34th Annual

THOMAS F. BOAT DAY OF SCHOLARSHIP

April 18, 2019

The Thomas F. Boat Day of Scholarship

"Since 1985, the intellectual highlight of the Department of Pediatrics has been its annual Evening of Scholarship. This was conceived by Dr. Boat as a festive evening in which pediatric residents and fellows could present the results of their scientific studies to their peers and faculty members. Although initially proposed as an Evening of Research, several members of the faculty suggested that Evening of Scholarship be substituted, since the term "research" often conjured up a limited image of wet bench research. Such a perception might have a negative effect on the main goal of the exercise, which was to encourage each of our pediatric house staff to engage in some sort of scholarly activity beyond their usual ward and clinic assignments. Such activities might range from a case report and review of the literature about some disease to a sophisticated laboratory or epidemiologic study. Since its inception the Evening of Scholarship has developed into a showcase event in the Department's spring calendar."

- Taken from "From Infancy to Maturity: The History of the Department of Pediatrics, The University of North Carolina at Chapel Hill, 1952-1995".

Awards Presented at the Day of Scholarship

The primary intent of the Thomas F. Boat Day of Scholarship is to acknowledge and honor residents and post-doctoral fellows in the Department of Pediatrics for their scholarly efforts during the academic year. While all the presentations offered at this event are meritorious, a committee has been appointed to identify presentations and posters of particular distinction. The committee is charged with evaluating these presentations and awarding the best basic science presentation, the best clinical science presentation, the best QI presentation, the best overall presentation, and the best presentation by a pediatric resident. In 2002, the decision was made to name the basic science award in honor of Dr. Jud Van Wyk and the clinical science award in memory of Dr. Walter Tunnessen. In 2009, a new prize was established, the Johnny L. Carson Award. This award is non-categorical and is given to the presenter showing significant scholarly contribution to the Day of Scholarship. The award is named in the spirit of Johnny L. Carson, a leader in promoting scholarship in the Department of Pediatrics. In 2010, the best Quality Improvement presentation was named after Gerald Fernald, MD, an advocate for resident education. Even in his retirement, Dr. Fernald was active in resident recruitment and the Day of Scholarship. In 2012, the Best Resident Award was named in honor of Alan Stiles, former Chair of the Department of Pediatrics. During Dr. Stiles' 11 year tenure as department chair, resident scholarship was emphasized and expanded, and always at the forefront of research initiatives. The recipients of these honors receive an individual plaque, a monetary award, and have their names added to a large plaque that is permanently displayed in the Curnen-Denny Conference Room. All monetary awards come from the generous gift of Dr. and Mrs. Jack Lynch that originally established the London-Lynch Learning Center. Dr. Lynch passed away November 16, 2010. The London-Lynch Learning Center has contributed to promoting the Evening of Scholarship and in funding these awards.

On behalf of the London-Lynch and the Resident Scholarship Support Committees, we congratulate all the participants in this year's event and welcome you to a lifetime of learning for 21st century pediatricians.

Schedule

8:00am **Oral Presentations**
Kirkland Auditorium (Dental School)

The Arresting Vertical Transmission of Hepatitis B Virus (AVERT-HBV) Study in the Democratic Republic of the Congo: Preliminary Results
Peyton Thompson, MD
Infectious Disease Fellow
Mentor: Steven Meshnick, MD, PhD
Abstract available on page 22

House dust promotes sensitization to environmental peanut through the airways
Peter Balogh
Undergraduate Student
Mentor: Tim Moran, MD, PhD
Abstract available on page 12

Quality Improvement (QI) for Universal Screening of Gonorrhea and Chlamydia (GC/CT) in a Resident Primary Care Clinic
Bianca Allison, MD, MPH
Pediatrics Resident
Mentor: Martha Perry, MD
Abstract available on page 26

Institutional Gender Symposium
Amalia Lee
MD Candidate
Mentor: Nina Jain, MD
Abstract available on page 37

12:00pm **Noon Conference presented by Stephanie Davis, MD**

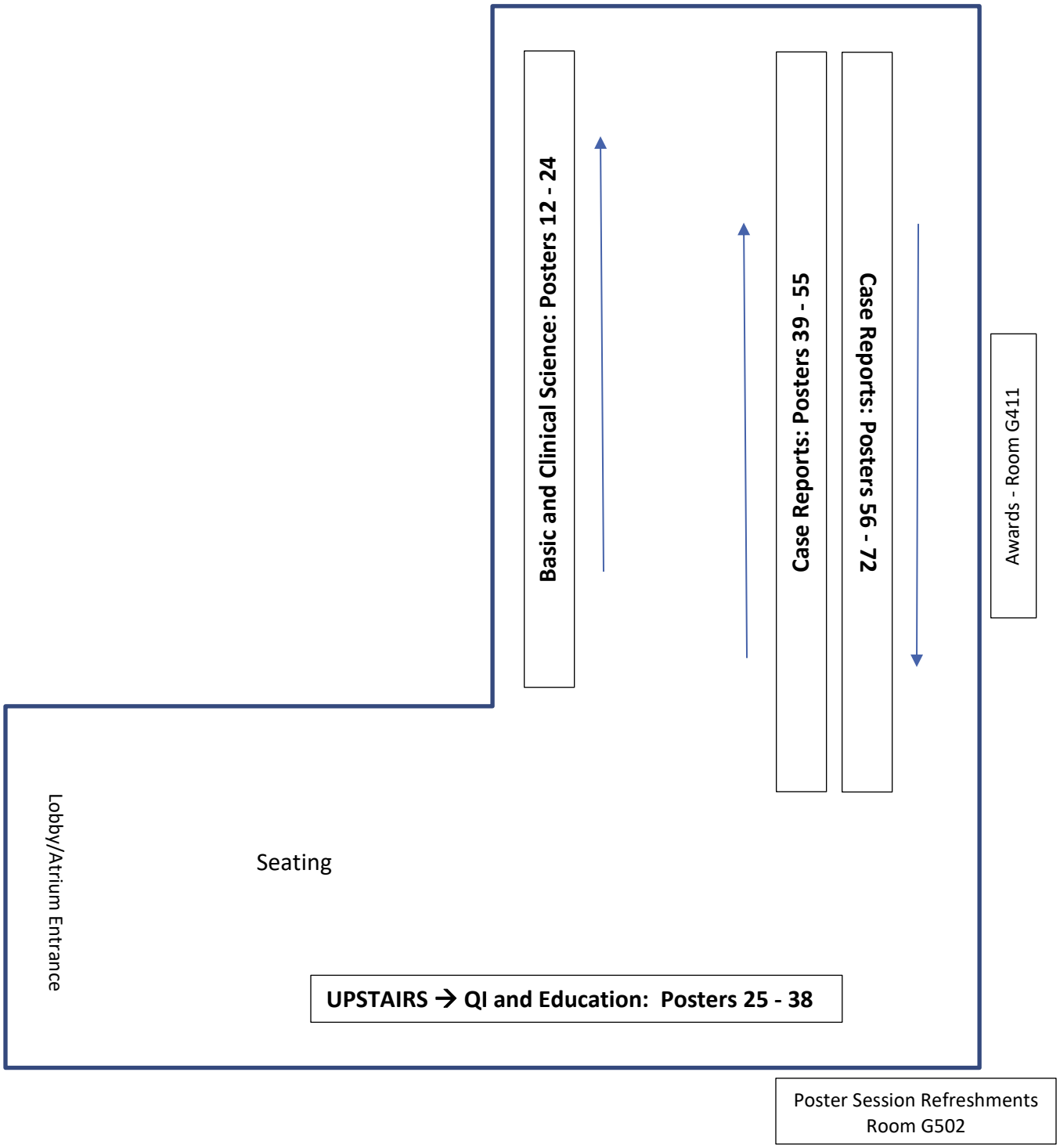
Let Your Passion Define Your Path: Choosing the Road Less Traveled
Curnen Denny Conference Room
Lunch Provided

4:00pm **Poster Session**
Dental School Lobby/Atrium Level
Refreshments Provided in Room G502 (lobby level; near posters)

5:00pm **Award Ceremony**
Dental School - Room G411 (lobby level; near posters)

Lobby/Atrium Level - UNC School of Dentistry

***Booklet page numbers correspond with poster numbers.**



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House dust promotes sensitization to environmental peanut through the airways

Peter A. Balogh, Johanna M. Smeekens, Robert M. Immormino, Michael D. Kulis, Timothy P. Moran

Peanut allergy affects 2% of the population and is the leading cause of life-threatening anaphylaxis. There is growing evidence that environmental peanut exposure through non-oral routes, such as the skin and respiratory tract, results in allergic sensitization to peanut. Peanut allergen is detectable in house dust, and there is a direct correlation between environmental peanut exposure in the home and risk of peanut allergy in children. In addition to peanut allergen, house dust contains a mixture of environmental agents with immunostimulatory properties, such as endotoxin, fungal-derived polysaccharides and proteases. By stimulating innate immune responses, these environmental agents can act as adjuvants in the respiratory tract and promote allergic sensitization to inhaled antigens. Whether exposure to these indoor environmental adjuvants contributes to peanut sensitization is unknown.

We have developed a mouse model that mimics natural inhalational exposure to environmental peanut. We have found that inhalational exposure to environmentally-relevant levels of peanut in combination with house dust extract (HDE), but neither alone, induces peanut-specific IgE and IgG1. Consistent with the development of peanut allergy, sensitized mice experienced anaphylaxis upon peanut challenge. Inhaled HDE stimulated production of innate cytokines (IL-33, IL-1α/β) in murine lungs and enhanced the maturation and migration of peanut-laden lung conventional dendritic cells to draining lymph nodes. Furthermore, inhaled HDE induced the differentiation of peanut-specific T helper 2 and T follicular helper cells, which are critical for directing allergen-specific IgE production. Finally, HDE triggered IL-1α/β production by primary human bronchial epithelial cells, suggesting that environmental adjuvants in house dust can stimulate innate responses in the human respiratory tract.

Collectively, these findings suggest that by promoting sensitization to environmental peanut, adjuvants in house dust contribute to peanut allergy development. Identifying the key components of house dust that promote peanut sensitization may lead to novel environmental interventions aimed at reducing peanut allergy development in at-risk children.

Gut Microbiome Composition Prior to Sensitization Predicts Reaction Severity in Peanut Allergic Mice

Andrew Hinton, Johanna Smeekens, Kelly Orgel, Darla R. Miller, Jeff Roach, M. Andrea Azcarate-Peril, Peter J. Mucha, Fernando Pardo-Manuel de Villena, A. Wesley Burks, Mike Kulis, Martin T. Ferris.

Rationale

Aberrant colonization of the gut by commensal microbes in early-life is associated with increased susceptibly to allergic diseases. Here we evaluated the influence of the gut microbiome on anaphylaxis to peanut in mice. Specifically, we retrospectively examined the relationship between the pre-sensitization composition of the gut microbiome and post-sensitization reactivity when challenged with peanut.

Methods

CC027/GeniUnc X C57BL/6J F1 and F2 mice were sensitized with peanut and cholera toxin for 4 weeks and challenged with peanut the following week. Fecal pellets were collected before and after sensitization and sequenced using 16S rRNA. Mice were characterized as strongly reactive (n=22) and non-reactive (n=23). Phylogenetic investigation of communities by reconstruction of unobserved states (PICRUSt) was used to predict each sample's gut metagenome from its 16S data.

Results

Using supervised learning, we identified 5 clusters of metagenomic ratios important for predicting non-reactive versus strongly reactive mice (AUC = 0.85). Cox regression identified a significant association between the log-ratio of glutamyl-tRNA reductases (K02492) to dipeptide transport system permease proteins (K12369) and reaction severity hazard. Strongly reactive mice with a log-ratio of K02492 to K12369 in the top 50th percentile had a mean predicted reaction time of 13.90(11.14–16.66) minutes compared to mice in the bottom 50th percentile with mean predicted reaction time of 8.67(6.47–10.87) minutes. Further, a log-ratio of K02492 to K12369 in the top 50th percentile versus bottom 50th percentile (HR=0.34, p=0.025) is associated with a reduction in reaction severity hazard.

Conclusion

Metagenomic log-ratios derived from gut microbiome data may be important for predicting anaphylaxis.

Abnormal neonatal ultrasounds are associated with neurodevelopmental burden at school age among children born extremely preterm

Campbell H, Check J, Mokrova I, Kuban K, Joseph RM, Allred E, O'Shea TM

Background: Impaired neurodevelopment (ND) remains one of the most concerning potential morbidities faced by survivors of extremely preterm (EP) birth. Neuroimaging via cranial sonography is often used to identify neonates who may be at increased risk of ND burden and who may benefit from increased surveillance and early intervention and therapies. The association between neonatal cranial ultrasound (CUS) abnormalities and long-term outcomes of EP survivors assessed at school age has not been well described.

Objective: To examine the association between neonatal CUS abnormalities and adverse ND outcomes of children born EP assessed at 10 years of age.

Design/Methods: We collected data prospectively from 1506 children born between 2002 and 2004 before the 28th week of gestation; 899 of 966 eligible children from this cohort returned for follow up at 10 years of age and completed ND assessments. Neonatal CUS abnormalities were grouped into the following categories: (0) no intraventricular hemorrhage (IVH) or white matter lesions (WML); (1) IVH only; (2) WML only; and (3) presence of both IVH and WML. Based on assessments performed at 10 years of age, 3 groups of children with ND burden were defined: (1) children without ND impairment; (2) children with normal cognitive ability (IQ =>70) but with one of the following morbidities: cerebral palsy (CP), autism spectrum disorder (ASD), and/or epilepsy; (3) children with cognitive impairment (IQ < 70) both with and without other ND comorbidities.

Results: Children with IVH alone had the same likelihood of having ND burden at school age as children with no CUS abnormalities (OR = 1.3, 95% CI 0.6-2.9 for CP/ASD/epilepsy; OR = 1.3, 95% CI 0.7-2.2 for cognitive impairment). Children with WML were more likely to have CP/ASD/epilepsy (OR 4.7, 95% CI 2.6-8.6) and more likely to have cognitive impairment (OR = 2.3, 95% CI 1.3-3.8) compared to children without CUS abnormalities. Similarly, children with both IVH and WML were at increased risk for CP/ASD/epilepsy (OR = 3.3, 95% CI 1.8-6.3) and cognitive impairment (OR 3.1 95% CI 1.9-5.1) compared to children with normal neonatal head ultrasounds.

Conclusion: EP survivors with evidence of white matter injury on neonatal CUS have an increased risk of experiencing ND burden at school age compared to EP children with a normal neonatal CUS. The presence of IVH alone or in addition to WML does not appear to confer an increased risk of ND burden.

Dosing and Safety of Peanut Food Equivalents After Immunotherapy Trials

Quindelyn Cook, MD[1], Luanna Yang, MD[1], Ahmad Hamad, MD[1], Holly Barber BSN[2], Lauren Herlihy, CPNP[1], A. Wesley Burks, MD[1], and Edwin Kim, MD, MS[1]. [1]Department of Pediatrics, Division of Allergy, Immunology, and Rheumatology, University of North Carolina Chapel Hill, Chapel Hill, NC. [2]School of Nursing, University of North Carolina Chapel Hill, Chapel Hill, NC.

RATIONALE:

Regular peanut consumption after immunotherapy may provide continued protection against accidental exposures, however limited data exists regarding its long-term safety.

METHODS:

Past participants in peanut immunotherapy trials were enrolled in a longitudinal observational study. Those desensitized to ≥300mg of peanut were instructed to incorporate dietary peanut. We reviewed peanut food equivalent dosing and associated reactions in 55 subjects who completed oral (OIT) or sublingual immunotherapy (SLIT) studies between 2010-2017.

RESULTS:

The majority of subjects were male (55%), Caucasian (94%), and participated in an OIT trial (73%). Peanut consumption was continued in 49/55 (89%) subjects. Adverse reactions, including EoE in one subject, and taste aversion were causes for discontinuation. Median peanut consumed was 600 mg (mean 808 mg, 100-4800 mg). Thirty-one subjects (74%) consumed peanut daily. Lower peanut consumption correlated with older age (r = -0.17). Ten (23.8%) subjects reported reactions with urticaria, gastrointestinal symptoms, and oropharyngeal pruritus, the most common. The majority of reactions were treated with antihistamines; however 1 reaction required epinephrine and 2 required EMS. There was no correlation between peanut dose and reactions. Participants in SLIT trials consumed less peanut (median 500 mg, mean 543 mg) compared to OIT participants (median 600 mg, mean 955 mg); however more reactions with dosing were reported with OIT (7/10).

CONCLUSIONS:

The majority of subjects continued dietary peanut up to 8 years after study completion. Food equivalents may be a safe option for maintaining desensitization. Further study is needed to understand the impact of age and types of immunotherapy on peanut consumption and reactions.

Too many checks? Evaluating the optimal number of glucose measurements in a neonatal asymptomatic hypoglycemia screening protocol

Christopher Cummings, Victor Ritter, Sherry Leblanc, Jason Fine, and Ashley Sutton

Introduction: Neonatal hypoglycemia (NH) is a common problem. Approaches to screening vary. At our institution, asymptomatic infants with a known risk factor (RF), including late preterm gestation, large or small for gestation age, or infant of a diabetic mother undergo screening. A minimum of 3 consecutive acceptable pre-prandial blood glucose measurements (BGM) are required to pass. Many infants do not experience NH, yet undergo a minimum of 3 BGM. Few infants are identified who ultimately require IV dextrose (IVD) administration, suggesting opportunity to better tailor screening protocols to risk of NH.

Objective: To determine which RFs are predictive of asymptomatic NH; secondarily, to determine if infants requiring IVD can be identified in less than 3 BGM.

Methods: We analyzed a retrospective cohort of infants with $\geq$ 1 RF for NH between 5/2017-4/2018. Infants were excluded if BGM was for reason other than screening or if given intravenous dextrose (IVD) for reason other than NH. Data included demographics, delivery mode, gestational age, RF, birth weight, and use of IVD. Association between categorical and continuous variables and first and second BG were determined using Pearson's Chi-square test and one-way ANOVA models, and two-sample t-tests and simple linear regression models, respectively. The cohort was also assessed for total number of at-risk infants with NH detected on first, second, or third BGM.

Results: 1037 infants underwent chart review; 830 infants met inclusion criteria, of which most (n=510, 61%) did not develop NH. Of infants with NH, only 3.7% (n=31) required IVD. Nearly all (n=30, 97%) had NH on initial BGM, and the remaining infant had NH on second BGM. In analyzing RFs for protocol initiation, no single variable showed strong prediction capabilities of NH, although multiple variables demonstrated an association. Delivery mode was identified as a novel potential RF.

Conclusion: Despite screening only 'at-risk' infants for asymptomatic NH, many did not develop NH. Of those that did, few required care beyond feeding. All at-risk infants who required IVD developed NH within 2 BGM. Accepted RF were not strongly predictive of NH, suggesting that further refinement of RF to initiate screening could reduce unnecessary BGM. Together, these findings suggest multiple avenues of optimization to reduce BGM in the newborn nursery.

Pediatrics Patients' Preferences About Sexual Orientation and Gender Identity Questions during Routine Pediatric Clinic Visits

Amalia Lee, MPH; Sue Tolleson-Rinehart, PhD; Emily Vander-Schaaf, MD MPH; Martha Perry, MD

Background: LGBTQ youth are at increased risk for substance use, suicide, homelessness, HIV and STDs, and psychological distress. Consistently asking youth about sexual orientation and gender identity (SOGI) in the clinical setting permits providers to screen for, prevent, and mitigate conditions that disproportionately affect LGBTQ youth. There are many recommendations identifying the importance of asking SOGI questions. At present, these guidelines are based on best practice while identifying the ideal timing and manner in which youth are asked these questions has not been well studied.

Methods: We surveyed a convenience sample of English-speaking adolescents at a general pediatrics resident clinic, an academic adolescent medicine clinic, and a pediatric endocrinology gender clinic. The survey asked respondents at what age and to whom they would be most comfortable disclosing SOGI status (physician, nurse, receptionist, or other), and in what manner they prefer these questions to be asked (multiple choice, sexuality and gender scales, or another manner). Additionally, the survey asked respondents to rank their preferred method of delivery for SOGI question (on paper, electronically, or spoken, with or without a caregiver present). This study was approved by the University of North Carolina at Chapel Hill's Institutional Review Board.

