8th Annual
Pediatric Science Days

April 3 & 4, 2014

Department of Pediatrics
Saint Louis University

Visiting Professor

M. Michele Mariscalco, MD

Regional Dean
University of Illinois
College of Medicine
Urbana, IL

We thank the following for their valued and continued support of Pediatric Science Days
Cardinal Glennon Children’s Medical Center

This program is supported in part through restricted educational grants from the following:

Abbott Nutrition, a division of Abbott Laboratories, Inc.

Mead Johnson Nutritionals

Congratulations to the Presenters and their Mentors!
7:30 AM  Breakfast and Poster Viewing

7:55 AM  Welcome, Dr. Robert W. Wilmott
IMMUNO Professor and Chair

Opening Remarks, Dr. Joyce Koenig
Professor, Pediatrics and Molecular Microbiology & Immunology
Coordinator, Pediatric Science Days 2014

8:00 AM  PLATFORM SESSIONS

1. Basic Science Research
   Moderator: Robert E. Fleming, MD (Neonatology)

8:00  Regulation of Gene Transcription and Development by Sister Chromatid Cohesion Proteins
      Dale Dorsett, Ziva Misulovin, Maria Gause, Amanda Koenig, Cheri A Schaaf

8:15  The Development of a Less Immunogenic Protein for Enzyme Replacement Therapy of
      Morquio A Disease Alexandria Lee, Catalina Sosa, Adriana M Montaño

8:30  Vitamin D Receptor Deficiency Underlies Genomic Instability and Premature Senescence
      in Hutchinson Gilford Progeria Syndrome Arindam Das, Martin A. Neumann,
      Ray Kreienkamp, Adriana Dusso, Susana Gonzalo

8:45  Normal Regulation of Hepatic Atoh8 MRNA by Dietary Iron in Mice Requires Transferrin
      Receptor 2 And Hfe Yihang Li, Qi Feng, Molly Rozier, Mary C. Migas, Robert E. Fleming
9:00 Increased Prostaglandin D2 is Associated with Delayed Dendritic Cell Maturation in Neonates during Respiratory Viral Infection. Brunda Tumala, Somashubra Bhattacharya, Laurie Shornick

9:15 Age-Dependent Analysis of Inflammatory Markers in Cartilage Tissue of Morquio A Mice. Shane Grace, Catalina Sosa, Adriana Montano

9:30 Coffee Break and Poster Session

2. Educational and Social Science
Moderator: Donna R. Halloran, MD (General Academic Pediatrics)

10:00 Social Skills Training Reduces Victimization and Ostracism Experiences in Adolescents with ASD, ADHD and Behavior Problems - A Preliminary Study. Jennifer Heithaus, Morgan White, Barbara Braddock, Kimberly Twyman

10:15 Happy Mothers, Healthy Families: Feasibility of Text Messaging as an Adjunct Treatment for Postpartum Depression. Matt Broom, Amy Ladley, Gene LaBarge, Elizabeth Rhyne, Rashmi Narayan, Ashley Borawski, Donna Halloran

10:30 Text4peds: Text Messaging Curriculum for Pediatric Residents. Lauren Draper, Matt Broom, Greg Adams


11:00 Rapid Bedside Triage in a Pediatric Emergency Department. Robert G Flood, Paula Szwargulski, Nadeem Qureshi, Mary Bixby, Steven Laffey, James M Gerard

11:15 Feasibility Report of the Palliative and End-of-Life Communication Intervention for Parents of Children with a Brain Tumor. Verna Hendricks-Ferguson, Joan Haase, Javier Kane, Kamnesh Pradhan, Shih Chie-Schin, Karen Gauvain
11:30 AM Symposium Luncheon

3. Clinical Research I
Moderator: Ajay Jain, MD (Gastroenterology & Hepatology)

1:00 Skin and Soft Tissue Infections in Children: Analysis of Risk Factors for Admission and Clinically Important Complications Jaime L Velasco Masson, Nirupama Kannikeswaran, Usha Sethuraman, Marwan Daoud, and Gina Nieto

1:15 Clinical Co-Morbidities, Ablative Site Placental Calcification, and Vascular Remodeling in Twin-To-Twin Transfusion Syndrome With and Without Selective Fetoscopic Laser Photocoagulation (SFLP) Sarah E Starnes, Philip Fitchev, Constance Thorpe, Emanuel Vlastos, Suwan Mehra, Mona Cornwell, Susan E Crawford


1:45 A 20 Sec Sustained Inflation did not Alter Markers of Lung Injury in Surfactant-Treated Fetal Lambs Noah Hillman, Matthew Kemp, Yuichiro Miura, Suhas Kallapur, Alan Jobe

2:00 PM Coffee Break

4. Clinical Research II
Moderator: Debra H. Zand, PhD (Developmental Pediatrics)

2:30 Administration of Iron-Replete Lactoferrin to Pre-Weanling Mice Increases Liver Hepcidin Expression Jeffrey A Cooper, Elif Memisoglu, Princy Prasad, Yihang Li, Robert E Fleming

2:45 Deformational Scaphocephaly Results in Increased Therapy Duration and Less Effective Cranial Index Correction than other Types of Deformational Plagiocephaly Sarah A. Donigian, Emma M. Kulig, Chelsea R. Horwood, Alexander Y. Lin
3:00  CPAP Therapy Adherence in Children with Obstructive Sleep Apnea
       Shalini Paruthi, Angela Orlando, Raman Malhotra, Eric Armbrecht

3:15  Evidence Based Recommendations for the Treatment of Children with Hydrocephalus
       Ann Marie Flannery, Catherine Mazzola, Paul Klimo, David Limbrick, Lissa Baird, Tina
       Duhaime; Joanna Kemp, Mandeep Tamber, Dimitios Nikas, Mark Van Poppel, Laura Mitchell

9:30 AM   POSTER SESSION

1. Determination of Specificity and Sensitivity of a New Lc/Ms/Ms Method for Newborn
       Screening of Mucopolysaccharidoses Qi Gan, Mary Campbell, Adriana M Montaño

2. A Novel, Multidisciplinary Approach to Treatment of a Case of Cellular Neurothekeoma of
       the Lip in a Pediatric Patient Kyle Y. Xu, and Christina Plikaitis

3. Wolman Disease – Developing Management Approaches. A Case Report and Literature
       Review. José A. Torres Garcia, Stephen R. Braddock, Jeremy S. Garrett

4. Chemical Genetics Identifies Bet Proteins as Potential Therapeutic Targets in
       Facioscapulohumeral Muscular Dystrophy. Francis M. Sverdrup, Matthew P. Yates,
       Jon Oliva, Marshal Huang, Stephen J. Tapscott

5. Vitamin D Rescues DNA-Repair Defects in a Mouse Model of Progeria. Ray Kreienkamp,
       Arindam Das, Martin Neumann, Susana Gonzalo

6. Reported Communication Ability in Persons With Trisomy 18 and 13
       Cheryl A. Liang, Barbara A. Braddock, Jennifer L. Heithaus, Katherine M. Christensen,
       Stephen R. Braddock, John C. Carey

7. Engaging Residents in Pediatric Hospital Medicine – A Novel Elective
       Marta King, Victoria Wilkins, Mara Chavolla Calderon

8. Comparing Two Ambulatory Pediatric Clerkship Scheduling Models
       Rochelle M. Remus, Marta King, Jamie Sutherell

9. Resident Perception of an Audience Response System in Graduate Medical Education
       Amol Purandare, Angela Delecaris, Laura Holt, Matt Broom

10. Does the Degree of Lymphopenia Induced by Atgam Impact Outcomes in Pediatric Lung
       Transplant Recipients with Bronchiolitis Obliterans?
       Josie Vitale Lorbert, Charles Huddleston, Stuart Sweet, Albert Faro
11. Isoprostanes as Physiological Mediators of Transition to Newborn Life: Novel Mechanisms Regulating Patency of the Term and Preterm Ductus Arteriosus
   Jian-Xiong Chen, Patrick W. O'Mara, Stanley D. Poole, Naoko Brown, Noah J. Ehinger, James C. Slaughter, Bibhash C. Paria, Judy L. Aschner, Jeff Reese

12. Horizontal Transmission of Neonatal Hsv: A Unique Case Involving Eczema Herpeticum
   Hayley Friedman, Marya Strand, Justin Josephsen, and Jeffrey Cooper

   Aaron Pitzele, Eric S Armbrecht, Mohammad Rahimi, and Thomas Havranek

14. Development and Validation of a Novel Ultra-Mobile Ambulatory Total Parenteral Nutrition Model
   Ajay K. Jain, Keith S. Blomenkamp, Jonathan Rodrigues, Timothy A. Blaufuss, Mike A. Carl, Victor D. Liou, Joy X. Wen, Barbara Stoll, Douglas Burrin, John P Long, Jeffrey Teckman

15. Infant Sleep Safety on a Pediatric General Medical Unit. David A. Wathen

16. Voiding Symptoms in Pediatric Patients with Elevated Calcium-Creatinine Ratios
   Kyle Spradling, Barry Duel

17. Chemotherapy as a First Line Treatment for a Subtotally Resected Rosette-Forming Glioneuronal Tumor of the Fourth Ventricle in a 6-Year old child: Case Report and Literature Review of Pediatric Cases Cynthia Morris, Yongxin Chen, MD, PhD, Vilaas Shetty, MD, Samer Elbabaa, MD, Miguel Guzman, MD, Mohamed S. Abdel-Baki

18. The Report of a Case Series: Pediatric Pseudoaneurysm with Absent to Minimal History of Trauma Arya Namin, Christina Plikaitis

19. Resolution of Precocious Puberty Following Surgical Resection of Fourth Ventricular Medulloblastoma: Case Report and Review of Literature
   Renata G Medina, David P Dempsher, Karen M Gauvain, Thomas J Geller, Samer K Elbabaa

20. Age of Initiation of Helmet Therapy Does Not Improve Duration of Treatment, Rate of Improvement or Final Outcome in Children with Deformational Plagiocephaly
   Chelsea R. Horwood, Emma M. Kulig, Sarah A. Donigian, Alexander Y. Lin
21. Effects of Torticollis on Helmet Therapy for Deformational Plagiocephaly
   Emma M. Kulig, Sarah A. Donigian, Chelsea R. Horwood, Alexander Y. Lin


23. Perceptions and Impacts of Childcare Responsibilities on Healthcare Workers Following a Disaster Rachel L. Charney, Terri Rebmann, Robert G. Flood

24. A Fixed-Dose Ketamine Protocol for Adolescent Sedations in a Pediatric Emergency Department Megan Street, James Gerard

25. The Use of a Pediatric Migraine Practice Guideline in an Emergency Department Setting Albert Nakanishi, Courtney Kaar, and James Gerard

26. Critical Care Interventions for Asthmatic Patients Admitted from the Emergency Department to the Pediatric Intensive Care Unit Kristen Cundiff, James Gerard, Robert Flood

27. Analysis of Pediatric Atopic Dermatitis: The Relationship Between Disease Progression and Total IgE/Percent Positive Immunocaps, Year 2 Experience-Combined Results Courtney Tobin, Hala Adil, Niraj Butala, Eric Armbrecht, Elaine Siegfried

28. Analysis of Immune Function in Children with Eczema Stephanie Frisch, Michael Ansstas, Elaine C. Siegfried


30. Charge Syndrome Clinical Database Project (CSCDP): Early Experience of a Web-Based, Parent-Report Database and Registry Meg Hefner, Emily Fassi, Kevin Ballard
Platform Session I.
Basic Science

Moderator:

Robert E. Fleming, MD
Professor
Department of Pediatrics
REGULATION OF GENE TRANSCRIPTION AND DEVELOPMENT BY SISTER CHROMATID COHESION PROTEINS

Cornelia de Lange syndrome (CdLS) is characterized by diverse developmental deficits, including limb and organ structural abnormalities, intellectual impairment, and autism. CdLS is caused by sporadic dominant mutations in genes that affect the function of the cohesin protein complex. Cohesin is a ring-like complex that holds sister chromatids together until cell division in order to ensure accurate chromosome segregation. CdLS is caused by partial decreases in cohesin function that do not noticeably alter sister chromatid cohesion or chromosome segregation. Studies in model organisms and human cell lines show that these partial decreases in cohesin function alter expression of several key genes that control growth and development. Using a combination of genetic, molecular and genomic methods in Drosophila, we find that cohesin binds and controls the transcription of many genes. Cohesin has multiple roles in regulating gene transcription. These include facilitating activation by transcriptional enhancers, and controlling the levels of Polycomb epigenetic silencing complexes. The numerous effects on gene transcription caused by minor changes in cohesin function likely underlie the severe birth defects associated with CdLS.
THE DEVELOPMENT OF A LESS IMMUNOGENIC PROTEIN FOR ENZYME REPLACEMENT THERAPY OF MORQUIO A DISEASE, Alexandria Lee¹, Catalina Sosa², Adriana M Montaño²,³
¹ School of Medicine, ² Dept of Pediatrics, ³ Dept of Biochem. and Mol. Biol., Saint Louis University.

Introduction: Preclinical trials of enzyme replacement therapy (ERT) in Mucopolysaccharidosis IVA (Morquio A disease, MPS IVA) has been shown to successfully treat the disease, but is accompanied by a large immune response that significantly decreases the efficacy of this treatment.

Aims: The aims of this study are to bioengineer a GALNS protein with reduced immunogenicity without affecting the biological activity for effective ERT of MPS IVA; analyzed via *in silico* predictions and *in vitro* studies.

Materials and Methods: The original GALNS amino acid and cDNA sequences were compared to the corresponding altered sequences in a variety of *in silico* programs. Upon creating different combinations of amino acid substitutions, considerations included (1) polarity, (2) amino acids with unique structures, and (3) already known mutations of Morquio A disease. We evaluated predictions of the immunogenicity, post-translational modifications, physico-chemical properties, and 3D structural evaluation of molecular docking. *In silico* analyses were performed in the new sequences harboring various combinations of substitutions on various immunodominant peptide regions within the whole GALNS protein.

