between residency classes was done using Fisher's Exact Test. The most recent intervention implemented is a revision of the templates for well child visits used in our primary care clinic with the goal of standardizing the templates with the Bright Futures recommendations. Further chart reviews will assess the impact the template revision has on resident documentation.

RESULTS: 42 PGY 1-3 residents completed the baseline survey of resident comfort with best practice guidelines. Residents reported being at least Moderately Comfortable on all aspects for the 2 year old well visit except for autism screening (mean score=44.1). Resident comfort was found to positively trend with greater experience. In the initial pre-intervention chart review, adherence to all recommendations was good (> 70% total completion) with the exception of resident documentation of HC (54%), dental home (21%), developmental screening (12%), and autism screening (0%). Documentation was also poor for risk assessments including lipid (61%), vision (3%), hearing (18%), and BP (3%). Comparison between classes did not reveal differences in any expected or predictable pattern. Interval chart reviews as interventions were employed did result in a statistically significant change in overall documentation of performance of developmental screening (p=0.03) and autism screening (p=0.05). Templates are currently being revised to adhere to best practice guidelines as recommended by Bright Futures and subsequent chart reviews will assess the impact of the template revision on resident assessment and documentation.

CONCLUSIONS: Though resident comfort with adhering to best practice guidelines is high in most areas, actual documentation is not optimal. Our portable reference document and resident education have demonstrated positive results in documentation in areas with the least amount of resident comfort and initial baseline performance. We hypothesize that revision of the templates for well child visits will further improve resident documentation of performance of best practice guidelines.
IMPLEMENTATION AND REVISION OF COMMUNICATION SKILLS CURRICULUM IN A PEDIATRIC RESIDENCY PROGRAM: PEDICOMM

Colby L. Day MD, Sarah Mennito MD, Conrad Williams MD, Amanda Price MD, Michael Bowman MD, Olivia Titus MD, Pediatrics, Medical University of South Carolina, Charleston, South Carolina

BACKGROUND: The importance of communication skills in effective health care interactions cannot be overstated and should be an integral component of pediatric training. In spite of this, formal communication training is frequently not emphasized in residency training. In 2011, a communication skills curriculum (PediComm) spanning the three years of pediatric residency training was created during which residents practice communication techniques in standardized encounters. Our objective was to assess the efficacy of this curriculum in teaching useful communication skills for both resident participants and faculty mentors. Our secondary objective was to obtain specific feedback to be used in revision of the current scenarios.

METHODS: At the end of the 2012-2013 academic year, all current pediatric residents were anonymously surveyed and asked to rate how helpful the PediComm curriculum was in improving their communication skills, whether the scenarios teach better communication in the healthcare field, whether use communication skills learned in PediComm during patient interactions in residency, if they planned to apply learned skills to their future practice, and what specific suggestions they had for improvement of the curriculum. Faculty and fellow mentors were also anonymously surveyed with similar questions. For the 2013-2014 academic year, the entire PediComm curriculum, including required readings, standardized scenarios, and overall format, was reviewed and updated to better conform to recommendations given by mentors and residents. Hand-off practice was incorporated into the intern sessions and upper level scenarios were revised to include more complex interactions with inherent communication difficulties.

RESULTS: Of the residents who participated in the initial survey reviewing the 2012-2013 PediComm curriculum (20 out of 45 residents), 9 had participated in 1 session, 5 had participated in 2 sessions, and 6 had participated in 1 session during residency. Overall, 60% of residents found the PediComm sessions to be helpful in improving their communication skills and 70% found the standardized scenarios to be effective in teaching better communication in the healthcare field. At least half of the residents also believed that the presence of other residents during sessions was beneficial to their learning. Of the 10 out of 30 faculty and fellow mentors who participated in the survey, 100% agreed that the sessions were useful in resident education (with 70% indicating strong agreement). Additionally, 80% of mentors believed that the PediComm sessions helped to improve their own communication skills (with 40% indicating strong agreement). Specific feedback on individual scenarios was also reviewed from the surveys. Analysis of current literature was performed to ensure educational teachings were most current.

CONCLUSIONS: Anonymous surveys of residents and faculty in our pediatric residency program indicate a positive association between the PediComm curriculum and comfort with communication skills in difficult health care interactions. Based on survey results we have optimized the structure of the program to meet the needs of both learners and mentors. As we continue to improve our PediComm curriculum through additional surveys of residents and faculty, we anticipate increasingly positive feedback regarding resident preparedness to handle difficult communication scenarios in their medical practice both during and beyond residency.
Our future lies not in the hands of our children, but in the hands of their parents.
How do parents and families deal with our changing culture? How do pediatricians deal with it? How can we, or do we, advise our patients’ parents to handle problems like, lying, cheating, stealing, social media, video games, sexuality, peers, peer pressure, and violence? How can we address the rising rate of child poverty, sub-optimal schools, unemployment and the ever present threat of terrorism?

Statistics consistently show society is failing to meet these challenges and our kids are not thriving. Because of these challenges today’s parents are about as stressed as they can be. Many admit they do not enjoy being parents. Former Arkansas Governor Huckabee told viewers last year on Father’s Day that parenting was the hardest thing he had ever done. Harder, he said, than being governor, running for president, or producing his daily radio show and his weekly television show. What’s going on? How can we help parents, families, and ultimately children?

I will discuss some of the readily available tools that help parents meet these challenges. Many don’t consider dinner together, the “Off” button, or parental attitudes as implements for parenting, but I will explain how parents can use them to advance character development. Likewise organized religion, peers, chores, and money management are all everyday things which help kids grow into the adults we hope them to become. In short, parents need to be the person they want their kids to become, because as we all know kids become their parents.

In the spirit of full disclosure this discussion is taken in part from my newest parenting book, *Tools for Effective Parenting*.
NEONATAL OUTCOMES IN ELECTIVE CAESAREAN SECTIONS AT TERM
Finn, O’Neill, Collins, Khasan, Dempsey

Introduction:
The gold standard gestational age for elective caesarean section is 39 weeks, however rates of elective caesarean section prior to 39 weeks remain high.

Objective:
To assess neonatal outcome following elective caesarean delivery at term (≥ 37 weeks gestation) including rates of NICU admissions, reasons for admission and medical interventions in the newborn period.

Methods:
A retrospective review of all elective caesarean sections over a four-year period from August 2008 to July 2012 was performed. Details were obtained from theatre ward section books and cross-referenced with maternal notes where required. Newborn details were obtained from neonatal intensive care unit electronic record system where applicable. Two authors (DF, AC) performed the data entry. Elective caesarean section was defined as a caesarean section performed in the absence of labour, without any foetal or maternal indications for delivery. Data were analysed using SAS software®, version 9.2 (SAS Institute Inc., Cary, NC, USA). The incidence rates of each adverse outcome were calculated using the Cochran-Armitage test for trend for the entire study cohort (n=4,995) and separately for infants admitted to the NICU only (n=445).

Results:
4,495 women had elective caesarean sections at term, accounting for approximately 15% of all term deliveries. Among the women undergoing elective caesarean delivery almost one fifth (18.5%) were delivered prior to 39 weeks gestational age. The most frequent indication for caesarean section was a previous caesarean section (72%) followed by an abnormal lie (17%). A higher percentage of women who delivered at 37 or 38 weeks gestational age were over 35 or had private health insurance compared with women who delivered at 39 weeks or later. The admission rates at 37 weeks gestation was 16.1% versus 5.5% at 40 weeks gestation (P for trend < 0.0001). Overall composite measure for any outcome was significantly less likely as gestational age at birth increased from 37 to 40 weeks (22.6% at 37 weeks and 6.7% at 40 weeks; P for trend < 0.0001). When singleton pregnancies were analysed alone the significant trend for decreasing NICU admissions and any adverse outcome for greater gestational ages persisted. Medical interventions including intubation, antibiotics, phototherapy or intravenous fluids combined were more commonly performed in those delivered at earlier gestational ages (with rates of 10.5% at 37 weeks decreasing to 3.6% at 40 weeks; P value for trend 0.0001). The adjusted odds ratio for admission to the unit at 37 weeks was 2.35 (1.33, 4.14) and 1.37 (1.05, 1.79) at 38 weeks compared to 39 weeks.