Results: Twenty-two adolescents completed the survey, thus far. Thirty-two percent identified as transgender or gender diverse and 35% identified as sexual minorities. Respondents identified 13-16 years of age as the time frame they wanted to be asked about their gender identity (71%) and sexual orientation (67%). They preferred that their physician ask about SOGI status (88%). Participants preferred to answer questions "on paper without parent present" (78%) followed by "electronically without parent present" (75%). Participants preferred multiple choice compared to a continuous scale method for both gender identity (71%) and sexual orientation (59%) questions. There were no statistically significantly differences in preferences related to SOGI questions based on participants' reported SOGI status.

Conclusions: Adolescents in our small pilot study were willing to answer questions about preferred method of collecting SOGI data. Although we have promising preliminary data about pediatric patients' preferences for SOGI questions, these results emphasize the need for future larger scale studies in order to examine whether these preferences persists in a broader population or differs by LGBTQ status.

An Apple iPad a Day Brings the Doctor to you: Virtual Family Centered Communication in a Pediatric Intensive Care Unit – Phase 1

Shannon Solt DO, Danielle Stolfi RN, Meg Kihlstrom MD

Introduction: In 2003, the American Academy of Pediatrics stressed in multiple policy statements that family centered rounds should be the standard of pediatric health care. Family centered rounds works wonderfully for families who are able to be present at the bedside of their child. However, it is difficult for many families to be present Monday through Friday in the morning due to a number of different factors. We hypothesize that for these families, a virtual family centered rounds approach would improve communication between the healthcare team and the family in a busy pediatric intensive care unit. The initial phase of our study was to determine length of time spent on rounding with and without families at the bedside so that this could be compared to the length of time spent rounding virtually with families not present at the bedside.

Methods: This observational study of a convenience sample was conducted over a 10-day period (July-August 2018). The individual patient rounding times for 174 patient encounters were recorded. The presence of family members, subspecialists, intubation status, patient census, patient length of stay and whether the family was English speaking or not was recorded.

Results: Of 174 patient encounters, family was present on rounds for 58 encounters (33%). Family was at the bedside but did not participate on rounds for 47 encounters (27%) and family was not present for 69 patient encounters. (40%). Rounds took on average 9 minutes and 40 seconds (range of 50 seconds to 27 min and 51 seconds) when families were not present on rounds, compared to an average of 10 minutes and 49 seconds (range of 4 minutes and 36 seconds – 20 minutes and 6 seconds) when family was present on rounds.

Discussion: On average, rounds lasted approximately one minute longer with family presence. However, there is significant variability in rounding times without family presence, and interestingly the longest rounding encounter was without family members present. The next phase of this study is underway and will incorporate virtual family centered rounds into the daily rounding process. Further analysis will focus on family and provider satisfaction with this rounding option as well as further documentation of rounding times to assess the effect of virtual family rounds.

Contamination rates using sheathed versus unsheathed syringe plungers

Hillary B Spangler[1], M Concetta Lupa[2], Monika Chatrath[2], Melissa L Veneracion[2], Kevin Alby[3], Robert D Valley2

Internal Medicine-Pediatrics,[1] University of North Carolina Departments of Anesthesiology,[2] Chapel Hill, USA; University of Pennsylvania, Department of Pathology and Laboratory Medicine, Perelman School of Medicine, Philadelphia, USA[3]

Background:

Nosocomial infections contribute significantly to morbidity and increase health care costs for both pediatric and adult patients in the United States.1 Intravenous access remains a main source of nosocomial infections from environmental and (non-intentional) aseptic technique violations by healthcare providers via contact with stopcocks, IV tubing, and syringes.2,3 Stopcock contamination and repeated syringe strokes have been associated with increased risk of nosocomial infection, which is of particular concern in immunocompromised hosts.2,4-7 Syringe reuse frequently occurs by anesthesia and critical care providers with "push-pull" technique. We hypothesize that standard syringes can introduce risk for bacterial contamination with this technique and the use of modified sheathed syringes can decrease this contamination risk.

Methods:

20 modified sheathed syringes (Figure 1) were sterilized and 20 standard 10 mL syringe packs were opened under a filtered-air hood with sterile gloves. Sterile gloves were "contaminated" by introducing Staphylococcus aureus (S. aureus) via a dropper and used to handle the syringes using an otherwise aseptic technique. Each syringe was filled and emptied five times from a test tube containing a solution of sterile 1% peptone culture medium, incubated for 8 hours, and then plated. The Fisher Exact test was used to analyze results.

Results:

Six out of 20 culture mediums from the standard syringes were positive for S. aureus while none of the culture mediums from sheathed syringes were positive, $p = 0.02$ ($p<0.05$).

Conclusion:

Our results support the hypothesis that standard syringes can be a source of bacterial contamination and the use of sheathed syringes can reduce this contamination. Closed syringe sheaths provide improved aseptic technique adherence and minimize contamination, particularly during clinical scenarios requiring syringe reuse.6,8 This is directly applicable to the use of syringes by anesthesia providers during the intraoperative period as well as pediatric critical care providers using a "push-pull" technique. The use of sheathed syringes should be considered in a setting where the provider might use a syringe multiple times, improving aseptic technique adherence while minimizing resource use and infection risk.

Comparison of 2 and 10-Year Neurodevelopmental Outcomes of 23-25 Week Gestational Age Infants

G Taylor MD, R Joseph PhD, L Douglas MD, J Laux PhD, W Price MD, TM O'Shea MD MPH

Background:

Two-year neurodevelopmental outcomes of extremely preterm infants are used in prenatal counseling, clinical trials, and early intervention. There have been concerns about the stability of these early childhood outcomes compared to later neurodevelopmental outcomes.

Objective:

To compare 2 and 10-year neurodevelopmental outcomes for infants born 23-25 weeks gestational age.

Methods:

We classified NDI at 2 years as defined by the Neonatal Research Network Estimator into moderate-to-severe (MDI 50-70, PDI 50-70, GMFCS 3-4, bilateral legal blindness, or bilateral hearing loss requiring amplification) and profound (MDI<50, PDI<50, or GMFCS 5). We classified NDI at 10 years using criteria developed by an expert panel in a cohort born at 23 to 25 weeks gestation (the ELGAN cohort) in moderate (IQ 55-70, GMFCS 3, bilateral hearing loss requiring amplification, bilateral legal blindness, autism level 2, and epilepsy), severe (IQ 35-54, GMFCS 4, or autism level 3), and profound (IQ<35, GMFCS 5, or autism level 3 combined with IQ 35-54). We compared the accuracy of NRN Estimator predictions of outcomes at 2 and 10 years and evaluated NDI at 2 years as a predictor of NDI at 10 years.

Results:

Of 679 infants, 70% survived. Data sufficient to classify severity of NDI at 2 and 10 years were available for 65% of survivors (n=315). The positive predictive value (PPV) for moderate-to-severe NDI at 2 years was 40%; the PPV for profound NDI at 2 years was 39%.

Conclusion:

The majority of children improved on 10-year neurodevelopmental assessments compared to 2-year assessments.

Empiric MRSA Coverage: Opportunities for Improvement

Peyton Thompson,[1] William Wilson,[2] and Zachary Willis[1]

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Background and Aims:

Empiric coverage for methicillin resistant Staphylococcus aureus (MRSA) is critical in certain clinical situations such as sepsis. Targeting of antimicrobial therapy based on clinical course and microbiologic cultures is equally important in the promotion of antimicrobial stewardship. The purpose of this retrospective chart review was to determine the frequency, indications, duration, and toxicity of empiric MRSA coverage at North Carolina Children's Hospital (NCCH).

Methods:

This was a retrospective cohort study of patients at NCCH who received antibiotics targeting MRSA over a four-month time period (October 1st, 2018 to January 31st, 2019). We documented: demographics, inpatient service, indication, underlying medical conditions, code sepsis order set use, length of therapy, culture results, Pediatric infectious disease (ID) consultation, medication allergies, and toxicities.

Results:

In total, we reviewed 150 patient charts, including nine (6.0%) who received ceftaroline, four (2.7%) who received daptomycin, 29 (19.3%) who received linezolid and 108 (72.0%) who received vancomycin. Six of nine ceftaroline recipients were treated on the pulmonology service (5 for CF bronchopneumonia and 1 for chronic suppurative bronchitis); the remainder were treated for bone/joint infections. All daptomycin recipients had bloodstream infections. Indications for treatment were heterogeneous for linezolid and vancomycin. Of linezolid recipients, 19 (65.5%) received therapy for >48 hours, 13 (44.8%) were culture-confirmed, and Pediatric ID was consulted on 14 (48.3%). No documented cases of neutropenia occurred among the linezolid cohort. Among vancomycin recipients, 13 (12.0%) had culture-confirmed infections requiring vancomycin, five of which were documented MRSA infections. Seven patients (6.5%) received vancomycin for contaminated blood cultures. Seven patients (6.5%) received vancomycin for code sepsis. Fifty-five patients (50.9%) received vancomycin for >48 hours. Pediatric ID was consulted on 55 vancomycin recipients (50.9%). Of 74 vancomycin recipients with troughs measured, supratherapeutic troughs were observed in 11 patients (10.2%), two of whom developed acute kidney injury (AKI). Subtherapeutic initial troughs occurred in 43 patients (58.1%).

Conclusion:

Overall, 150 patients received empiric MRSA coverage over a four-month time period at NCCH. As expected, vancomycin was most commonly used, followed by linezolid, ceftaroline, and daptomycin. Vancomycin and linezolid were generally used empirically while ceftaroline and daptomycin use represented directed therapy. Fewer than half (44.8%) of patients who received linezolid and only 12.0% of patients who received vancomycin had culture-confirmed indications, and over half of these patients received therapy for greater than 48 hours. Avoiding prolonged anti-MRSA courses for patients with negative cultures presents an important improvement opportunity.

The Arresting Vertical Transmission of Hepatitis B Virus (AVERT-HBV) Study in the Democratic Republic of the Congo: Preliminary Results

Peyton Thompson,[1] Jonathan B. Parr,[2] Kashamuka Mwandagalirwa,[3] Noro Lantoniaina Rosa Ravelomanana,[3] Martine Tabala,[3] Malongo Fathy,[3] Patrick Ngimbi,[3] Bienvenu Kawende,[3] Charles Mbendi,[3] Jérémie Muwonga,[4] Ravi Jhaveri,[5] Gavin Cloherty,[6] Marcel Yotebieng,[7] and Steven R. Meshnick[8]

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Background and Aims:

Despite the widespread availability of effective vaccination against hepatitis B virus (HBV), it remains endemic throughout sub-Saharan Africa. Mother-to-child transmission (MTCT) of HBV is preventable through birth dose vaccination and identification and treatment of "high-risk" pregnant women (those with a high viral load and/or HBV e antigen [HBeAg] positivity). The purpose of this study was to show the feasibility of adding HBV testing and treatment of pregnant women, as well as birth dose vaccination of exposed infants, to the existing infrastructure of an HIV prevention of MTCT (PMTCT) program in the Democratic Republic of the Congo (DRC).

Methods:

We enrolled pregnant women at two maternity centers in Kinshasa, DRC that have ongoing HIV PMTCT programs. We screened women for HBV using point-of-care HBV surface antigen (HBsAg) testing (Abbott Alere Determine™ HBsAg, Abbott Park, IL). Women with positive HBsAg testing who presented at <24 weeks' gestation were eligible for enrollment. Enrolled women were evaluated for high-risk disease using HBV viral load (Abbott RealTime, Abbott Park, IL) and HBeAg (Abbott ARCHITECT, Abbott Park, IL), and were tested for baseline liver function and kidney function abnormalities. Pregnant women with high-risk disease – viral load >200,000 IU/mL and/or HBeAg positivity – were initiated on tenofovir antiviral therapy between 28- and 32-weeks' gestation, which was continued through 12 weeks' post-partum. Exposed infants were given a birth dose of monovalent HBV vaccine within 24 hours of life.

Results:

Of 4,016 women screened, 109 (2.7%) were HBsAg-positive and 91 women were enrolled in the study. Nine (9.9%) of the enrolled women were found to have high-risk disease and were started on (or will soon receive) tenofovir therapy. None of the high-risk women had baseline liver or kidney function abnormalities. Of the 20 infants who have been born to date, 14 (70.0%) received HBV birth dose vaccine within 24 hours of life.

Conclusion:

This study demonstrated the feasibility of adding HBV screening and treatment of pregnant women, as well as infant birth dose vaccination, to the existing HIV PMTCT platform in Kinshasa, DRC. Fewer women had high-risk disease than expected (9.9% observed, 30% expected), which is consistent with data from other studies of MTCT of HBV in Africa. Challenges exist in administering timely birth dose vaccination to Congolese infants; these implementation issues must be addressed prior to roll-out of universal birth dose vaccination in Africa.

Advanced Care Planning in Cystic Fibrosis

Todd, Sarah H, Sonntag, Elizabeth A, Howe, Katie L, Buchanan, Marianne L, Jones, Brooke W, Goralski, Jennifer L

RATIONALE

Advanced care planning (ACP) is recommended for all patients with cystic fibrosis (CF). No clear guidelines exist and practice varies widely among institutions. Retrospective studies demonstrate that many patients with CF have not engaged in ACP discussions at all, while others wait until the final weeks of their life, when decisions are made during times of high stress. This study was designed to elicit the CF patients' perceptions of end-of-life care and advanced care planning and to investigate how that perception changes in response to formalized ACP meetings.

METHODS

A multidisciplinary group at the University of North Carolina Adult CF Center completed a quality improvement project focusing on the implementation of outpatient ACP meetings for patients with CF. Any CF Center patient was eligible. Interested patients had one-hour, billable ACP meeting with CF provider, LCSW, Dietician, Nurse Coordinator, and designated support people. A note documenting wishes and outcomes of the meeting was placed in the EMR. A survey was given before and after the meeting via RedCap to assess attitudes toward end of life (EOL) care and ACP.

RESULTS

Of 53 patients approached, 22 have elected to complete surveys. Of these 22 study participants, 15 have an FEV1 ≤40%, 12 are between 20 and 29 years old. Seventeen pre-surveys and thirteen post-surveys have been completed so far.

Prior to their meeting over 90% of patients felt that patients with cystic fibrosis have unique needs when it comes to ACP with 88% of patients wanting the opportunity to talk more about their wishes at end of life with their loved ones. Prior to the meeting only 18% had engaged in these discussions. Only 56% of patients felt comfortable discussing EOL care prior to the meeting with an increase to 69% in post-meeting surveys.

Only 29% of patients reported having an advanced directive prior to their meeting. After the meeting 77% of patients stated they had completed or were in the process of completing advanced directives.

CONCLUSIONS

In a multidisciplinary care model at a large academic CF Center, we successfully implemented sustainable ACP for CF patients and continue to evaluate the effect of the program. Results shows an increase in patient comfort with and documentation of end-of-life goals after ACP meetings. It is important for healthcare providers to meet the needs of this special patient population by ensuring that ACP is in place for patients prior to crisis situations.

Back to Life, Back to Reality – What Happens after Peanut Immunotherapy?
A Long-Term Follow up Study on Perceptions of Safety and Lifestyle

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Abstract # 41691

INTRODUCTION

- Food allergy is a growing health problem worldwide that impacts millions of individuals.
- Treatment options are limited and strict dietary avoidance remains the standard of care.
- Prevention and treatment of allergic reactions to food(s) places a significant financial and emotional burden on patients and their families.
- Immunotherapy has shown promise in increasing tolerance in peanut allergic individuals; however, little is known about long-term effects of therapy on perceptions of safety and lifestyle behaviors.

OBJECTIVE

- To examine the long-term effects of peanut sublingual immunotherapy (SLIT) or oral immunotherapy (OIT) on patient and parent perceptions of safety, quality of life, and lifestyle.
- Analyze any differences in perceptions of safety and changes in lifestyle in SLIT vs OIT subjects.

METHODS

- Past participants in either peanut OIT or SLIT studies at the UNC Food Allergy Study Center were enrolled in a longitudinal observational study.
- Individuals demonstrating tolerance to ≥ 300mg of peanut were instructed to incorporate daily peanut ingestion at home after study completion.
- A Qualtrics telephone survey was administered to fifty-five families up to 8 years after study completion.

RESULTS

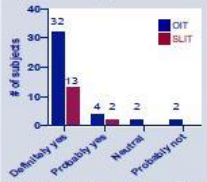
Baseline Subject Characteristics

Table 1: Subject Characteristics	
Total number of participants	55
Caucasian (%)	52 (94%)
Male (%)	30 (55%)
Patients who completed OIT (%)	40 (73%)
Patients who completed SLIT (%)	15 (27%)
Total number of subjects who continued home peanut ingestion	49 (89%)
OIT subjects who continued home peanut ingestion (%)	33 (83%)
SLIT subjects who continued home peanut ingestion (%)	15 (100%)
Median time since study completion	2.9 years (range 1.2 mo – 7.5 years)

Q1: Do you feel your child is safer on peanut treatment?

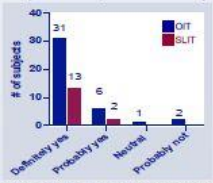
- 100% of OIT and 93% of SLIT parents "definitely" or "probably" felt their child was safer after therapy

Q2: Is your child's QOL better after peanut treatment?



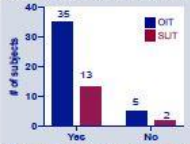
- 90% of OIT and 100% of SLIT parents "definitely" or "probably" felt their child's QOL improved after therapy

Q3: Is parental QOL better after peanut treatment?



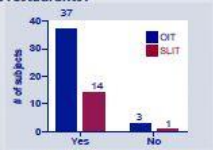
- 93% of OIT and 100% of SLIT parents "definitely" or "probably" felt their own QOL improved after therapy

Q4: Do you consume foods labeled "may contain peanut" after being on treatment?



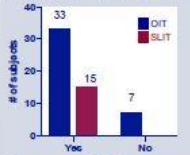
- 88% of OIT and 87% of SLIT subjects consumed foods labeled "may contain peanut" following peanut therapy

Q5: Yes or No - we feel more comfortable eating out at restaurants?



- 93% of OIT and 93% of SLIT subjects were more comfortable eating out following peanut immunotherapy

Q6: Yes or No - we feel more comfortable with our child in unsupervised social settings?



- 83% of OIT and 100% of SLIT parents felt more comfortable with their child in unsupervised social settings following peanut therapy

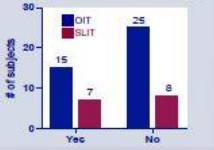
Q7: Does your child have an epinephrine autoinjector with him/her or carried by an accompanying adult?

- Only 83% of OIT and 93% of SLIT subjects carried an epinephrine autoinjector "often"

Q8: Does your child have any other food allergies?

- 63% of OIT and 73% of SLIT subjects reported allergies to at least 1 other food

Q9: Does your child see an allergist at least yearly?



- Only 38% of OIT and 47% of SLIT subjects reported seeing an allergist at least once a year.

RESULTS SUMMARY

- 89% of subjects continued to tolerate daily peanut ingestion at time of survey.
- 98% of all parents "definitely" or "probably" felt their child was safer after peanut therapy.
- 93% and 95% of all subjects "definitely" or "probably" felt their child's quality of life AND parental quality of life improved following immunotherapy, respectively.
- A majority of all subjects reported improvements in QOL parameters.
- 50% of subjects who had stopped home peanut still had "definite" or "probable" improvement in all survey parameters.
- Parents and subjects reported positive effects on safety and lifestyle even in the presence of other food allergies.
- Only 85.5% of subjects carried autoinjectors "often" and only 60% saw an allergist annually.

DISCUSSION & CONCLUSIONS

- Regular ingestion of peanut after completion of food immunotherapy is associated with improved perceptions of safety and life satisfaction in the majority of both SLIT and OIT patients and their parents up to 8 years after study completion.
- Benefit was seen even in a notable portion of patients who reported other food allergies or who had discontinued home peanut ingestion due to intolerance or side effects.
- Increased risk-taking behavior (ie: not carrying an epinephrine autoinjector or seeing an allergist regularly) may be an unintended consequence of immunotherapy.