Results: 324 mutated sequences were created and narrowed to 7 sequences after analyzing (1) the immunogenicity predictions, (2) the predictions of phosphorylation sites, N-glycosylation sites, and physico-chemical properties, and (3) three-dimensional visualization of molecular docking. Sequences were selected if the altered sequence showed zero or insignificant changes when compared to the original GALNS sequence. The distances between the active site residues and the corresponding ligand were measured in angstroms. Mutated sequences were chosen if their measured distances within the active site were very close to or even smaller than the measured distance in the original GALNS sequence, indicating a greater likelihood to bind. The cDNA sequence was used for the determination of restriction sites and mutagenesis of the 7 selected sequences. Restriction enzymes for the selected sequences were chosen if they differed from the restriction enzymes at the corresponding sites on the original GALNS sequence; this allows slicing of a different number of fragments with the application of the same restriction enzyme to both the original and altered GALNS for use in *in vitro* studies.
VITAMIN D RECEPTOR DEFICIENCY UNDERLIES GENOMIC INSTABILITY AND PREMATURE SENESCENCE IN HUTCHINSON GILFORD PROGERIA SYNDROME

Arindam Das,1 Martin A. Neumann1, Ray Kreienkamp1, Adriana Dusso2, Susana Gonzalo1

1Edward A. Doisy Department of Biochemistry and Molecular Biology, St Louis University School of Medicine
2Institute of Biomedical Research in Lleida (IRBLLEIDA). Universidad de Lleida (UdL), Spain

A-type lamins are components of the nuclear lamina, a proteinaceous meshwork underlying the inner nuclear membrane. Mature lamin A/C proteins arise through alternative splicing of the LMNA gene and post-translational modifications. Over 300 mutations have been identified in the LMNA gene that cause human degenerative diseases ranging from muscular and adipose tissue dystrophies to premature aging syndromes such as Hutchinson Gilford Progeria Syndrome. HGPS is a rare but devastating disease, with patients developing severe growth abnormalities by 2 years of age. These children continue to exhibit aging characteristics, and die due to atherosclerosis and cardiovascular complications in their teens. In HGPS patients, a truncated lamin A protein is produced, known as Progerin, which is toxic for the cells. HGPS cells show nuclear morphological abnormalities, epigenetic alterations, sensitivity to mechanical stress, and increased genomic instability. Our studies aim to understand the molecular mechanisms responsible for genomic instability in HGPS patients. Earlier we reported that loss of lamin A/C triggers transcriptional downregulation of BRCA1 and RAD51, proteins that are critical for DNA repair by homologous recombination. Here we show that fibroblasts from HGPS patients have greatly reduced levels of the vitamin D receptor (VDR), and that this is responsible for the downregulation of BRCA1 and RAD51. Vitamin D treatment of cells from progeria patients restores BRCA1 and RAD51 levels in a VDR-dependent manner, rescuing DNA repair deficiencies and premature entry into senescence. Thus, vitamin D emerges from this study as an exciting new therapeutic strategy for laminopathies such as progeria that are characterized by genomic instability, due to the loss of key factors in DNA repair, and premature senescence.
NORMAL REGULATION OF HEPATIC ATOH8 mRNA BY DIETARY IRON IN MICE REQUIRES TRANSFERRIN RECEPTOR 2 AND HFE

Yihang Li1, Qi Feng1, Molly Rozier1, Mary C. Migas1, Robert E. Fleming1,2
Departments of Pediatrics1 and Biochemistry & Molecular Biology2, Saint Louis University, Saint Louis, MO 63104, USA.

Background: Atoh8 is a member of the bHLH family of transcription factors. Hepatic expression of Atoh8 mRNA in mice is upregulated by dietary iron and down-regulated by stimulation of erythropoiesis, in association with parallel changes in Hamp1 mRNA. Atoh8 exogenously expressed in cell lines activates hepcidin promoter-reporter constructs by interacting with E box elements in the Hamp1 promoter. Transferrin receptor 2 and Hfe each contribute in the regulation of liver hepcidin expression by iron. The mechanism of signaling between Hfe, Tfr2 and Hamp1 is unknown, but interfaces with the BMP/Smad pathway. Whether Hfe and Tfr2 affect Atoh8 expression is unknown.

Objective: We analyzed the relationship between dietary iron and hepatic expression of Atoh8 mRNA in wild type mice and with disruption of Hfe, Tfr2, and both genes.

Design/Methods: Wild type Hfe knockout mice, Tfr2 (Y245X) mutant mice, and HFE/Tfr2 mice (n=3-5, female) on an FVB background were placed on diets containing 60 ppm or 25,000 ppm iron from the time of weaning at 21 days for two weeks. mRNAs for Hamp1, Atoh8, Bmp6, and Id1 were measured by real-time RT-PCR and normalized to β-actin. Tissue iron concentrations (liver, spleen) were measured.

Results: Wild type mice demonstrated the expected increases in Bmp6, Hamp1, and Atoh8 expression in response to dietary iron loading. Hfe knockout and Tfr2 knockout mice each demonstrated lower Hamp1 and Atoh8 expression than did wild type mice, despite hepatic iron loading. Hamp1 was increased in the HH mice with dietary iron loading; however Atoh8 expression was not statistically different. Mice with combination of Hfe knockout with Tfr2 knockout demonstrated further suppression of Hamp1 expression, but no combined effect was observed on Atoh8 expression.

Conclusions: These results are consistent with a role for Hfe and Tfr2 in normal signaling between iron status and Atoh8 as well as Hamp1 gene expression. The observations moreover suggest that dietary iron can at least in part regulate Hamp1 independent of a change in Atoh8 expression.
INCREASED PROSTAGLANDIN D2 IS ASSOCIATED WITH DELAYED DENDRITIC CELL MATURATION IN NEONATES DURING RESPIRATORY VIRAL INFECTION

**Brunda Tumala¹, Somashubra Bhattacharya¹, Laurie Shornick¹⁻²**

¹Department of Biology, ²Department of Molecular Microbiology & Immunology Saint Louis University, St. Louis, MO

During respiratory viral infection in adults, inflammatory cytokines stimulate lung conventional dendritic cells (cDC) to increase expression of MHC class II molecules, co-stimulatory molecules, and chemokine receptors. Activated cDC then migrate to the draining lymph nodes where they present antigen to T-lymphocytes. Our previous studies demonstrated that neonatal mice exhibit reduced inflammation and inflammatory cytokine expression during paramyxoviral infection. Thymic stromal lymphopoetin (TSLP) is a cytokine known to participate in cDC activation. We observed significantly lower expression of TSLP in neonatal mice during respiratory viral infection compared to adults. Adult cDC showed increased expression of MHC Class II and Chemokine Receptor 7 (CCR7) post infection days 1, 3, and 7. However, these events were delayed in neonates. Interestingly, studies in geriatric mice suggest that high levels of Prostaglandin D2 (PGD2) may inhibit cDC maturation and migration to lymph nodes. Thus, we hypothesized that increases in PGD2 expression may be associated with delayed cDC maturation and migration to the lymph nodes in neonates post paramyxoviral infection. Adult and two-day old neonatal C57BL/6 mice were inoculated intranasally with Sendai virus (500 pfu/g body weight). Expression of PGD2 in bronchoalveolar lavage fluid (BAL) was measured post infection days 1, 2, 3, and 5. We observed higher PGD2 expression in neonatal BAL fluid compared to adults during paramyxoviral infection, thus supporting our hypothesis that higher PGD2 in neonates may contribute to the impairment of cDC maturation in neonates.
**AGE-DEPENDENT ANALYSIS OF INFLAMMATORY MARKERS IN CARTILAGE TISSUE OF MORQUIO A MICE**  
*Shane Grace, Catalina Sosa, Adriana Montano*

**Abstract:** The mucopolysaccharidoses (MPS) are inherited metabolic disorders resulting from defective catabolism of glycosaminoglycans (GAGs). MPS IVA, also called Morquio A syndrome, involves a mutated or otherwise deficient lysosomal N-acetylgalactosamine-6-sulfate sulfatase (GALNS) enzyme needed to breakdown two GAGs, keratan sulfate and chondroitin-6-sulfate. Clinical manifestations and previous biochemical studies indicate cartilage tissue is a main source of pathology in Morquio A patients. We used a Morquio A mouse model to investigate the relationship of autophagy, apoptosis and inflammation within cartilage tissue. We performed gene analysis using qRT-PCR and found that inflammatory, apoptotic and autophagic markers were differentially expressed in an age-dependent manner. Specifically, we saw a significant up-regulation of *Dapk1*, a positive mediator of interferon-gamma induced cell death in our 12-month-old Morquio A mice compared to age-matched wild-type controls. Several other autophagy- and apoptosis-related genes displayed significant up-regulation with increasing age. These findings suggest that several inflammatory cytokines could modulate autophagy and apoptosis in cartilage tissue of Morquio A mice. These data could be used in developing future treatment modalities for bone growth abnormalities and/or biomarkers in Morquio A patients.
Platform Session II.
Education and Social Sciences

Moderator:

Donna Halloran, MD
Associate Professor
Department of Pediatrics
SOCIAL SKILLS TRAINING REDUCES VICITMIZATION AND OSTRACISM EXPERIENCES IN ADOLESCENTS WITH ASD, ADHD AND BEHAVIOR PROBLEMS – A PRELIMINARY STUDY
Jennifer Heithaus, Morgan White, Barbara Braddock, Kimberly Twyman.
Department of Pediatrics, Saint Louis University School of Medicine, St Louis, MO.

Background: Adolescents that have ASD, ADHD and other communication obstacles have been shown to be more at risk for peer bullying, victimization and ostracism (PBVO) [Sterzing et al 2012; Twyman et al 2010]. Social skills training by the UCLA Program for the Education and Enrichment of Relational Skills (PEERS) has been shown to improve social interaction in this population [Laugeson et al 2011]. It is unknown if such intervention impacts PBVO.

Objective: To determine if social skills training reduces PBVO in adolescents with ASD & other behavior problems.

Design/Methods: 30 adolescents (12-18 y/o), 29 with ASD and co-morbidities of ADHD (n=24), depression (n=4), anxiety (n=8) and communication problems (n=3), completed PEERS in 4 cohorts from 9/2012-7/2013. Group means on the Children's Communication Checklist-2 (Bishop 2006) included Communication Composite Score of 74.2 and Social Interaction Difference Index of -9.29. Pre- and post-intervention assessment was by standardized self-report of 1) bully/victim experiences on the Bully Victimization Scale (BVS, Reynolds 1993) and 2) ostracism on the Bullying & Ostracism Screening Scale (BOSS, Saylor 2012). Mean change due to intervention was measured by paired t-tests.

Results: Post intervention, participants showed significant reduction in ostracism experiences on the BOSS (pre M=8.77, SD=2.56; post M=7.80, SD=3.09; t(29)=2.276, p=0.03) and in BVS victimization percentile (pre M=64.57, SD=29.76; t(29)=2.468, p=0.02). By BOSS report, participants also felt less ignored (pre M=2.77, SD=1.14; post M=2.27, SD=1.11, t(29)=2.812, p=0.009) and less disconnected from others (pre M=2.87, SD=1.07; post M=2.27, SD=1.17; t(29)=3.17, p=0.004). Also, a subset clinically significant for victimization pre-PEERS showed significant improvement in BVS victimization t-score (pre M=79.14, SD=10.65; post M=62.86, SD=18.87; t(6)=3.30, p=0.016), BOSS ostracism belonging factors (pre M=9.43, SD=3.41; post M=6.71, SD=2.69; t(6)=2.55, p=0.043) and BOSS total score (pre M=45.86, SD =15.09; post M=37.14, SD=10.33; t(6)=2.53, p=0.045).

Conclusions: The PEERS social skills training program shows promise in improving participants’ PBVO experiences. Additional data collection/analysis with current program participants is in progress.
HAPPY MOTHERS, HEALTHY FAMILIES: FEASIBILITY OF TEXT MESSAGING AS AN ADJUNCT TREATMENT FOR POSTPARTUM DEPRESSION

Matt Broom1, Amy Ladley1, Gene LaBarge1, Elizabeth Rhyme1, Rashmi Narayan1, Ashley Borawski1, Donna Halloran1

1Department of Pediatrics, Saint Louis University.

Background: Postpartum depression (PPD) occurs in 13-20% of women and is the most common medical problem in new mothers. Common barriers to care include affordability of and access to mental health services, transportation concerns, and child care during therapy. Text messaging is an established medium to assist with behavioral change with the potential to further reinforce the therapeutic message between counseling sessions.

Objective: In an urban, academic pediatric clinic, assess the feasibility of sending text messages to mothers with PPD.

Design/Methods: Prospective study of mothers 1 week to 6 months postpartum in a large pediatric clinic (16,000 visits/year) from February-October 2013. Mothers at risk for PPD (≥10 on the Edinburgh Postnatal Depression Scale [EPDS]) enrolled to receive counseling and optional text messaging for 6 months. Texts were informational, motivational, or reflective statements sent out 4 days/week. One-third of texts offered an option for mothers to respond or request a call back. Individual text messaging use and consistency of access to cellular service was evaluated pre-intervention; the effect of the texting program was surveyed post-intervention.

Results: 2007 women completed an EPDS and 7.5% (n=167) scored “at risk.” Among mothers at enrollment (86% Black, 91% with income of < $25K/year), mean EPDS was 13.5 +/- 3.4 and Beck Depression Inventory-II was 23.1 +/- 10.4 (moderate depression). 35 women enrolled, and 91% (n=32) received text messages. 1520 texts were sent, with 9.1% (n=139) having delivery failure. 49% (n=17) experienced message failure at least once; however one mother accounted for 44% (n=61) of failed messages (due to inability to afford consistent cellular service). Although only 2.2% (n=34) of all texts were responded to, 71% of the responses requested a call back. Individual text messaging use and consistency of access to cellular service was evaluated pre-intervention; the effect of the texting program was surveyed post-intervention.

Conclusions: Text messaging is a feasible and welcomed method of communication among minority, low-income mothers suffering from PPD and holds promise as a low-cost, adjunctive mental health therapy.
TEXT4Peds: TEXT MESSAGING CURRICULUM FOR PEDIATRIC RESIDENTS
Lauren Draper1, Matt Broom1, Greg Adams1
1Pediatrics, St. Louis University, St. Louis, MO, United States

Background: The duty-hour restrictions implemented in 2011 by the Accreditation Council for Graduate Medical Education have raised considerable concerns regarding the duration and quality of education for residents. It is imperative for residency programs to address educational concerns related to duty-hour restrictions and to find viable alternative methods to educate residents. One potential avenue of maximizing educational yield is to cater to residents' preferred methods of communication. Text messaging is a powerful potential avenue for reaching residents quickly, but its utility in educating residents has hardly been evaluated. The high prevalence of text messaging among residents creates an opportunity for a new approach to resident education, particularly for board review purposes.

Objective: Evaluate the feasibility of texting educational messages to residents, and resident satisfaction with, and perceived usefulness of, a text message-based curriculum to teach pediatric board-relevant information.

Methods: A prospective study with pediatrics and combined medicine-pediatrics residents. Text4Peds messages were derived from the most-missed pediatric in-training exam questions and were sent once daily to enrolled residents, Monday-Friday. Residents completed pre- surveys examining their text messaging habits. After three months, residents completed surveys that gauged their perception on the educational value of the text messages and impact on pediatric board preparation.

Results: A total of 35 residents participated in the program, 26 (74%) of whom completed the pre-survey. The overwhelming majority of responders own a smart phone (96%) and have unlimited text-messaging plans (88%). Prior to starting, the majority of residents (92%) believed that Text4Peds would be a useful addition to their pediatric education. Of the 2534 messages sent out to participants, 2437 (96.2%) were delivered successfully. Among those residents completing the post-survey (n=18), 100% believed that the Text4Peds messages were relevant to their education, and the large majority (89%) enjoyed receiving the messages. The majority (68%) of residents felt the messages were useful for their pediatric board preparation.

Conclusions: A text message-based curriculum for pediatric resident education, Text4Peds, was successfully implemented in our residency program. Messages were delivered with a high success rate and residents found educational value in the messages. Text messages may be a useful adjunct intervention to improve the pediatric board preparation of residents.
VALIDATION OF GLOBAL RATING SCALE AND CHECKLIST INSTRUMENTS FOR THE INFANT LUMBAR PUNCTURE PROCEDURE AMONG RESIDENT RATERS

Colleen Braun¹, David Kessler², Marc Auerbach³, Renuka Mehta⁴, Anthony Scalzo¹, James Gerard¹

Departments of Pediatrics, Saint Louis University¹; Columbia University²; Yale University³; Medical College of Georgia at Georgia Regents University⁴

Background: The International Network for Simulation-based Pediatric Innovation, Research and Education (INSPIRE) has developed tools for the assessment of competency to perform the infant lumbar puncture (ILP) procedure. We previously reported evidence to support the validity and reliability of these instruments when used by expert raters in a simulated setting. [Gerard JM, et al. Validation of global rating scale and checklist instruments for the infant lumbar puncture procedure. *Simul Healthc* 2013; 8:148-54.]