Discussion:
One in 5 elective caesarean deliveries occur at less than 39 weeks completed gestation. Repeat elective caesarean sections accounts for the largest proportion of elective deliveries. Parents and physicians need to be aware of increased neonatal morbidity when delivery occurs electively at less than 39 weeks, including neonatal intensive care unit admission and medical interventions, increased risk of respiratory problems and a prolonged length of stay.
Inflammatory myofibroblastic tumor (IMFT) is rare, most frequently affecting soft tissue and lung, with fewer than 20 reported cardiac cases, predominantly in the young. Cardiac involvement from infancy to 21 years shows an average age presentation of 10 years without gender bias. IMFT mostly arises as an endocardial-based intraluminal mass, attached to valves (especially mitral). The mobile lesion may obstruct ventricular outflow or coronary ostia causing sudden death, embolic myocardial infarction and rarely seizures with cerebral embolic infarction. IMFT has limited malignant potential with low incidence of recurrence once successfully surgically resected. We report sudden cardiac death in a three-year-old as the initial presentation of IMFT. She was on the sofa when her mother heard her make a “whining” sound followed by immediate collapse. Resuscitation attempts were unsuccessful. Gross cardiac examination revealed smooth, multilobulated polypoid endocardial and valvular masses within the left ventricle inlet and outlet, attached to the mitral valve apparatus and apex. Tumor extended through the left ventricular outlet, across the aortic valve outflow tract and into the proximal ascending aorta. Embolic occlusion of the left coronary ostium which saddled the bifurcation of the left main and left circumflex coronary arteries was observed. Microscopically, peripheral stellate and spindle myofibroblasts were admixed with mononuclear cells in a myxoid background. This is the third recorded case of IMFT embolism to the coronary arteries and the second recorded case of previously asymptomatic sudden cardiac death due to this phenomenon.
CONSERVED REGULATION OF THE TINMAN GENE NKX2.5 DURING CARDIAC OUTFLOW TRACT DEVELOPMENT: A WINDOW INTO NOVEL CONGENITAL HEART DISEASE ENTITIES.

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Background: Congenital heart disease is the most common form of congenital anomaly and the leading cause of death from birth defects. Insight into the underlying causes has been provided by studies of key genes regulating signaling and transcriptional events during heart development. The Nkx2.5 transcription factor is commonly mutated in familial congenital heart disease, and graded loss of its expression in outflow tract precursors in mice results in right heart and outflow tract (OFT) hypoplasia of varying severity. Critical regulation of Nkx2.5 expression levels in these precursors is achieved through a balance of inducing or inhibiting BMP and FGF signals, but whether altered expression of Nkx2.5 plays a role in human heart disease is yet unknown. The ability to identify genetic abnormalities leading to altered regulation of an otherwise normal Nkx2.5 gene product requires a basic understanding of its transcriptional regulation mechanisms in critical cardiac progenitor populations.

Objective: To identify and define important regulatory elements of the Nkx2.5 in OFT precursors and assess their relationship to signaling pathways regulating normal heart development.

Design/Methods: An in silico analysis of Nkx2.5 genes from multiple vertebrate species was used to identify conserved Nkx2.5 non-coding regions potentially regulating expression in OFT progenitor cells, and consensus transcriptional regulator binding sites within these regions. The functional role of candidate consensus binding sites for regulators downstream of developmentally important BMP and FGF inducing factors in archetypal mouse and chicken regulatory regions were tested by in vitro analysis in cell culture and in vivo analysis in transient transgenic mouse experiments.

Results: We found conserved but evolutionarily distinct conservation of non-coding elements regulating expression of Nkx2.5 in OFT progenitors in mammalian vs. non-mammalian vertebrates. This conservation includes functionally important binding sites for BMP-transducing Smad1/4 and YY1 transcriptional regulators, Nkx2.5/Mef2c binding sites necessary for long term expression of Nkx2.5 in OFT and right ventricular cells, and for Pea3/Ets factors mediating signaling from the FGF pathway. In vitro studies show surprisingly nuanced interaction of the Ets factors with mechanisms regulating long-term gene expression or silencing relevant to stability of cardiac fate.

Conclusions: Basic investigation of early cardiac gene regulation in animal models may be of great value in enabling the targeted investigation of novel, non-coding genomic variations leading to human congenital heart disease.
STRUCTURED PROMOTION OF BREAST MILK EXPRESSION IS ASSOCIATED WITH A REDUCTION IN THE LENGTH OF HOSPITALISATION FOR THE VERY PRETERM INFANT.

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Background: Human milk provides nutritional and immunological benefits to the very preterm and very low birth weight (VLBW) infant. Much work has been done on the benefits human milk confers on premature infants\textsuperscript{1-6}, including reduction of morbidities prevalent in neonatal units such as necrotising enterocolitis and neonatal sepsis. Although expressed breast milk rates vary significantly, studies have demonstrated effective methods of presenting this information to mothers or premature infants to help increase expressed milk volumes and promote early expression\textsuperscript{7,8}. A 2008 study\textsuperscript{9}, carried out in Utah, USA assessed the effects of introducing the ‘BEST Program’ in a Level III Neonatal Intensive Care Unit. The aim of this study was to assess neonatal outcomes after the implementation of a structured, practical approach designed to promote breast milk expression and the early initiation of human milk feeding in mothers of very preterm/VLBW infants. Outcomes assessed included the incidence of necrotising enterocolitis, sepsis, re-attainment of birth weight, attainment of full feeds and duration of hospitalisation.

Methods: A multi-modal approach between lactation consultant, neonatal dietician, medical, midwifery and neonatal nursing staff resulted in the development of a protocol focussed on enhancement of expression of human milk by mothers of very preterm infants. Following implementation of the intervention a prospective chart review was performed for 6 months. All eligible infants were included. The group of infants enrolled in this time period was compared to a pre-intervention group from 2010. Statistical analyses were carried out using SPSS 18.

Results: 82 infants (39 prospective; 43 retrospective) were included. Initiation of enteral feeding with EBM (median = 2 days) and earlier achievement of full feeds (median = 12 days) resulted in a reduced length of stay in the prospective cohort (mean = 50 days; $p = 0.021$) Birth weight was regained earlier in the prospective cohort (mean = 10.42 days; $p = 0.038$).

Conclusions: This work has shown the potential for producing positive end-outcomes with focussed interventions in mothers of very preterm infants, in particular a significant reduction in the duration of hospitalisation, thereby decreasing patient stressors, parent anxiety and hospital expenditure.


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Background: In the late 1980’s, the Guidelines for Perinatal Care set the framework for Regionalized Perinatal Care in South Carolina. Included in these guidelines were specific notations that defined the geographic boundaries of perinatal regions; designated all delivery hospitals as either Level I, II, or III; and specifically recommended that all very low birth weight (VLBW) babies be delivered in the Level III Regional Perinatal center. Since that time the environment surrounding the overall perinatal care system has changed significantly with a greater number of neonatologists in community hospitals and improved financial coverage for all pregnant women and their newborns. There is discussion regarding the elimination of the Certificate of Need process as well as the challenge that hospital designations be revisited to allow for care of certain high risk babies in community hospitals. These discussions can be informed by a comparison of the patterns of care and associated outcomes between the earlier periods and the recent past.

Specific Aim: To compare the patterns of care and outcomes for very low birth weight (VLBW) newborns in the LowCountry Perinatal Region between the periods of 1998-2000 (T1) as compared to 2009-2011 (T2).

Methods: Data from the South Carolina Live Birth files; Live Birth / Infant Death Cohort files; Medical University of South Carolina (MUSC) Perinatal Information System (PINS) files, and the MUSC Meducare transport files were used to assess the characteristics of VLBW live birth in the eight county LowCountry Perinatal Region for the periods 1998-2000 and 2009-2011.

Results: While the total number of VLBW births increased from 651 to 755 (increase of approx. 35 babies per year), the incidence remained 1.9% for both periods. Community hospitals decreased their expected inborn VLBW deliveries by 36% and 32%, respectively, by transferring those deliveries to MUSC as in-utero transports. Including these maternal transports, MUSC was the location of delivery for 82% of regional VLBW in T1 as compared to 81% in T2. Of the community hospital VLBW inborn deliveries, 74% and 79% were transported to MUSC for care. Including these neonatal transports, MUSC was the hospital of care for over 95% of regional VLBW babies in both periods. The number of VLBW babies cared for by MUSC increased by 15% between the two periods with a 60% concomitant reduction in neonatal mortality. Overall, there was a 45% reduction in the number of regional VLBW neonatal deaths and all three levels of hospitals experienced an improvement in VLBW neonatal survival. Still, optimal survival occurs among babies delivered in the regional perinatal center where babies have a survival rate over three times of those delivered in community hospitals.