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Healthy Lifestyle Needs Assessment: Foundations of Advocacy

Bianca Allison MD MPH, LeeAnne Flygt MD, Blaire Hanvey MD, Alex Turek MD, Anna Liles

University of North Carolina, Chapel Hill

Introduction:

We created a two-pronged (quality improvement and advocacy) project to help improve the care of pediatric patients with obesity at our resident continuity clinic, UNC Children's Primary Care. The advocacy arm has two components: a needs assessment and a series of advocacy projects informed by the needs assessment. The needs assessment aims to evaluate parental knowledge of obesity and identify barriers to healthy living for families with children who are overweight or obese. Understanding parents' perceptions, beliefs, and attitudes is important for childhood obesity prevention and intervention.

Methods:

The study population included parents or legal guardians of any patient >24 months old whose BMI was greater than the 85th percentile (overweight or obese by pediatric standards). After a literature review, an anonymous survey was created which contained questions to assess knowledge, perceptions, behaviors, and barriers related to healthy lifestyles. The survey was translated into Spanish by the UNC Interpreter Services. The survey was distributed to the study population during well visits or follow-up weight visits from 8/2018 to 1/2019. Responses were entered into Excel, and results were analyzed using frequency distribution charts.

Results:

Sixty families completed the survey. The majority of families did not think their doctor had ever told them their child was overweight/obese. The majority of parents who answered the survey thought their child was the right weight. Parents identified not knowing enough about nutrition as their #1 barrier (lack of healthy snacks outside the home was #2). Parents thought doctors should provide them with more information about how much and what to eat. Parents have tried to cut down on fast foods or high fat/sugar snacks (#1 and #2) as ways to help with their child's weight.

Conclusions:

It is important for residents to explicitly identify children in our clinic who are overweight and obese and communicate this information to parents. Current literature shows that parents are unlikely to modify behaviors or seek interventions for weight management when they do not perceive a problem with their child's weight. Moreover, residents should provide additional guidance to families on healthy lifestyle modifications, especially around healthy nutrition. These results will be formally presented to residents at UNC Children's Primary Care in 5/2019 to provide guidance on issues affecting our population.

Quality Improvement (QI) for Universal Screening of Gonorrhea and Chlamydia (GC/CT) in a Resident Primary Care Clinic

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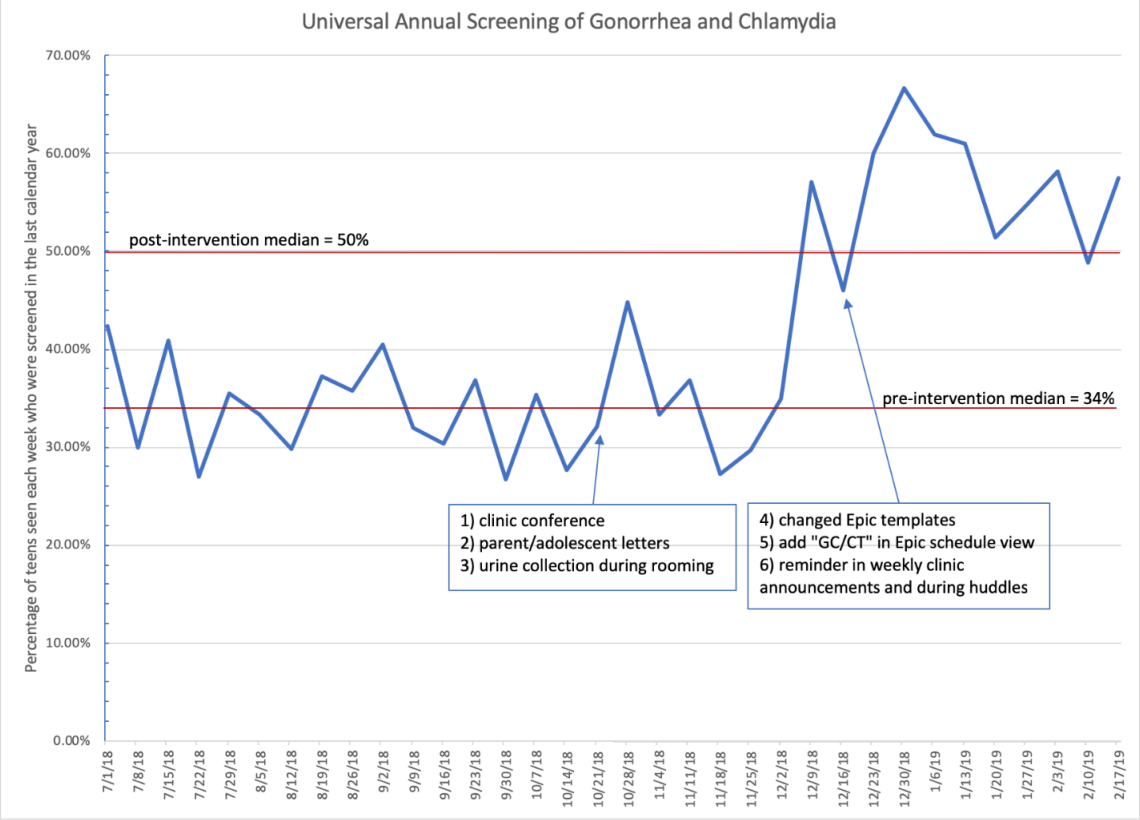
Introduction: The prevalence of both gonorrhea and chlamydia in North Carolina for 15- to 19-year-olds was higher than the national average, and 15- to 29-year-olds comprised nearly 71% of gonorrhea and 85% of chlamydia cases in North Carolina in 2017. The American Academy of Pediatrics recommends annual screening for GC/CT in sexually active females <=25 years old; however, data suggest that adolescents may not consistently report sexual activity to their health care providers, and current billing practices for private insurance hinder confidentiality for adolescents as GC/CT testing appears on EOBs. This QI intervention implements universal annual (once per year, regardless of visit type) GC/CT screening of all male and female adolescents (>/= 13 years) in the UNC Children's Primary Care Clinic. The aim of the project was to increase GC/CT annual screening to 100%, thereby increasing detection and treatment of GC/CT, reducing stigma associated with screening, and mitigating concerns regarding confidentiality.

Methods: This study was exempted by UNC IRB. The project was introduced to residents and preceptors via a weekly clinic newsletter and during a conference about GC/CT screening and treatment (10/21/18). The clinic staff assisted with re-designing clinic workflow to include "introduction to the adolescent visit" letters to adolescents and parents at clinic check-in, and residents placing a note in the Epic schedule view if adolescents required GC/CT testing which triggered clinic staff to collect urine during the rooming process. Additional changes made included Epic template revisions to include questions about last GC/CT screening and reminders during clinic huddles twice daily. Data were extracted from Epic to examine GC/CT screening frequency, and were examined pre-interventions (7/1/2018 - 10/21/2018) and post-interventions (10/22/2018 to present) using a run chart.

Results: See run chart copied below. The median annual screening percentage for the period prior to the interventions was 34%, and for the 5 months after the interventions was 50%.

Conclusions: Our intervention has improved GC/CT screening in the clinic, however more work is needed to reach the goal of 100%. Additional PDSA cycles will be conducted over time including improving identification of adolescents needing annual GC/CT screening, adding visual cues for staff, providers and patients regarding GC/CT screening, and providing more feedback to individual residents about their GC/CT screening practices.



Social Determinants of Health Screening and Intervention in Complex Care Pediatric Patients

Marie Anderson, Dr. Neal deJong, Heidie Tkach, Madlyn Morreale, Darragh Davis

We performed a quality improvement study to assess screening of social determinants of health and intervention among patients in the UNC Children's Complex Care Program. In this program, children with medical complexity are provided with resources and a team to address and manage health and social factors that can impact adherence to treatment, access to resources and health outcomes. We hypothesized that most patients in our program would have additional non-medical needs due to increased number of hospitalizations, expenditures, stress and family expenses compared to the average child. Patients were screened using telephone interviews and a screening tool created by the complex care team. Data was collected on the amount of screenings performed, number of positive screens and number of interventions made weekly. After four weeks of data collection, 53% of members in the program were screened and 59% of screenings were positive, meaning that families indicated a "yes" to one of ten questions on the UNC Complex Care Program Social, Resource, and Legal Problems Screening Form. We found that 48% of screenings indicated problems with learning and/or school, 34% of positive screens were notable for food insecurity, and 34% of families have an adult parent in need of health insurance. Additionally, intervention rates increased from 19% to 45% over the course of 4 weeks. Currently, we can conclude that most children with medical complexity in our program are faced with non-medical factors that may impact their health and health outcomes. More screening will be done in addition to continued intervention and follow-up. This highlights the importance of infrastructure building and design of complex care coordination teams for children with medical complexity.

Results are preliminary.

Time to Tip the Scales: An Obesity QI Project in the Primary Care Setting
LeeAnne Flygt, Bianca Allison, Alex Turek, Blaire Hanvey, Martha Perry, Colin Orr

Background:
Approximately 17% of 2- to 19-year-old children and adolescents are obese (defined as having an age- and gender-specific BMI at 95th percentile) according to a 2014 US Preventive Task Force (USPSTF) study and the number is only rising. Childhood obesity is one of the largest epidemics our nation is facing today. While the American Academy of Pediatrics and the USPSTF have issued guidelines for treatment of obese and overweight pediatric patients, many clinics do not have the resources, and many clinicians do not have the training to adequately address these needs.

Goals:
Our goal is to standardize the approach to overweight and obese patients in our resident clinic by increasing:

- Recognition of the problem through documentation of BMI status on patient problem lists
- Frequency of follow-up visits to the AAP recommended 1-3 months
- Residents' knowledge of screening labs recommended for overweight and obese patients.

Through these small, measurable changes we hope to have a positive impact in our patients lives and eventually see a downward trend in obesity in our clinic.

Methods:
We surveyed residents and preceptors in clinic about the management of overweight and obese patients. We used this data to create:

- A protocol to standardize visits
- A four-week conference series reviewing basic definitions, associated comorbidities, and motivational interviewing
- An EPIC template to help facilitate follow-up visits

Results:
While we are still receiving patient data following our interventions, baseline data shows that of our patient population aged 2-20, 36% are overweight or obese. Of these patients, 37% had their BMI status documented on their problem list and 12.4% had follow-up visits scheduled for weight management.
In looking at resident data, 91% of residents agreed or strongly agreed that the interventions were helpful in their management of overweight and obese patients.

Discussion:
Our resident clinic has more overweight and obese children than the general population which makes it even more important to have a standardized approach to managing these patients. While the protocol, note template and conferences seem to have helped resident comfort, there is still work to be done. Our next steps involve increasing preceptor buy-in, creating SMART sets, and increasing visibility of our goals in clinic.

Accuracy of the After Visit Summary Discharge Instructions from an Academic Children's Hospital

Meera Jairath BS, Kathy K. Bradford MD, Sheryl Galin, Heidi Troxler MSN RN CPN

Introduction

Transitions between sites of care occur in most all pediatric hospitalizations. Previous work has identified hospital to home transition record quality as a key quality measure to access transition for Hospitalized children. The printed After-visit summary (AVS) is typically given to families and patients at the time of hospital discharge as a communication tool to inform and reiterate relevant and actionable information and instructions regarding hospitalization dates, discharge diagnose/s, hospital course, updated medications list, PCP and subspecialty follow up, and emergency call back instructions and contact numbers.

Objective

The purpose of our study was to determine the completeness and accuracy of the after visit summary from an academic childrens hospital.

Methods

We prospectively evaluated the AVS of Children being discharged from an academic Children's hospital using nurse survey. Following entry of a patient discharge order into our EPIC EMR system, a single research assist was notified of the discharge and personally asked the bedside nurse 3 questions to understand the accuracy and clarity of the AVS to address medications and follow up appointment accuracy and clarity of discharge instructions. Nurses were surveyed after patients were physically discharged from the hospital and after they had fully reviewed the discharge paperwork with families, had time to identify discrepancies and clarify inconsistencies with the medical team. Survey question were; 1) Were there any problems with the medication section of the AVS?, 2) Was the follow up appointment scheduled or plan indicated? and 3) Were the discharge instruction clear to nursing and the patients? Nurses who were not able available for a face to face survey were called or emailed shortly after the discharge process was completed.

Results

Two hundred and six consecutive weekday discharge AVS were prospectively studied during two time periods: May 22, 2017 - 2017 and June 27-August 1 , 2017 for accuracy and clarity at the time of discharge for patients leaving an academic Children's hospital. Overall, 16% of AVS had an inaccurate medication/s instructions, follow up appointment/plan, or clarity of discharge instruction/s. The most common inaccuracy was medication related.

Discussion/Conclusion

Hospital discharge and transition of care is a complex and important process. AVS documentation often includes EMR templates and auto population. Our study found that inaccuracies identified at the time of discharge by nursing and or nursing/ family communication was 16% and the most common inaccuracy is medication related. Further research is needed to eliminate AVS mistakes.

Pediatric Residency Transgender Education Initiative

Elizabeth Sandberg, MD, Amalia Lee, MPH, Steven Weinberg, MD, Emily Vander Schaaf, MD, Sue Tolleson-Rinehart, PhD

Background: Pediatricians play a critical role in the lives of transgender youth, making a gender-health curriculum essential in residency training. Before June 2017, our institution's pediatric residency did not have a formal curriculum about the health needs of transgender youth.

Objective: Develop a residency curriculum to achieve 80% of residents reporting (1) comfort (2) familiarity, and (3) feelings of preparedness to care for transgender patients.

Design/Methods: Using a multidisciplinary team and formal quality improvement (QI) methods, we redesigned content and format of the curriculum based on results from key measures and Plan-Do-Study-Act (PDSA) cycles. Primary outcome measures included resident feelings of comfort, familiarity with, and preparedness for caring for transgender youth, which we evaluated every 6 months for 18 months.

Results: Residents strongly agreed/agreed with the following statements: 50% felt familiar with the unique health issues affecting transgender people at baseline, improving to 86% at 6-months, 76% at 12-months and 89% at 18 months; 23% felt prepared to meet the clinical needs of transgender patients at baseline, improving to 52% at 6-months, changing to 41% at 12 months and 57% at 18-months; 58% felt comfortable providing services to transgender people at baseline, while 70% felt comfortable at 6-months, 58% at 12-months, and 92% at 18-months. At baseline, 11% of residents reported being very comfortable talking to patients about gender identity, 49% were somewhat comfortable. At 6-month follow-up, 10% were very, 86% were somewhat comfortable, at 12-months, 11% were very, 72% were somewhat comfortable, and at 18 months, 37% were very, and 52% were somewhat comfortable talking to their patients about gender identity.

Conclusion: During the initiation of a wide variety of educational and experiential learning opportunities for pediatric residents, we have achieved our goal of 80% of residents reporting familiarity and comfort providing services to transgender patients, but we have failed to achieve our goal of 80% of residents feeling prepared to do this work. More exposure and education may have helped residents recognize gaps in their knowledge and reassess their feelings preparedness in the context of a more nuanced understanding of what this care involves. However, the significant improvement in resident comfort discussing gender identity with patients, suggests we have taken a critical steps toward better care for transgender patients.

A Quality Improvement Project to Improve Social Determinants of Health Screening and Referral in a General Pediatrics Resident Clinic

Katherine Schroeder, MD; Martha Perry, MD; Colin Orr, MD MPH; Emily Vander Schaaf, MD MPH; Rushina Cholera, MD PhD; Christian Lawrence, MD; Darragh Davis, BA; Eric Bonilla Reyes; Madlyn Morreale JD MPH; Kori Flower, MD MS MPH

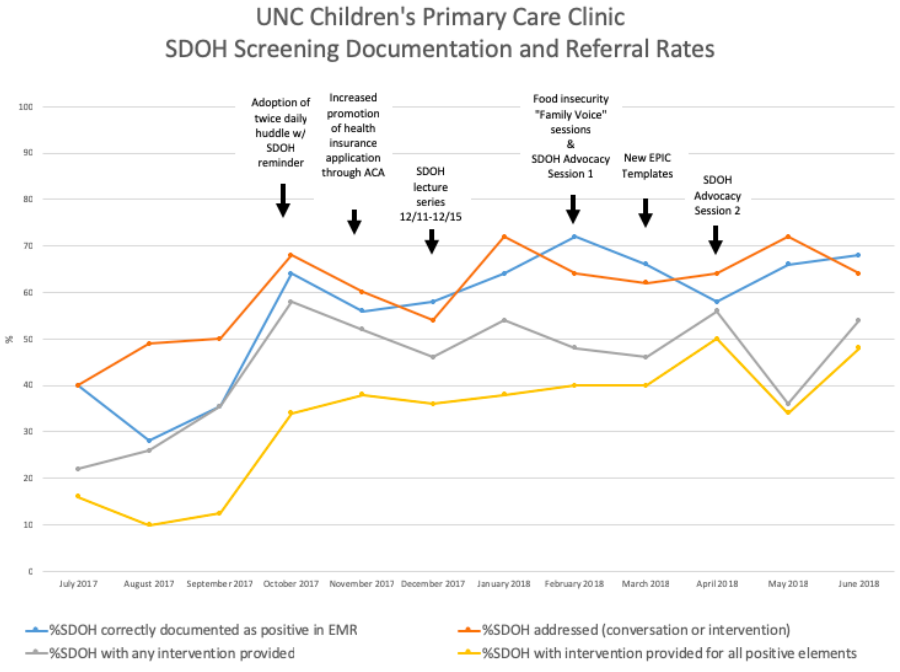
Background: According to the American Academy of Pediatrics (AAP), half of children live in or near poverty and are impacted by social determinants of health (SDOH) such as food insecurity and exposure to domestic violence. To better address patient needs, the AAP recommends regular screening for SDOH during patient encounters.

Objective: We designed a quality improvement project to improve SDOH screening, documentation, and resource provision during well-child checks.

Methods: We created a multidisciplinary team to identify drivers for change within the existing SDOH screening and referral process in an academic medical center clinic. We collected baseline screening data over three months via review of monthly samples of randomly-selected positive SDOH paper screens (N=50). We compared these screens to electronic medical record (EMR) documentation of screen results and associated resources/referrals provided within a standardized well-child visit EMR template. Using Plan-Do-Study-Act methodology, the team applied seven interventions related to SDOH screening and workflow over a 7-month period. Interventions included a lecture on SDOH for all providers, increased promotion of available SDOH resources, and refining EMR visit templates to prompt discussion with families and correct documentation of SDOH screen results. We tracked progress monthly by reviewing a random sample (N=50) of positive paper screens and associated EMR documentation. A parent/caregiver survey (N=50) was conducted after the intervention period to elicit family experiences with improved SDOH screening.

Results: Proportion of positive SDOH paper screens correctly documented in the EMR increased from 35% at baseline to 68% after all interventions. Discussion about positive screens increased from 46% to 64%. Positive screens with any resource/referral provided improved from 28% to 54%. Positive screens with resource/referral provided for all identified needs improved from 13% to 48%. On parent/caregiver survey, 36% of families reported having SDOH needs, 73% found the resources provided to be helpful, and 44% reported wanting to discuss SDOH at every clinic visit.

Conclusions: Changes to education and workflow improved SDOH documentation, discussion, and resources/referrals at well-child visits. The majority of families found resources/referrals provided to be helpful and many would like SDOH to be addressed at every visit.



Reducing Acute Kidney Injury from Nephrotoxic Medications in Hospitalized Patients

William Stoudemire, Nicole Damari, Nour Akil, Meredith Cuthriell, Carmen Echols, Yuting Liao, Cameron McKinzie, Erica Bjornstad, Katie Westreich

Problem Statement:

Acute kidney injury from nephrotoxic medications for pediatric patients is common and can lead to short- and long-term morbidity and mortality.

Aim:

To reduce the rate of acute kidney injury from nephrotoxic medications by 50%.

Introduction:

Acute kidney injury (AKI) from nephrotoxic medications in hospitalized patients is common. AKI can lead to both short term (increased duration of hospitalization, need for dialysis) and long term complications (hypertension, increased risk for chronic kidney disease, increased risk of mortality). Prior efforts have shown that AKI from nephrotoxic medications can be significantly reduced with screening and increased monitoring.