Objective: To evaluate the validity and reliability of the ILP scoring tools when used by resident raters to assess simulated ILP performances.

Methods: Video recordings of 60 subjects performing an unsupervised LP on an infant bench top simulator were collected prospectively; 20 performed by subjects in each of three categories (beginner, intermediate experienced, and expert). Six blinded resident raters independently scored each subject's video recording [3 via a global rating scale (GRS), 3 via a checklist instrument].

Results: For the resident raters, across all subject groups, higher GRS scores were found with advancing level of experience (P < 0.01). Total checklist scores were similar between the expert (80.0%) and intermediate experienced (76.6%) groups (P = 0.68). Both groups scored higher than the beginner group (50.4%) on the checklist instrument (P < 0.01). Cronbach's alpha coefficient for the checklist was 0.77. The intraclass correlation coefficients among raters for the GRS and total checklist scores were 0.49 and 0.47 respectively.

Conclusions: Similar to that found for expert raters: 1) acceptable internal consistency was found for the checklist instrument, and 2) The GRS instrument outperformed the checklist in its discriminant ability. Only moderate agreement among raters was found for each of the scoring tools. Compared to expert raters, we found the level of agreement among resident raters to be poorer for both of the scoring instruments.
RAPID BEDSIDE TRIAGE IN A PEDIATRIC EMERGENCY DEPARTMENT
Robert G Flood, Paula Szwargulski, Nadeem Qureshi, Mary Bixby, Steven Laffey, James M Gerard

Background: Crowding and long wait times in the emergency department can increase risks for patients and decrease patient satisfaction. Thus, the Centers for Medicare and Medicaid Services (CMS) has identified throughput and patient satisfaction measures as quality indicators, which, in the near future, will be tied to annual payments made to hospitals.

Objective: To 1) improve Door to Provider (DTP) times to < 30 minutes for the majority of our patients; and 2) improve patient satisfaction.

Design/Methods: On November 1, 2011, we changed from our traditional Waiting Room Triage (WRT) to a new Rapid Bedside Triage (RBT) in patient care rooms in our 30 bed, academic, urban pediatric level 1 trauma center. Both outcome and balancing measures were established a priori; the 6 months prior were compared to the 6 months following the implementation of this new process. As an ongoing quality improvement initiative, these same measures are continually tracked. All of the analyses were 2-sided with an alpha less than 0.05 considered significant.

Results: More patients were cared for in the 6 months post initiation of the process change than before: (pre: 19,871, post: 23,303, p < 0.001). The percent DTP < 30 minutes significantly improved (pre: 33.8%, post: 60.5%, p = 0.004). For the current year (through October 31, 2013), 83% (33,377/40,204) have been seen within 30 minutes of arrival. Our mean Press Ganey (PG) benchmark satisfaction score of “Likelihood to Recommend” remained strong in both time periods but was not statistically different (pre: 88.4, post: 91.1, p = 0.07). Year to date, our mean benchmark PG score is 90.7 (90th percentile). During the study period, DTP time < 30 minutes was associated with significant improvement in patient satisfaction [Spearman's rank correlation (rs) coefficient 0.75, p = .005]. As for balancing measures, we found: 1) a small increase in the percentage of total patients seen by our pediatric nurse practitioners, caring for lower acuity patients throughout the department, in the post period (pre: 39.2%, post 41.3%, p < 0.001); and 2) no significant decrease in compliance with our nurse driven extremity pain protocol for the one month (preliminary data) pre and post initiation of the process change (pre: 54.6%, post 57.5%, p = 0.50).

Conclusions: RBT leads to improvements in DTP times, and this is correlated with patient satisfaction. As such, rapid triaging in patient care rooms should be considered as a "best practice" in pediatric emergency medicine.
FEASIBILITY REPORT OF THE PALLIATIVE AND END-OF-LIFE COMMUNICATION INTERVENTION FOR PARENTS OF CHILDREN WITH A BRAIN TUMOR

Verna Hendricks-Ferguson, Joan Haase, Javier Kane, Kamnesh Pradhan, Shih Chie-Schin, Karen Gauvain

Background/Significance: A national health care priority is to initiate early and compassionate communication about palliative and end-of-life care (PC/EOL) to parents of children with a poor prognosis.


Methods: A one-group feasibility study to evaluate the efficacy of a tailored PC/EOL communication intervention titled Communication Plan: Early through End of Life (COMPLETE) for a total of 12 families of children with a brain tumor during clinic visits at two sites. COMPLETE featured: a physician-nurse (MD/RN) team approach to initiate and sustain PC/EOL communication; tailored parent resource forms to review treatment and PC/EOL options; and inclusion of hope-sustaining and non-abandonment messages, tailored to the communication style of MD/RN teams and parents’ information preferences.

Results: Eleven families received COMPLETE. Significant parental findings over time included: hope increased (p<.0001); uncertainty decreased (p=0.04); guilt & worry decreased (p=0.05); perceived emotional support (p=0.007); marginal differences in decision regret (p=0.07); and marginal differences in perceived satisfaction with child’s delivery of care (p=0.07). Non-significant parental findings over time included: (a) parents’ preferences to receive advance care planning information about their child’s future care did not change and (b) parental appraisal of communication by health care providers, inclusion of family members, the perceived amount of time spent talking with parents, and technical skills related to delivery of the child’s care did not show any differences.

Conclusions: Our findings provide evidence to support development of a future multi-site randomized clinical trial to evaluate the efficacy of the COMPLETE intervention with a larger sample of parents of children diagnosed with a brain tumor and other types of cancer with a poor prognosis.

Funding acknowledgement: NIH-NINR - Grant Number: 1R21NRO11071-01A1 (PI: Hendricks-Ferguson, V).
Platform Session III
Clinical Research I

Moderator:

Ajay Jain, MD
Assistant Professor
Department of Pediatrics
SKIN AND SOFT TISSUE INFECTIONS IN CHILDREN: ANALYSIS OF RISK FACTORS FOR ADMISSION AND CLINICALLY IMPORTANT COMPLICATIONS

Jaime L Velasco Masson, Nirupama Kannikeswaran, Usha Sethuraman, Marwan Daoud, Gina Nieto

1Department of Pediatrics, Saint Louis University, Saint Louis, 63104, United States; 2Pediatric Emergency Medicine, Children's Hospital Of Michigan, Detroit, 48201, United States; 3Carman and Ann Adams Department of Pediatrics, Wayne State University, Detroit, 48201, United States and 4Pediatrics, St. Joseph's Children's Hospital, Paterson, 07503, United States.

Background: Skin and soft tissue infection(SSTI) related visits to the Emergency Department(ED) have increased in recent years and information regarding characteristics of pediatric SSTIs associated with admission to the hospital is scant in literature.

Objective: To determine the characteristics of pediatric SSTIs that are associated with hospital admission and help generate a clinical decision rule that can identify those who are at very low risk of adverse outcomes or clinically important complications [increased length of stay (LOS) >48hr, incision and drainage (I&D) in the operating room (OR) or ICU admission].

Design/Methods: This was a prospective study of children 0-17 years with SSTI who presented to a tertiary care pediatric ED between May 2012 to June 2013. Children with immunosuppression, indwelling catheters, prosthetic devices and pregnancy were excluded. Once informed consent was obtained, the ED attending filled a form with patient demographics, features of SSTI (size, erythema, induration, fluctuance), work up, treatment and disposition. Those admitted were monitored until discharge while those discharged from the ED were followed up via phone call.

Results: During the study period, 358 children were enrolled, with a median age of 73 months (interquartile range, 0-248). The overall admission rate was 43%. In the multivariable logistic regression model, 5 factors were associated with admission to the hospital: Age <12 mos [OR] 2.06; (95%C.I) 2.84-21.74 p<.000; Temperature ≥39°C 3.53; 8.2-144.0 p <.000; Size of erythema >5 cm 2.04; 2.24-26.3 p<.001; Face/Neck location 1.34; 1.30-11.33 p<.015 and Currently on antibiotics 1.39; 1.99- 8.10 p<.000.

None of these risk factors were associated with adverse outcomes or clinically important complications [increased length of stay (LOS) >48 hr, incision and drainage (I&D) in the operating room (OR) or ICU admission]. Of those followed by phone call, 53.9% returned to a pediatrician or an ED. Of these group 13(8.2%) had antibiotics changed and 6(3.7%) had progression of symptoms requiring higher level of care (admission, I&D).

Conclusions: In children with SSTI, factors associated with admission were age <12 months, fever ≥39ºC, erythema>5cm, face/neck location and previous antibiotic use. However, none of these factors were associated with adverse outcomes or clinically important complications. Further studies to implement guidelines for disposition would help reduce admission rates.
Background: Twin-to-twin transfusion syndrome (TTTS), imbalanced shunting of blood from one twin to the other via placental vascular anastomoses, complicates approximately 10 to 15% of monochorionic pregnancies and carries a high risk of fetal mortality. The current treatment of choice for TTTS is selective fetoscopic laser photocoagulation (SFLP) and ablation of the vascular anastomoses. Recurrence of TTTS after SFLP occurs in up to 16% of cases and is associated with an adverse outcome. Previous studies of placental pathology in TTTS cases have primarily focused on angioarchitecture in placentas not treated by SFLP with scant evaluation of other microscopic findings.

Design: Charts of 9 twin pregnancies affected by TTTS were reviewed; 6 underwent SFLP and 3 had no intervention. All cases were assessed by ex vivo vascular injection studies with gross and microscopic evaluation. Sections were taken at SFLP sites, remote from sites, and of grossly abnormal regions. Studies including immunohistochemistry were performed to highlight the vasculature (VEGF, PEDF, ATGL) and to evaluate cell proliferation (PCNA).

Results: Of TTTS cases (n=9), mean maternal age was 24 yrs (range 20-32 yrs). For this young age group, there was a high rate of co-morbidities including obesity, diabetes, biliary disease, smoking, and hematopoietic abnormalities. Fetal demise of one twin occurred in 67% (2/3) of the untreated group, but in those with SFLP, the rate was only 33% (2/6). Vascular studies showed residual anastomoses in one case. There was abnormal umbilical cord insertion in 44% (4/9). Histologic analysis revealed vascular pathology including intravascular calcifications in 67% (6/9), two of which had no SFLP. In 6/7 cases with SFLP treatment, fine concentric calcifications surrounded areas of villous fibrosis underlying SFLP sites and in 3/7, there was a wedge-shaped infarct subjacent to the SFLP site.

Conclusions: This cohort of women with TTTS-complicated gestations was young, with a high number of metabolic co-morbidities (30%). There was accelerated mineralization of choriovilous vasculature in TTTS placentas regardless of SFLP status, suggesting a new disease association. The wedge-shaped infarcts associated with SFLP sites indicate that superficial chorionic intervention can significantly alter subjacent placental perfusion.
EXTREMELY PRETERM NEONATES BORN AFTER HISTOLOGIC CHORIOAMNIONITIS EXHIBIT PRO-INFLAMMATORY ALTERATIONS OF ADAPTIVE IMMUNITY

Daniel Rito¹, Luke T Viehl¹, Ryan P Weisert¹, Justin B Josephsen¹, Mohamad Al-Hosni¹, Joyce Marie Koenig¹,²

¹Pediatrics and ²Molecular Microbiology & Immunology, Saint Louis University.

Background: Histologic chorioamnionitis (HCA) is a common gestational antecedent of preterm delivery. Extremely preterm infants with HCA exposure are also at greater subsequent risk for chronic inflammatory disorders, such as bronchopulmonary dysplasia, through poorly understood mechanisms. We recently reported that murine antenatal inflammation exaggerates postnatal inflammatory innate and adaptive immune responses, including enhanced expression of pro-inflammatory lung T-helper (Th) subsets (Pediatric Research, 2014).

Objective: Our goal was to test our hypothesis that HCA promotes a gestational age-dependent, postnatal inflammatory adaptive immune phenotype in humans by characterizing circulating T regulatory (Treg) and CD4 T helper subsets in exposed preterm and term neonates.

Design/Methods: We prospectively collected placental umbilical cord blood from extremely preterm (EP, 23-29 weeks, n=18) and term (>37 weeks, n=18) gestations with HCA (or age-matched controls) and obtained correlative demographic data. Treg (CD4⁺CD25⁺CD127⁺) and Th17 (CD4⁺CD161⁺CCR6⁺) cells were characterized for co-expression of various chemokine receptors, Th-related cytokines, and Th-specific transcription factors by multicolor flow cytometric analysis of surface and intracellular antigens.

Results: EP-HCA neonates had higher circulating proportions of Treg, Th17 and CD4⁺IL17⁺ cells and Th17:Treg ratios compared to T-HCA neonates (all, p<0.01). 67% of EP-HCA neonates were ventilator dependent at 3 weeks of life vs. 18% of age-matched controls. HCA was clinically silent in 30% of EP gestations; in contrast, HCA correlated with maternal symptoms in 100% of term gestations.

Conclusion: Our data suggest that often-indolent in utero exposure to HCA biases developmental immune programming towards pro-inflammatory Th17-type immunity. This bias may set the stage for hyper-inflammatory neonatal immune responses with long-term health consequences. Our study, the first to characterize neonatal adaptive immunity in the context of HCA, may help identify cellular therapeutic targets to modulate neonatal inflammation.
A 20 SEC SUSTAINED INFLATION DID NOT ALTER MARKERS OF LUNG INJURY IN SURFACTANT-TREATED FETAL LAMBS
Noah Hillman, Matthew Kemp, Yuichiro Miura, Suhas Kallapur, Alan Jobe

Background: Sustained inflations (SI) use a prolonged positive pressure inflation to rapidly recruit functional residual capacity (FRC) and may decrease the need for mechanical ventilation in preterm infants. We have previously demonstrated that surfactant-deficient preterm lambs require high pressures at birth to recruit a variable FRC and that SI alone created a modest acute phase response with no alteration in lung injury from subsequent mechanical ventilation.

Hypothesis: A 20 sec SI at birth will decrease lung injury from mechanical ventilation in surfactant-treated preterm fetal lambs.

Methods: Ewes at 126±1 day GA were anesthetized and a sterile hysterotomy performed. The head and chest of each fetal lamb was exteriorized, tracheostomy performed, fetal lung fluid gently removed and replaced with surfactant (300 mg diluted in 15ml of saline). With placental circulation intact, fetal lambs were then randomized to one of four 15 minute interventions (n =4-8/group): 1) PEEP 8 cmH2O, 2) 20 sec SI at 40 cmH2O, then PEEP 8 cmH2O, 3) mechanical ventilation with a tidal volume (VT) of 7 ml/kg, or 4) 20 sec SI at 40 cmH2O then VT 7 ml/kg (SI+VT). Recruitment volume and ventilation variables were recorded. Fetal lambs remained on placental support for 30 min after the intervention. Fetal lung fluid (FLF), bronchoalveolar lavage fluid (BAL), and lung tissue were evaluated.