Conclusion: These data suggest the regionalization of perinatal care in the LowCountry perinatal region is stable and achieving a high level of accomplishment in risk appropriate care (VLBW care consolidated in the regional tertiary center).
ACADEMICS AND PHYSICAL ACTIVITY IN TITLE ONE ELEMENTARY SCHOOLS: CHANGING WHAT WE CAN

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Background: Title One schools struggle to find innovative ways to improve the academic achievement of their students who are high risk for failure. Evidence for interventions aimed at more malleable factors, including in-school physical activity programs, has been mixed in this population. A unique program that combines physical activity with classroom lessons was piloted at one Charleston, SC elementary school. Pilot data suggested that this program had positive impact on academic achievement and the school system elected to expand the program to six additional Title One schools two years later. However, successful large-scale implementation of such programs requires an understanding of the intractable factors that contribute to complexity of this issue as well as attention to program fidelity.

Objective: Understand the real world impact of a physical activity program on academic achievement and the underlying factors that modify this impact.

Design/Methods: Improvement in reading and math test scores during the first year of implementation was compared for the intervention schools (n=2377) and nine control schools (n=2,346). Underlying factors were examined to determine their impact on test scores. A generalized linear mixed model was used with change score as the outcome variable and baseline score, intervention group, age, gender, ethnicity, and free lunch status as covariates. A random school effect was included to account for the correlation between students in a particular school.

Results: Mean reading score improvement was 12.4 points and mean math score improvement was 13.3 points over the course of the year. Intervention group was not associated with test score improvement. The intervention group improved 0.7 points less in reading and 1 point less in math than the control group. However, neither difference was statistically significant (p=0.361 and 0.291 respectively). The generalized linear mixed model was driven by demographic variables with the change in test score significantly associated with baseline score, age and ethnicity (all p<0.01). Gender had an impact on math score improvement (p=0.01).

Conclusions: Contrary to pilot data, data from this large scale intervention does not show an association between the intervention and test score improvement. The success of programs aimed at addressing malleable factors, such as physical activity, is likely tied to underlying factors and impacted by program fidelity. Further study including a more closely matched school and subgroup analysis as well as the impact of program fidelity and the intervention’s effect on students’ BMI is ongoing. These results from the first year of intervention are useful in the planning of this and future interventions.
Term female infant with Pierre-Robin sequence (micrognathia and cleft palate), dysmorphic facial features (short palpebral fissures, flat nasal bridge, small nose and anteverted nares), 2-3 toe syndactyly and hypotonia presented at 48 hours of life with acute respiratory decompensation, profound hypotension and severe metabolic acidosis (base deficit of -22) associated with mild hyponatremia (Na+ of 133) and hyperkalemia (K+ of 6.6). She recovered with mechanical ventilation, catecholamine support, stress dose of steroids, and aggressive fluid resuscitation suggestive of adrenal insufficiency. Transthoracic echocardiography revealed large patent ductus arteriosus with supra-systemic pulmonary hypertension. Genetic/Metabolic workup showed 46XX karyotype, elevated lactic academia and hyperammonemia. She was diagnosed with Smith-Lemli-Opitz syndrome (SLOS) based on markedly elevated 7-dehydrocholesterol (48 mcg/ml; normal 0.04-0.36 mcg/ml) and low serum cholesterol (8 mg/dl; normal >50 mg/dl). She had mandibular sagittal osteotomy with external distractors for correction of Pierre-Robin sequence with severe obstructive sleep apnea. She required treatment with sildenafil for idiopathic persistent pulmonary hypertension. For her SLOS, she was treated with cholesterol supplementation and statin therapy. She received stress dose steroids prior to all surgical interventions and did not present with any further decompensations during the hospital stay.

SLOS is an autosomal recessive disorder due to deficient activity of 7-dehydrocholesterol reductase which results in cholesterol deficiency and toxic accumulation of cholesterol precursors, such as 7-dehydrocholesterol. SLOS is a multiple malformation syndrome and its phenotypic spectrum is very broad, ranging from a mild disorder to a lethal malformation syndrome. The 2-3 toe syndactyly is the most common physical finding in SLOS patients. As cholesterol is a precursor in the pathway to steroids, interruptions in this synthesis pathway may result in deficient production of steroids. Case reports have demonstrated adrenal insufficiency in younger patients with severe phenotype of SLOS. Infants with severe phenotypes of Smith-Lemli-Opitz Syndrome may be at risk of adrenal insufficiency and early administration of steroids in time of stress may prevent decompensation. Persistent pulmonary hypertension of the newborn remains a challenging condition to diagnose and treat. It has been reported in infants with SLOS and typically there is evidence of pulmonary hypoplasia. We report the case of PPHN in the absence of pulmonary hypoplasia or other parenchymal disease. Although no randomized controlled trials are available so far, multiple observational studies suggest that cholesterol supplementation and long term statin therapy showed beneficial effects. Long term prognosis and life expectancy of infants with SLOS determined by the severity of malformations and survival correlates strongly with higher plasma cholesterol concentrations.
ACUTE CHILDHOOD ASTHMA IN GALWAY CITY, WEST OF IRELAND FROM 1985-2005: RELATIONSHIP TO AIR POLLUTION AND CLIMATE.

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We examined the relationship of black smoke levels and climatic factors (rainfall, temperature, wind speed, humidity and sunshine) to asthma admissions of children resident in Galway city over a 21-year period.

Methods: Paediatric asthma admissions were recorded from 1985 to 2005, and admission rates per thousand calculated for preschool age (1-4), school age (5-14), and all (1-14) children on a monthly and annual basis. These data were compared to average black smoke levels and climatic data using both correlations and a Poisson GAM analysis, which modelled smooth fitted relationship trends.

Results: Admission rates and pollution levels were significantly correlated in all age groups (r=0.22, p<0.001 for 1-14 year olds). The smooth trends, both in time and due to black smoke, were significant. In particular there was convincing evidence of a positive association between particulate concentrations and asthma admissions for all subjects aged 1-14 years (p<0.0001), and for the 1-4 year olds (p<0.0004) and 5-14 year olds separately (p<0.0004).

When the age groups are separately analysed, the effects of pollution appears greater in the pre-school children, and some of the more recent reduction in admissions of school age children may be attributable to improved prophylactic medication.

Conclusions: We have shown a strong association between a measure of childhood asthma morbidity and black smoke in Galway city. We hypothesise that improved air quality as a result of changes to domestic and vehicular emissions have contributed to reduced hospital admissions since 1995; that the effect has been more marked in pre-school children; and that improved therapy in primary care has contributed to the reduction in hospital admissions in school age children.
AUDIT ON GASTRO-JEJUNAL (GJ) TUBES PLACED OVER A TWO YEAR PERIOD

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Background: GI tubes are feeding tubes inserted through a previously established gastrostomy site and positioned by a radiologist into the proximal jejunum. They are used in cases where neither oral or gastric feeding is tolerated.

Aims: To identify the number of GJ tubes placed or replaced between April 2011 and April 2013 and to identify the indications for placement and replacement. Secondary aims included looking at the total number of procedures per patient, cumulative radiation dose per patient and complications secondary to GJ placement.

Methods: A retrospective review using the online radiology coding system to identify all children who presented to the radiology department between April 2011 and April 2013 for GJ tube related procedures.

Results: 17 patients (10 male and 7 female) who underwent a total of 80 GJ tube procedures were identified in the study period. These included 4 children with pre-existing GJ tubes and 13 who had GJ tubes placed during the study period. The most common reason for placement was reflux and related problems including aspiration and recurrent respiratory infections. The most common reason for replacement of tubes was broken or blocked of the tubes.