Methodology:

We created a multidisciplinary team of attending and fellow physicians, pharmacists, and nurses. We first analyzed data to gather baseline data for the project and confirm that our rate of AKI from nephrotoxic medications was unacceptably high. We then used quality improvement methods to identify key drivers leading to nephrotoxic kidney injury, define current processes, and identify possible interventions for improvement. Successful interventions implemented included automated screening for patients exposed to high nephrotoxic medications, increased monitoring for signs of early kidney injury, and a pathway to improve hydration of these patients. These interventions were first piloted on the pediatric pulmonology service and then sequentially expanded to other services in the children's hospital optimization with PDSA cycles.

Results:

Our baseline data revealed that 25% of hospitalized patients were exposed to 3 or more nephrotoxic medications, and that 23% of those patients developed acute kidney injury during their hospitalization. For pediatric pulmonology patients, the baseline AKI rate for exposed patients was 18.8%. In the 6 months following implementation of our project, the mean rate of AKI for patients on pediatric pulmonology was 9.0% (a 52% relative reduction), and analysis revealed a statistically significant shift in our process ($p < 0.05$). No adverse events including central line associated bloodstream infections or volume overload occurred.

Conclusions:

Interventions to screen for and improve monitoring and hydration of patients exposed to nephrotoxic medications led to reduced rates of AKI for pediatric pulmonology patients. Spreading these interventions to other patients in the children's hospital will help reduce this risk in other patients as well.

Enhancing Antibiotic Selection through Inpatient Penicillin Allergy Evaluation

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Rationale

Penicillin allergy testing has historically been underutilized in the inpatient setting where it may directly impact treatment. We expanded penicillin allergy testing to hospitalized patients to determine the rate of penicillin hypersensitivity, the barriers of inpatient testing, and the subsequent use of beta-lactams or cephalosporins.

Methods

We evaluated 55 hospitalized patients from 9/2017-8/2018 with reported penicillin allergy that were referred by primary teams or specialists if beta-lactams or cephalosporins were desired for prophylaxis or to treat underlying infection. Qualifying patients then underwent skin testing followed by oral Amoxicillin challenge.

Results

37 patients (67%) underwent testing with 35 (94%) proving to be tolerant of beta-lactams or cephalosporins. Of the patients that were tested, direct drug challenges were administered to 19 (51%) patients based on clinical history, with no reported adverse outcomes. Of the 35 tolerant patients, 25 (71%) had a beta-lactam added to their treatment, 9 (26%) received a cephalosporin, whereas 4 (11%) received other antibiotics. Barriers to testing included patients with incompatible medication use (4), clinical instability (5), or delayed symptoms (4); 4 patients declined testing.

Conclusions

Our results suggest that 6% of patients with reported penicillin allergy exhibit true sensitivity, consistent with previous reports. A majority of patients were able to tolerate direct drug challenge, indicating that improved utilization of clinical history can simplify testing protocols and may prove sufficient for many patients. Additionally, most subsequent treatment plans for tested patients included a beta-lactam or cephalosporin. Despite barriers, inpatient penicillin allergy testing may decrease reported allergies and broaden antibiotic options.

A Professional Coaching Program for Residents at the University of North Carolina

Hannah Y Coletti, MD, MPH; Joanna M Hales, MD; Alyssa Tilly, MD; Kathryn C Adair, PhD; Christine Gladman, MD, MPH

Purpose:
To describe our experience and early outcomes from the first 2 years following implementation of a coaching program for residents

Background:
Coaching is a successful tool to combat burnout and promote resilience through a model of positive psychology. Learners who are being coached are taught to identify positive traits and experiences in order to counter manifestations of burnout.

Methods/Process:
In 2017, the UNC Pediatrics, Internal Medicine, and Medicine-Pediatrics residency programs instituted the Professional Development Coaching Program. Faculty members from both medicine and pediatrics were recruited and attended a 2 hour training session in coaching and positive psychology. In our first year, 43 interns participated (70% of eligible interns) and 20 coaches were trained; and 88%oftrainees and 85% of coaches elected to continue in the program. In our second year, 48 additional interns joined the program; an additional 22 new coaches were trained. Our next step is to quantitatively assess residents' wellness outcome using survey tools including indices of burnout (adapted from the Maslach Burnout Inventory), as well as validated measures of work-life balance, depression, resilience, and subjective happiness.

Conclusions:
Anecdotally, residents who have participated in the coaching program have expressed extremely positive feedback and feel that the program has helped with overall wellness and resiliency. We are looking forward to analyzing the data from the survey results in the near future. In addition, given persistently high demand within the departments of Pediatrics and Medicine we hope to expand participation throughout the hospital in the future.

Procedural Competency in Neonatal Chest Tube Placement

Jacob K Johnson, Matthew M Massaro, Wayne A Price, Sofia R Aliaga

Background:
The achievement and maintenance of procedural competency is an important aspect of pediatrics residency and fellowship training. High fidelity simulation has been increasingly used to accommodate for a lack of procedural experience. A recently proposed teaching method (LSPPDM) includes the following process: learn the procedure (Learn), observe the procedure (See), practice the procedure on a simulator (Practice), competency-based assessment and feedback on the simulator (Prove), perform the procedure on a human (Do) and maintain the skill with clinical practice and simulation (Maintain). However, the interval for maintenance of skills is unknown.

Objective:
Compare retention of competence, as defined by a scoring tool, at 3 and 6 months among pediatric residents after learning to place a neonatal chest tube on a simulator.

Methods:
We modified and validated a chest tube placement scoring tool. We used the borderline groups method to reach a consensus cut-off score for competence. Twenty-one pediatric residents were recruited and completed a standardized training process using the "learn, see, practice, prove" teaching method. Each resident attained 2 consecutive competence scores and then an additional competence score 1-2 weeks later. Each learner was then stratified by postgraduate training year and randomized to re-test after 3 or 6 months. Competence score and procedure completion time were recorded at each time point. We used Fisher's exact test and paired t-test to determine statistical significance.

Results:
The completion rate for this study was 95% (20/21 participants). The mean pretest score, competence score and procedure completion time were similar between each group prior to randomization. The percentage of participants who attained a competent score after 3 months and the actual score was higher than the percentage and actual score after 6 months, although the differences did not reach statistical significance (90% vs 60%, p=0.30; and 33 vs 31.5, p=0.052). Procedure completion time was lower in the 3-month group (368±42 seconds) compared to the 6-month group (402±71 seconds) but did not reach statistical significance (p=0.24).

Conclusions:
Data suggests there is no difference in percent of trainees who maintain competence at 3 vs 6-month intervals of evaluation using a neonatal chest tube simulator. Limitations of this study include the smaller sample size. Therefore, based on this data, biannual training and assessment for infrequent procedures such as neonatal chest tube placement is likely sufficient.

Medical Student Chiefs: An Innovative Role in Undergraduate Medical Education

Marni Krehnbrink, Samantha Robin, Karina Javalkar, William Mills, M.D., Richard Hobbs, M.D.

Background
Medical student chiefs are senior medical student leaders who serve as educators and mentors and who help to improve clerkships and pre-clinical education in their chosen specialty. A literature review indicates that this type of specialty specific role for senior medical students has never been published in a peer-reviewed journal, and that the Pediatric Medical Student Chief (PMSC) position at the University of North Carolina School of Medicine is one of the first of its kind.

Objectives
Throughout the 2018-2019 school year, the PMSCs initiated many educational projects. Post-orientation bedside teaching for pediatric clerkship students aimed to help new learners in pediatrics gain comfort with examining pediatric patients. Senior leadership at a free, student-run Well Child Clinic, assisted other student doctors who oftentimes had limited pediatrics experience. Writing and distributing daily case-based questions to clerkship students helped emphasize and refine knowledge of important topics. Updating guides for clerkship students and residency applicants based on student experience helps advise other student doctors. Creating a central resource of pediatric research opportunities helps students access and become involved in scholarly projects. Future directions for the PMSC position include student-organized morning report conferences for junior medical students that can focus on core pediatric topics. Other future projects include refining and expanding existing pediatric simulations.

Summary
Medical student chiefs are passionate about teaching, mentorship, and leadership and can contribute significantly to the clinical and pre-clinical education of junior medical students. This role offers a novel opportunity for senior students to contribute to the education of junior students while developing their clinical teaching skills. Due to their recent experiences as medical students, they are able to provide constructive feedback on medical curriculum as well as relate to junior students to provide support and advice. This role has the potential to be expanded to other specialties to both improve the education of medical students and help senior medical students to develop teaching and leadership skills amongst their peers.

Institutional Gender Symposium

Amalia Lee MPH, Nina Jain MD, Elizabeth Sandberg MD

Background:

Our institution has providers across multiple departments and subspecialties working to provide care for transgender (TG) patients. To date, there have been no formalized gatherings of these individuals to offer opportunity to collaborate, educate, and share resources.

Objective:

Organize a symposium on the current state of TG health at our institution, and evaluate the effectiveness of this symposium to educate attendees about key components of care for this at-risk patient population and resources available at the institution.

Design/Methods:

We planned a full-day conference, including speakers specialized in bioethics, law, endocrinology, infectious diseases, and surgery. Invitees included care managers, social workers, students, physicians, nurses, and administrative staff. An anonymous baseline survey and follow up survey were administered to evaluate change in knowledge after attending the conference.

Results:

Participants strongly agreed/agreed with the following statements – 33% felt knowledgeable about resources for TG patients at UNC at baseline, 100% at follow-up (p=0); 77% were aware of basic ethical concepts associated with care of TG individuals, 96% at follow-up (p=0.104); 81% understood barriers faced accessing transition-related healthcare at baseline, 100% at follow-up (p=0.062); 84% were knowledgeable about disparities experienced by TG patients at baseline, 100% at follow-up (p=0.129); 77% understood the basic elements needed to create a gender affirming clinical environment at baseline, 100% at follow-up (p=0.044); 60% understood the role of the pediatric endocrinologist at baseline, 100% at follow-up (p=0.002); 47% were aware when to refer to pediatric endocrinology at baseline, 81% at follow-up (p=0.023); 20% were familiar with surgical treatment options available at our institution at baseline, 95% at follow-up (p=0); and 61% were familiar with the role of HIV pre exposure prophylaxis in TG care at baseline, with 100% at follow-up (p=0.011). The participants requested future conferences including ethnic/racial disparities, perspectives from a mental health provider, discussion of other surgeries, case management tips for navigating health insurance and legal issues, techniques for supporting patients, as well as more involvement of TG patients, including a patient panel.

Conclusion:

A one-day conference increases provider awareness of resources available at an institution, and is an effective way to disseminate information amongst healthcare professionals caring for TG patients.

PREVALENCE OF FEEDING DIFFICULTIES ACROSS NORTH CAROLINA COUNTIES PRESENTING TO THE INTERDISCIPLINARY FEEDING TEAM AT THE NC CHILDREN'S HOSPITAL

Kristen F. Cole, MSN, RN, CPNP, Victoria M. Powell, MSN, RN, CPNP, Kristen A. Brackett MS, CCC-SLP, Derica N. Sams, MD, Maureen M. Kelly, DNP, RN, CPNP

Feeding difficulties are increasingly common in children, with an estimated 25-40% of typically developing children and up to 80% of those with chronic medical conditions affected. Feeding problems span socioeconomics, ethnicity, culture and demographics, making feeding problems applicable to all people and children. Many studies have investigated the reason for this phenomenon while numerous feeding teams have been assembled to address the growing need for care in this specific population.

The UNC Feeding Team has grown from its inception in 2009 with three feeding team visits per week to seventy-five per week at present. We have collected data from the feeding team visits in 2017 (the feeding team's ninth year providing coordinated appointments) that reveals the widespread distribution of patients accessing our feeding team services from across the state of North Carolina.

North Carolina encompasses one hundred counties that span from the coast to the mountains. Its citizens are equally as diverse as its geography, making it a prime ground for studying patterns and trends in children with feeding difficulties. The NC Children's Hospital takes pride in providing specialized care for over 70,000 children across the state per year.

The UNC Feeding Team is unique in its approach, offering an interdisciplinary team, consisting of physicians, a pediatric nurse practitioner, a speech and language pathologist and a pediatric dietitian who see each patient together, developing a synthesized plan of care for each unique patient. Our team treats underlying gastroenterological (GI) issues and rules out GI disorders before moving on to teach children to eat. We use medications and formulas to prepare the GI tract to be the most comfortable and prepared to receive and absorb nutrients for proper growth and development. We then use a behavioral feeding therapy approach to engage children to progress in oral skills for most efficient eating and growing.

Because our team is unique in its approach and we have established positive outcomes, we now have a three-month waitlist for new patient appointments. The distribution of our patient population demonstrates the growing need for an interdisciplinary approach to pediatric feeding problems along with associated difficulty patients have in accessing these services locally. We have identified strategies from our success in both providing medical care and educational outreach to other professionals and families in North Carolina, to address the growing need for information and comprehensive services for our children with feeding difficulties.

Salvation from Inflammation: An 11-year-old boy with fever and acute kidney injury
Amber Beg MD, Roman Melvin MD, Eric Zwemer MD

Initial History/Presentation

An 11-year-old boy presented with three weeks of body aches, fatigue and intermittent emesis. He was previously treated for presumed UTI with amoxicillin. He continued to have daily fevers and developed periorbital swelling and conjunctival erythema without purulence, along with headaches and abdominal pain. Despite excessive thirst, he had low urine output and dark urine.

Physical Exam

Vital signs were normal for age. Exam revealed an alert but tired appearing child with dry mucous membranes. Abdominal exam showed significant distention and hepatosplenomegaly.

Diagnostic Evaluation

Initial chemistries showed BUN/Cr of 32/2.15, Na 126, and K 3.7. LFTs demonstrated albumin 2.6, AST 331, and ALT 153. CBC showed WBC 5.2, Hgb 8.6, and platelets of 128. CRP was elevated at 29.8 (normal <0.5). Urinalysis showed mild protein and 2 RBCs. Workup for acute kidney injury (AKI) showed normal C3/C4, normal renal ultrasound, and negative urine culture. Blood culture and stool studies were negative. Abdominal ultrasound confirmed significant hepatosplenomegaly without other masses. Given cytopenias and hepatosplenomegaly, ferritin was checked and significantly elevated to 16,600. EBV IgM and IgG testing was positive, with negative CMV titers. Bone marrow studies showed no evidence of malignancy, or other marrow infiltrative process.

Diagnosis

Patient met criteria for EBV-induced Hemophagocytic Lymphohistiocytosis (HLH), with fever, cytopenias, hyperferritinemia, hepatosplenomegaly, low fibrinogen, elevated triglycerides, transaminitis, and positive EBV titers. HLH gene panel later revealed a mutation in the XIAP gene consistent with X-linked Lymphoproliferative Disease.

Discussion/Conclusion

Pediatricians should consider the diagnosis of HLH in any patients presenting with multiple cytopenias and hepatosplenomegaly, and ferritin is an effective screening test. AKI is an uncommon presenting symptom in pediatric patients with HLH, although it is frequently seen later in the illness/treatment course. In adults, the incidence of AKI in HLH is higher, with common causes including acute tubular necrosis, hypoperfusion, tumor lysis syndrome, or HLH-associated glomerular nephropathies. While the same multifactorial causes likely contribute to HLH-associated AKI in children, it has also been proposed to be a consequence of renal tissue infiltration by the disease process itself. Pediatric nephrology was consulted during admission, and the AKI was attributed to a predominantly pre-renal insult, as supported by a FeNa of 0.7%, secondary to dehydration and renal hypoperfusion from splanchnic dilation due to systemic inflammation.

A Twisted Fate: A 16-Year-Old Girl with Severe Abdominal Pain
Roman Melvin MD, Amber Beg MD, Ola Akintemi

Initial History/Presentation:
A 16 year-old girl presented with one day of abdominal pain she initially attributed to menstrual cramps. The pain escalated into intermittent severe and sharp abdominal pain, localized to bilateral lower quadrants and worsened by movement. She reported subjective fever and non-bloody, non-bilious emesis, along with constipation for 1 week. Despite menarche 2 years prior, she had not menstruated in the past year. She denied any sexual activity.

Physical Exam:
Vital signs were within age appropriate limits. Exam revealed a visibly uncomfortable teenager, with tenderness to palpation in the bilateral lower quadrants and guarding. No rebound tenderness, masses or hepatosplenomegaly were noted. External genitourinary exam was normal.

Diagnostic Evaluation:
Initial laboratory evaluation was notable for WBC of 16.5 with left shift. CMP, lipase, serum bHCG, urine pregnancy and urine GC/Chlamydia were negative. Abdominal ultrasound did not visualize the appendix. CT abdomen/pelvis did not visualize the appendix, but no right lower quadrant inflammatory process was seen. Transabdominal and transvaginal pelvic ultrasounds noted no evidence of ovarian torsion with Doppler flow to both ovaries. The right ovary measured at 6.1 x 3.9 x 3.9 cm, and the left ovary at 4.2 x 2.7 x 2.9 cm. Small free fluid in the pelvis was read as likely physiologic, and cystic changes within the endometrium noted as likely due to premenstrual changes.

Diagnosis:
She continued to have severe pain throughout admission and after discussion with OB/GYN and pediatric surgery, MRI abdomen/pelvis was obtained to rule out Mullerian duct abnormalities, abscess or other potential etiologies. Torsion of the right ovary was diagnosed on MRI, with a twisted pedicle along the right lateral aspect of the right ovary and marked enlargement of the right ovary (7.0 x 6.4 x 7.7 cm).

Discussion/Conclusion:
Diagnosis of ovarian torsion was elusive in this patient due to negative transabdominal and transvaginal pelvic ultrasounds. While studies have shown that sonography has a relatively high accuracy, in cases where ovarian torsion is not visualized on ultrasound, a high clinical suspicion should be maintained if a patient has 2 or more combined sonographic findings (ovarian edema, abnormal ovarian blood flow, relative enlargement of the ipsilateral ovary, free fluid around the ovary or in the pouch of Douglas, ovarian cyst/mass, or abnormal ovarian location). In this case, the patient had free fluid noted in the pelvis as well as relatively enlarged ipsilateral ovary. Once MRI findings were available, she underwent emergent surgical detorsion and the ovary was salvaged.

Successful Allogeneic HCT for W.H.I.M. Unexpectedly Complicated By Hypersensitivity Pneumonitis

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Intro

Warts, hypogammaglobulinemia, infections, myelokathexis (WHIM) syndrome is a rare primary immunodeficiency defined by its symptoms and CXCR4 gene mutation. This leads to myelokathexis in which mature neutrophils cannot exit the bone marrow, causing neutropenia. Use of hematopoietic cell transplant (HCT) to correct a primary immunodeficiency is well accepted; yet, due to the rarity of WHIM, data in this setting are limited. We present a successful matched sibling donor (MSD) bone marrow (BM) transplant in a WHIM patient who later developed unexpected respiratory complications.

Results

The patient was initially followed for congenital neutropenia and therapy consisted of weekly filgrastim. At age 5, he was diagnosed with WHIM by CXCR4 sequencing. Treatment was changed to plerixafor, but he required frequent dose escalation. HLA typing revealed his brother was an 8/8 allele match. Patient's history was significant for asthma, but no recent flares. At 6 years old he received a MSD BM graft after myeloablative conditioning. Myeloid engraftment ocurred on day +20 with >95% donor chimerism after day +55.

On days +76, +87, and +104, he was admitted for respiratory distress. Infectious evaluations only revealed EBV PCR in a BAL specimen. For this he received four weeks of rituximab. On each admission he improved quickly with albuterol and steroids, but symptoms recurred after returning home. During his third admission it was found that he was exposed to an African Grey Parrot in his home. Due to the close association of his symptoms with returning home, and his exquisite response to steroids, Pulmonology consultation lead to a diagnosis of hypersensitivity pneumonitis. The bird was relocated, the home extensively cleaned, and while staying in temporary housing the patient did not relapse. After returning home the patient had no further respiratory events. The bird was allowed to return on day +367, and patient has had no further respiratory events.

Conclusion

Hypersensitivity pneumonitis is a type IV hypersensitivity reaction that can be linked to bird exposure. It is T cell mediated and IgE independent. Acquisition of IgE mediated allergies has been reported after HCT, but there are no reports of new type IV hypersensitivity development. Neither patient nor donor had previous hypersensitivity to the bird. Due to the transient nature of the pneumonitis in our patient, this likely represents an unusual case of abnormal T cell maturation post HCT. This case also emphasizes the importance of environmental exposures in post HCT respiratory symptoms.