Results: SI recruited a mean volume of 6.8±0.8 mL/kg with 75% of the volume achieved in the first 2 sec. SI did not alter respiratory physiology during mechanical ventilation. Heat shock protein 70, HSP60, and total protein were recovered in FLF and BAL in both ventilation groups, with no difference with SI. A modest pro-inflammatory cytokine (IL-1, IL-6, IL-8) and acute phase response was found in both VT groups, but were not altered by SI. SI alone did not alter markers of lung injury.

Conclusion: In the setting of surfactant replacement, a 20 sec SI at 40 cmH2O recruited 7 mL/kg but did not alter ventilation physiology or markers of lung injury from mechanical ventilation. In comparison to surfactant-deficient preterm lambs, these lambs had a milder acute phase response and no effect of SI alone.
Platform Session IV
Clinical Research II

Moderator:

Debra H. Zand, PhD
Associate Professor
Department of Pediatrics
ADMINISTRATION OF IRON-REPLETE LACTOFERRIN TO PRE-WEANLING MICE INCREASES LIVER HEPcidIN EXPRESSION

Jeffrey A Cooper¹, Elif Memisoglu², Princy Prasad¹, Yihang Li, PhD¹ and Robert E Fleming¹,³
¹Pediatrics, ²Nutrition and Dietetics, ³Biochemistry & Molecular Biology. Saint Louis University, St. Louis, MO, 63104 USA.

Background: Hepcidin is a hepatocellular peptide hormone which regulates dietary iron absorption and systemic iron distribution. Expression of the hepcidin gene is regulated by signals which reflect body iron status and erythropoietic activity. Regulation by iron status includes signaling from circulating transferrin (Tf) to hepatocellular transferrin receptors (TfRs). Lactoferrin (Lf) is a member of the Tf family of iron-binding glycoproteins and found in high concentrations in milk. Lf has a well described role in host defense; however, its role in iron metabolism is less clear. Like Tf, Lf delivers iron to hepatocytes but unlike Tf, Lf cannot deliver iron to erythroid cells or interact with TfRs.

Objective: We administered iron-replete Lf to mice to determine if delivering iron to the liver but not erythron, in a form not dependent on Tf, would up-regulate hepcidin.

Methods: We advantaged the low iron and low hepcidin state of pre-weanling mice, and administered Lf equivalent to 2 mg/kg iron. Some mice were administered Lf (n=10) or carrier (n=10) intraperitoneally and sacrificed 6 h later. Other mice were administered IP Tf (n=5) or carrier (n=4). Lf (n=8) or carrier (n=7) was instead administered enterally (gavage) to other mice, and sacrificed 24h later. Liver hamp1 mRNA was measured by real-time RT-PCR. Liver iron distribution was determined histochemically in tissue sections by modified Perls' staining.

Results: Parenteral (IP) administration of Lf to pre-weanling mice was associated with a 7.3-fold increase in liver hamp1 expression after 6 h (P<0.001), compared with a 4.5 fold increase in hamp1 (P<0.01) after a comparable dose of Tf. Enteral Lf was associated with a ~30-fold increase in hamp1 (P<0.005) after 24h. Iron delivered by IP Lf mainly distributed to sinusoidal lining cells, whereas iron delivered by enteral Lf mainly distributed to hepatocytes.

Conclusions: Exogenously administered iron-replete Lf increases liver Hamp1 expression in pre-weanling mice. These observations demonstrate that iron delivery to the liver in a form not dependent upon transferrin or its receptors up-regulates hepcidin expression. We speculate that Lf-mediated changes in hepcidin expression may contribute to reported benefits of supplemental Lf on host defense.
DEFORMATIONAL SCAPHOCEPHALY RESULTS IN INCREASED THERAPY DURATION AND LESS EFFECTIVE CRANIAL INDEX CORRECTION THAN OTHER TYPES OF DEFORMATIONAL PLAGIOCEPHALY
Sarah A. Donigian, Emma M. Kulig, Chelsea R. Horwood, Alexander Y. Lin

Background & Purpose: Scaphocephaly, or long head shape, can be due to deformational or craniosynostotic etiologies. If the etiology is purely deformational, some patients will undergo helmet orthotic therapy. Our aim was to compare helmet therapy outcomes of infants with deformational scaphocephaly.

Methods: Patients with deformational plagiocephaly who completed helmet orthotic treatment from 2006 to 2013 were included. Cranial index (CI) was defined as calvarial width/length, with mesocephalic range 0.76-0.81, scaphocephalic less, brachycephalic greater. Asymmetry was measured by transcranial difference (TCD) between frontozygomatic-to-eurion diagonals (millimeters). Continuous variables were compared with nonparametric tests reported as median (range).

Results: 209 patients with deformational plagiocephaly met our criteria: scaphocephalic 9 (4.3%), mesocephalic 14 (6.7%), brachycephalic 186 (89.0%). The duration (in months) of helmet therapy: scaphocephalic 4.87 (3.27-11.67), non-scaphocephalic 3.45 (1.13-15.87), **P=0.007. Initial CI: scaphocephalic 71.23, mesocephalic 79.00, brachycephalic 91.39. Initial TCD: scaphocephalic 5.9, mesocephalic 12.8, brachycephalic 10.3, P=0.001. Difference between final and initial CI: scaphocephalic 0.90 (-1.19, 4.70), brachycephaly 3.57 (-9.80, 19.77), ***P<0.003. Rate of change in CI per month: scaphocephalic 0.185 (-0.20, 0.59), mesocephaly .068 (-1.34, 2.18), brachycephaly .995 (-2.62,4.59) ***P<0.001. Final TCD: scaphocephaly 1.0 (0.0, 4.0), mesocephaly 3.0 (1.0, 10.0), brachycephaly 4.0 (0, 11.0), **P=0.002. Final CI: scaphocephaly 71.68 (69.0, 75.7), mesocephaly 78.79 (74.58, 82.58), brachycephaly 87.19 (74.86, 100.0), ***P<0.001.

Conclusion: Since deformational scaphocephaly patients appear to have a different response to helmet therapy than non-scaphocephalic, parents and providers need to refer these patients early to improve their chances of response to helmet therapy.
CPAP THERAPY ADHERENCE IN CHILDREN WITH OBSTRUCTIVE SLEEP APNEA
Shalini Paruthi, Angela Orlando, Raman Malhotra, Eric Armbrecht

Introduction: Many children receive their first hands-on introduction to continuous positive airway pressure (CPAP) therapy on the night of the in-lab titration. The experience in the sleep lab may influence 90-day adherence to PAP therapy. We hypothesize children with positive mask fittings and pre-trials prior to lights-out on a polysomnogram will show improved adherence. We also hypothesize children with perceived immediate benefit following the titration (post-sleep morning questionnaire) will show improved adherence.

Methods: Retrospective chart review performed on data from 104 children initiated on CPAP therapy in 2012. Adherence was defined as use of CPAP >4 hours per night for >70% of nights in a consecutive 30-day period during the first 90 days of CPAP therapy. Chi square analyses and logistic regression analysis were performed to determine associations of adherence with polysomnogram processes and patient-level attributes.

Results: Of 104 children (49 girls), 58 children achieved criteria for 90-day adherence. There were no univariate associations between adherence and mask fitting/pre-trial tolerance (p=0.599), type (split-night vs full-night) of titration (p=0.323), baseline apnea-hypopnea index (AHI) on polysomnogram (p=0.07), gender (p=0.471), age (p=0.872), or body mass index (p=0.067). In comparison to others, patients who perceived the titration to be a ‘worse’ night of sleep had lower rates of adherence (p=0.006). The logistic regression model revealed that patients who reported a ‘worse’ than usual night of sleep were about 80% less likely (OR=0.195, 95%CI: 0.042-0.903) to achieve 90-day adherence when controlling for insurance status (Medicaid vs commercial), age, and AHI. Conclusion: Children who had subjectively poor experiences during the in-lab titration had lower 90-day CPAP adherence rates. These factors may help to identify children who require additional clinical interventions to achieve optimal adherence.
ABSTRACT FORM
Pediatric Science Days 2014 Research Symposium
Thursday, April 3, 2014

Presenting Author:  Ann Marie Flannery M.D

Please check one:  X Faculty Fellow  □ Resident  □ Post-Doc  □ Student  □ Other ____

Faculty Sponsor (required if trainee and not faculty): ____

Faculty sponsor e-mail:  flanneam@slu.edu

School/College/Dept/Division:  Neurological Surgery

Contact E-mail:  flanneam@slu.edu  Contact Phone: 577-8715

Presentation topic (Please select one most relevant):

☐ Disease-Oriented Research: Basic

☐ Disease-Oriented Research: Clinical/Translational/Trials

☐ Disease-Oriented Research: Outcomes/Epidemiology

☐ Other: _____ (eg., developmental biology, signaling mechanisms, physiology, etc.)

EVIDENCE BASED RECOMMENDATIONS FOR THE TREATMENT OF CHILDREN WITH HYDROCEPHALUS

Ann Marie Flannery, Catherine Mazzola, Paul Klimo, David Limbrick, Lissa Baird, Tina Duhaime, Joanna Kemp, Mandeep Tamber, Dimitios Nikas, Mark Van Poppel, Laura Mitchell

Introduction: Evidence-based management has become important in Neurosurgery. Guidelines exist in Pediatric Neurosurgery, but not for management of hydrocephalus.

Methods: Members of the Pediatric section and other selected experts discussed and selected topics and questions relevant to the management of Pediatric hydrocephalus. After extensive systematic review and vetting, nine review topics were determined to be of significance and likely to have some relevant and reviewable literature to support the topics. MeSH terms were generated and a research librarian conducted the searches over databases that included PubMed, and The Cochrane Central Register of Controlled Trials. The abstracts were reviewed by topic subcommittees, and relevant papers were selected for full-text review and if appropriate, analysis. Those papers found to have relevance were analyzed and evidence tables were created, using evidence-based methods approved by the Joint Guidelines Committee of the American Association of Neurological Surgeons (AANS) and the Congress of Neurological Surgeons (CNS).

Results: Reviews were conducted for: timing of shunts in premature infants, effect of shunt entry point, usefulness of technical adjuvants for positioning catheters, effectiveness of endoscopic third ventriculostomy, effect of shunt design on outcome, effectiveness of pre/periparative antibiotics, effectiveness of other shunt infection preventions, treatment of shunt infections, and the effect of ventricular size on outcome. The review resulted in a total of 19 recommendations in the 9 areas including 6 recommendations with sufficient strength to be Class I

Conclusions: There is evidence of sufficient strength to make strong recommendations in some key areas of hydrocephalus management. The areas where there is insufficient or weak evidence are potential targets for future studies
POSTERS
DETERMINATION OF SPECIFICITY AND SENSITIVITY OF A NEW LC/MS/MS METHOD FOR NEWBORN SCREENING OF MUCOPOLYSACCHARIDOSES
Qi Gan1, Mary Campbell2, Adriana M Montaño1,3

Introduction: Mucopolysaccharidoses (MPS) are caused by excessive GAG accumulation from deficiencies of degradative enzyme activities. There are 11 known enzyme deficiencies, resulting in seven distinct forms of MPS with a collective incidence of approximately 1 in 25,000 live births. Accumulation of undegraded metabolites in lysosomes gives rise to distinct clinical syndromes. MPS disorders are potentially treatable with enzyme replacement therapy or hematopoietic stem cell transplantation. For maximum benefit of available therapies, early detection and intervention is critical. To overcome the current problems of MPS screening, we have developed a highly sensitive, specific, accurate and inexpensive strategy in which the disaccharides produced from DS, HS, and KS are analyzed simultaneously using LC/MS/MS in plasma and serum samples. A significant advantage of our method is that it can predict all types of MPS in the first-tier screening, with further confirmation by determining enzyme activity in the second-tier screening.

Aim: The goal of this project is to develop a highly specific, sensitive, and simple newborn screening (NBS) system for mucopolysaccharidoses (MPS).

Materials and Methods: Dried blood spot (DBS) samples were digested by chondroitinase B, heparitinase, and keratanase II to yield disaccharides of DS, HS, and KS. After digestion the samples were injected into the LC/MS/MS (API 4000) and results were analyzed.

Results: For method development we have found that there are no matrix or carryover effects in our system. In addition, the original method had some limitations regarding specific detection of the analyte ΔDiHS-6S because it co-eluted with ΔDi-4S (DS). As a result ΔDiHS-6S and ΔDi-4S were reported as ΔDi-4S/6S. We have now refined the LC/MS/MS method to obtain chromatographic separation of ΔDi-4S and ΔDiHS-6S analytes. Finally we have studied precision of the methodology and cut-off values by analyzing DS, HS, and KS levels of 701 DBS of newborns.

Conclusion: Our methodology can be tested further for disseminated use to supplement current NBS, leading to earlier treatment, improved prognosis and quality of life for affected individuals.
A NOVEL, MULTIDISCIPLINARY APPROACH TO TREATMENT OF A CASE OF CELLULAR NEUROTHEKEOMA OF THE LIP IN A PEDIATRIC PATIENT Kyle Y. Xu, Christina Plikaitis

Cellular neurothekeoma is a rare benign soft tissue neoplasm with risk of local recurrence. It presents a challenge to diagnose because of its histologic similarities to other dermal tumors. This distinction is important because each of these tumors has a different behavior pattern and risk of recurrence and distant metastasis. In addition, it is difficult to obtain clear margins surgically in this tumor especially in the setting of incomplete resection or recurrence because of its similarity to scar tissue on routine pathologic staining, thereby requiring the assistance of specialized staining techniques. Finally, obtaining accurate, clear resection margins is important prior to proceeding with definitive facial reconstruction as methods of tissue rearrangement often distort or obscure the original tumor bed.

Mohs micrographic surgery provides a method of improving clearance of skin and soft tissue tumors by obtaining accurate margins while sparing normal tissue. It is especially useful in treating tumors located in cosmetically sensitive areas with limited like tissue for reconstruction. However, the usual technique of same day pathologic clearance under local anesthesia is not possible to use with immunohistochemical staining and is also difficult to perform on a young child while awake.