Conclusion: GJ tubes can be technically difficult to place and the length of time per procedure varies for each child. Although the tubes are meant to last for up to six months, they can easily become blocked, broken or pulled out. Several children underwent multiple procedures. The next step will be to investigate the reasons behind non-scheduled replacements and see if it is possible to reduce the number of blockage/breakages.
Case Report

MORGANELLA MORGANII AN UNUSUAL CASE OF A SORE THROAT IN A 2 YEAR OLD GIRL

Michael A. Mandarano, DO, MS

BACKGROUND: Morganella morganii is a commensal Gram-negative bacillus of the intestinal tract of humans and other mammals and reptiles. Few reports exist in the literature regarding infections caused by this organism. This case report reviews the clinical course of a 2 year old girl who developed a tonsillitis caused by a rare pathogen, Morganella morganii. This gram-negative enteric bacterium, within the Enterobacteriaceae family, has most commonly been a nosocomial pathogen in debilitated, postsurgical patients. Like many other Enterobacteriaceae, M. morganii has an inducible β-lactamase and is resistant to multiple antibiotics. When caring for children with culture-proven M. morganii infections, one must select an appropriate antibiotic.

METHODS: An upper respiratory culture was obtained from the posterior pharynx of a 2 year old child to identify and analyse tonsillitis caused by M. morganii. The patient in whom the organism was isolated from the clinical specimen was identified by the Microbiology laboratory and susceptibility was expressed.

RESULTS: Upper respiratory throat culture revealed heavy growth by Morganella morganii in this 2 year old child. Antimicrobial susceptibility of this organism showed resistance to: amoxicillin/clavulanic acid, ampicillin, cefazolin, cefuroxime and tetracycline and susceptibility to: cefepime, ceftriaxone, ciprofloxacin, ertapenem, gentamicin, levofloxacin, piperacillin, tobramycin and trimethoprim/sulfa.

CONCLUSIONS:

*Morganella morganii* is a gram-negative enteric bacterium, within the Enterobacteriaceae family. It has most commonly been described as a nosocomial pathogen in immunocompromised patients or those with chronic urinary catheterization. *M. morganii* typically has an inducible beta-lactamase and is resistant to multiple antibiotics.

Therefore, the author concludes that prompt diagnosis and treatment of this organism is imperative to reduce the patient's overall morbidity and mortality. *Morganella morganii* is an unusual cause of tonsillitis. Affected patients are usually immunocompromised. The use of the appropriate antibiotic is critical to eradicating this organism to avoid sepsis and reduce patients overall morbidity and mortality.
The current generation of America’s children is in trouble. Among its many problems are rising rates of obesity, falling school performance scores, high rates of teenage pregnancy, lack of physical fitness, binge drinking, violence, and more.

Pediatricians must help parents set goals for their children and to develop and implement a strategic plan for achieving these goals. It should be stressed must be that at different developmental ages (windows of opportunity) there are jobs that must be accomplished. Also important is parents who are actively engaged parents teaching, guiding, and setting clear rules. These rules and much of what the parent does with and for the child must be explained to the child with increasing attention paid to children’s interests and skills as they emerge and to the child’s developing ability to make comment. The end product should be the release into responsible adulthood of children with the life skills and direction needed.

The issue of “best parenting” practices is discussed in some detail. One chapter called “The Child’s Other Parents”, deals with the strong 21st Century influences that media and peer groups have on children. Media literacy for child and parent are emphasized.

In summary: our children are in trouble and they need help. Parents are the best sources of this help but they need to know more, considerably more, about the whys, hows, whats, and whens of their very important job. This is especially true in a time when traditional parental helps from community and family are often lacking and when the influence of media and peers can be huge. This book has been written to provide pediatrician’s with a guidebook and support tool to recommend to their patient’s parents. The book’s title is A Pediatrician’s Blueprint – Raising Happy, Healthy, Moral, and Successful Children.

“This book is must reading for parents who want their children to grow into healthy, productive and caring adults who have a clear minded and principled perspective.” Hoover Adger, Jr., M.D., Professor of Pediatrics, Johns Hopkins University School of Medicine,
RARE CAUSE OF PEDIATRIC ACUTE ABDOMEN, CURED BY SURGICAL RESECTION: A CASE REPORT OF BLEEDING INTO A RETROPERITONEAL LYMPHANGIOMA.

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Background: Lymphangiomas are rare multiloculated cystic congenital malformations, most occurring in the head and neck, with retroperitoneal location accounting for only 1% of all “abdominal” sites (3-9% of lymphangiomas). Retroperitoneal lymphangiomas which can be confused with other cystic tumors of childhood usually cause a vague chronic condition from compression of nearby structures. Symptoms may include abdominal pain, fever, fatigue, weight loss, and hematuria. Rarely, lymphangiomas may acutely enlarge due to intracystic hemorrhage or rupture and we report such a case. Surgery is needed for symptom relief and definitive diagnosis, as imaging alone cannot differentiate this entity from other more ominous cystic growths.

Clinical Summary: A 5-year-old previously healthy girl presented to an outside emergency department for abdominal pain (epigastric) of two days duration. Her symptoms included pain and nausea without vomiting, but she continued to have normal bowel movements. Labs including CBC, complete metabolic panel, lipase, and urinalysis were unremarkable. CT scan of the abdomen showed a large 17 x 21 cm thin-walled, septated cystic mass within the left abdomen and pelvis of unclear etiology, and she was transferred to the University of Kentucky. The mass was thought to be ovarian in origin, representing a cystadenoma. Prominent lymph nodes were found within the retroperitoneal fat inferior to the left kidney. Physical examination revealed only a firm and distended abdomen and mild epigastric discomfort. The mass was palpable through the left side of the abdomen. Surgical intervention via laparoscopic approach showed a large cystic tumor arising from the pericolic gutter. The cyst wall was adherent to the transverse colon and left fallopian tube, encased the left gonadal vessels, extended past the psoas muscle and displaced the left ureter medially. The mass was isolated and decompressed of 1400mL of clear fluid sent for cytologic examination. The cyst wall was removed with an endocatch bag and morcealated for pathologic examination. The patient showed no cyst recurrence at follow-up.

Pathology Findings: Received in surgical pathology was a multiloculated mass in multiple pieces. Microscopically the cysts had fibrous walls and a bland flat endothelial lining with intervening scattered foci of lymphoid cells, within a vascular stroma. Other connective tissue elements included adipose tissue showing serous atrophy and smooth muscle bundles. Recent and old hemorrhage was present. No mesothelial lining was seen on H&E stain. Immunohistochemical staining demonstrated a lack of Calretinin (mesothelial) marker and confirmed lymphatic endothelial lineage with D2-40 positivity.

Discussion: This case highlights the need for complete surgical resection in this pediatric patient, in order to establish the correct diagnosis and treatment. Patient prognosis following incomplete surgical resection is less favorable. The main complication is recurrence, which can happen when resection is incomplete. Patients with recurrence of retroperitoneal lymphangiomas experience compromise of adjacent vital structures such as the ureter.
MINOR NEURODEVELOPMENTAL MORBIDITIES IN LATE PRETERM INFANTS
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**Purpose:** Identify the minor neurologic dysfunction in preterm infants of 34-36 wk. gestation; explain the challenges from the nursery to age 5-7 years and recommend strategies to intervene and advocate for the late preterm survivor.

**Background Infants:** Infants born at 34-36 weeks gestation, who escape major medical or developmental disabilities, often have mild problems in the newborn period and neurologic dysfunction at school age. The most common problems in the nursery are inability to latch on and to coordinate sucking, swallowing and breathing. GERD is another issue affecting growth and feeding.

Muscle tone is mixed with hypotonia in the head and trunk and increased flexor tone with arching and extensor thrust. The strong palmar grasp, typically seen in term infants, is lacking. Parents comment that they are unusually strong and precocious in their ability to "flip over" from their backs to front. Attempts to place them in an infant seat are difficult due to resistance and increased extensor tonus.

**Case Studies:** On review of several pairs of preterm twins and a high-risk boy who had been exposed to drugs in utero, there were common problems in the newborn period which resulted in an extended stay for observation in the NICU. All infants of 34-36 week gestation were unable to latch on successfully and take adequate volumes of formula to gain weight without gavage feedings of 22 calorie formula.

At discharge, following 10-14 days in the NICU, they were taking a maximum of 3 - 4 ounces of high caloric density formula slowly. Weight gain improved after 2-3 months of age. Two infants developed significant feeding disorders. One had persistent hypotonia with impaired oral-motor coordination and excessive drooling. At 9 to 12 months of age, he was taking only pureed solids and gradually advanced to Stage 2 baby foods. Aversion to certain textures complicated his willingness to accept a variety of table foods. In addition, there was a strong behavioral overlay. As a result of his preterm birth and high-risk history, his mother was convinced that unless she gave him the few pureed solids he would eat that he would lose weight and become susceptible to infection. At daycare, the sitter was able to get him to try a variety of table foods and he imitated the feeding behaviors of other children his age.