Call the Butcher: A Pig-Tail Catheter Was Not on the Menu

Darius Byramji MD and Zach Pettigrew MD; Robert Gardner MD, Alana Painter MD, Tom Belhorn MD, and Ashley Sutton MD

PRESENTATION: An 11-year-old previously healthy female presented with three weeks of abdominal pain and intermittent high fever. Previous evaluations resulted in presumptive diagnoses of constipation or viral gastroenteritis. On admission to our hospital, patient had lower abdominal cramping, loose non-bloody stools, and night sweats associated with an eight-pound weight loss. She had no known sick contacts or exposures. Initial working diagnosis was malignancy given prolonged systemic symptoms.

EXAM: Patient appeared generally unwell. The pertinent significant finding was diffuse abdominal tenderness with normal bowel sounds. Lymphatic, cardiac, respiratory and neurologic examination were normal.

LABS/IMAGING: Abdominal radiograph showed an abnormal paravertebral stripe. MRI abdomen revealed 5.8 cm left paravertebral mass at T10-T12, concerning for neuroblastoma, ganglioneuroblastoma, or lymphoma. PET scan showed no other lesions. Vertebral mass needle biopsy showed histiocytic infiltrate with negative AFB stain and no malignancy on flow cytometry. She was initially discharged pending final biopsy results, but returned three days later with new left sided chest pain, dyspnea, and high fevers. Laboratory evaluation was significant for negative blood cultures, HIV Ab and QuantiFERON gold. MRI of the thoracic spine for ongoing fever showed T10-12 vertebral osteomyelitis with adjacent paraspinal abscess. Lung ultrasound showed complex left pleural collection prompting VATS and thoracostomy tube and pig-tail catheter. Pleural rind culture grew non-typhoid *Salmonella*. Fecal *Salmonella* PCR was negative. The patient responded well to a four-week course of intravenous ceftriaxone followed by two weeks of oral cefdinir therapy.

DIAGNOSIS: Non-typhoid *Salmonella* vertebral osteomyelitis and discitis with associated paraspinal phlegmon and secondary empyema.

DISCUSSION: We present a case of non-typhoid *Salmonella* gastroenteritis with hematogenous spread to the spine and pleural space in a healthy adolescent. Further history revealed the family ate meat from a local butcher and later had self-resolved diarrhea. It is unclear if the pleural space infection was due to the primary infectious process or related to a communicating tract following needle biopsy with spread from the paraspinal infection. Invasive *Salmonella* infection affecting either the bones or pleural space is rare in immunocompetent hosts, with multisite infection being even rarer. Our patient's vague symptoms, complexed with an initially negative infectious work-up and imaging concerning for malignancy, likely resulted in anchoring bias that delayed further evaluation of infectious etiologies for her fevers. Ultimately, this case highlights the importance of maintaining a broad differential in pediatric patients with fever and contributes to a growing caseload of immunocompetent children with invasive *Salmonella* disease.

A Case Series of Arterial Cannula Thrombosis in Pediatric Extracorporeal Life Support

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Introduction

Patients on extracorporeal life support (ECLS) require anticoagulation to prevent thrombosis within the circuit. Thrombotic events on ECLS can be catastrophic. Children have a weaker capacity to generate thrombin, yet in clinical practice infants and young children often require higher doses of heparin to remain anticoagulated while on ECLS. This physiology may make achievement of optimal anticoagulation a challenge. In addition to anticoagulation, maintaining adequate circuit flow is imperative for avoiding thrombotic complications, as low flow states can lead to rapid fibrin accumulation, particularly at low shear zones. We report 3 cases of arterial cannula thrombosis requiring emergent intervention in infants cannulated for venoarterial ECLS.

Case Presentation

1) A neonate with pulmonary hypertension and respiratory failure secondary to congenital diaphragmatic hernia was cannulated on day of life 3. Heparin infusion was delayed for several hours secondary to elevated ACT. Approximately 14 hours after cannulation he developed a short run of tachyarrhythmia, resulting in the cessation of flow to the ECLS circuit with a clot soon discovered in the arterial cannula, which had to be replaced.

2) A neonate with postnatally diagnosed congenital heart disease and hypertrophic cardiomyopathy was cannulated for ECLS due to profound hypoxemia. He had multiple failed trials off support in attempt to decannulate prior to palliative cardiac surgery. He was maintained in a low flow state for a few hours each day, with progressive fibrin accumulation to the arterial cannula, resulting in emergent transition to the OR for next stage cardiac repair.

3) Five month old with Trisomy 21, chronic lung disease and unrepaired atrioventricular canal defect, cannulated for ECLS for gram negative rod sepsis. She was moved during routine care that lead to complete loss of flow and required CPR. During circuit rescue, a large occlusive thrombus was noted in the arterial cannula, which was able to be manually evacuated.

Discussion

These cases demonstrate how quickly thrombosis can occur during periods of low circuit flow. Small clots are common and typically lead to no harm to the patient or circuit, however in infants with small cannulas an average sized clot can have devastating effects. Well established flow in combination with adequate anticoagulation are essential for avoiding thrombosis in ECLS patients, especially in pediatric patients as the cannula sizes and flow rates are much less than that in adults. This series of thrombotic events has prompted an evaluation in practice to direct immediate attention to cannula patency during periods of no to low flow. Our institution has performed a thorough investigation of anticoagulation practices and management leading to the initiation of heparin infusions at a higher rate to help achieve therapeutic anticoagulation in a shorter time frame since ECLS initiation.

Meddling Meckel's Obstructs Diagnosis

Robert Gardner[1], Roman Melvin[1], Ashley Sutton[1]
[1]UNC Pediatric physicians

Initial History/Presentation: A 16-month-old boy with history of inguinal hernia repair presented with emesis, abdominal pain and decreased stooling. His last stool was three days prior to admission. He had increasingly frequent non-bloody, non-bilious emesis despite ondansetron prescribed by his pediatrician. Parents noted new episodes of waking from sleep agitated and screaming, occurring almost every hour and overall increased fussiness. He had no associated fevers and no sick contacts.

Physical Exam: Afebrile, vital signs normal. Exam notable for an inconsolable, non-toxic appearing toddler who would draw his legs up toward his chest during pain episodes. His abdomen had hypoactive, but present bowel sounds and was mildly distended with diffuse tenderness, but remained soft without peritoneal signs.

Diagnostic Evaluation: Labs including CMP, CBC and CRP were normal. Intussusception was suspected clinically. KUB showed a non-obstructive gas pattern with an abnormality in the right upper quadrant. Abdominal ultrasound revealed an indeterminate aperistaltic tubular structure in the left upper quadrant, possibly a duplication cyst vs. segment of bowel. UGI with SBFT was performed with normal rotation, but after several hours contrast did not progress to the colon. Serial KUBs demonstrated increasing gaseous intestinal distension with multiple foci of abrupt tapering, concerning for obstruction. Repeat ultrasound demonstrated new free fluid in the abdomen. Given radiographic findings, exploratory laparotomy was performed. Intraoperatively, a 3.5 cm Meckel's diverticulum with an adhesive mesodiverticular band and internal hernia of 20 cm of strangulated terminal ileum were identified. Bowel was viable after division of the band, and the diverticulum was resected.

Diagnosis: Intestinal obstruction secondary to internal hernia of the terminal ileum through Meckel's diverticulum and an adhesive band.

Discussion/Conclusion: This case is an uncommon initial presentation for a Meckel's diverticulum, which often is asymptomatic or has associated painless rectal bleeding. In our case, internal herniation through an associated band resulted in intestinal obstruction/strangulation and etiology was not clearly evident on initial imaging. Meckel's diverticulum should be considered as a possible etiology for obstruction in cases without other risk factors for obstruction.

Presenting symptoms of vomiting and abdominal pain in a previously healthy toddler were initially diagnosed as gastroenteritis, then thought to be most likely due to intussusception. Though there was concern for intestinal obstruction, the child's overall well appearance and lack of conclusive imaging initially delayed surgical intervention.

Imaging:

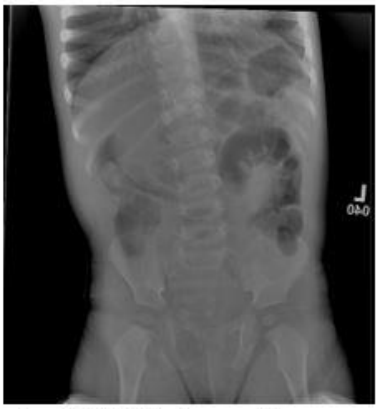


Figure 1. Initial KUB with non-specific gaseous distension of colon and RUQ abnormality

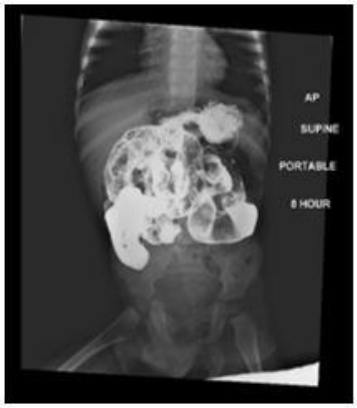


Figure 2. Repeat KUB 8 hours after start of UGI, showing delayed passage of contrast

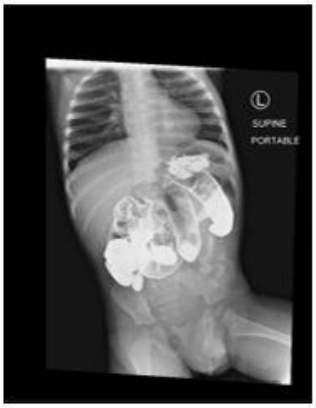


Figure 3. Final KUB with numerous foci of abrupt tapering, and continued retention of contrast consistent with high-grade bowel obstruction

A Trick Candle: A 27-day-old with fever and subcutaneous nodules
Blaire Hanvey MD, Eric Zwemer MD
University of North Carolina, Chapel Hill

A 27-day-old female presented with one day of fever and three weeks of subcutaneous nodules on her extremities. She did not have cough, congestion, vomiting, or diarrhea. Pregnancy and delivery were unremarkable. She did not require cooling at birth, and there were no concerns for ischemic injury. Growth was appropriate.

Admission vitals were 38.8C, pulse 175, and blood pressure 90/45. She was fussy but consolable. Cardiac exam showed 2/6 systolic murmur that radiated to the back. There was no hepatomegaly. Lungs were clear. She had palpable, erythematous nodules over her extremities.

Differential diagnosis for her fevers and subcutaneous nodules included subcutaneous fat necrosis (SCFN), Langerhans cell histiocytosis, and sclerema neonatorum. Neonatal sepsis workup was unrevealing, with negative blood, urine, and CSF cultures. Calcium and renal function labs were normal. CRP was mildly elevated (1.47, normal < 1). 25-OH-Vitamin D and 1,25-OH-Vitamin D were normal.

A lower extremity ultrasound was normal. A lower extremity MRI showed superficial nodules within subcutaneous fat, but no osteomyelitis.

Due to recurrent fever and new nodules, she underwent skin biopsy, which revealed histiocytic infiltration in the dermis and subcutis. Results were not indicative of Langerhans cell disease. Neutrophil function panel was normal. Bone marrow biopsy showed mild increase in blasts, but no significant dysplasia. Whole exome sequencing showed a splice variant in *PSMC5* and a rare second variant in *PSMD11,* consistent with CANDLE syndrome, or Chronic Atypical Neutrophilic Dermatosis with Lipodystrophy and Elevated temperatures.

The general pediatrician should be familiar with the differential for newborn skin nodules. This differential includes SCFN, Langerhans cell histiocytosis, congenital self-healing reticulocytosis, and sclerema neonatorum. The absence of cooling history, perinatal ischemic events, and hypercalcemia made SCFN unlikely. Skin biopsy ruled out the remaining disorders.

CANDLE syndrome is a rare, newly-described, autoinflammatory disease that results from mutations in proteasome subunits. Affected infants have a sustained production of type I interferons and present with daily fevers, fat loss, and skin lesions. Whole exome sequencing can help practitioners diagnose this rare syndrome. There are limited treatment options for CANDLE syndrome, as these patients do not respond to corticosteroids or anti-cytokine agents.

Early consideration of an autoinflammatory disorder in newborns with persistent fevers and negative infectious workup is important to allow for prompt diagnosis and treatment.

A Wolf in Cat's Clothing: An 8-year-old with recurrent fever and lymphadenopathy

Kirabo Herbert, MD; Nikkan Das, MD; Caroline Newman, MD; Joanna Hales, MD; Hannah Y Coletti, MD, MPH

Presentation:
An 8-year-old girl presented with 2 weeks of fever. She had also complained of neck pain and her mother had noticed swollen lymph nodes. She denied weight loss, night sweats, or respiratory or gastrointestinal symptoms. Of note, she presented similarly 7 months prior and was diagnosed with Bartonella infection (IgM+) for which she was treated with azithromycin and had resolution of symptoms at that time.

Physical Exam:
Vital signs were T 36.8C, HR 105, RR 19, BP 110/55, with normal oxygen saturation. Exam was notable for mild blepharedema, bilateral tender submandibular and cervical lymphadenopathy, and an erythematous papular rash with central crusting on her cheeks.

Diagnostic Evaluation:
Initial labs showed normocytic anemia (Hb 10.5 g/dL), thrombocytopenia (platelets 47 10*9/L), and elevated inflammatory markers (CRP 50 mg/L, ESR 73 mm/h). Infectious workup including Bartonella, parvovirus, EBV, CMV, TB, and HIV was negative. Lymph node and bone marrow biopsies were negative for malignancy. Rheumatologic workup revealed normal C3 and C4, negative dsDNA, positive ANA (>1:640), and ENA screen notable for positive anti-RNP and anti-SSA antibodies.

Diagnosis:
At 1 month follow up, the patient was noted to have developed oral ulcers. She was diagnosed with systemic lupus erythematosus (SLE) on the basis of meeting 1 immunologic criterion (positive ANA) and 3 clinical criteria (acute cutaneous lupus, oral ulcers, and thrombocytopenia).

Discussion:
SLE is an autoimmune disease that affects 5,000-10,000 children in the US with a female predominance (8:1). The clinical manifestations of SLE are broad and variable; thus, classification criteria have been developed such that diagnosis is confirmed when a patient meets 4 or more criteria (including at least 1 clinical and 1 immunologic criterion) or has biopsy-proven lupus nephritis. Nevertheless, diagnosis remains difficult due to the nonspecific nature of initial presentation. The most common initial symptoms of fever, weight loss, and malaise can also be seen in a variety of infections and malignancies. Our patient underwent an extensive workup and was initially diagnosed with Bartonella during a prior hospitalization. Furthermore, patients may not initially demonstrate sufficient findings to meet diagnostic criteria, but later develop new signs and symptoms. Therefore, close outpatient follow up may be key to subsequently making the diagnosis of SLE, as was the case for our patient.

STONES, BONES & GROANS: PARATHYROID ADENOMA ASSOCIATED WITH PEDIATRIC PANCREATITIS

Kane, Joelle, MD; Willis, Elizabeth, DO; Coletti, Hannah, MD, MPH; Jordan, Katherine, MD; Levenson, Amy, MD

Initial History: A 9 year old previously healthy female presented with non-specific abdominal pain for 4 days. She had not experienced fever, vomiting, diarrhea, constipation, or trauma. She had decreased appetite and increased pain with forward bending.

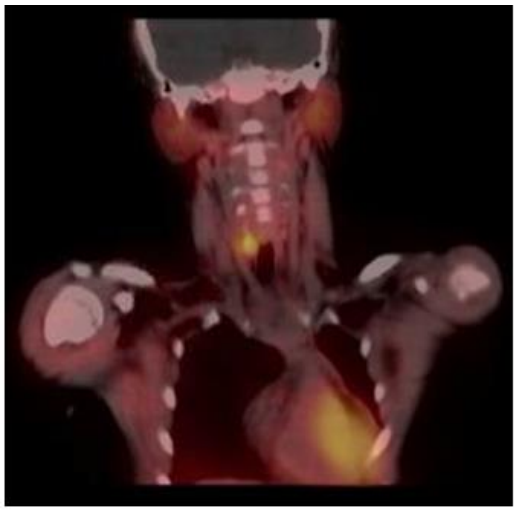
Physical Exam: Vitals were normal for age. BMI is 98th percentile for age. She was overall well appearing with a diffusely tender abdomen, worst in the right upper quadrant. She had voluntary guarding but no rebound tenderness.

Diagnostic Evaluation: Initial labs demonstrated elevated AST (54 U/L), ALT (92 U/L), and lipase (2,456 U/L). Abdominal ultrasound revealed cholelithiasis without evidence of cholecystitis or common bile duct dilation. Further workup showed normal triglycerides, elevated calcium (12.1 mg/dL), low phosphorus (3.9 mg/dL), and elevated PTH (295 mg/dL) suggesting primary hyperparathyroidism.

Diagnosis: With suspicion for a hyperfunctioning parathyroid, an ultrasound was performed revealing a mass overlying the right lobe of the thyroid. Tc-99m Sestamibi scan revealed a 1.1 x 0.9 cm soft tissue nodule consistent with a parathyroid adenoma. The patient had no family history or prior concerns for endocrine tumors.

Discussion/Conclusion: Primary hyperparathyroidism from a parathyroid adenoma is rare and challenging to diagnose in children and adolescents. The most common presentation, as in this patient, includes vague symptoms of hypercalcemia such as polyuria, abdominal pain, nausea, emesis, weight loss, and fatigue. When severe, hypercalcemia can also cause pancreatitis – a diagnosis most often related to trauma or biliary disorder in this age group. Clinicians should have high index of suspicion for primary adenoma in the setting of pancreatitis with an elevated calcium level. If suspected, localization of the adenoma with ultrasound and Tc99m Sestamibi scan is warranted.

A rare childhood parathyroid adenoma was likely the cause of this patient's hypercalcemia and subsequent pancreatitis. The parathyroid adenoma was excised, and the patient's calcium levels normalized. MEN testing was negative; however, other potential mutations in CDKN1B, Cyclin D1, and Prad1 genes have not yet been evaluated.



SPECT/CT: A 1.1 x 0.9 cm soft tissue nodule is noted along the inferior aspect of the right thyroid lobe demonstrating increased radiotracer uptake.

Fig 1: Spect CT demonstrating parathyroid adenoma in the right lower parathyroid gland.

Baby, why are you so blue?
Laura W. Lemley MD, Cherrelle Smith-Ramsey MD, Ashley Sutton MD.

Initial History/Presentation: A 7-day-old term infant, product of uncomplicated pregnancy and delivery, presented with cyanosis and lethargy worsening for 3 hours following elective ankyloglossia frenectomy. The procedure was performed with topical benzocaine applied to her frenulum. The infant was noted to develop immediate bluish discoloration of her oral mucosa that later generalized to her face, trunk, and extremities prompting presentation to the emergency department.

Physical Exam: Vital signs showed tachycardia and an SpO2 of 88%. Infant was lethargic, in no respiratory distress and had marked, generalized cyanosis and mottling (Figure 1).

Diagnostic Evaluation and Treatment: Venous blood gas significant for pH 7.39, pCO2 48, pO2 <30, HCO3 29, Lactate 3.15 (normal 0.05–2.0mmol/liter). Oxygen saturations were unchanged with administration of supplemental oxygen. Co-oximetry was notable for methemoglobin level of 32.4% (normal <3.0%) and venous fractional oxygenated hemoglobin 51.2% (normal 65.0–79.0%) diagnostic for methemoglobinemia. Intravenous methylene blue 1mg/kg was administered to the infant after which cyanosis resolved. Subsequent methemoglobin level was 1.0%.