We describe a unique, multidisciplinary approach for the treatment of a recurrence of cellular neurothekeoma in the upper lip of a 3 year old involving pediatric plastic surgery, Mohs dermatologic surgery, pathology, dermatopathology and anesthesia departments in order to achieve an accurate diagnosis, tumor clearance and aesthetic facial reconstruction. A modified, staged “slow Mohs” procedure under general anesthetic was used to obtain clear margins prior to proceeding with final facial reconstruction. This approach emphasizes the importance of innovation and teamwork when dealing with complex clinical challenges.
WOLMAN DISEASE – DEVELOPING MANAGEMENT APPROACHES. A CASE REPORT AND LITERATURE REVIEW José A. Torres Garcia, Stephen R. Braddock, Jeremy S. Garrett

We report the case of a 5-month-old male who initially presented for failure to thrive, returning for concerns of abdominal distension and vomiting with fever. He was found to have large calcified adrenal gland suggestive of Wolman Disease. This rare disease, with an incidence of less than 1 in 100,000 live births, previously has had treatment options limited to bone marrow transplantation or palliation. New enzyme replacement therapies may contribute to alternative approaches in care. We review the English-language medical literature and share our experience managing this rare disease.
CHEMICAL GENETICS IDENTIFIES BET PROTEINS AS POTENTIAL THERAPEUTIC TARGETS IN FACIOSCAPULOHUMERAL MUSCULAR DYSTROPHY

Francis M. Sverdrup¹, Matthew P. Yates, Jon Oliva, Marshal Huang, Stephen J. Tapscott²

¹ Center for World Health & Medicine, Saint Louis University, Saint Louis, Missouri USA
² Division of Human Biology, Fred Hutchinson Cancer Research Center, Seattle, Washington USA

Facioscapulohumeral Muscular Dystrophy (FSHD) is characterized by epigenetic changes resulting in the aberrant expression of the DUX4 gene in muscle. The long term objectives of this research are to identify druggable targets that regulate DUX4 expression and to evaluate the therapeutic potential of the corresponding inhibitory compounds. We have used a unique cell based assay that detects DUX4 expression in FSHD muscle cells to screen a library of compounds that target epigenetic modifier proteins. This "chemical genetics" approach identified the bromodomain and extra-terminal domain (BET) family of proteins as key targets involved in DUX4 expression. Selective inhibitors of BET proteins (BETi) blocked the induction of DUX4 and its downstream targets ZSCAN4 and TRIM43 during differentiation of FSHD1 and FSHD2 muscle cells in vitro. BETi also blocked DUX4 expression in undifferentiated FSHD myoblasts as evidenced by decreased ZSCAN4 and TRIM43 mRNA levels 48 to 72 hours after compound addition. Cells grown continuously in the presence of low dose BETi JQ1 exhibited a greater than 95% reduction in steady state levels of ZSCAN4 mRNA and this effect was sustained, requiring longer than 6 days to recover after removal of JQ1 from the cultures. These data suggest that BETi may have therapeutic potential in FSHD and that further investigation into the role of BET proteins in FSHD is warranted.
VITAMIN D RESCUES DNA-REPAIR DEFECTS IN MOUSE MODEL OF PROGERIA
Ray Kreienkamp, Arindam Das, Martin Neumann & Susana Gonzalo Department of Biochemistry & Molecular Biology, SLU

Hutchinson-Gilford Progeria Syndrome (HGPS) is a rare but devastating pediatric disorder. Patients present with signs of premature aging and usually die in their teens from cardiovascular complications. The disease is caused by mutations in the LMNA gene, which encodes the structural nuclear proteins lamin A/C. Cells from patients show profound genomic instability, which is thought to contribute to the pathophysiology of the disease. However, the molecular mechanisms underlining this genomic instability are largely unknown in patients and in the mouse model of progeria (Zmpste24−/−). Our previous studies in lamin A/C-depleted mouse embryonic fibroblasts (MEFs) revealed some mechanisms behind genomic instability in these cells. Loss of lamin A/C results in downregulation of BRCA1 and RAD51, proteins that are critical for DNA repair by homologous recombination. Interestingly, we also found that these cells harbored low levels of vitamin D receptor (VDR), and that treatment with vitamin D not only increased levels of VDR, but also rescued BRCA1 and RAD51 levels. This suggests that VDR deficiency might underlie the loss of DNA repair factors in lamin A/C-depleted cells, and that vitamin D could represent a new strategy to overcome genomic instability in some laminopathies.

Here, we tested whether cells from the progeria mouse model exhibit similar DNA repair deficiencies as lamin A/C-depleted cells, and if these can be rescued by vitamin D. We demonstrate that Zmpste24−/− mice show profound genomic instability and deficiencies in BRCA1 and RAD51, as well as very low levels of VDR at both the transcriptional and protein level. Importantly, treatment of Zmpste24−/− fibroblasts with vitamin D for 24 hours resulted in increased levels of VDR and both BRCA1 and RAD51 at the transcriptional and protein level. This rescue also had functional consequences, as Zmpste24−/− fibroblasts treated with vitamin D presented with lower levels of DNA damage than untreated fibroblasts, as measured by γH2AX foci by immunofluorescence. These findings implicate low levels of VDR in the genomic instability of Zmpste24−/− fibroblasts. Furthermore, we show that vitamin D can reduce this genomic instability by rescuing levels of BRCA1 and RAD51. As a result, these findings support vitamin D as a novel therapeutic strategy for reducing genomic instability in Zmpste24−/− mice, which might eventually translate into a novel approach for rescuing DNA damage in HGPS patients.
REPORTED COMMUNICATION ABILITY IN PERSONS WITH TRISOMY 18 AND 13

Cheryl A Liang, Barbara A Braddock, Jennifer L Heithaus, Katherine M Christensen, Stephen R Braddock, John C Carey

Objective: The aim of this study was to describe the communication ability of individuals with trisomy 18 and trisomy 13 syndromes.

Methods: This is the first known prospective study of communication ability in a larger group of individuals ascertained with trisomy 18 (n=17) and trisomy 13 (n=15). Parents reported on children’s potential communication acts, words, spontaneous gesture, and augmentative and alternative communication (AAC) using a parent report inventory (n=32; age range 3 - 35 years). The use of a parent report tool is designed to examine participants’ communicative abilities over a wide range of functions. Parents are able to report on their children’s communicative abilities within everyday contexts. Potential Communicative acts are defined as behaviors produced by an individual that may be interpreted by others to serve communicative functions.

Results: Potential communicative acts categorized as body movement displayed the highest median rank for reported occurrence followed by vocalization and facial expression. Although symbolic forms were ranked lower, more than half of parents (66%) reported that their children produced at least one word, gesture or AAC form. Symbolic forms followed in rank as spontaneous gestures and manual signs were reported in the communication repertoires of more than half of the participants. For this subgroup of individuals who were reported to use symbols (n=21), the most frequently reported symbolic form was a spontaneous gesture for a head-shake used to represent “no” (85.7%). To examine the difference in the number of reported symbolic forms between subgroups of persons with trisomy 18 and those with trisomy 13, a Mann-Whitney non-parametric statistical test was used. Results indicated no statistically significant difference in the reported number of different symbolic forms between subgroups of persons with trisomy 18 and trisomy 13 (U=121.5, p=ns). This indicates that subgroups of individuals with trisomy 18 and 13 looked more similar than dissimilar in the amount of symbolic forms produced. This study adds to the literature on the natural history and development of children with trisomy 18 and 13 who have survived past the first year of life and beyond, which may be helpful to parents experiencing a new diagnosis. This study may also assist parents as they plan for future educational and therapeutic needs of their children.

Conclusions: Results are discussed in terms of communication potential and the need to address AAC in trisomy 18 and 13.
ENGAGING RESIDENTS IN PEDIATRIC HOSPITAL MEDICINE – A NOVEL ELECTIVE

Marta King, Victoria Wilkins, Mara Chavolla Calderon

Educators have proposed several methods to address the gap between residency training and the practice of hospital medicine. Electives within residency allow those considering careers in hospital medicine to sample curricular components of the field. The purpose of this project was to design a learner-centered pediatric hospital medicine resident elective. We used American College of Graduate Medical Education core competencies, pediatric hospital core competencies, and expert consensus to design clinical, administrative, research, teaching, practice-based learning, and systems-based practice elective activities. Pediatric hospitalists, as well as pediatric, combined program, and family medicine residents and graduates were asked to rate the proposed elective using an anonymous web-based survey. 23 hospitalists, 61 residents, and 30 program graduates completed the survey (n=101). 41% spent or were planning to spend ≥25% of their time practicing in an inpatient setting. 84% rated the proposed elective as valuable or very valuable. Those focused on inpatient work ranked the rotation as significantly more valuable than those who were not (93% vs. 67%, p=0.003). Activities receiving highest interest were: 1) general inpatient care and consults, 2) conscious sedation and airway management, 3) rapid response and palliative care teams; 4) patient teaching sessions with asthma and diabetes educators. Activities receiving lowest interest were: 1) shadowing ancillary hospital staff (interpretation, education, child life), 2) care of specific patients (tracheostomy/ventilator dependent and sedation observation), 3) hospital medicine list-serve; 4) administrative meetings. Significant differences were seen between activities practicing hospitalists and residents interested in hospital-based careers rated as valuable. Participants endorsed the hospital medicine elective with freedom to choose activities most suited to individual resident regardless of current or future career plans. As a variety of activities were deemed valuable, a learner-centered approach with a menu of activities to choose from may be most beneficial.

Future studies will focus on elective implementation and evaluation.
COMPARING TWO AMBULATORY PEDIATRIC CLERKSHIP SCHEDULING MODELS
Rochelle M. Remus, Marta King, Jamie Sutherell

Student experience and education are influenced by clerkship structure and the resultant student-attending relationship. Although working with multiple attendings allows for diversity of experience, previous studies demonstrate more student clinical responsibility and greater faculty motivation to teach with increased student-attending continuity. The purpose of our study was to compare two ambulatory pediatric clerkship scheduling models; an experimental model (emphasis on continuity) with existing control model (emphasis on diversity of experience). All students (N=59) enrolled in the pediatric clerkship between July and October 2013 were assigned to experimental or control scheduling models during alternating months. Experimental group students were each matched with 1-3 attendings, while control group students worked with 5-8 attendings each. Students and attendings were invited to complete anonymous surveys based on prior month’s ambulatory experience regarding: scheduling preference, ability to provide or receive meaningful feedback and evaluation, and comfort in asking for or in writing letters of recommendation. Number of ambulatory evaluation comments, NBME exam scores, student final clerkship, and ambulatory grades were compared.

73% (N=43; 21 experimental) students completed the surveys. More students (38 vs. 5, p<0.01) and attendings (9 vs. 1, p=0.02) preferred the continuity model. Continuity group students felt comfortable asking more attendings (20 vs. 3, p<0.01) for a letter of recommendation. Similarly, attendings endorsed being comfortable writing a letter of recommendation (23 vs. 2, p<0.01) and able to provide more meaningful feedback and evaluation (58 vs. 24, p<0.01) to more continuity students. Although continuity students rated their overall ambulatory experience higher and believed attendings were able to provide meaningful feedback and evaluation, these differences were not statistically significant. There were no differences in the overall clerkship, NBME exam, or ambulatory grades, however, continuity students received more ambulatory evaluation comments (11 vs. 6, p<0.01).

Continuity is valued over diversity of experience by both students and attendings. While a scheduling model focused on continuity did not alter students’ overall grades, it did create an environment that allowed for improved ability to provide meaningful feedback and evaluation.
RESIDENT PERCEPTION OF AN AUDIENCE RESPONSE SYSTEM IN GRADUATE MEDICAL EDUCATION

Amol Purandare1, Angela Delecaris2, Laura Holt2, Matt Broom2

1Department of Pediatrics, Saint Louis University

Background: Studies show didactic lecture as a learning style is susceptible to low participation, decreased attention, poor retention and recall. A solution for better engagement is through the use of an audience response system (ARS).

Objective: To determine resident perception regarding the use of an ARS in graduate medical education to better define educational benefits in training. We believe use of an ARS will advance education by allowing anonymous participation and instant feedback within the Saint Louis University Pediatric Residency Program.

Design: A prospective study evaluating perceived educational value of residency program conferences with and without use of an ARS. Over a 16-week period, lectures utilizing an ARS were intermittently incorporated into the residency program’s conference schedule. Residents were surveyed prior to formal incorporation of the ARS and again after the 16-week evaluation period. The evaluations aimed to assess resident perception about the educational value of the current resident conference schedule. Also during the 16-week period, residents completed evaluations to provide specific feedback for conferences conducted with and without the ARS.

Results: Pre-survey results include 38/61 (62%) resident responses in the program (including pediatrics and combined medicine-pediatrics residencies), with 15 PGY1, 8 PGY2, 12 PGY3, 1 PGY4; 95% (36/38) of residents consented to the use of their program evaluation data. The mean educational value scores for the 5 conferences (scale 1-5) ranged from 2.56 (Ambulatory) - 3.86 (Board-focused PREP). Similarly in regards to conference participation, mean scores ranged from 2.86 (Ambulatory) - 3.47 (Board-focused PREP). Of the responses to date, 29 (80%) have indicated neutral or unhappy sentiments regarding the current conference format. Of those surveyed, 27 (75%) specifically felt that answering questions improves their focus; 28 (78%) felt that adding participation in the form of questions via an ARS would help with learning. Additionally, they would similarly be more willing to participate if able to give input anonymously. As the study is ongoing, post-survey results remain forthcoming.

Conclusions: The ARS is a welcomed method of interactive learning among residents and may help augment traditional teaching styles and standard lecture formats.
DOES THE DEGREE OF LYMPHOPENIA INDUCED BY ATGAM IMPACT OUTCOMES IN PEDIATRIC LUNG TRANSPLANT RECIPIENTS WITH BRONCHIOLITIS OBLITERANS?
Josie Vitale Lorbert, Charles Huddleston, Stuart Sweet, Albert Faro

Purpose: Antithymocyte globulin (ATGAM) is a polyclonal antibody preparation that decreases the number of circulating lymphocytes and is frequently used to treat post-transplant bronchiolitis obliterans (BO). However, little is known about the overall efficacy of the drug for this purpose and no guidelines exist as to what extent of immunosuppression optimizes treatment benefits without placing patients at further risk for acquiring life-threatening infections or malignancies. We investigated the degree to which ATGAM induced lymphopenia correlated with treatment responses.

Methods and Materials: We retrospectively analyzed data from lung transplant patients who received ATGAM therapy at our center between 2000 and 2011 (n = 53). Data were collected from electronic medical records and our center’s database. We compared groups with ATGAM induced absolute lymphocyte counts (ALCs) below 500 to those with ATGAM induced ALCs over 500. We analyzed time to death, time to retransplantation, graft survival, development of PTLD, episodes of acute rejection, development of infections, changes in FEV1, and changes in chest CT and VQ scans following ATGAM.

Results: We found no significant associations. Of note, although not statistically significant, all three patients who developed PTLD following ATGAM therapy had ALCs less than or equal to 100.

Conclusions: Our findings suggest that the degree of lymphopenia induced by ATGAM does not affect outcomes. However, as the presumed mechanism of action of ATGAM in treating BO is augmented immunosuppression, the lack of a difference in outcomes between the two groups raises concerns as to the efficacy of ATGAM in treating BO. A limitation of this study was sample size. Multi-centered controlled trials are needed to further investigate the efficacy of ATGAM and its potential association with increased risk of infection and malignancy. This may be particularly important in evaluating the risk for PTLD.
ISOPROSTANES AS PHYSIOLOGICAL MEDIATORS OF TRANSITION TO NEWBORN LIFE: NOVEL MECHANISMS REGULATING PATENCY OF THE TERM AND PRETERM DUCTUS ARTERIOSUS

Jian-Xiong Chen, Patrick W. O’Mara, Stanley D. Poole, Naoko Brown, Noah J. Ehinger, James C. Slaughter, Bibhash C. Paria, Judy L. Aschner, Jeff Reese

1Department of Pharmacology and Toxicology, University of Mississippi Medical Center, Jackson, Mississippi; 2Department of Pediatrics, 3Department of Biostatistics, 4Department of Cell and Developmental Biology, Vanderbilt University, Nashville, Tennessee;

Background: Increased oxygen tension at birth regulates physiologic events that are essential to postnatal survival, but the accompanying oxidative stress may also generate isoprostanes. We hypothesized that isoprostanes regulate ductus arteriosus (DA) function during postnatal vascular transition.

Methods: Isoprostanes were measured by gas chromatography–mass spectrometry. DA tone was assessed by pressure myography. Gene expression was measured by quantitative PCR.