With regard to the 34 week preterm girls, who were seen at 6 - 7 years of age, they both had motor incoordination, resulting in clumsiness and difficulty with fine and visual-motor skills and dyspraxia. A 4 year old girl was 1.5 years behind her peers in balance and gross motor milestones. As a result, they had problems keeping up with their peers on the playground and in athletics as well as performing paper and pencil skills in the classroom. One twin had a 20-point discrepancy between her verbal IQ, which was in the high average range, and in her non-verbal skills. This represents a specific learning disability which affects her ability to perform written tasks at school. Even at 6 years of age, her level of frustration is significant because she knows the information but is unable to communicate it on paper (as expected in first grade). Her twin has less of a problem with visual-motor integration but shows mild features of attention deficit disorder of the Inattentive type. Because of their inability to keep up with peers in sports and complete academic tasks in a timely manner, the twins are beginning to experience problems with self-esteem. The school, unfortunately, has not found them eligible for special education services. Strategies to improve their functioning included alternatives use of a computer keyboard, participation in a social play group or activities such as Scouts (where they can experience success), a 504 Plan to provide accommodations for their mild attention deficit disorder and either OT or a tutor to improve visual-motor skills.
Conclusions: The mild impairments seen in 34-36 week preterm infants in the newborn period need to be followed up to prevent secondary problems at school age in the areas of academic skills and self-esteem. With this type of intervention, they can reach their potential without undue frustration or academic under-achievement.
EARLY IDENTIFICATION & INTERVENTION FOR BOYS WITH KLINEFELTER’S SYNDROME

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INTRODUCTION: Klinefelter Syndrome is the most common sex chromosomal disorder and affects approximately one in 660 boys. It is characterized by varying of cognitive, social, learning and behavioral disorders. The phenotype varies from "near-normal" appearance and development to significant abnormalities, including delays in speech/language and tall stature for age and small testes often not identified until after puberty.

Decreased awareness of this syndrome among health professionals and a general perception that all affected males with 47 XXY result in lack of diagnosis prior to puberty. As many of 75 % of the affected males are undetected until puberty or later, when their condition is identified in the course of a fertility work-up.

Early identification and treatment of KS is recommended in order to offer treatment of the medical endocrine and developmental aspects of this syndrome. For example, speech delays leading to behavioral and social maladjustment in the preschool period can be avoided through early intervention programs and therapy. A variety of special education services are available at the appropriate ages and stages of Klinefelter Syndrome.

In addition, metabolic treatment can be provided to prevent osteoporosis, metabolic syndrome and other medical conditions associated with hypogonadism. These can also reduce the impact and severity of potential learning and psychosocial problems.

Conclusions: Children with 47 XXY, Klinefelter Syndrome, have a broad range of phenotypes which can result in a variety of clinical findings. These include developmental delays, learning and behavioral problems, hypogonadism, tall stature and endocrine/metabolic disorders. Through early identification and intervention, these boys can receive special education and therapy in addition to medical treatment to reduce the secondary clinical (e.g. metabolic) disorders.

Recommendations: When a 2 year old boy presents with speech/language delay and tall stature, it is important to check a chromosome analysis to determine whether he has 47XXY as well as a DNA for Fragile X syndrome. This will enable the family to pursue both developmental intervention and medical treatment for a metabolic disorder.
A HISTORICAL PERSPECTIVE OF PEDIATRIC PUBLICATIONS – A BIBLIOMETRIC ANALYSIS

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Background:
Bibliometric tools can be used to identify highly cited articles in a given speciality, which can provide insight into what types of article are of importance and interest to the readership. The aim of this study was to identify and analyse the top cited articles in pediatric literature over the past 65 years.

Method:
A bibliometric search of the Web of Science database from 1945-2010 in the category of “Pediatrics” was undertaken. A comprehensive list of the most-cited articles was generated and analysed.

Results:
There were 497,240 articles published in pediatric journals in the time period under study. Just under 50% of these articles were published in the last 16 years. The mean citation count of the top 100 cited articles was 782 (range 457-2284). Of the top 100 cited articles published, 94 were original articles with two of these proceeding papers. Only two of the articles were laboratory based studies. Endocrinology, neonatology and psychiatry accounted for 28, 22 and 15, publications respectively in this top cited list. The 100 top cited publications are to be found in only 16 journals. There were 89 of these articles published in just five journals - Pediatrics (40), Journal of Pediatrics (22), American Academy of Child and Adolescent Psychiatry (10), Archives of Disease in Childhood (9) and JAMA Pediatrics (8). The top 100 cited articles originated in 12 countries, with USA, UK and Canada contributing 59, 14 and 7, articles respectively.

Conclusions:
Citation analysis of pediatric publications reveals a sustained interest in articles that are based around criteria or guidelines rather than clinical trials. Most of the studies in the list have similar characteristics in that they describe criteria or definitions which can be used as tools by pediatricians. All but one of the top ten in the list belong to this category. That there is a lack of clinical trials in the top 100 citations may be attributed to the ethical constraints of conducting research in pediatrics. Notwithstanding, medical research is important to ensure optimal evidence based care in children.

Most of the older articles describe important syndromes or observations rather than breakthroughs in medical science. The only such example in the top 15 is Avery’s article on hyaline membrane disease. Despite this the older papers continue to be cited, particularly because their findings were novel, yet pertain to pediatricians in current medical practice.

Although older articles remain popular there is a strong bias towards recently published articles and the trends in pediatric publications have changed over time. It is not surprising that the top 100 cited articles in pediatrics were published in higher impact journals. The internet has an ever rapidly expanding role in evidence based medicine, which provides a manuscript for directing patient care, exploring new ideas and guiding the path for future research.
Background:
Sweet’s syndrome is a reactive neutrophilic dermatosis which is rare in children. We report a case of childhood SLE that presented with Sweet’s syndrome. This atypical, diagnostically challenging case is the first case of Sweet’s in association with SLE to be reported in a Caucasian child.

SLE is a chronic auto-immune disorder characterised by multi-system organ involvement and marked clinical heterogeneity. Although the underlying aetiology remains to be fully elucidated, it is thought that SLE may be triggered by several factors including environmental pathogens and infection in those with an underlying genetic predisposition. Typically, ANA and anti-ds-DNA are found, and are routinely tested for in those with suspected SLE.

Sweet’s syndrome is a reactive neutrophilic dermatosis which is rare in children and has been reported to occur in association with a variety of systemic and autoimmune disorders. It was first described in 1964 by Dr. Robert Douglas Sweet, who documented an acute inflammatory skin eruption associated with fever and leukocytosis in eight women. A diagnosis of Sweet’s syndrome requires the presence of two major and two of four minor criteria (Table 1). Characteristically, Sweet’s responds rapidly to corticosteroids.

Case
A 12 year old female presented with widespread arthralgia, myalgia and anorexia. She was systemically unwell, dehydrated and drowsy. Examination revealed a hot, swollen right wrist and a painful, swollen left ankle with restricted movement. On the skin there was an erythematous rash on the nose and eyelids and a papulo-vesicular rash on the buttocks, elbows and ankles (Figure 1).

Initial investigations are shown in Table 2. Biopsy of the papulo-vesicular lesion revealed a florid neutrophilic infiltrate with apoptotic debris smeared between collagen, confirming severe neutrophilic dermatosis (Figures 2 and 3). Subsequent bone marrow aspirate revealed reactive features. Further immunological profile demonstrated a strongly positive homogenous ANA (1:2560) and anti-ds-DNA antibodies were also evident at 26IU/ml (Table 2). Thus, she was commenced on intravenous methylprednisolone. Her symptoms resolved rapidly.

Subsequently the dsDNA crithidia titre was found to be significantly elevated at a titre of ≥1:160 and DNA ELISA was elevated at 16IU/ml confirming a diagnosis of SLE and she was commenced on Azathioprine. At her most recent review she was much improved. The rash and arthralgia had resolved but she had ongoing fatigue and low mood.