Diagnosis: Methemoglobinemia resulting from topical benzocaine application

Discussion: Neonatal cyanosis can result from life threatening conditions including ductal dependent congenital heart defects, sepsis, and hemoglobinopathies. Acquired methemoglobinemia, an oxidized form of hemoglobin, is a result of oxidative stress and a rare cause of presentation for cyanosis. Benzocaine use in this case resulted in acute onset impairment of oxygen delivery to tissues, shifting the oxygen hemoglobin dissociation curve to the left. Methemoglobinemia, as in this case, results in cyanosis out of proportion to the inaccurate oxygen saturation measured by pulse oximetry. Co-oximetry should be utilized for accurate diagnosis of oxygen saturation given the presence of elevated methemoglobin. Methylene blue is the treatment of choice and results in rapid resolution of symptoms. The United States Food and Drug Administration (FDA) issued an initial advisory in 2006 warning of the risk of methemoglobinemia associated with topical benzocaine sprays and a black box warning in 2018 on over the counter oral products containing benzocaine for children younger than two years of age.

Figure 1 and 2: Infant upon initial presentation with marked, generalized cyanosis and mottling with slight jaundice.

A Novel Gli-similar 3 (GLIS3) Mutation with Concomitant Trisomy 21 resulting in a Unique Genotype-Phenotype Interaction

Lemley, Laura W. [1]; Jetten, Anton M. [2]; Levenson, Amy [1]

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Background: GLIS3 (Gli-similar 3) is a multifunctional transcription factor nuclear protein that plays an integral role in embryogenesis and is expressed primarily in cells of the pancreas, thyroid, liver, heart, skeletal muscle, and bone tissues. Although the clinical spectrum associated with GLIS3 mutations is variable, many individuals may exhibit neonatal diabetes, congenital hypothyroidism, and proportionate intrauterine growth restriction. Here, we present a case report of an infant female with Trisomy 21 and a novel mutation in GLIS3, resulting in a unique phenotype.

Design/Methods: The patient was a dichorionic, diamniotic twin female born at term with symmetric intrauterine growth restriction. The pregnancy was complicated by advanced maternal age and gestational diabetes mellitus. Postnatally, the infant was diagnosed with Trisomy 21, insulin dependent neonatal diabetes, nonsyndromic paucity of intrahepatic bile ducts, atrioventricular septal defect, and congenital hypothyroidism.

Results: In evaluating genetic etiologies of neonatal diabetes, a Maturity Onset Diabetes of Young (MODY) Neonatal Diabetes Panel was obtained at the sixth week of life and revealed a heterozygous GLIS3 variant c.328G>T (p.Val110Phe), an autosomal recessive defect reported as a variant of uncertain clinical significance. Interestingly, the neonatal diabetes was transient, and she no longer required insulin therapy after the fifteenth week of life. Our patient's mutation of GLIS3 was recapitulated in a gene variant model which demonstrated consistently diminished transcriptional activity compared to wildtype.

Conclusion(s): This case represents the overlay and likely interaction of phenotypes of the GLIS3 mutation and Trisomy 21. Specifically, these genetic syndromes are individually associated with congenital hypothyroidism, transient neonatal diabetes, hepatic fibrosis, cardiac defects, and growth restriction. In isolation, this novel variant of GLIS3 may not have a significant effect on the thyroid, pancreatic beta cells, or other affected tissues, but may act in combination with effects from the Trisomy 21 genotype producing the constellation of findings from the genotype-phenotype interaction demonstrated in our patient. This case illustrates that our patient's mutation may be a clinically significant or pathologic variant when in combination with Trisomy 21.

Patent Omphalomesenteric Duct Fistula
Rachael Lester, MD; Rebecca Smith, MD; Sean McLean, MD; Jacob Lohr, MD

A newborn boy, who had been born at 39 weeks and 1 day of gestation via primary cesarean delivery due to transverse lie, was noted at delivery to have an enlarged umbilical cord.

Physical examination showed a large umbilical cord insertion with two translucent yellow-colored areas near the proximal end of the cord; the underside had darker areas but without evidence of bowel contents. The patient had a normal newborn nursery course and was discharged on day 2 of life with a recommended 24- to 48-hour follow-up visit. At the newborn follow-up with the pediatrician, the umbilical cord was noted to be very thick without surrounding erythema.

At 13 days of age, the patient presented to his pediatrician for evaluation of a residual thick umbilical stump (**Figure 1**).

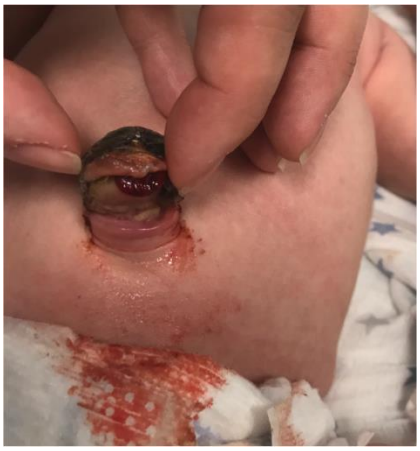

Figure 1. *The newborn presented to his pediatrician at 13 days of age for a residual thick umbilical stump.*

On examination, the cord appeared to be drying on the dorsal aspect, with the skin-abutting ventral aspect remaining soft and oozing rust-colored fluid, thought to be blood. The mother had noted a malodor but no surrounding erythema. At the office visit, the provider attempted to cinch the umbilical stump just above the skin; but application of an umbilical tie elicited immediate crying and thus was removed. The patient was sent to a pediatric surgery clinic for further evaluation (**Figure 2**).

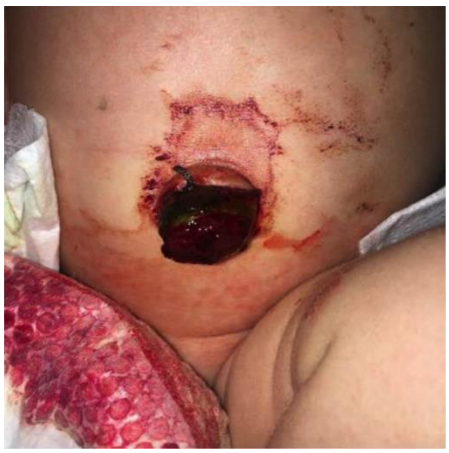

Figure 2. *Umbilical stump at presentation to a pediatric surgeon due to persistent bleeding.*

The patient later underwent surgical correction of the umbilical stump. Intraoperatively, it was noted that there was residual patent omphalomesenteric fistula connected to the skin. Pathological test results confirmed small bowel mucosa tracking to the surface of the specimen. Based on clinical presentation, intraoperative findings, and pathology findings, the newborn received a diagnosis of a patent omphalomesenteric duct (OMD) fistula.

The boy was seen 4 weeks postoperatively, at which time he was treated for surrounding cellulitis that was believed to have resulted from infected suture material, but otherwise he was doing well. He had an uneventful recovery with no further sequelae to date.

Discussion

The OMD is a temporary structure essential to normal fetal development, connecting the yolk sac to the primitive gastrointestinal tract in the developing embryo. Normally, the OMD completely involutes by week 9 of intrauterine life.[1,2] OMD remnants are uncommon, with an occurrence rate of 2% and with the rarest being an OMD fistula,[3,4] as in this patient's case. There are no known risk factors, although many reports show a male predominance in symptomatic cases.[3,4]

The presentation of OMD varies depending on the degree and location of duct patency. Complete patency results in the OMD connecting the umbilicus to the terminal ileum. This can lead to intestinal drainage from the umbilicus, similar to a stoma, as in this patient's case. A persistent OMD at the umbilicus but with no intestinal connection results in an umbilical sinus. Persistent tissue at the ileum with no connection to the umbilicus results in a Meckel diverticulum. Finally, patent OMD with closure at both ends results in an OMD cyst, which can lead to small bowel obstruction given that both ends are fixed.
Often, ultrasonography or a fistulogram may be required to visualize the anatomy. Symptomatic OMD remnants require surgical intervention, which is curative.

Given the risk of catastrophic outcome with ileum incarceration or strangulation, it is important that physicians remain vigilant and diagnose this abnormality to allow prompt surgical intervention.

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Don't hold your breath: how an apneic episode led to a timely diagnosis
C Liu, C Atkins, M Kihlstrom

Presentation:
6 week old term male infant presented to the ED for a 30-second apneic episode followed by perioral cyanosis that resolved with positioning. Since birth, he had frequent episodes with rhythmic arm and leg movements, eye deviation, and labored breathing but no apnea. A Pediatric Neurology appointment was scheduled for further evaluation. Other than a VSD on prenatal ultrasound, both pregnancy and delivery were uncomplicated.

Physical Exam:
On presentation his initial vitals were within normal range. Physical exam was notable for his small size, posteriorly rotated low-set ears, and generalized hypotonia. No murmur was appreciated on exam. During his ED visit, 4 episodes of apnea with desaturation and tachycardia were witnessed, along with extension of his extremities and eye deviation.

Diagnostic Evaluation:
Head CT and brain MRI ruled out intracranial abnormality. Infectious workup, including blood, urine and CSF studies, were unremarkable. Echocardiogram revealed a PDA, PFO, small VSD, and subsequent CTA found a right-sided aortic arch with an aberrant left subclavian artery. EEG showed multifocal epileptiform discharges, confirming seizures. His ionized calcium was low at 2.4 mg/dL, with a total calcium of 4.4 mg/dL. PTH low at 10.7 pg/mL with normal vitamin D and phosphorus levels. Karyotype, FISH and microarray revealed a 22q11.2 deletion.

Diagnosis:
DiGeorge Syndrome

Discussion:
DiGeorge syndrome is caused by chromosome 22q11.2 deletion, occurring in approximately 1:4000 births.[1] It accounts for 2.4% of individuals with intellectual disabilities, the second most common syndromal cause after Down syndrome.[2] It contributes to approximately 1.5% of all major congenital heart diseases.[3] It is also found frequently in infants with velopharyngeal insufficiency (64%) and hypocalcemia (74%).[1]

Despite its clinical significance, diagnosis of DiGeorge syndrome is often delayed or missed due to its high variability in expression of velocardiofacial features: the majority of individuals with the syndrome have palatal and facial anomalies, 77% have heart defects, and 77% have immunodeficiencies.[1] In our patient, his clinical features were subtle: he had posteriorly rotated and low-set ears but no other overt facial anomalies. His cardiac defects and aberrant left subclavian artery, though found in DiGeorge syndrome frequently, were only revealed after extensive cardiac workups. His apneic episode, presumably secondary to his seizure, was what brought him to medical attention. DiGeorge syndrome may not present overtly with characteristic features; seizure secondary to hypocalcemia should prompt endocrine and genetic workups.

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Seize the diagnosis…with caution

Pamela Londres, Robert Corty, William Mills

Objectives:

1. Recall importance of responding in timely manner to rapidly changing patient presentations in the emergency department
2. Recognize need for frequent reassessment of differential diagnosis
3. Review presentation of SSRI overdose

Case:

A thirteen year-old female with a history of type 1 diabetes mellitus, anxiety, depression, and suicide attempt presented with emesis and altered mental status. She was in usual state of health until she woke abruptly from sleep with nausea, emesis, and weakness. Parents state she takes insulin, fluoxetine, and PRN hydroxyzine, which she has reportedly been compliant with. She has no access to other medications. On arrival to ED, the patient appeared uncomfortable, but was alert and communicative. Initial exam completed 30 minutes after arrival was significant for tachycardia, diaphoresis, mydrosis, and weakness of right upper and lower extremities, along with difficulty following commands and communicating verbally, which acutely worsened during the exam. Patient was immediately taken to CT, where she had back-to-back first time generalized tonic-clonic seizures that resolved with lorazepam. Labs remarkable for glucose of 288 pH 7.33, lactate 2, and urine glucose >1000. Her CBC, BMP, ethanol, acetaminophen, and salicylate levels, urinalysis, and pregnancy test were otherwise unremarkable. EKG revealed sinus tachycardia. Head CT was unremarkable. During admission, patient had a normal brain MRI and an EEG which was consistent with post-ictal changes. Her clinical status improved throughout admission, and she was ultimately discharged with neurology follow up.

Discussion:

The differential diagnosis of emesis and altered mental status is broad. Evaluating for DKA and cerebral edema were initial priorities in this presentation. Other differentials include primary seizure disorder, autoimmune encephalopathy, meningitis, metabolic derangement, and SSRI overdose. Ultimately, access to SSRI in the setting of past suicide attempt, gastrointestinal disturbance, altered mental status, resolved seizure activity, and normal head imaging made it the most likely etiology. Interestingly, even severe SSRI overdose does not lead to serotonin syndrome.

Conclusion:

It is critical to keep clinical status foremost in your considerations when evaluating patients in the ED. Although it is tempting assume a diagnosis based on history and presentation—and these are often correct—doing so is always a potential danger to the patient. The initial presentation was most consistent with DKA, however the patient's clinical status acutely changed during evaluation. This demanded a rapid change of plan and broadening of differential diagnosis, ultimately consistent with SSRI overdose.

Diagnosing at breakneck speed: A 2-year-old with fever and neck pain

Alexandra Lorentsen, MD; Nikkan Das, MD; Eric Zwemer, MD

A 2 year-old previously healthy girl presented with one week of fever and neck pain. Three weeks prior, she had fallen off a stool, but did not complain of pain at that time. She initially complained of neck pain while playing and subsequently began turning her whole body to look around. Fevers were daily with Tmax 38.9 C. Review of systems was negative for facial swelling, redness, difficulty breathing, recent URI or GI illnesses, or rash. There was no recent travel or sick contacts. The family has a cat and dog at home.

Vitals were temperature 36.8 C, blood pressure 95/60, pulse 134, respiratory rate 30, and oxygen saturation 100% on room air. She was well-appearing and well-nourished. She had neck stiffness without lymphadenopathy, erythema, or pain on palpation of neck. She had normal tone, 5/5 strength in all extremities, intact sensation and reflexes, and no focal neurologic deficits.

Labs were notable for WBC 17.7, ESR 15, and CRP 43.5 (normal <0.5). CSF showed 2 WBCs and normal glucose/protein, with negative blood and CSF cultures. Infectious workup was negative for mononucleosis, CMV, Flu/RSV, Ehrlichia, and Rickettsia. CT was read as normal. MRI spine showed a small joint effusion of the left C3-4 facet with enhancement extending into the posterior left epidural space consistent with septic arthritis with epidural phlegmon.

The general pediatrician should be familiar with causes of fever and neck pain in children to facilitate early diagnosis. The differential diagnosis typically includes soft tissue abscess, meningitis, retropharyngeal/peritonsillar abscess, reactive lymphadenitis, and Bartonella. This patient's presentation was inconsistent with the above diagnoses given her normal CSF studies, absence of lymphadenopathy, and CT without evidence of abscess.

Septic arthritis is relatively common in children, usually presenting with fever, joint pain, swelling, and limited range of motion. The most common joints affected are the knee and hip. Cervical facet joint septic arthritis is rare, with only a few case reports in the literature, but can lead to neurologic impairment if not diagnosed and treated. Surgical intervention for drainage is risky in cervical spine septic arthritis. The patient was treated with 6 weeks of IV Ceftriaxone and Vancomycin with full recovery and no lasting deficits to date.

Not exactly a Petit(e) Hernia: An infant with abdominal distention
Melissa K. Moore, MD; Ashley G. Sutton, MD

Initial History/Presentation
A term male infant presented at 2 hours of life with a large right-sided abdominal mass following SVD. The prenatal history was unremarkable. He required no resuscitation in the delivery room. Approximately 2 hours after delivery, the patient developed an enlarging right-sided abdominal mass. Notable for a well-appearing term infant in no distress, III/VI systolic murmur at left lower sternal border, femoral pulses 2+. Abdominal exam showed a mildly tender, large mass on right side of abdomen extending around the patient's back past the midline; with bowel sounds present in the abdominal mass (Image 1). Testicles descended with easily reducible bilateral inguinal hernias. Anus patent.

Diagnostic Evaluation
CBC was normal. Plain radiographs did not show rib or vertebral abnormalities, though large protruding mass with bowel loops was noted (Image 2). An abdominal ultrasound demonstrated peristalsing bowel loops just deep to the skin surface. A lumbar triangle hernia through a 1.8 cm wall defect along the right flank was present, containing multiple non-distended bowel loops, and extending around the lateral superficial abdominal wall to the infra-umbilical region. Midline hernia also noted with intermittent protrusion of the left hepatic lobe. Also noted was P2 urinary tract dilatation with concern for right ureter obstruction. MRI abdomen/pelvis confirmed the findings (Image 3). Echocardiogram showed a PDA.

Discussion/Conclusion
Congenital lumbar hernia is rare with case reports identifying fewer than 60 cases reported in the literature. There are many possible locations for defects. The two most common are described by eponyms. The Grynfelt-Lesshalt hernia is a protrusion through the superior lumbar triangle as bordered by the 12th rib, internal oblique and quadratus lumborum muscles. The Petit hernia is defined by the iliac crest, the external oblique and the latissimus dorsi muscles. The median age of diagnosis is 3 months. Many hernias are described in the setting of lumbo-costovertebral syndrome with associated rib, spine and muscular anomalies. Other associated anomalies reported include anorectal malformations and heart defects. The majority of these hernias are repaired with open surgery without recurrence.This case is represents an unusual cause of an abdominal mass in a neonate and emphasizes as the importance of looking for other anomalies with congenital lumbar hernias.

Life or Limb: Compartment Syndrome as Presentation for Vitamin K Deficiency

Authors: Patrick O'Shea MD MBA, Mousumee Shah MD, Rebecca Smith, MD, Daniel Lercher, MD, Brendan Kleiboer, MD

Introduction:
Severe vitamin K deficiency (VKD) classically presents as hemorrhagic disease of the newborn or in cystic fibrosis patients through pancreatic insufficiency. Less frequently, it occurs in older children due to dietary deficiency or malabsorption from anatomic or physiologic changes that impair vitamin K absorption.

Case:
We report a case of a 10-year-old girl with a remote history of jejunal atresia status post jejunal and partial ileal resection and central line associated thromboses with a negative thrombophilia work up in infancy, who presented with lower extremity pain and epistaxis. After numerous ED visits for those complaints, she was diagnosed with deep vein thrombosis (DVT) and normocytic anemia. Soon after admission, she had an acute hemoglobin drop from 11.9 to 6.1 g/dL. She subsequently developed signs and symptoms consistent with compartment syndrome in her lower extremities and was transferred to the pediatric intensive care unit. Workup revealed profound coagulopathy with prothrombin time (PT) 282, international normalized ratio (INR) 27, partial thromboplastin time (PTT) 161, with deficiencies in factors II, VII, IX, X, and S. Factor V and mixing studies were normal. Protein C levels were insufficient quantity for testing. Her severe coagulopathy rendered her unsafe for immediate surgical intervention. Fresh frozen plasma, vitamin K, and prothrombin complex concentrate corrected the coagulopathy, allowing for emergent fasciotomies. Further investigation revealed deficiencies in vitamins A, D, and E. She recovered well with preserved function of her extremities.

Discussion:
There is sparse literature regarding VKD due to malabsorption in the setting of short gut syndrome. Our patient's deficiencies in fat-soluble vitamins suggest an acquired problem of absorption stemming from her history of jejunal and ileal resection. This case highlights the potential for life-threatening complications related to acquired vitamin K deficiency. It also serves as a reminder that patients with severe VKD are at risk not only for bleeding, but also development of thrombosis because of concomitant protein S and protein C deficiency.

Congenital Mesoblastic Nephroma Complicated by PTHrP-Mediated Severe Hypercalcemia

Amira Ramadan, MD, MSc, Derek Hoerres, MD, Jennifer Law, MD, MSCR

Background:
Congenital Mesoblastic Nephroma (CMN) is the most common nonencapsulated solid renal tumor in the neonatal period. It comprises 3-10% of all pediatric renal tumors and typically occurs in the first 6 months of life. Clinical presentation ranges from asymptomatic abdominal mass to paraneoplastic syndrome with hypercalcemia or hypertension. Management of hypercalcemia in a premature infant is rarely described.

Objective:
To present a case of the management of PTHrP-mediated hypercalcemia secondary to congenital mesoblastic nephroma in a premature infant.

Design/Methods:
This case report was developed from chart review.