Results: Oxygen exposure was associated with increased 8-iso-prostaglandin (PG)F2α in newborn mouse lungs. Both 8-iso-PGE2 and 8-iso-PGF2α induced concentration-dependent constriction of the isolated term DA, which was reversed by the thromboxane A2 (TxA2) receptor antagonist SQ29548. SQ29548 pretreatment unmasked an isoprostane-induced DA dilation mediated by the EP4 PG receptor. Exposure of the preterm DA to 8-iso-PGE2 caused unexpected DA relaxation that was reversed by EP4 antagonism. In contrast, exposure to 8-iso-PGF2α caused preterm DA constriction via TxA2 receptor activation. Further investigation revealed the predominance of the TxA2 receptor at term, whereas the EP4 receptor was expressed and functionally active from mid-gestation onward.

Conclusion: This study identifies a novel physiological role for isoprostanes during postnatal vascular transition and provides evidence that oxidative stress may act on membrane lipids to produce vasoactive mediators that stimulate physiological DA closure at birth or induce pathological patency of the preterm DA.
ABSTRACT FORM
Pediatric Science Days 2014 Research Symposium
Thursday April 3, 2014

Presenting Author: Hayley Friedman, MD, MS

Please check one: □ Faculty Fellow □ Resident □ Post-Doc □ Student □ Other □

Faculty Sponsor (required if trainee and not faculty): Marya Strand, MD, MS

Faculty sponsor e-mail: mstrand2@slu.edu

School/College/Dept/Division: St. Louis University School of Medicine, Dept. of Pediatrics

Contact E-mail: hfriedman1@gmail.com Contact Phone: 310-351-2449

Presentation topic (Please select one most relevant):
- Disease-Oriented Research: Basic
- Disease-Oriented Research: Clinical/Translational/Trials
- Disease-Oriented Research: Outcomes/Epidemiology
- Other: (eg., developmental biology, signaling mechanisms, physiology, etc.)

HORIZONTAL TRANSMISSION OF NEONATAL HSV: A UNIQUE CASE INVOLVING ECZEMA HERPETICUM Hayley Friedman, Marya Strand, Justin Josephsen, Jeffrey Cooper

Background: Disseminated neonatal Herpes Simplex Virus (HSV) infection is a serious illness, most commonly transmitted through a vertical pathway, with the maternal genital tract as the primary conduit of infection during time of delivery. While there have been a few reported cases of horizontal transmission via contact with maternal herpetic whitlow or breast lesions, we report a case of transmission via direct contact with maternal, upper extremity eczema herpeticum (HSV infection of atopic dermatitis). Neonatal HSV infection is life threatening and can lead to severe neurologic sequelae.

Case: A mother with history of atopic dermatitis presented with her 6-day old former 36 weeks’ gestation infant, to an outside Emergency Department. This mother was concerned about two acute episodes of arm stiffening, jaundice, and notable lethargy of her neonate. A full sepsis evaluation was completed, including CBC analysis, blood, urine and CSF cultures, and the infant was treated with meningitic dosing of cefotaxime and ampicillin. The infant’s clinical status worsened, resulting in transfer to our NICU, and initiation of vancomycin, gentamicin, and acyclovir therapy. Shortly thereafter, the mother revealed eczematosus lesions on her right upper extremity, which she thought was atopic dermatitis. This finding prompted immediate consideration of horizontal transmission of HSV via eczema herpeticum as the underlying etiology of the fulminant hepatic failure, pulmonary hemorrhage, and status epilepticus that led to this infant’s demise within 12 hours of presentation. Subsequent test results were positive for HSV in CSF, eye, and mucous membrane samples.

Conclusion: In consideration of the significantly high morbidity and mortality rate associated with neonatal HSV infection, physicians must promptly consider and identify alternative modes of HSV infection. HSV can be transmitted through horizontal exposure to eczema herpeticum, as evidenced by this case. Current AAP Redbook recommendations should include prevention guidelines regarding eczema herpeticum, to ensure appropriate early intervention and therapy that is needed to prevent catastrophic outcomes.
PACKED RED BLOOD CELL TRANSFUSION ATTENUATES INTESTINAL BLOOD FLOW RESPONSES TO FEEDINGS IN VERY LOW BIRTH WEIGHT NEONATES WITH NORMALIZATION AT 24 HOURS Aaron Pitzele¹, Eric S Armbrecht¹, Mohammad Rahimi¹, Thomas Havranek¹
¹Pediatrics/Neonatology, SLU/Cardinal Glennon Children's Medical Center, St.Louis, MO, United States and ²Center for Outcomes Research (SLUCOR), Saint Louis University, St.Louis, MO, United States.

Background: Packed red blood cell (PRBC) transfusion has been associated with necrotizing enterocolitis and altered superior mesenteric artery blood flow velocity (SMABFV) responses to feedings in larger, uncomplicated preterm neonates.

Objective: To determine whether PRBC transfusion affects postprandial SMABFVs in VLBW neonates and if so, at what time point after transfusion restoration of previous SMABFV patterns occurs.

Design/Methods: VLBW preterm neonates, older than 14 days and tolerating bolus enteral feedings administered every 3 hours were enrolled in this prospective observational study. Pulsed Doppler was used to measure pre and postprandial (at 45 min) time-averaged mean, peak and end diastolic velocities (TAMV, PSV, EDV) immediately before and after 15 ml/kg of PRBC transfusion given over 3 hours; patent ductus arteriosus (PDA) status was evaluated as well. Subsequent pre and postprandial SMA BFVs were recorded 24 and 48 hours after the transfusion. Paired t-tests assessed statistical significant changes in SMABFVs pre and postprandially for each measurement period, alpha 0.05. Effect modification was measured for PDA.

Results: Of the twenty-five infants studied, 21 completed the study. Postprandial TAMV, PSV, EDV were attenuated during the feedings immediately after transfusion; there was no significant change in pre and postprandial SMABFVs during this measurement period while other changes were significant in the other measurement periods. At 24 and 48 hours, postprandial velocities displayed change patterns similar to the one prior to transfusion; presence of the PDA did not affect results in this sample. Refer to the figure for an illustration of TAMV results.

Conclusions: PRBC transfusion blunted SMABFV responses to feedings immediately after the transfusion with normalization observed 24 hours post transfusion.
DEVELOPMENT AND VALIDATION OF A NOVEL ULTRA-MOBILE AMBULATORY TOTAL PARENTERAL NUTRITION MODEL
Ajay K Jain, Keith S. Blomenkamp; Jonathan Rodrigues, Timothy A. Blaufuss; Mike A. Carl, Victor D. Liou, Joy X. Wen, Barbara Stoll, Douglas Burrin, John P. Long, Jeffrey Teckman

Background: Total parenteral nutrition (TPN) involves providing all of a patient’s nutritional needs intravenously and is life-saving in clinical settings where enteral based delivery of nutrition is not possible. Though TPN therapy has grown enormously over the last few decades; it is associated with significant complications including mortality and amelioration of side effects remains a major research focus. Past studies have used rodents and recently piglets due to their homology in both form and function to the human liver and gastrointestinal tract. However, such models employ tethering and limit animal mobility resulting in stress. In humans TPN is delivered in an ambulatory fashion allowing complete mobility. The lack of such reproducibility remains a major drawback of current TPN based studies. We describe a novel model for long term TPN infusion in piglets using ambulatory pumps which allow complete animal mobility thus replicating clinical TPN delivery in humans. We also tested enteral drug delivery using this model to establish access for planned therapeutic interventions.

Methods: 7 day old piglets were surgically implanted with jugular vein (JV) and duodenal catheters (DC) which were exteriorized between the scapulae as described previously (Jain AK et al). The DC was additionally secured using a duodenal patch. The animals were fitted in jackets with large pockets on both sides. An ambulatory infusion pump (AMBIT Summit Medical, UT) was placed in one pocket. TPN (Clinimix E, Baxter, IL) and Intralipid (Fresenius Kabi, Germany) or swine replacement milk (LitterLife, Merrick’s Inc., WI) in EVA bags (Medtec Medical, IL) was placed in the other pocket.

Results: 4 piglets were successfully implanted with both JV and DC for nutritional support spanning 14 days. TPN piglets received via the jugular vein fluids at 260ml/kg/day with 26 g/kg dextrose, 11.05 g/kg protein and 5 g/kg fat along with electrolytes, trace minerals and vitamins for a total of 182 kcal/kg/day. Enteral Nutrition (EN) was delivered continuously via the DC. Nutrient bags were replaced every 8 hours. Mean daily weight gain (kg) for TPN vs EN animals was 0.116 vs 0.095. As expected small bowel density (g/cm) was less for TPN vs EN (0.183 vs 0.254). Successful enteral drug delivery via the DC as small volume injections followed by saline flushes was established. All animals remained healthy. Catheter sites and surgical wounds healed well. Due to non-tethered nutrition delivery the animals remained completely mobile.

Conclusion: Our successful infusion of TPN and EN using ambulatory pumps establishes a novel, ultramobile strategy for nutrition delivery and we believe fills a large lacunae of an absent model closely replicating nutrition delivery in humans.
INFANT SLEEP SAFETY ON A PEDIATRIC GENERAL MEDICAL UNIT

David A. Wathen

**Background:** In 2011, the AAP released updated more recent statement and provided the recommendation that all "Primary Care Providers...actively participate in the program" and that "healthcare professionals...should endorse the SIDS reduction recommendations from birth. There are no studies evaluating safe sleep adherence and education in a pediatric general medical.

**Objective:** To assess staff and parent compliance with AAP's safe sleep recommendations for hospitalized infants in a pediatric general medical unit.

**Design/Methods:** An observational study evaluating infants admitted to a general medical unit in a children’s hospital was conducted. Infants aged 0-12 months, on the general pediatric team were observed for adherence to the AAP's Safe Infant Sleep Environment from September 2012 until February 2013. Observations were performed daily during morning rounds by a single clinician and may have included the same subject on multiple days. Nursing staff, caregivers, residents and students were not aware of the ongoing observations until the conclusion of the study. Patients were excluded if they were admitted to the intensive care unit, intermediate care, the subject was awake, or the subject was being held by an awake caregiver.

**Results:** 234 observations were conducted and 163 observations met criteria to be included in the study. 94% (n=154) of subjects were noted to have at least 1 risk factor. Loose blankets accounted for 92% (n=150) of observed risks and pillows or soft positioning rolls accounted for 26% (n=42) of risks. All infants with a pillow or soft roll also had a loose blanket noted. 7% of observations (n=11) were co-sleeping with a caregiver.

**Conclusions:** Adherence to the AAP's Safe Infant Sleep Environment was found to be lacking in inpatient general medical units. This represents a great opportunity for improvement in role modeling and education of safe sleep practices in a large population of infants and their caregivers. Children’s hospitals should be encouraged to develop safe sleep programs for their general medical units. A future study is underway, evaluating compliance to a safe sleep program and the effects on caregiver education and future practice.
ABSTRACT FORM
Pediatric Science Days 2014 Research Symposium
Thursday, April 3, 2014

Presenting Author: Kyle Spradling

Please check one: ☑ Faculty Fellow ☐ Resident ☐ Post-Doc ☑ Student ☐ Other _____

Faculty Sponsor (required if trainee and not faculty): Barry Duel, M.D.

Faculty sponsor e-mail: duelbp@slu.edu

School/College/Dept/Division: Cardinal Glennon Children's Hospital/Department of Pediatric Urology

Contact E-mail: spradlkd@slu.edu Contact Phone: (417) 316-0094

Presentation topic (Please select one most relevant):

☐ Disease-Oriented Research: Basic
☑ Disease-Oriented Research: Clinical/Translational/Trials
☐ Disease-Oriented Research: Outcomes/Epidemiology
☐ Other: _____ (eg., developmental biology, signaling mechanisms, physiology, etc.)

VOIDING SYMPTOMS IN PEDIATRIC PATIENTS WITH ELEVATED CALCIUM-CREATININE RATIOS Kyle Spradling, Barry Duel

Purpose: The purpose of this study is to describe the voiding symptoms associated with elevated urinary calcium-creatinine ratios in pediatric patients.

Methods: From 2010 – 2013, 90 pediatric patients at one institution were found to have an elevated random urinary calcium-creatinine ratio (>0.200). Presenting voiding symptom (reason for visit) and patient calcium-creatinine laboratory values were recorded for these patients, and a one-way ANOVA analysis was used to compare calcium-creatinine ratios in patients grouped according to voiding symptom.

Results: Median age at the time of presentation was 8 years. Fourteen patients (15.6%) with elevated urinary calcium-creatinine ratios presented with symptoms of dysuria, 31 patients (34.4%) presented with urinary incontinence, 40 patients (44.4%) presented with nocturnal enuresis, and 5 patients (5.6%) presented with symptoms of daytime urinary frequency. There was no statistically significant difference (p = 0.16) between urinary calcium-creatinine ratio values in pediatric patients grouped according to voiding symptom.

Conclusion: Elevated urinary calcium-creatinine ratios are seen in pediatric patients with a variety of voiding symptoms. Among patients with elevated urinary calcium-creatinine ratios, there does not appear to be a relationship between the value of calcium-creatinine ratios and the type of voiding symptom in pediatric patients.
CHEMOTHERAPY AS A FIRST LINE TREATMENT FOR A SUBTOTALLY RESECTED ROSETTE-FORMING GLIONEURONAL TUMOR OF THE FOURTH VENTRICLE IN A 6-YEAR OLD CHILD: CASE REPORT AND LITERATURE REVIEW OF PEDIATRIC CASES

Cynthia Morris, Yongxin Chen, Vilaas Shetty, Samer Elbabaa, Miguel Guzman, Mohamed S. Abdel-Baki

Background:
Rosette-forming glioneuronal tumor of the fourth ventricle (RGNT) is a rare WHO grade 1 neoplasm. A total of 91 cases have been reported in English language literature, of which 21 are pediatric cases. Recurrence has been reported in only 6 cases of all ages so far. Gross total resection (GTR) is the treatment of choice, and there is no firm evidence base for other treatment options when GTR is not feasible. Radiation has been used in a few cases, but chemotherapy was used just once before, on a recurrent tumor with extensive leptomeningeal dissemination.

Methods:
After initial subtotal resection of the tumor, we administered an individualized chemotherapy protocol with Vincristine, Etoposide, and Carboplatin for three cycles on a 6-year-old boy with a rosette-forming glioneuronal tumor of the fourth ventricle. Each three-week cycle consisted of Vincristine 1.5mg/m² weekly, Etoposide 100mg/m² for the first three days of each cycle, and Carboplatin 640mg/m² for the first day of each cycle.

Results:
Our patient had no clinical or radiographic evidence of tumor progression for almost 2 years after the subtotal resection. Gross total resection was successfully performed later, and at 18 months after surgery the patient currently does not show any evidence of tumor recurrence.

Conclusion:
We report the first case where chemotherapy has been administered as a first line treatment for a subtotally resected RGNT. Further trials are needed to explore chemotherapy as an adjuvant treatment for rosette-forming glioneuronal tumors where gross total resection cannot be achieved.
THE REPORT OF A CASE SERIES: PEDIATRIC PSEUDOANEURYSM WITH ABSENT TO MINIMAL HISTORY OF TRAUMA

Arty Namin, Christina Plikaitis
Saint Louis University School of Medicine 1; Saint Louis University School of Medicine, Department of Surgery, Division of Plastic Surgery 2

**Background:** Head and neck traumatic pseudoaneurysms are reported in the literature following significant history of trauma to the region. Pseudoaneurysms tend to present 2-4 months after trauma, thus many patients do not associate their symptoms with trauma. The overlying skin is sometimes discolored, and may at times be mistaken as an abscess. The mass is typically pulsatile, and a bruit can occasionally be auscultated.