Conclusions
SLE is a chronic multi-system autoimmune disease. Recent studies have suggested that childhood-onset has a more severe disease course than in adults. Aggressive treatments incorporating biological agents have improved survival in childhood SLE, but the disease is still associated with significant morbidity. Early diagnosis is essential if disease-associated complications are to be prevented and quality of life optimised. Our patient was diagnosed with SLE having presented with Sweet’s syndrome emphasising the importance of clinician-awareness of the association between them. Although Sweet’s
syndrome is rare, it is important that practicing clinicians are familiar with its presenting features, as its onset may often herald a serious underlying disorder requiring prompt diagnosis and treatment.

IS VERY LOW BIRTH WEIGHT INFANT FIRST POSTNATAL WEEK ENERGY AND PROTEIN INTAKE ASSOCIATED WITH GROWTH PARAMETERS AT TERM AGE EQUIVALENT?

Jacqueline Razzaghy (Presenter), Carolyn Finch, Myla Ebeling, Sarah Taylor. Medical University of South Carolina Department of Pediatrics, Charleston, SC.

Background: For very low birth weight (VLBW) infants, aggressive nutritional practices including early and high protein delivery have been shown to benefit growth during hospitalization, but less emphasis is given to energy intake and to growth until term age equivalent (TAE). Therefore, the objective was to assess the role of first week (168 hour) protein and energy delivery in growth at TAE.

Methods: From a vitamin D health study of VLBW, appropriate-for-gestational age at birth born <34 weeks’ gestation of black or white race infants; parenteral and enteral intake for the first 168 postnatal hours, birth growth parameters, and TAE growth parameters and growth status were collected. Data were tested for normality and then Spearman Correlations and regression models and T tests were performed. Results are given as median (interquartile range).

Results: 107 infants with birth weight 1045 (865,1235)g and gestation 27 (26,29) weeks received protein 21 (18.4,23.5)g and energy 428 (365,481) kcal for the first 168 hours. At TAE, gestational age was 38.6 (37.5,39.3) weeks and weight 2.88 (2.61,3.1) kg, length 46 (45,47) cm, and head circumference (HC) 34 (33,35) cm. Growth velocity from birth to TAE 20 (20,30) g/day. 22 infants (21%) were <10th%tile for weight at TAE. First week protein and energy intake were both significantly but weakly correlated with TAE weight (r=0.22 and 0.2 and p= 0.02 and 0.4 respectively). No significant correlation was seen for TAE length, HC, or growth velocity and first week nutritional intake. TAE weight was significantly negatively correlated with birth weight (r=-0.2, p=0.4) and positively with gestational age at TAE visit (r=0.4, p<0.0001). When controlling for birth weight and TAE visit gestational age, protein intake remained significantly associated with TAE weight (p=0.02), but energy intake was no longer significantly associated with TAE weight. Infants <10th%tile for weight at TAE did not have significantly different first week protein or energy intake when compared to infants with appropriate weight for age.

Conclusions: For VLBW infants, first postnatal week protein and energy intake were associated with TAE weight. When controlling for the week of gestational age at the TAE (range 37-41 weeks) visit and weight at birth, the association between early energy intake and TAE was no longer significant, but the significant association for early protein intake and TAE weight persisted. Early protein was not associated with TAE length which may be the preferred marker for lean mass. Also, no significant association with TAE HC was seen. Further investigation into how early nutrition is related to weight gain, i.e. perhaps by returning to birth weight more quickly, and whether TAE weight is associated with neurodevelopment for these subjects is warranted.
MOLECULAR BASIS FOR RACIAL DISPARITY IN PREECLAMPSIA

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Background: Preeclampsia (PE) affects 5-8% of pregnancies and is a significant source of perinatal morbidity and mortality. PE results in hypertension and proteinuria after 20 weeks of pregnancy whose increasing severity leads to premature delivery. Mechanisms underlying PE are poorly understood as are risk factors mediating observed racial disparity. The developmental gene Nkx2.5 is expressed in trophoblast cells and abnormal amnions are observed in mice bearing null Nkx2.5 mutations. We assessed Nkx2.5-related gene expression in placenta from normal and PE pregnancies.

Objective: To examine expression levels of Nkx2.5/Nkx2.5 target genes in placenta of normal vs PE patients

Design/Methods: IHC assay of Nkx2.5 and target gene expression and qPCR of mRNA from normal term and PE placenta.

Results: Nkx2.5 and its targets are expressed in syncytiotrophoblast of human placenta with a strong correlation between early onset severe PE (EOSPE) and high levels of placental Nkx2.5 expression more prevalent among whites vs. blacks (n=32). A highly significant correlation was found between high Nkx2.5 and sFt-1 mRNA expression levels (Pearson's r=-.73). Nkx2.5/sFlt-1 levels were also highly correlated to levels of Nkx2.5 target gene Sam68 (Pearson's r=.64). Sam68 regulates mRNA splicing of the type generating sFlt-1 mRNA, suggesting a functional link between Nkx2.5 and induction of PE via sFlt-1, an inducer of PE. Expression of two other Nkx2.5 targets, Ccdc117 and the stress response factor Xbp1 is elevated in white (Ccdc117) or black (Xbp1) PE patients with high Nkx2.5 levels.

Conclusions: Nkx2.5 may play a direct role in the genesis of PE via regulation of Sam68/sFlt-1, and serve as a common link to two other stress-related modifiers of EOSPE.
VARIANT TRICUSPID ATRESIA: A CIRCULATORY DILEMMA

John Romond, MD, Resident-Medicine and Pediatrics, Carol M. Cottrill, MD, Douglas Schneider, MD and William N O’Connor, MD

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In 1979, Freedom reported 6 cases of tricuspid atresia that seemed different from the usual form of this disorder. Rather than a “dimple” in the floor of the right atrium representing the atretic tricuspid valve, this variant had a tricuspid valve, along with its tensor apparatus, but was imperforate. These individuals did not have the left axis deviation seen on the electrocardiograms of the usual type of tricuspid atresia. In addition, children with variant tricuspid atresia also had absence of the pulmonary valve, and “curious” distortion of the left ventricular septum. The septum bulged into the LVOT with varying degrees of obstruction. Upon sectioning the bulging septum, the vascular pattern was distinct from IHSS, in that large sinusoids from the right ventricle were seen in the thickened septum of variant tricuspid atresia. Since this report additional cases have emerged, mandating understanding of the type of tricuspid atresia one is dealing with prior to any surgical intervention. Echocardiography with or without cardiac catheterization should yield the information needed to surgically approach all cases of tricuspid atresia.

Case: We present a 10 year old boy who has this variant of tricuspid atresia. He had a Blalock-Taussig shunt in early infancy, followed by a Glenn operation at 9 months of age. His “Fontan” operation, done at 3 years of age, consisted of a 12mm Gore-tex tubular graft from the inferior vena cava to the right pulmonary artery. He did well with this operation and his growth and development have been normal. His exercise tolerance is mildly impaired, but he does tae kwon do. His echocardiogram documented variant-type of tricuspid atresia. He underwent cardiac catheterization to assess whether or not the 12mm Gore-tex was adequate at his present size.

His cardiac catheterization data and angios will be presented along with the following questions:
1. What is the sequelae of insufficient venous drainage from the lower body?
2. What is the next surgical option? (Is surgery the best option?)
3. What can be done now with his RV and absent pulmonary valve?
4. Does his outlook differ from the “usual” type of tricuspid atresia?
“SPELLS” IN AN OTHEWISE HEALTHY INFANT
Presenter: Dr. Tamika Katherine Rozema, M.D. PGY 2 Pediatric Resident
Co-Authors: Dr. Carol Cottrill, M.D., Dr. William O’Connor M.D.
Institution: University of Kentucky, Lexington, Kentucky

Background
Apparent Life-Threatening Events, ALTE, are what the title describes, events in a infant that are significant to both the child and the caretaker and are comprised of three components: apnea, color change and changes in the muscle tone. Some studies suggest an association or link between history of ALTE and SIDS, particularly when there is an underlying medical condition. With the cause of SIDS still unknown, much research on the causes ALTEs and the work up of ALTEs is very important to not only general pediatricians but also certain subspecialties. An unusual case and cause of an otherwise healthy infant with ALTEs is discussed.