Results:
A male neonate born at 29 weeks' gestation was found postnatally to have a large abdominal mass along with Ca of 13.8 mg/dl (reference range 7.6-10.4); ionized calcium 7.2 mg/dl (4.4–5.4); Phosphorus 4.5 mg/dl (4.3-10.5); 25-OH-vitamin D 34.7 ng/ml (20.0-80.0), PTH 4.1 pg/ml (12.0–72.0), PTHrP at 4.8 pmol/L (<2.0). EKG was normal. Immediate surgical resection of the renal mass was not possible due to the patient's low weight (1200 g). Hypercalcemia management was challenged by his prematurity and lack of literature to guide any particular treatment strategy. Lasix and hydration failed to manage his calcium which reached 15.7 mg/dl in day of life 5. We restricted his calcium intake while remaining mindful of the risk of provoking or aggravating osteopenia of prematurity. We initiated calcitonin 4 units/kg every 6 hours on day of life 7. Calcium decreased to 11.7 mg/dl on the following day. We anticipated tachyphylaxis of calcitonin but patient's calcium level remained 11.2-12.1 mg/dl. Calcitonin was then discontinued on day of life 24 when calcium dropped to 8.4 mg/dl with improving PTH and PTHrP levels, 59.6 pg/ml and 1.6 pmol/L respectively. He underwent right nephrectomy and ureterectomy at the age of 6 weeks. Correction of his calcium level preoperatively suggested the possibility that the tumor demonstrated a significant degree of regression; however ultrasound demonstrated increase in size of the mass (6.6 x 5.4 x 5.0 cm from 6.1 x 4.5 x 4.2 cm). Postoperative labs revealed normal Ca, PTH, and Phosphorus. Calcium levels remained normal (10.4 mg/dl) two months post surgically.

Conclusion(s):
Though it is unclear why this patient's PTHrP levels eventually normalized, CMN is a renal tumor that may be associated with PTHrP-mediated hypercalcemia, which may be successfully managed by calcitonin in a premature infant.

Avoiding the temptation to blame it on constipation: A 13-year-old boy with left-sided abdominal pain
Derica Sams, MD; Eric Zwemer, MD

Case presentation: A 13-year-old male presented with 3 days of abdominal pain. Pain was intermittent, sharp in character and in the left upper and lower quadrants. Bowel movements had been less frequent and associated with straining. He was seen in the ER twice, received enemas and was discharged with diagnosis of constipation. PMHx was notable for 1 year of episodic LUQ pain associated with fluid intake. He denied any current or prior dysuria or hematuria. Since discharge from the ER, he had Miralax as prescribed. His abdominal pain worsened, and he had 10 episodes of emesis. He was seen by his pediatrician who recommended admission for dehydration and constipation.

On admission, his abdominal exam showed severe tenderness of the LUQ > LLQ, left CVA and hypoactive bowel sounds. Labs showed a benign urinalysis, BUN 13 mg/dL and creatinine of 0.9 mg/dL. Abdominal x-ray demonstrated LUQ soft tissue density and minimal stool burden. Renal ultrasound showed severely enlarged left kidney with urinary tract dilation and loss of corticomedullary differentiation. Right kidney was enlarged with normal architecture. Non- contrast CT confirmed renal anomalies but showed no nephrolithiasis or hydroureter. He was diagnosed with left ureteropelvic junction (UPJ) obstruction and compensatory right renal hypertrophy. He was discharged after stenting of his left kidney. Mag-3 scan at follow-up demonstrated reduced function of the left kidney, and pyeloplasty was performed with discovery of a crossing vessel extrinsically compressing the ureter.

Discussion: UPJ obstruction is a common cause of obstructive uropathy in children, particularly males. Etiology involves either congenital intrinsic narrowing of the ureteral entry or extrinsic compression of the ureter from aberrant vessels. Most cases are detected during postnatal imaging for antenatally detected hydronephrosis. Older children may present with intermittent flank or abdominal pain (known as Dietl's crises), often with nausea and vomiting. Delays in diagnosis are common due to intermittent symptoms and treatment of other causes of abdominal pain (e.g, constipation). In a case series of 8 pediatric patients, all 8 were misdiagnosed and had symptoms for at least 1 year prior to correct diagnosis. A key feature is worsening of pain with intake of substances that induce diuresis. If UPJ obstruction is suspected, renal ultrasound should be performed.

Conclusion: UPJ obstruction should be considered in any child with intermittent abdominal pain, even in the absence of flank pain or urinary symptoms. In particular, pain that worsens with brisk diuresis should raise suspicion.

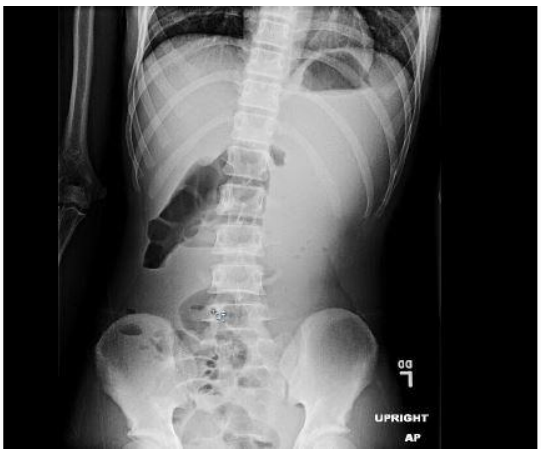

Image 1: XR Abdomen

Nonspecific bowel gas pattern, with prominent loops of gas-filled colon. Developing obstruction is a consideration. Apparent soft tissue density within the left hemiabdomen.

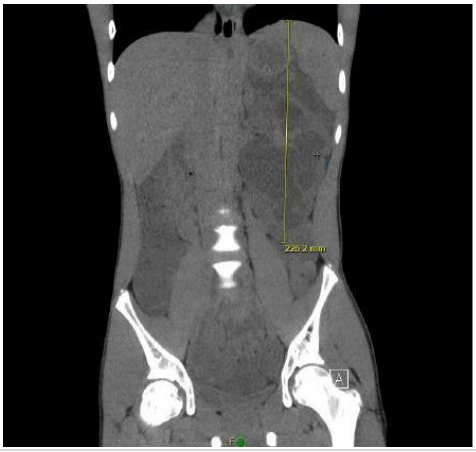

Image 2: CT Abdomen Pelvis Wo Contrast

Severe left-sided hydronephrosis with associated cortical thinning. UPJ obstruction is suspected.

Enlarged, otherwise unremarkable right kidney.

Small to moderate volume free fluid is seen in the left upper quadrant, likely from forniceal rupture.

No evidence of bladder, ureteral, or kidney stone.

Severe Respiratory Failure in a Patient with Hunter Syndrome

Sara Sanders, MD, Mousumee Shah, MD, Terry Noah, MD, Robert Wood, MD, PhD, Michael Rutter, MD, Trista Reid, MD, MPH, Tina Schade Willis, MD, Paul Shea, MD, Joseph Muenzer, MD, PhD

Case Report:
Hunter syndrome (MPS II) is a rare genetic disorder with accumulation of glycosaminoglycan resulting in short stature, hepatosplenomegaly, joint stiffness, cardiac valvular disease and airway involvement including macroglossia, tracheal deformities, laryngomalacia, and obstructive sleep apnea.

Description:
A 19-year-old man with attenuated MPS II and known critical airway, but no previous illnesses, presented with respiratory distress secondary to influenza and presumed status asthmaticus. He was treated with maximal asthma therapies which did not provide much benefit. Given his extremely difficult airway and the high likelihood of cardiac arrest with induction, the decision was made to place ECMO sheaths preemptively then fiberoptically intubate him in a controlled setting in the operating room with only local anesthesia. He tolerated the procedures well, however, he continued to have worsening ventilation and hemodynamic instability. Inhaled anesthesia was attempted but unsuccessful so he was cannulated to veno-venous ECMO. He successfully separated from ECMO after 10 days, but required high ventilatory pressures to prevent airway collapse. A tracheostomy attempt was unsuccessful as his cricoid was substernal and unable to be mobilized. Bronchoscopy revealed nearly 95% collapse of the tracheal lumen. After four weeks of no progress, he was transferred to Cincinnati Children's Hospital where he underwent a partial manubriectomy and tracheostomy. Due to significant collapse of the distal trachea and bronchi, the tracheostomy tube was positioned at the carina and two left bronchial stents were employed. He was weaned from the ventilator, transferred back two months later, and eventually discharged home with intact neurologic function.

Discussion:
Our patient had severe airway abnormalities that were not previously clinically significant until this acute illness triggered airway decompensation. Patients with MPS II in respiratory distress should be presumed to have severe airway anomalies until proven otherwise, and many cannot be intubated by conventional means. Earlier interventions including intubation and even ECMO should be anticipated as in our patient, whose care required not only multidisciplinary but multi-institutional coordination.

Vitamin K Deficiency Compartmentalized
Sean Shannon, MD; Jennifer Vincent, MD

Patients with short bowel syndrome are at significant risk for various nutritional deficiencies based on the extent and specific segments of absent bowel. We present a case of a 10-year-old female with a history of congenital jejunal atresia and short bowel syndrome who initially presented with several days of bilateral calf pain. She then developed acute swelling of her lower extremities after a minor fall. Studies revealed left lower extremity deep vein thromboses, followed by an acute, severe drop in her hemoglobin with worsening pain and leg swelling. This ultimately led to left lower extremity compartment syndrome. Further evaluation showed extreme prolongation in clotting times that resolved with mixing studies, along with multiple factor deficiencies, confirming a diagnosis of Vitamin K deficiency. Initial DVT may have been related to low levels of protein C and protein S, also vitamin K-dependent anti-coagulant factors.

She was treated with emergent infusion of FFP and prothrombin complex concentrate with normalization of coagulation parameters prior to operative intervention for compartment syndrome. Following stabilization of coagulopathy and bleeding, she was started on IV Vitamin K and therapeutic heparin. Other nutritional deficiencies were also identified. She did well following her fasciotomy and was finally discharged on vitamin supplements and low molecular weight heparin. This case highlights the challenges of adequate nutrition in patients with short bowel syndrome and the risks of severe bleeding, hypercoagulability, and corresponding complications in those with Vitamin K deficiency.

Don't Get Rusty on the Basics: Complications of Severe Iron Deficiency Anemia

Bobby Slater, Dan Lercher

Presentation

MM is a 12-year-old premenstrual girl with a remote history of ileal resection secondary to necrotizing enterocolitis who presented after a fall without loss of consciousness. She had fatigue, bilateral leg pain, and palpitations for two weeks. She denied hematemesis, hematochezia, melena, easy bleeding or bruising. She lived with 6 people, including her mother, who had mental health and substance abuse problems along with a history of incarceration. Patient ate school-provided meals, but denied eating at home, milk, or vegetables. She did not have health insurance or receive food stamps.

Physical Exam

Vitals upon admission were T 36.3, HR 116, RR 25, BP 113/76 and SpO2 100%. She appeared thin and pale. Exam revealed tachypnea with significant work of breathing. Breath sounds were diminished at the lung bases. Capillary refill was prolonged with cool extremities. No organomegaly.

Evaluation

Hgb 1.4 g/dl, MCV 41, platelets 947 109/L and reticulocytes 2.5%. Serum sodium 130 mmol/L, albumin 3 g/dL, AST 640 U/L and ALT 137 U/L. Her anemia workup revealed iron level 15 ug/dL, TIBC 365 mg/dL, transferrin 289 mg/dL, iron saturation 4% and LDH 4000 U/L. With the exception of low Factor VII, she had normal factor levels, nutrient labs and coagulation studies. Peripheral smear was unremarkable. Her CT abdomen and CXR were notable only for cardiomegaly. Cardiac studies revealed a pro-BNP 4970 pg/mL and troponin 1 ng/mL. Echocardiogram revealed moderately diminished ejection fraction, enlarged chambers and a pericardial effusion without tamponade. Infectious workup including viral studies were all negative.

Diagnosis

Congestive Heart Failure (CHF) secondary to Severe Iron Deficiency Anemia (IDA)

Discussion

IDA is the most common nutritional anemia, particularly among the most vulnerable members of society. Food insecurity can double the risk of IDA. IDA progresses to true anemia along a continuum and the effects can include poor cardiomyocyte function, oxygen delivery and oxygen extraction leading to CHF. This CHF is generally correctable with iron supplementation and slow administration of blood products. IDA can be dismissed as commonplace, but it is a reminder of the dire consequences of the negligence of the social determinants of health. MM’s heart failure was acutely managed with milrinone and diuresis with administration of packed red blood cells and iron supplementation. She continues to improve in an outpatient setting, but limited resources remain a significant problem.

Vaccinate against SVT: in-utero SVT secondary to transplacental influenza
Bobby Slater, Erin Finn, Jennifer Whitham

Presentation
On a routine 25-week prenatal ultrasound, a fetal male was noted to be tachycardic to 200 beats per minute (BPM). Pregnancy was normal with no complications until mother recently developed influenza. She was being treated with Oseltamivir at the time of the exam.

Physical Exam
Initial newborn vital signs included T 36.5 C, HR 138 bpm, RR 64, BP 53/21, SpO2 99%, weight 3045 g, and Apgars 8 and 9. Term, appropriate for gestational age, well-appearing male with a normal exam and no evidence of tachycardia or fetal hydrops.

Evaluation
Initial echocardiogram at 25 weeks gestation revealed HR 200bpm with supraventricular tachycardia (SVT) and small pericardial effusion. Mother was started on Digoxin and Fleicanide initially with discontinuation of Digoxin after conversion to sinus rhythm and resolution of the pericardial effusion in the fetus. The infant developed SVT by one week of life and atenolol was initiated for rate control. He had a patent foramen ovale and small pericardial effusion remained.

Diagnosis
Persistent SVT secondary to transplacental influenza

Discussion
Fetal tachycardia is present in 1% of all pregnancies, though fewer than 10% are attributed to arrhythmias. Up to 90% are due to SVT without an underlying cardiac defect. It is diagnosed with fetal echocardiogram on m-mode. The mortality rate of prenatal SVT is between 6-8.9% due to complications such as non-immune fetal hydrops, defined by 2 or more fluid collections in fetal compartments. This can become resistant to therapy and leads to worse neurologic outcomes and increased risk for prematurity. Influenza is a common illness that causes fevers and respiratory symptoms, though can cause more systemic illness and cardiac disease. Studies show that about 50% of adults with the flu exhibit ECG changes, though these are transient and usually insignificant. A small percentage develops reversible cardiac dysfunction. Transplacental transmission of has been associated with neonatal SVT previously. Antiarrhythmic agents are the mainstay of therapy with Digoxin and Fleicainide recommended as first and second line agents, respectively. Most fetal SVT is survivable, but recurs in 50-82% ex-utero. This typically resolves by one year of life, but persists in 20%. This case is a classic presentation of fetal SVT, with the twist of transplacental influenza as the likely cause. It is important to inquire about vaccination status of mothers and to be aware that SVT can recur.

Fluff it up first: Undifferentiated hypotension in an 11-year-old girl
Bobby Slater, Hillary Spangler, Mark Piehl

Presentation
An 11-year-old healthy girl presented from her pediatrician's office with fever of 106 F, cough and epigastric pain that had migrated to her left flank over the past day. She endorsed similar pain in the past, and currently had palpitations and pleurisy. She denied change in pain with position. No recent history of emesis or diarrhea.

Physical Exam
On arrival to the ED, her vital signs were T 100.8, HR 137, RR 22, BP 83/44 and normal oxygen saturation. She was initially well-appearing, conversant and ambulating, with faint wheezing in her left upper lobe and diminished aeration at the right base, and taking shallow breaths. No abdominal tenderness or reproducible chest pain.

Evaluation
A CXR and renal ultrasound were normal. Her white blood cell count was 22,000 and urinalysis revealed moderate ketosis. Over three hours she received two 20 ml/kg boluses for hypotension, with transient improvement. She was treated with empiric Ceftriaxone for presumed sepsis and admitted. She again became hypotensive to 73/35, was unresponsive, and developed O2 desaturation to 84% prompting the addition of high flow nasal cannula oxygen. Bedside ultrasound demonstrated a left lower lobe pneumonia with small effusion, normal heart function, and complete inspiratory collapse of the IVC. A 500 ml fluid bolus was pushed rapidly and she briefly improved before recurrence of hypotension while US showed minimal IVC collapse, prompting a norepinephrine infusion. Repeat CXR demonstrated a left lower lobe pneumonia. Within 12 hours, she was on room air and off norepinephrine within 72 hours.

Diagnosis
Decompensated septic shock secondary to pneumonia

Discussion
Point of care ultrasound "POCUS" is becoming an important adjunct to the clinical exam. In our case, POCUS detected the pneumonia and persistent hypovolemia prompting additional fluid and subsequent initiation of a vasopressor while monitoring for volume overload. Undifferentiated hypotension has mortality rates between 5-25% and doubles with each delayed hour of care. Identifying septic shock is particularly important, as each delayed intervention leads to an increase in mortality by 8%. POCUS is particularly helpful by observing IVC respirophasic collapsibility and preventing over-resuscitation. Pneumonia is the leading cause of non-neonatal pediatric death worldwide and can be missed on radiography prior to fluid resuscitation. This case highlights the importance of early shock recognition and intervention as well as using a Bayesian approach when tests are negative.

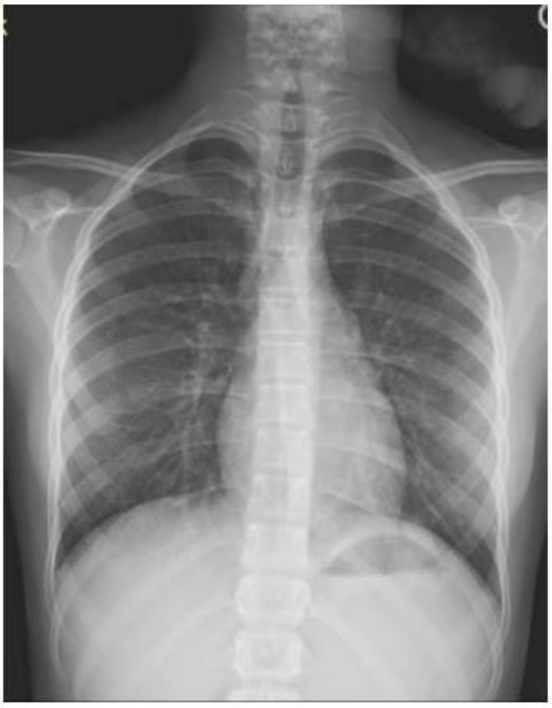

Figure 1: Initial chest x-ray without evidence of consolidations

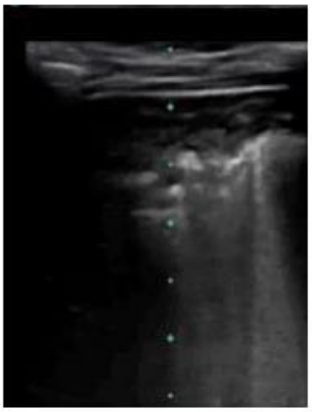

Figure 2: Ultrasound of left lower lobe of lung showing subpleural consolidations and B-lines

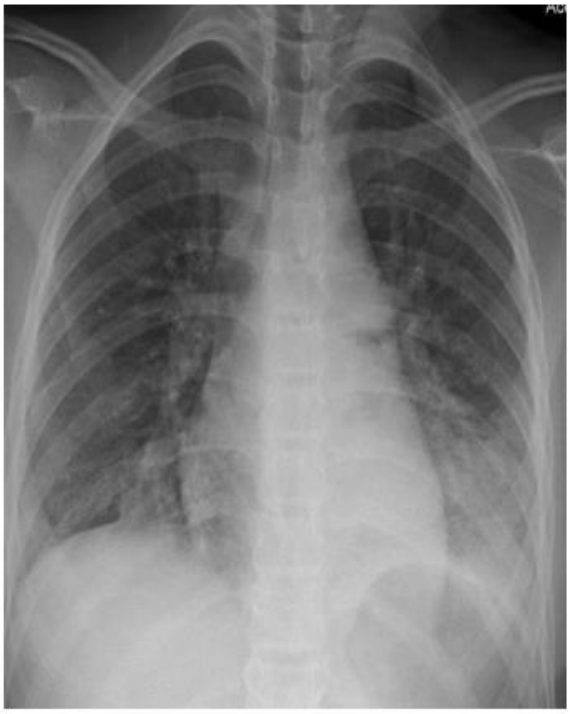

Figure 3: Follow-up chest x-ray showing evidence of left lower lobe consolidation

Slicing back on calories: A 1-month-old girl with an unusual cause of failure to thrive

Bobby Slater, Hillary Spanger, Nikkan Das

Presentation
A 1-month-old ex-term infant presented to the hospital with failure to thrive (FTT). She was born at the 3rd percentile for weight, with admission weight at the <1st percentile. Her pediatrician had trialed ranitidine for reflux, formula changes, and fortification of feeds with persistent weight loss. Review of systems was notable for heavy breathing while feeding, recent oral aversion, and increased sleepiness. Urine and stool output were appropriate for age. She had no recent fevers or illnesses.