**Objective:** Describe a diagnostic protocol for cystic appearing masses in the head and neck of pediatric patients. Describe a therapeutic protocol for pseudoaneurysms of the head and neck. Emphasize the importance of considering pseudoaneurysm in the differential diagnosis of cystic masses in the head and neck, as well as stress the importance of palpation and auscultation in the examination of cystic masses in the head and neck.

**Methods:** Chart and literature review.

**Results:** Four pediatric patients presented with cystic masses in the head and neck area. Although one of the patients did report a traumatic event to the region of the pseudoaneurysm, three of the patients could not recall a traumatic event. Upon palpation, all four of the cystic masses were pulsatile. All four cases were treated with surgical excision, with excellent outcomes.

**Conclusions:** Pediatric patients who present with cystic masses in the head and neck region must be carefully assessed. A thorough history regarding traumatic events in the past months is important. Careful physical exam including palpation and auscultation is extremely important. The differential diagnosis should include lipoma, hematoma, epidermoid inclusion cyst, abscess, and inflamed lymph node. Surgical excision of pseudoaneurysms provides excellent outcomes.
RESOLUTION OF PRECOCIOUS PUBERTY FOLLOWING SURGICAL RESECTION OF FOURTH VENTRICULAR MEDULLOBLASTOMA: CASE REPORT AND REVIEW OF LITERATURE
Renata G Medina, David P Dempsher, Karen M Gauvain, Thomas J Geller, Samer K Elbabaa

Objective and Importance: Medulloblastoma is a malignant embryonal tumor that arises in the cerebellum and invades the fourth ventricle, often resulting in obstructive hydrocephalus. Patients typically present with symptoms related to increased intracranial pressure and cerebellar dysfunction. We report a rare case of classic medulloblastoma presenting with central precocious puberty as its only presenting symptom.

Clinical Presentation: A seven-year-old male with no prior history of medulloblastoma, presented with Tanner Stage IV testicular enlargement, and a four-month history of acne and pubic hair. Blood labs demonstrated highly elevated LH, FSH, and testosterone. MRI of the brain revealed a mass in the posterior fossa, which bordered and compressed the fourth ventricle. The patient also exhibited mild lateral and third ventriculomegaly. Surgical options were discussed with the neurosurgical department.

Intervention: A suboccipital craniotomy and C1 laminectomy was performed. A large mass was seen arising from the inferior surface of the vermis, and lying within the fourth ventricle. Microsurgical resection of the mass was performed until the midline and the brainstem could be appreciated up to the aqueduct of Sylvius, and CSF could be seen flowing from the third ventricle into the fourth ventricle. Histopathology characterized the tumor as classic medulloblastoma. Follow-up blood labs demonstrated a reduction of LH, FSH, and testosterone back to pre-pubertal levels. The patient then began radiation and chemotherapy.

Conclusion: This report demonstrates that mild obstructive hydrocephalus due to a posterior fossa tumor may present with unexpected symptoms, such as central precocious puberty. To our knowledge, precocious puberty has not yet been associated with medulloblastoma, though it has been found with other posterior fossa tumors. [6] Extensive imaging of the CNS for patients presenting with central precocious puberty is recommended.
AGE OF INITIATION OF HELMET THERAPY DOES NOT IMPROVE DURATION OF TREATMENT, RATE OF IMPROVEMENT, OR FINAL OUTCOME IN CHILDREN WITH DEFORMATIONAL PLAGIOCEPHALY  Chelsea R. Horwood, Emma M. Kulig, Sarah A. Donigian, Alexander Y. Lin

Background: It is commonly thought that helmet therapy for deformational plagiocephaly is more effective the earlier the age of initiation, but this did not seem the case for our patients. We therefore sought to compare age of initiation with helmet therapy outcomes.

Methods: Patients with deformational plagiocephaly who underwent helmet orthotic treatment from 2006 to 2013 were retrospectively reviewed. Asymmetry was measured by transcranial difference (TCD) between frontozygomatic-to-eurion diagonals in millimeters. Analysis of variance was used to compare groups; results in means (standard deviations).

Results: Patients were stratified into 6 groups (G1,5,6,7,8,9), by age in months: G1, <5 (n=13); G5, 5 (n=40); G6, 6 (n=53); G7, 7 (n=45); G8, 8 (n=23); G9, 9 (n=35). Initial TCDs: G1 11.1 (±4.0), G5 10.3 (±3.6), G6 10.8 (±4.1), G7 11.0 (±3.2), G8 9.30 (±4.7), G9 9.00 (±4.5), P=0.165. Final TCDs: G1 2.39 (±1.7), G5 3.74 (±1.98), G6 3.72 (±2.0), G7 3.51 (±1.6), G8 3.83(±2.5), G9 3.49 (±2.5), P=0.391. Change between final and initial TCD: G1 8.69 (±3.7), G5 6.72 (±3.3), G6 7.0 (±3.1), G7 7.49 (±3.1), G8 5.68 (±3.6), G9 5.51 (±3.5), **P=0.010; Tukey posthoc pairwise comparisons showed G9 significantly less than G1. Duration of treatment in months: G1 3.87 (±.88), G5 4.24 (±2.1), G6 3.97 (±1.99), G7 3.68 (±1.36), G8 3.85 (±1.5), G9 3.92 (±2.4), P=0.777. Rate of change (change in TCD over duration, mm/month): G1 2.22 (±.86), G5 1.87 (±1.2), G6 2.11 (±1.2), G7 2.29 (±1.3), G8 1.92 (±1.7), G9 1.61 (±1.0), P=0.201.

Conclusions: When patients were stratified by their initial age of therapy, we found that duration of helmet therapy, final TCD, and rate of change to be similar without significant differences. There was slightly less TCD change in G9, but no differences in initial or final TCDs. One possibility is that our patients had longer durations of helmet therapy, and perhaps this helped standardize our results. This study cautiously suggests that age at initiation of helmet therapy may not make as much of a clinical difference as previously thought.
ABSTRACT FORM
Pediatric Science Days 2014 Research Symposium
Thursday April 3, 2014

Presenting Author: Emma M. Kulig, BA

Please check one: ☐ Faculty Fellow ☐ Resident ☐ Post-Doc ☐ Student ☐ Other _____

Faculty Sponsor (required if trainee and not faculty): Alexander Y. Lin, MD
Faculty sponsor e-mail: aylin@slu.edu

School/College/Dept/Division: School of Medicine/Surgery/Pediatric Plastic Surgery
Contact E-mail: aylin@slu.edu Contact Phone: 314-678-2171

Presentation topic (Please select one most relevant):
☐ Disease-Oriented Research: Basic
☐ Disease-Oriented Research: Clinical/Translational/Trials
☐ Disease-Oriented Research: Outcomes/Epidemiology
☐ Other: Clinical case report and literature review

EFFECTS OF TORTICOLLIS ON HELMET THERAPY FOR DEFORMATIONAL PLAGIOCEPHALY
Emma M. Kulig, Sarah A. Donigian, Chelsea R. Horwood, Alexander Y. Lin

Background/Purpose: Children with deformational plagiocephaly frequently have some degree of torticollis. It is commonly thought that concomitant torticollis makes deformational plagiocephaly treatment more refractory. Among patients undergoing helmet therapy for deformational plagiocephaly, we compared those with torticollis and those who did not have torticollis.

Methods/Description: Patients with deformational plagiocephaly treated by helmet orthotic from 2006 to 2013 were retrospectively reviewed. The helmet orthotist recorded standard cranial measurements at each clinical visit, and only patients who completed their treatment course with final measurements were included. Parametric tests (t-tests) and chi-square tests were used for variable analyses.

Results: 157 patients met the inclusion criteria. Torticollis (T) was seen in 59.9% (94/157), P=0.098. Helmet therapy was initiated at age in months adjusted for prematurity: T 6.14, NT 6.7, P=0.069. Asymmetry was measured by transcranial difference (TCD) between frontozygomatic-to-eurion diagonals in millimeters, with initial TCD: T 11.8, NT 8.3, ***P<0.001; final TCD: T 3.9, NT 3.1, *P=0.013; change in TCD: T 7.9, NT 5.2, ***P<0.001. The duration of therapy in months was: T 3.91, NT 3.85, P=0.819; with rate of TCD change (mm/month) being: T 2.33, NT 1.60, *P<0.001. 47.3% (70/148) of torticollis patients underwent physical therapy (PT) for neck exercises, and their average final TCD (in mm) was 3.8, compared to those who did not get PT at 3.3, P=0.212.

Conclusions: Our data suggest that torticollis does not significantly affect effectiveness or duration of helmet therapy. Although torticollis patients had greater initial transcranial differences (TCD), they ended with statistically different but clinically similar final TCD measurements (3.9 vs. 3.1mm). Helmet therapy duration was similar despite the worse initial asymmetry. This greater rate of TCD change is not explained by the similar age at initiation of helmet therapy. Infants with torticollis who received physical therapy did not have improved final transcranial differences as compared to those who did not receive physical therapy. These results suggest that although torticollis and deformational plagiocephaly often occur hand in hand, helmet therapy outcomes appear to proceed independently.


INTERNAL CAROTID ARTERY BYPASS TO EXTERNAL CAROTID ARTERY BYPASS IN AN 18 MONTH OLD INFANT; A CASE REPORT, DISCUSSION OF A SINGLE INSTITUTION’S EXPERIENCE IN NON-VEIN OF GALEN PEDIATRIC ANEURYSMS AND REVIEW OF LITERATURE

Catherine F McClung Smith, Samer K Elbabaa, Ann Marie Flannery, Jeroen Coppens, Tonya Quinn, Randal Edgell, Saleem I Abdulraulf

Background:
Non-vein of Galen intracerebral aneurysms are a rare entity in the pediatric population.

Methods:
The authors present 4 cases of primary intracerebral aneurysm diagnosed in children under the age of 24 months from March 2010 through March 2013. The presentation, location, treatment strategies and follow up of the aneurysms are discussed.

Results:
In two cases a good outcome was obtained with the infant being discharged with a good functional status. The first infant presented with new onset seizure activity resulting in an MRI and diagnosis of a complex left MCA bifurcation aneurysm. This infant underwent elective external carotid to internal carotid bypass of the MCA bifurcation aneurysm utilizing a saphenous vein graft with subsequent discharge home at her baseline functional level 8 days postoperatively. The second infant was diagnosed secondary to a parietal hemorrhage and the aneurysm was treated endovascularly. This infant’s outcome was directly related to the location of the hemorrhage and prompt treatment of the ruptured aneurysm; the infant has managed a good recovery. Of the 2 infants with poor outcomes, one infant was discharged devastated after rupture of a fusiform right A2 aneurysm and the second infant was discharged deceased, secondary to rupture of a partially thrombosed basilar tip aneurysm.

Conclusion:
While cerebral aneurysms in the infant population are exceedingly rare, they remain a treatable disease with potential for good outcomes depending primarily on the presentation of the patient. Based on our institution’s experience, even complex aneurysm requiring bypass are potentially curable.
PERCEPTIONS AND IMPACTS OF CHILDCARE RESPONSIBILITIES ON HEALTHCARE WORKERS FOLLOWING A DISASTER

Rachel L. Charney, Terri Rebmann, Robert G. Flood

Background: In the spring of 2011, an EF5 tornado swept through the city of Joplin, MO, destroying a section of the city up to a mile wide and directly hitting one of the city’s major hospitals. Objective: To measure the perception of having childcare responsibilities on healthcare workers ability and willingness to work during the week following a natural disaster.

Methods: A survey was distributed to all healthcare workers at both the evacuated hospital and a nearby intact hospital in May 2013. With respect to those with children and their ability and willingness to work, the following predictors were considered in the survey: 1) providers’ alternative childcare (AC) plans before and after this disaster; 2) additional costs accrued by the providers for AC; 3) impact of altered work schedule; and 4) the presumed impact of a theoretical hospital-provided AC facility. Chi-square was used for comparisons. A McNemar test was used to compare those with AC plans pre and post tornado.

Results: Most of the 1234 respondents were female (83.8%), white (96.9%), and worked the week following the tornado (88%, n = 1083). About half (48.5%, n = 598) had children, and this cohort was the focus for most of the study. Many (61.2%, n = 366) reported having childcare needs the week of the tornado, approximately half of which (54%, n = 198) required the use of AC, that, for 20.3%, cost up to $100/week extra, and, for 25%, caused the provider to be very concerned for their child’s wellbeing. More have a personal AC plan now compared to before the tornado (61% then vs 82% now; p < .001). Of those with a plan at the time of the tornado, 60% executed their plan; those without plans were no less likely to have actually worked, although 46% reported they would have been more able and/or willing to work if they had AC plans beforehand. Altered shifts made it more difficult to find childcare for 12%. If their hospital had provided AC: 51% of those needing childcare would have used it, including 45% of those with existing AC plans; 42% would have felt less concerned for their children’s wellbeing; and 47% and 42% felt they would have been more able and willing to work, respectively. Eleven percent (11%) believe that childcare during a disaster is at least jointly the hospital’s responsibility to provide.

Discussion: The overwhelming percentage of respondents reported for work following this disaster. Yet, their perception of childcare needs, especially AC, appears to have placed a psychological and financial burden on these workers. Disaster planners should be aware of these perceptions as they formulate their own emergency operation plans.
A FIXED-DOSE KETAMINE PROTOCOL FOR ADOLESCENT SEDATIONS IN A PEDIATRIC EMERGENCY DEPARTMENT Megan Street¹, James Gerard²
Departments of Pediatrics, UT Southwestern School of Medicine¹; Saint Louis University².

Objectives: To assess provider and patient satisfaction with a fixed-dose ketamine protocol for procedural sedation of adolescent subjects. We further compared data for normal weight [body mass index (BMI) ≤ 25] versus overweight/obese subjects (BMI > 25).

Study Design: Prospective, observational cohort study of adolescent patients undergoing procedural sedation in a pediatric emergency department. Adequate sedation was defined as a Ramsay sedation score (RSS) ≥ 5. Subjects received an initial 50 mg IV ketamine dose followed by 25 mg IV doses to maintain an RSS ≥ 5. The sedating physician, procedural physician and sedating nurse independently rated the sedations on a 100 mm visual analog scale (0 - “very unsatisfied”, 100 - “very satisfied”). Subjects and their guardians were contacted 12 - 24 hours post-sedation.

Results: Forty-three subjects (26 normal weight, 17 overweight/obese), ages 12 - 17 years, were enrolled in the study. An RSS ≥ 5 was observed in 35 (81.4%) of the subjects following the initial 50 mg ketamine dose and in the remaining 8 subjects following the first additional 25 mg dose. The median combined provider satisfaction score for the sedations was 92.7 (IQR 83.7, 95.0) and was similar for the normal weight and overweight/obese groups [93.1 (IQR 84.6, 95.9) versus 89.7 (IQR 83.7, 93.5) respectively, P = 0.27]. Subjects and guardians in both groups reported high rates of satisfaction.

Conclusion: The fixed-dose ketamine protocol resulted in an adequate level of sedation and high provider/patient satisfaction for the majority of patients regardless of weight or BMI status.
THE USE OF A PEDIATRIC MIGRAINE PRACTICE GUIDELINE IN AN EMERGENCY DEPARTMENT

Albert Nakanishi1, Courtney Kaar2, James Gerard3

1,3 Pediatrics, Saint Louis University School of Medicine, St Louis, MO 63104, United States; 2Saint Louis University School of Medicine, St Louis, MO 63104, United States.