Clinical Summary
A case of an 13 month old infant with “spells” of unconsciousness is presented. The usual workup including labs, EEG, ECG, ECHO, Brain MRI, Holter monitor and admission were unfruitful. The ECHO did show some trabeculations in her left ventricle, but was otherwise unremarkable. After a 2nd normal 24 hour Holter monitor done 6 months later from initial work-up and a normal 48 hours video EEG monitoring, a 3rd 24 hour Holter monitor revealed the etiology of the “spells:” to be a 20 second pause in heart rate with recovery into sinus rhythm. This was treated with a Divine ® pacemaker placement. Her prognosis is uncertain.

Discussion
The work up of “spells” or ALTE, Apparent Life-Threatening Events including: gastrointestinal, neurological, cardiac, metabolic, infection, and child abuse will be discussed. With the ECHO showing trabeculations in the left ventricle, a brief discussion about this newly diagnosed type of cardiomyopathy will be presented.
ECHOCARDIOGRAPHY PARAMETERS PREDICTING DUCTUS ARTERIOSUS PATENCY AT ONE MONTH IN PRE-TERM INFANTS

Name of Presenter: Aisling Mary Smith, MB
Other Authors: Dr. Gene Dempsey

Background
Early diagnosis and effective treatment of a significant Patent Ductus Arteriosus (PDA) remains contentious among clinicians.

Aims & Objectives
To evaluate what echocardiography (ECHO) parameters in the first 48 hours of life predict ductal patency at one month in very pre-term infants.

Methods
Eligible infants had an ECHO performed within 48 hours of birth (ECHO 1) and at one month of life (ECHO 2). Standard ECHO parameters and relevant clinical data were documented. Parental consent was obtained.

Results
52 babies were included in this study. Mean gestation was 29 weeks and birth weight 1141g. Within the first 48 hours of birth ECHO 1 demonstrated that 50 babies have a PDA (96%), of which 19 were large (>2mm) (36%) and 31 were small (60%), and 2 babies have no PDA. At one month ECHO 2 data reported 30 babies still have a PDA (58%), of which 10 were large (19%) and 20 were small (39%), and 22 (42%) of babies had no PDA. Statistical analysis identified that the parameters significantly associated with large PDAs at one month were gestational age, birth weight, ventilation support at 48 hours, ductal size on colour Doppler at 48 hours, systolic and diastolic flow velocity and systolic to diastolic flow ratio.

Conclusion
Very pre-term ventilated neonates with a large PDA and pulsatile flow pattern within 48 hours of life will have a persistent large ductus at one month.
INTESTINAL MICROBIAL COLONIZATION DIFFERS PRIOR TO DIAGNOSIS OF BRONCHOPULMONARY DYSPLASIA

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Background: Inflammation of the developing lung is thought to underlie the pathogenesis of bronchopulmonary dysplasia (BPD). Animal models have shown that the intestinal microbiota facilitate inflammatory response not only at a local (intestinal) level, but also remotely, including lung, and that intestinal commensals can regulate immunity in the respiratory mucosa through the activation of inflammasomes. Thus, microbial colonization of the premature gut may be relevant to BPD, but studies are lacking.

Objective: To examine differences in gut colonization of preterm infants with and without BPD.

Methods: Serial (days of life 4 to 9, 10 to 16, and 20 to 23) stool specimens were collected prospectively from infants ≤29 weeks gestational age. Infants who died or developed necrotizing enterocolitis were excluded. High throughput 16S rRNA sequencing was used to profile stool bacterial communities. BPD was diagnosed by physiologic-BPD criteria; controls were gestationally-age matched infants who survived free of BPD. The microbiome of BPD and control infants was compared for alpha- and beta-diversity, and the relative abundance of microbiota at all taxonomic levels. GEE models were used to examine confounding and analyze repeated samples per infant.

Results: Samples were analyzed from 39 infants: 11 who developed BPD vs 28 controls. The Simpson diversity index was significantly lower in BPD cases than controls for samples from 4 to 23 days (p=0.03). In days 4 to 9 samples, species richness ($S_{\text{chao1}}$) was significantly lower in the BPD infants compared to controls (p=0.02), while control samples had significantly greater relative abundance of the bacterial order Lactobacillales than cases (p=0.008). Results of these analyses were significant after controlling for birth weight, and were not confounded by race, gender or gestational age.

Conclusions: BPD is associated with decreased gut microbial diversity and species richness in the first weeks of life. We speculate that lack of beneficial colonization contributes to BPD by dysregulation of the developing immune system.
A BRIEF HISTORY OF CHRISTMAS DISEASE
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Hemophilia is a rare, sex linked inherited, disorder of coagulation, which probably has been recognized since antiquity. The name Hemophilia was coined by a German physician, F. Hopff. It was popularized since 1893 by Johann Lukas Schönlein, of Henoch Schönlein’s Purpura fame. In 1947 Professor Alfredo Pavlovski demonstrated that there was more than one form of hemophilia, because occasionally the plasma of one patient with hemophilia could correct the clotting defect in another person. In 1952 Drs. Paul Aggeler in the US and Rosemary Biggs in the UK described patients with a form of hemophilia that was distinct from classic hemophilia. Dr. Biggs’ patient had the surname “Christmas.” Dr. Biggs was editor of the British Medical Journal. She published her findings in the December issue of the BMJ and named the new disease “Christmas Disease.” Dr. Aggeler was not that good a salesman and his patient, who had the surname Kincaid, is forgotten. Christmas disease is now generally referred to as Hemophilia B, a defect in Factor IX. Queen Victoria is well known to have carried the gene for hemophilia, which she spread to several royal houses in Europe, most notably to the Russian Imperial family, the Romanovs. They were assassinated after the Russian revolution. Their remains have been found and individually identified. DNA has been extracted. Analysis showed that the gene defect in Tsarevitch Alexis was for Hemophilia B. His sister Anastasia was a carrier. Thus Queen Victoria spread Christmas disease and not classic hemophilia as had always been assumed.
PROGERIA – OLD DISEASE, NEW INSIGHTS.
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Background: Progeria is a very rare genetic disorder that affects 1 in 4-8 million children. There are currently 100 known cases in the world with just as many likely undiagnosed. The hallmarks of Progeria are the physical features, which lead the patient to appear to prematurely and rapidly aged. The diagnosis of Progeria is difficult before these features become prominent. Newborns with Progeria appear healthy with biophysical measurements well within normal. After a few months, these infants fall off their growth curves, develop characteristic skin changes, thinning hair, and loss of adipose tissue around the face causing prominent veins to be exposed on the forehead. These features parallel internal changes that also mimic those of a person far beyond their years with early cardiovascular disease and death by stroke or MI at an average age of 13. Progeria was first described in 1886 but research took an immense leap forward in 2002 when the gene mutation causing the disease was first isolated. This guided the way to understanding the biochemical pathway that produces Progeria from a mutation in the Lamin A gene.

Clinical Summary: Zach’s case is similar to that of others with the disease. He was born at term without complications. Mom described him as “somewhat small and veiny” but otherwise normal. She also noted that the skin on his abdomen was always “tight”. At 9 weeks of age he developed “bumps on his shins” and his doctor was concerned about failure to thrive. A full workup did not reveal any cause for these anomalies until a skin biopsy was done and finally the diagnosis was made. Zach is now 6 years old and fully manifests many of the unique phenotypic features of Progeria. The question for Zach’s family is how to halt the inward changes his body is undergoing. Zach has been treated under a research protocol with a triple combination of medications aimed directly at the pathway leading to Progeria in an effort to slow progression of the disease. Prior to the initiation of treatment, Zach’s coronary arteries and aorta were dilated. Now, after two and half years of treatment the size of his vessels have returned to normal. Zach and his Mom are now joyful advocates for the cause of Progeria and hope to have many more years of a happy, loving, active life for Zach.

Discussion: Over the last several years a research trial has been underway using 3 drugs known to inhibit various parts of the pathway in the production of Progeria, the abnormal protein which distorts the nuclear membrane of cells and leads to the manifestation of disease. These are a statin, bisphosphonate and farnesyltransferase inhibitor. It is difficult at this time to know whether the normal results from Zach’s last cardiac ECHO are a result of the therapy or just a normal part of the progression of the disease.
PERCEPTIONS OF DIABETES PATIENTS, THEIR PARENTS AND HEALTHCARE PROFESSIONALS ON DIABETES TRANSITION IN THE WEST OF IRELAND: THE MATER STUDY

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Background: A structured transition process for young adults with diabetes is increasingly advocated when they move from a paediatric to an adult service. The American Diabetes Association (ADA) recently produced guidelines on effective diabetes care transition from paediatric to adult diabetes care systems and Forbes et al. have identified seven dimensions of continuity which they consider to be relevant during the transition process. To date, there has been no formal evaluation of the transition process in the Irish context, where paediatric and adult diabetes services are often completely independent entities with little linkage. Research into perceptions of healthcare users and providers involved in this process also lacks formal investigation. This qualitative study describes healthcare users and providers’ perceptions on the current diabetes transition process and facilitators and barriers to smooth transition in hospitals in the West of Ireland.