Background: In the Emergency Department (ED), pain care for children with migraine headaches is often varied and evidence based treatment is inconsistently applied. We describe our experience with an evidence based, clinical practice guideline for the ED management of migraine headaches in children.

Objective: To evaluate the safety and efficacy of an ED pediatric migraine clinical practice guideline.

Design/Methods: A pediatric migraine protocol was established in collaboration between the Department of Neurology and Emergency Medicine utilizing evidence based data and best practice on the treatment of migraine headaches in children. The Migraine Clinical Practice Guideline (MCPG) was implemented for patients with an established diagnosis of migraines and with a numeric pain score (PS, 0-10) >6. All patients received intravenous saline, ketorolac, diphenhydramine, and either metoclopramide or prochlorperazine. After 40min a repeat PS was obtained and if not improved a repeat dose of metoclopramide or prochlorperazine was administered. If after 40min no significant pain relief was achieved, a Neurology consultation was obtained. A retrospective chart review identifying patients with the discharge diagnosis of migraine headache and enrolled in the ED-MCPG over a 4 yr period (4/09-4/13) was conducted. A data sheet was created recording demographic data, vital signs, ED length of stay, initial PS, last recorded PS, adverse events and if the patient required admission.

Results: A total of 533 charts were identified with the discharge diagnosis of migraine headache, of which, 266 were enrolled in our MCPG, 179 females (67.3%) and 87 males (32.7%) with an average age of 13.9y (5-20y). Mean initial PS were 7.8±2.0 and mean discharge PS were 2.1±2.8; representing a 73% reduction of pain. There were 20 patients admitted for status migrainous with an average age of 14y±3.5 and average PS=6.3±2.8 with an admission rate of 7.5%. Total length of stay in the ED was 283 min±107. No adverse events were identified.

Conclusion: We conclude, our clinical practice guideline appears to be safe and efficacious for children seeking migraine headache relief in an ED setting.
CRITICAL CARE INTERVENTIONS FOR ASTHMATIC PATIENTS ADMITTED FROM THE EMERGENCY DEPARTMENT TO THE PEDIATRIC INTENSIVE CARE UNIT

Kristen Cundiff, James Gerard, Robert Flood

Department of Pediatrics, Saint Louis University.

**Background:** Evidence-based pediatric intensive care unit (PICU) admission criteria for patients in status asthmaticus are lacking. Previous work suggests that clinical pathways appear to be effective in reducing length of stay and costs for asthma-related admissions to the pediatric wards. Most studies, however, have excluded the PICU from protocols and data collection.

**Objective:** To assess the frequency and predictors of critical interventions in asthmatic patients admitted to the PICU.

**Design/Methods:** Retrospective chart review of patients admitted from our emergency department (ED) to the PICU for treatment of status asthmaticus between January 1, 2008 and March 31, 2013. Patients were identified by ICD-9-CM discharge diagnosis code for asthma and ICU-specific UB-92 revenue codes. Data collected include demographics, clinical characteristics at the time of admission (e.g. clinical asthma score), hospital course, treatment course, and complications.

**Results:** A total of 568 patients were identified with a mean age of 7.6y ± 4.8 (59.5% male, 40.5% female). Seventeen (2.7%) required intubation, 3 (0.5%) received CPAP, 2 (0.3%) received BiPAP, 7 (1.1%) had a CVC placed, 1 (0.2%) developed circulatory collapse requiring chest compressions, 2 (0.3%) developed pneumothoraces. ED Predictors for patients requiring critical interventions are currently being explored.

**Conclusions:** Patients admitted to the PICU for status asthmaticus infrequently require critical interventions. Additional data collected in this study will be helpful in identifying patients with an increased risk of requiring critical interventions.
Objective: To evaluate the predictive value of commercially available biomarkers for progressive extrinsic atopic dermatitis (AD): total IgE and % positivity of the Atopic March Immunocaps Panel.

Methods: In this 3.6 year retrospective single center chart review, 74 children aged 9 mo - 18 yr with mild - severe AD were characterized as intrinsic versus extrinsic based on age-adjusted norms of total IgE. AD patients were included if they had 2 sets of total IgE and immunocaps panels and at least 2 visits to the dermatology clinic. Patients who failed to attend clinic visits within 3 months of the lab draws were excluded.

Results: The average age was 7.0 yrs with a range of 9 months to 18 years, and the average interval to follow up was 0.9 yr. AD was intrinsic in 18% (n=8) and extrinsic in 82% (n=37). Demographics and comorbidities were similar among the intrinsic and extrinsic groups. At baseline, 69% (n=42) of the extrinsic AD patients had severe disease compared to 31% (n=4) of intrinsic patients, p=0.019. Among the subset of severe extrinsic patients whose disease improved at follow-up, there was no significant baseline difference in total IgE (IU/mL) compared to those whose disease remained poorly controlled (9,235 vs 7,467; p=0.66). However, total IgE decreased significantly in those patients whose disease became controlled but increased in poorly controlled patients, (4,798 vs 12,916; p=0.01: a 4,437 decrease vs an 5,449 increase, p=0.001). Total IgE decreases were similar among patients treated with topical therapy versus systemic therapy patients, [absolute change of total IgE, (-)5,238.8 vs (-) 3,125.4, p=0.500]. Eczema herpeticum and failure to thrive occurred more frequently among the extrinsic poorly controlled group versus the controlled group, [eczema herpeticum (n=7) 24% vs (n=8) 62%; p=0.04: failure to thrive, (n=1) 3% vs (n=6) 46%; p=0.002].

Limitations: Retrospective chart review performed on a small sample size at a single center with no severity scoring system used.

Conclusions: This study supports the use of total serum IgE as a readily available laboratory test that may be a prognostic indicator of AD progression. It supported a relationship between poorly controlled extrinsic AD and progressively increasing total IgE and correlated a decrease in total IgE with disease control in extrinsic AD whether treated with systemic or topical therapy.
ABSTRACT FORM
Pediatric Science Days 2014 Research Symposium
Thursday, April 3, 2014

Presenting Author: Stephanie Frisch MD

Please check one: [ ] Faculty Fellow [x] Resident [ ] Post-Doc [ ] Student [ ] Other _____

Faculty Sponsor (required if trainee and not faculty): Elaine Siegfried MD

Faculty sponsor e-mail: esiegfri@slu.edu

School/College/Dept/Division: Dermatology and Pediatrics

Contact E-mail: sfrisch@slu.edu  Contact Phone: 310-466-6137

Presentation topic (Please select one most relevant):

[ ] Disease-Oriented Research: Basic
[ ] Disease-Oriented Research: Clinical/Translational/Trials
[ ] Disease-Oriented Research: Outcomes/Epidemiology
[ ] Other: _____ (eg., developmental biology, signaling mechanisms, physiology, etc.)

ANALYSIS OF IMMUNE FUNCTION IN CHILDREN WITH ECZEMA
Stephanie Frisch, Michael Ansstas, Elaine C. Siegfried

Background: Atopic dermatitis (AD) is the most common chronic inflammatory skin condition in children, characterized by red scaly skin and severe itch. AD is a phenotype. Genetically well-defined subsets include Netherton, Wiscott-Aldrich, and Hyper-IgE syndromes, but the majority of children with comorbidities have underappreciated abnormalities that do not fit into a genetically-defined syndrome. Current standard of care includes systemic treatment with immunosuppressive medications (cyclosporine, azothioprine, mycophenylate mofetil, methotrexate). Biologic immunomodulating agents are used less often, but may be a better option for children with immune dysfunction.

Objective: The objective of this retrospective analysis was to identify immunologic abnormalities in children with severe inflammatory skin disease, and correlate these with responses to immunosuppressive vs. immunomodulatory treatment.

Methods: Saint Louis University IRB-approved retrospective chart review of children seen in the Pediatric Dermatology clinic during a 5 year period.

Results: Thirty patients with severe inflammatory skin disease, microbiologic and laboratory evaluation were included. Recurrent skin infection with group A Strep and herpes simplex was common, including 4 patients with acyclovir resistant herpes. The most common immune abnormalities were selective antibody deficiency and decreased toll-like receptor levels followed by combined humoral and cellular immunodeficiency. Patients with an identifiable immune abnormality tended to respond less well to methotrexate compared with those whose immune evaluation was unremarkable. Immune-deficient patients treated with biologic immunomodulating agents were better controlled on IVIG than interferon gamma.

Limitations: Small number of patients, some with incomplete immune evaluations. Interpretation of laboratory results was complicated by concomitant treatment with methotrexate or interferon gamma in a subset of patients.

Conclusion: Evaluation of children with severe eczema should include a past medical history of frequent infections and screening for recurrent skin infections. Patients who fail to improve with aggressive topical therapy or systemic immunosuppressants may benefit from immune function evaluation and alternative treatment with biologic immunomodulatory agents. Selective antibody deficiency was the most common abnormality in this small cohort.
ENCEPHALOCRANIOCUTANEOUS LIPOMATOSIS: EVIDENCE FOR MOSAICISM AND EARLY BRAIN TUMORS

Stephen R. Braddock, Katherine M. Christensen, Erin E. Torti, Samer K. Elbabaa
Department of Pediatrics and Neurosurgery, Saint Louis University, Saint Louis, MO

Encephalocraniocutaneous lipomatosis (ECCL) is a rare neurocutaneous syndrome manifest by nevus psiloliparus, choristomas and CNS lipomas. Brain malignancies have been rarely reported. Moog (JMG, 2009) proposed that the etiology might represent mosaicism for genes involved in mesenchymal tumors and vasculogenesis. The purpose of this presentation is to describe 2 cases of ECCL, one with findings lending support to underlying mosaicism and another with an earlier onset of intracranial malignancy.

Case 1 was born at 36.9 weeks gestation with weight 2680 grams, length 44 cm and OFC 33 cm. Pregnancy complicated by fetal SVT and hydrops. Exam had occipital cutis aplasia congenita, corneal clouding, unilateral microphthalmia, pedunculated tags by the canthus, a mass under the eyelid, absent eyelashes, iris and optic nerve colobomas and absence of the superior iris. The skin had a raised yellowish waxy hypertrophic rash on forehead, face, trunk and extremities following Blaschko’s lines. There was bilateral alopecia. MRI showed ventriculomegaly, thin corpus callosum and small cerebellum. In addition there was a possible posterior fossa fat mass. Skeletal survey and microarray were normal. At 16 months the exam was essentially unchanged except for poor growth, microcephaly and the skin findings had progressed into a patchy hyperpigmented hyperkeratotic verrucous appearance over the face, trunk and extremities.

Case 2 was born at term weighing 3480 grams, length 51 cm, and OFC 35.3 cm. He had scalp cutis aplasia, preauricular skin tag, multiple eyelid tags and right parietal alopecia with a 10 cm pink fluctuant plaque. Microarray was normal. Initial brain MRI was normal except for possible parietal lipoma, but repeat at 3 months showed a 2 cm mass in the hypothalamic region with extension into the optic chiasm and a lipomatous lesion in the right parietal region with bony defect. Pathology revealed grade II pilocytic astrocytoma with mixed higher grade pilomyxoid components and high proliferating index.

These cases represent new findings in ECCL. The progressive hyperkeratotic changes seen in case 1 in such a distinct pattern suggest further evidence of mosaicism in these neural crest derived tissues. Comparative molecular analyses are pending. Case 2 is the fourth reported astrocytoma in ECCL, but is unique in the earlier asymptomatic presentation, rapid growth and higher grade. Therefore, early MRI monitoring of the asymptomatic patient with ECCL is indicated.
ABSTRACT FORM

Pediatric Science Days 2014 Research Symposium
Thursday, April 3, 2014

Presenting Author:  Meg Hefner, MS

Please check one:  X  Faculty Fellow  □  Resident  □  Post-Doc  □  Student  □  Other  ____

Faculty Sponsor (required if trainee and not faculty):  ____

Faculty sponsor e-mail: hefnerma@slu.edu

School/College/Dept/Division:  Dept of Pediatrics, Division of Medical Genetics

Contact E-mail: hefnerma@slu.edu  Contact Phone: 768-8371

Presentation topic (Please select one most relevant):

☐ Disease-Oriented Research: Basic
☐ Disease-Oriented Research: Clinical/Translational/Trials
☐ Disease-Oriented Research: Outcomes/Epidemiology
☐ Other:  ____  (eg., developmental biology, signaling mechanisms, physiology, etc.)

---

**CHARGE SYNDROME CLINICAL DATABASE PROJECT (CSCDP): EARLY EXPERIENCE OF A WEB-BASED, PARENT-REPORT DATABASE AND REGISTRY**  
*Meg Hefner, Emily Fassi, Kevin Ballard*

CHARGE syndrome (CS) is a complex genetic condition which can affect nearly every organ system, with effects ranging from mild to severe. Research on CS has been limited by the task of confirming the diagnosis (by CHD7 mutation or clinical criteria) and by the unique combination of features in each case. This database of CS individuals will facilitate CS research, aid in the search for other possible causes of the CS phenotype, and provide more information about natural history. CSCDP is a web-based, parent-completed database. Parent report was chosen because in a condition as complex as CS, multiple providers (medical and educational), hospitals and offices are utilized – there is often no medical home and it is the parents who have the most complete set of records, as well as the time and motivation to complete the survey. Qualtrics was chosen for CSCDP because of an existing contract with SLU and the versatility of the platform. CSCDP consists of ~500 questions in 12 sections, including birth history, genetic testing, diagnostic features, surgeries and hospitalizations, growth and development. Opportunities are provided for uploading photographs and selected medical records (e.g. genetic tests, audiograms). “Display logic” is used to conditionally display questions (e.g. if the child with CS is living, cause of death is not displayed). We estimate that each participant sees 30-50% of the questions. Total time for completing the 12 sections is 2-3 hours. This can be done in multiple “sittings” over several months. Periodic updates will be requested to collect longitudinal information. This sort of project could not have been done 10 years ago, but is now possible because of the evolution of the Internet and social media. The goals of CSCDP are to provide a more complete picture of CHARGE syndrome, its variation and natural history and to serve as a repository of baseline information and source of potential subjects to other CS researchers around the world. CSCDP was launched in May, 2013. In the first 6 months, of the 88 people who have expressed interest, 70 have consented and 50 have completed the diagnostic criteria surveys. The completed surveys have already provided valuable information about validity of parent-completed surveys.
PEDIATRIC SCIENCE DAYS 2014

THURSDAY, APRIL 3
7:30 AM – 3:30 PM
Danis Auditorium, Cardinal Glennon Medical Center

PEDIATRIC RESEARCH SYMPOSIUM

FRIDAY, APRIL 4
7:30 AM – 9:00 AM
Danis Auditorium, Cardinal Glennon Medical Center

PEDIATRIC GRAND ROUNDS

M. Michele Mariscalco, MD
Regional Dean, University of Illinois College of Medicine - Urbana
Urbana, IL

"Research...patient care...what, me worry? "

FRIDAY, APRIL 4
10:00 AM – 11:30 AM
Mund Room, Cardinal Glennon Medical Center

ACADEMIC ROUNDTABLE for Trainees/Jr. Faculty
Moderator: M. Michele Mariscalco, MD