Methods: On receipt of ethical approval, semi-structured interviews were conducted with 3 stakeholder groups (young adults, parents of young adults within 2 years of transitioning and healthcare professionals). Participants were purposively selected. Participation was voluntary and informed consent was obtained. To date, young adults (n=4), parents (n=5), and healthcare professionals (HCP) (n=9), have been interviewed at 2 of the 3 hospitals.

Results: Whilst accepting the transition process, participants expressed concerns with communication between healthcare users and providers, notably around helping young adults appreciate the transition process and helping both young adults and parents adjust to their changing roles in diabetes care. While parents appreciate their children’s right to autonomy, many felt their involvement was still needed “…a young adult doesn’t have the maturity…” but not accommodated within the current transition structure; “…they kind of don’t encourage us to come…” Users strongly perceived lack of acknowledgement by HCPs of the support still provided to the young adult by parents during diabetes transition; “…Goodbye Mother!” is the first thing.”

Communication between paediatric and adult diabetes teams was perceived to be facilitated by having a common key team member, most notably the diabetes nurse. Participants perceived that clinic structure should be age-banded within both paediatric and adult services and that the clinical environment should be age-appropriate and formally introduced to the users; “…they’re at a clinic with 2 year olds, and they’re - it really brings them down and they really hate it and it is wrong.” HCPs and parents perceived that psychological support would enhance a smoother transition; “the psychologist is really, really important.”

Conclusion: This study identified perceived barriers to the diabetes transition process in participating hospitals. There is a need to extend this study nationally to identify if these issues are replicated in diabetes services throughout Ireland. Additionally, this study informs the restructuring of the diabetes transition process in the West of Ireland, notably in the dimensions of effective communication between healthcare users and providers, identification of a key team member common to both diabetes teams, provision of age-banded clinics and services and possible provision of on-going psychological support throughout the transition process. The smooth transition of the parent as a key team member in their child’s transition from teen to young adult diabetes care requires further study.
Abstract:
Metabolic syndrome is becoming an increasingly prevalent disease in the pediatric population in South Carolina and is strongly associated with early hypertension and type II diabetes. Elevated fasting glucose (> 100mg/dL) and elevated fasting insulin (> 20 mcU/mL) are markers for glucose intolerance that may lead to type II diabetes. In this cross sectional study we analyzed the relationships between easily measured anthropometric data and fasting glucose and insulin levels. The anthropometric measurements used were waist circumference (cm) and body mass index percentile as calculated by an algorithm using CDC data. Our population was an obese pediatric cohort (n=269) aged 4 to 21 from the Pediatric Metabolic Syndrome Study at the Medical University of South Carolina in Charleston, SC. To analyze the relationships we used Pearson correlation coefficients that showed waist circumference had a significantly stronger correlation with fasting glucose and insulin than body mass index percentile. Specifically, the correlation value of 0.482 between fasting insulin and waist circumference was significant at the 0.01 level, and the correlation value of 0.145 between fasting glucose and waist circumference was significant at the 0.05 level. In comparison, the correlation value of 0.140 between fasting insulin and body mass index percentile was significant at the 0.05 level, and the correlation value of 0.035 between fasting glucose and body mass index percentile was not significant. The difference in correlation values between the relationships of body mass index percentile and waist circumference demonstrates that waist circumference may be a better indicator of glucose intolerance in an obese pediatric population. Further research is needed to develop waist circumference percentile data in an obese pediatric population so that more specific guidelines can be given to healthcare professionals.

Presentation Preferred: Poster

This project was supported by the South Carolina Clinical and Translational Research (SCTR) Institute at the Medical University of South Carolina, NIH/NCRR grants ULI RR029882 and ULI TR000062.
POST-TRANSFUSION RED CELL SURVIVAL OF NEONATAL AUTOLOGOUS AND ADULT DONOR BIOTIN LABELED RBCS, MEASURED CONCURRENTLY, ARE NOT DIFFERENT

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Background. Determination of red cell survival (RCS) in infants is important for its clinical relevance to the evaluation of erythropoiesis in defining mechanisms of neonatal anemia. Multi-density biotin labeling of RBCs in adults can be used to safely determine RCS of two or more RBC populations concurrently (Mock et al. Transfusion 51:158, 2011).

Objective. To compare RCS of neonatal autologous RBC with adult donor RBC in critically ill very low birth weight (VLBW) infants after clinical RBC transfusion. Our hypothesis was that RCS of neonatal autologous RBCs — measured by Bio-RBCs + pharmacokinetic (PK) modeling — is substantially shorter than RCS of adult donor RBCs.

Methods. Separate populations of neonatal autologous and adult allogeneic RBCs were labeled at two discrete biotin densities. Both RBC populations were transfused into five premature infants (mean ± SD: birth wt: 852±163 g, gest age: 26.0 ± 0.3 wk, study age: 2.4 ± 1.9 d). Serial samples (10 µL discarded blood) were analyzed for BioRBC enrichment by flow cytometry and RCS (ie, RBC life span) calculated using PK modeling adjusted for confounders: laboratory phlebotomy, RBC transfusion, and growth (assuming constant blood volume per kg). This lifespan model assumes disposition of transfused BioRBCs in the absence of phlebotomies, was lifespan-based (ie, due to senescence) and does not vary over time. Transfused BioRBCs were assumed to have been produced under steady state.

Results. Serial RCS results for a representative infant are shown. RCS for neonatal and adult RBCs were similar: 67 ± 7 and 74 ± 11 d, respectively, P=0.29.

Conclusions. Contrary to our hypothesis and to previous infant studies, RCS of adult and neonatal BioRBC, measured concurrently, were not different. This study demonstrates utility of concurrent multi-density BioRBC + PK modeling in determining in vivo RCS of multiple populations by flow cytometry. (Supported by NIH HL046925 & Thrasher Research Fund.)
ABSTRACT

Background:
Approximately every 1 in 600 people is a carrier of a balanced translocation. These individuals are at increased risk for infertility, miscarriage, stillbirth or a child born with physical and cognitive delays, including mental retardation. When the translocation involves a critical region regulating imprinting or an area of imprinted genes, the phenotype of the affected child varies according to maternal or paternal inheritance of the translocation.

Case:
We report a case of a paternally derived unbalanced translocation detected on microarray and confirmed with karyotyping. High resolution chromosome testing indicate 46,XX, der (13)t(11:13)(p15.4;q32.1) from amniocytes. Microarray analysis show the derivative chromosome 13 results in a gain of the short arm of 11 approximately 5.82Mb in size including 11p15.4-pter and a loss of the long arm of chromosome 13 approximately 18.05Mb in size including 13q32.1-qter. The duplication results in partial trisomy 11p, in a region containing at least 173 genes some of which are implicated in the etiology of Beckwith Wiedemann and beta thalassemia. The deletion results in partial monosomy of 13q, involving at least 89 genes, and aberrations in the area are implicated in holoprosencephaly. Our patient was delivered via emergent CS at 31 weeks and 4 days gestation and proved to be initially hemodynamically unstable with hypoglycemia and required mechanical ventilation. BWS was confirmed via methylation studies. The child has bony changes of her spine, including hemivertebrae and butterfly vertebrae, and 12 sets of ribs. She has no abdominal or liver tumors, but her AFP remained elevated. MRI of her brain shows multiple anomalies. At the time of submission, she is 5 months old, vent dependent via tracheostomy, and continues to be hospitalized.

Discussion:
This is a translocation not previously described and includes a medical literature review of duplication 11p and deletion of 13q. Potential effects of this unbalanced translocation are discussed, given our knowledge of the genes involved and the clinical phenotype observed. Our hope is to offer information regarding prenatal and neonatal findings associated with this novel translocation and the implications of the increased use of prenatal microarray.