Physical Exam
Vital signs on admission were T 36.9, HR 180, RR 56, BP 93/57, saturating appropriately on room air. Patient had minimal subcutaneous fat but no dysmorphic features. Cardiac exam with II/VI systolic outflow murmur loudest at lower left sternal border. Pulmonary exam with bibasilar coarse crackles-louder on the right than left-and mild subcostal retractions. No appreciable hepatomegaly. Tone and strength were appropriate for age.

Diagnostic Evaluation
Differential diagnosis on admission included non-organic FTT, metabolic disorder, reflux and anatomic malformation. Upper GI series was normal. CXR showed diffuse right lung field opacities that prompted an echocardiogram and CT scan. Echocardiogram revealed endocardial cushion defects, right pulmonary artery stenosis, dextrocardia, and possible partial anomalous pulmonary venous return (PAPVR). CTA demonstrated PAPVR to the IVC and atypical ground glass opacity more severe on the right than the left. Further workup for weight loss was abridged by her cardiopulmonary findings. Metabolic work-up was deferred based on normoglycemia and normal hepatic and thyroid function on admission.

Diagnosis
Scimitar Syndrome (SS) with Ventricular Septal Defect (VSD)

Discussion/Conclusion
SS should be considered when a patient presents with asymmetric pulmonary opacities and FTT, as SS is an unusual variant of the rare congenital heart disease PAPVR that often presents with FTT. It is classically associated with at least one pulmonary vein that drains from the right middle or lower lobe into the IVC that resembles a Turkish sword, or scimitar. The infantile presentation of Scimitar syndrome is associated with endocardial cushion defects, while recurrent pulmonary infections are typically observed in older patients. Prognosis is often worse in infants and patients with a VSD due to associated heart failure symptomology and required defect repair.

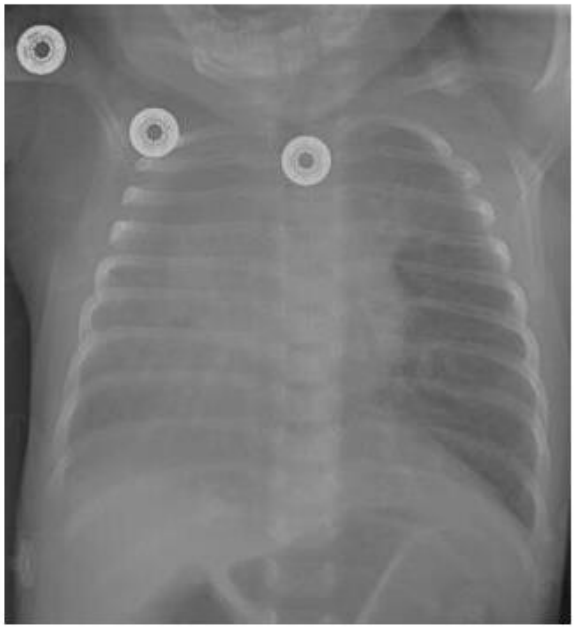

Figure 1: Chest X-ray of patient upon admission demonstrating diffuse right lung field opacification.

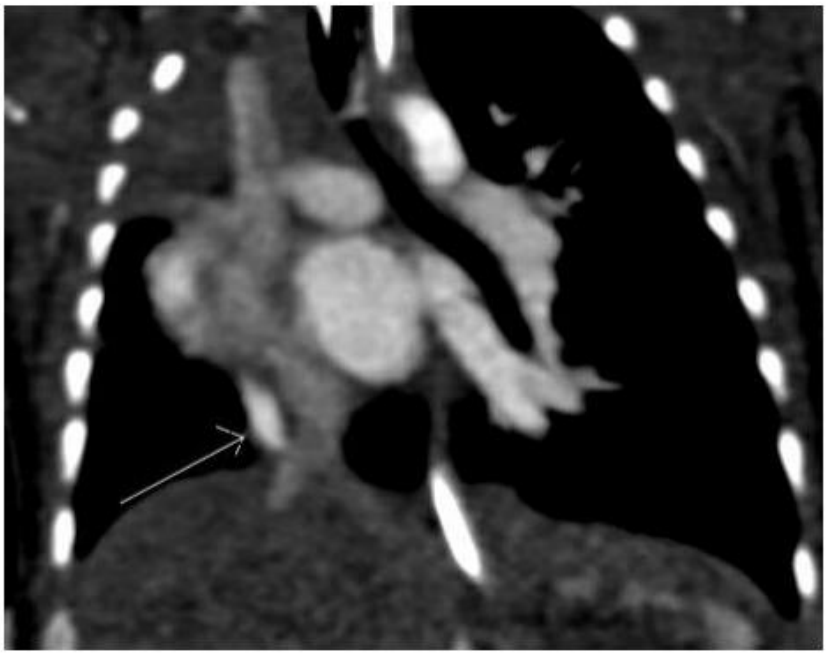

Figure 2: CTA of patient that diagnosed scimitar syndrome. Arrow demonstrating the pathognomonic finding. Also note the dextrocardia secondary to lung hypoplasia.

A short, scary nap; 4 month old presented with reduced consciousness and hyperglycemia
Bobby Slater, Matt Waters, Suresh Nagappan

Presentation
A previously healthy, full-term and fully-immunized boy presented with depressed level of consciousness. He had previously been in his normal state of health but was unusually fussy that morning before mom left him and returned to find him being unresponsive with perioral secretions. Mom denies any recent head trauma, sick contacts, fevers, or shaking.

Physical Exam
Afebrile, tachycardic, obtunded, without evidence of head trauma, pupils equal and reactive, normal cardiac exam with good peripheral pulses and perfusion, and a nonfocal neurologic exam. Within 2 hours of his initial presentation he was afebrile, awake, alert, and well-appearing.

Evaluation
Hyperglycemic to 467 with a lactate of 6.23, both which normalized within hours. He had a head MRI which showed branch occlusion in his left posterior cerebral artery with subsequent posterior watershed anoxia. This prompted a stroke workup including a normal ECG, echocardiogram and negative thrombophilic workup.

Diagnosis
Cryptogenic posterior cerebral artery stroke.

Discussion
Acute ischemic strokes (AIS) is rarely on the differential for altered mental status despite being among the top 10 causes of death in children and having an incidence between 2 to 13 per 100,000 children. Its mortality rate is between 3-15%, neurologic deficits between 35-74%, recurs in 10-30%. It commonly presents in infant males with depressed level of consciousness, apnea or seizures. This presentation leads to delayed diagnoses, but this largely does not change outcomes as acute interventions (such as TPA) are not currently recommended. Diagnosis should consist of a diffusion weighted MRI. AIS is commonly caused by non-atherosclerotic arteriopathies secondary to inflammation from infections, particularly varicella, or even mild trauma. Rarer causes, including MELAS or Sickle Cell may also be seen. Current best practice often includes daily aspirin but there is no clear evidence-based benefit. Long-term follow-up with developmental surveillance is vital as many of these children develop intellectual and emotional deficits as well as seizures. Overall, it is important to keep AIS in mind on the differential of a child with depressed level of consciousness, do a workup for arteriopathy and cardiac causes, weigh the consequences of antithrombotic or anticoagulant therapy and have frequent long-term follow-up.

That's not the "rule of twos": A case of Meckel's diverticulum in an adolescent

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Internal Medicine-Pediatrics, UNC Hospitals[1]; Emergency Medicine, UNC Hospitals[2]; UNC School of Medicine, WakeMed Hospital[3]

Initial History/Presentation:
A 13-year-old male presented to the emergency department with syncope in the setting of painless hematochezia during gym class. He later experienced three large, painless, bright-red bloody bowel movements and a syncopal episode. He denied any associated abdominal pain, emesis, fever, dyspnea, or chest pain. No personal history or family history of coagulopathy or inflammatory bowel disease.

Physical Exam:
On arrival to the emergency department, he was pale with dry mucous membranes. Vital signs were significant for blood pressure 71/41, respiratory rate 24 breaths/minute, heart rate 88 beats/minute, temperature 36.9 degrees Celsius. On physical exam, his abdomen was non-tender, non-distended, and without rebound or guarding. He was noted to have frank, bright-red blood on rectal exam.

Diagnostic Evaluation:
Admission hemoglobin and hematocrit were 8.4 g/dL and 24%, respectively, but due to a previous undetectable hemoglobin and hematocrit on a point of care venous blood gas, the patient received two liters of normal saline, four units of universally compatible O Rh- emergency release blood, one unit of fresh frozen plasma, and one unit of platelets. Repeat blood pressure was 106/51 and hemoglobin and hematocrit were 12.1 g/dL and 35%, respectively.

Initial abdominal computed tomography was unremarkable. He subsequently underwent a nuclear medicine Meckel's diverticulum scan, revealing an abnormal foci of radiotracer in the right lower quadrant consistent with ectopic gastric mucosa, suggestive of Meckel's diverticulum. He underwent a successful surgical resection of his Meckel's diverticulum and was discharged on hospital day two as he was tolerating oral intake, without hematochezia, and with stable hemoglobin and hematocrit levels.

Diagnosis:
Meckel's diverticulum in an adolescent

Discussion:
This case study is unique as acute gastrointestinal hemorrhage secondary to Meckel's diverticulum is atypical in the adolescent.[1] Meckel's diverticulum is classically described as following the "rule of twos": two percent of the population, two inches in length, two feet from the ileocecal valve, 2:1 male to female ratio, and in patients near two years of age. Therefore, even with initially negative imaging (e.g., CT), providers need to have a high suspicion for Meckel's diverticulum in all pediatric patients, as unexplained lower GI bleeding in is one of the leading symptom of Meckel's diverticulum in pediatric patients older than two years of age.[2,3,4] Therefore, even with a negative Meckel's scan, an exploratory laparotomy should be considered in adolescent with painless GI bleeding.

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Persistent Fever in the Neonate: When to Consider Zebras.
Erin Steinbach, MD, PhD; Paul Googe, MD; Eve Wu, MD; Raphaela Goldbach-Mansky, MD, MHS; Tim Moran, MD, PhD

A female African-American neonate born to non-consanguineous parents via uncomplicated repeat cesarean delivery presented at three weeks of life with fevers and rash. She subsequently developed elevated C-reactive protein. An extensive evaluation for infection, neoplasm, or rheumatologic disorder was negative. Initial skin biopsy was non-diagnostic. Ultimately, the fever and nodules improved with oral corticosteroids, prompting evaluation for an autoinflammatory disorder. Blood transcriptional analysis demonstrated markedly elevated expression of interferon response genes, and second biopsy of a skin nodule revealed a neutrophilic panniculitis. Whole exome sequencing identified novel mutations in the proteasome genes *PSMC5* and *PSMD11*; this with the clinical history is consistent with a diagnosis of *C*hronic *A*typical *N*eutrophilic *D*ermatosis with *L*ipodystrophy and *E*levated temperature (*CANDLE*) syndrome. The proteasome is a complex of multiple protein subunits that recognizes and targets ubiquitinated proteins for degradation. Proteasome dysfunction induces a cell stress response and production of type I interferons; a consequence is upregulation of inducible proteasomes. Patients with proteasome-associated autoinflammatory syndromes like CANDLE cannot overcome this cell stress response. Attempts to wean oral corticosteroids have been unsuccessful, and treatment with the JAK1/2 inhibitor baricitinib was started. Providers should consider interferonopathies, including CANDLE syndrome, in infants presenting with daily fevers and inflammatory skin lesions. Early in the disease course, the typical skin lesion histopathologic features may not be present, necessitating repeat biopsy for diagnosis. Care ultimately requires the coordination of multiple disciplines, both due to the manifestations that develop from chronic inflammation and from long-term use of systemic corticosteroids.

Ocular Involvement and Effective Use of Tacrolimus in Neutrophilic Dermatoses

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Case Presentation: RP is a 15-year-old female with juvenile idiopathic arthritis (JIA) treated with methotrexate and tocilizumab, who presented with two weeks of worsening joint pain and swelling and new rash. Her rash consisted of diffuse, indurated papules and plaques, a painless, violaceous nodule of the left eyelid, and right eye conjunctivitis. Biopsy of a leg lesion demonstrated "intense superficial and deep dermal perivascular and interstitial infiltrate of neutrophils" consistent with Sweet's syndrome. She was started on a prednisone taper and dapsone with improvement, but 6 months later had a persistent leg ulcer related to recalcitrant Sweet's syndrome. AV is a 14-year-old female with a history of JIA. Since age 5 years, she also had a history of presumed septic arthritis, cutaneous abscesses, and preseptal cellulitis all refractory to antimicrobials. Biopsy of a cutaneous lesion demonstrated "dense neutrophils in the subcutaneous fat" consistent with a neutrophilic dermatosis. She was ultimately diagnosed with genetically-confirmed PAPA syndrome (pyogenic arthritis, pyoderma gangrenosum, cystic acne). She was started on adalimumab therapy with topical clobetasol and dapsone and had some, but not complete, disease control. Due to treatment-refractory manifestations of neutrophilic dermatosis, RP and AV were ultimately started on adjunctive oral tacrolimus therapy with marked improvement. Specifically, both patients saw a drastic reduction in the development of lesions and improvement in ocular symptoms. In addition, oral tacrolimus was used safely in combination with other immunosuppressants.

Discussion: Ocular involvement in neutrophilic dermatoses is atypical and often a diagnostic challenge. Ocular manifestations are variable and include conjunctivitis, uveitis, scleritis, retinal vasculitis, and eyelid involvement. Due to such variable presentations, patients are often misdiagnosed with cellulitis, chalazion, or malignancy, thus delaying appropriate treatment. Delays in diagnosis and therapy increase the risk of poor outcomes and sequelae like vision loss. Ocular involvement is associated with a higher relapse risk and need for aggressive systemic therapy. Due to disease rarity, there is little data regarding optimal treatment of neutrophilic dermatoses, particularly ocular manifestations. There are few reports on the effective use of systemic tacrolimus in neutrophilic dermatosis.

Conclusions: Ocular manifestations of neutrophilic dermatoses like pyoderma gangrenosum and Sweet's syndrome are underrecognized and require a high level of suspicion to minimize delays in diagnosis and treatment and unnecessary exposure to antimicrobials. Oral tacrolimus is not only effective in the treatment of neutrophilic dermatoses, including co-morbid ocular inflammation, but can also be safely used in combination with other immunosuppressants.

Lessons Learned from a Languishing Limp
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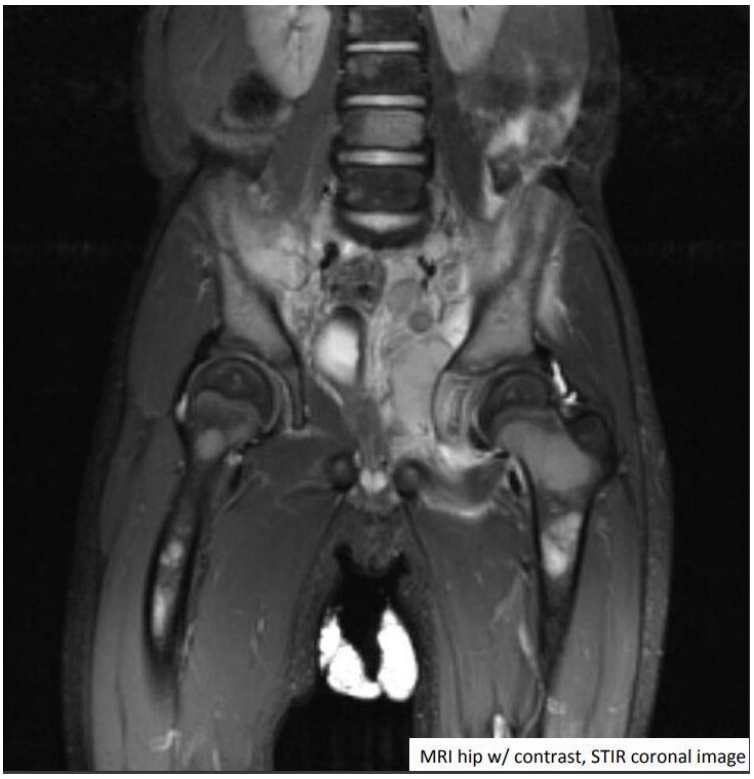
Case: A 3-year-old previously healthy male was transferred from an outside ED with a 2-month history of recurrent limp, left hip pain and fever. He was seen twice before by different providers, prescribed NSAIDs, and pain resolved. His mother sought care again with his new refusal to bear weight. There was no history of trauma, travel, pet exposure or sick contacts. He was living in a shelter. Prior to transfer, x-ray and CT images found left ischial osteomyelitis, obturator internus abscess, joint effusion and inguinal and pelvic sidewall lymph nodes.

He arrived to our hospital well appearing, with normal vital signs. He held his left hip in external rotation and had pain with internal rotation. There was full, painless active and passive range of motion at the left knee and ankle. He would not bear weight.

Initial workup included urgent left hip arthrocentesis, with 2,200 nucleated cells and monocyte predominance, inconsistent with septic arthritis. Initial CRP was elevated to 145.6 mg/L. He had a normal WBC count and slight hypochromic, microcytic anemia. A presumptive diagnosis of osteomyelitis was made considering usual pathogens, in addition to fastidious Kingella kingae and less likely mycobacterium. Following blood culture, antibiotics were started. The next day CRP decreased to 77.1 mg/L. MRI of the left hip and knee were obtained for further workup and to guide surgical debridement. MRI showed a large heterogeneously enhancing lesion of the left acetabulum and ischial tuberosity with periosteal reaction, large soft tissue component extending into the left hemipelvis and diffusely abnormal bone marrow signal intensity of lumbar vertebrae, left pelvis and proximal femur.

MRI impression suggested malignancy: leukemia, lymphoma, Ewing's sarcoma or histiocytosis, with concern for metastatic disease. Bone marrow and lymph node biopsies revealed neuroblastoma.

Discussion: Our patient is a rare presentation of neuroblastoma. Neuroblastoma can originate anywhere along the sympathetic chain, however only 1% originate in the pelvis. Primary neuroblastoma is most commonly in the adrenal glands, followed by non-adrenal abdomen, then thorax and neck. Our patient's metastatic presentation had limited constitutional symptoms. His waxing and waning limp was not typical of osteomyelitis, though initial imaging and down trending CRP on antibiotics supported the diagnosis. Admitting pediatricians must interpret imaging with clinical context and avoid anchoring. Additionally, we recognize socioeconomic challenges and inconsistency of care for this child may have delayed his diagnosis.



MRI hip w/ contrast, STIR coronal image

Invasive Cavitary *Aspergillus* Infection Complicated by ABPA

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Introduction:
Aspergillus can cause respiratory diseases ranging from invasive infection with cavitary lesions to saprophytic or allergic manifestations. We present a case of *Aspergillus* infection with cavitary lesions complicated by allergic bronchopulmonary apergillosis (ABPA).

Case Report:
A 9 year-old male with intermittent asthma presented with a 2-3 month history of persistent cough, 5-pound weight loss, and recent fever. His initial laboratory studies revealed peripheral eosinophilia (21%), and initial chest radiography showed a large cavitary lesion in the right upper lobe, with multiple smaller lesions observed on chest CT.

He was admitted and started on IV antibiotics, with empiric antifungals added on hospital day #2. Immune evaluation revealed normal IgM, IgG, and IgA, but significantly elevated IgE (13,203). Bronchoscopy demonstrated severe bronchitis with elevated eosinophils (26%) and cultures positive for influenza B and *Aspergillus fumigatus*. He was skin test positive for *Aspergillus* antigen. The following studies were negative/normal: sweat chloride test, HIV, PPD, AFB smears, echinococcus antibodies, toxocara antibodies, ACE, ANA, and ANCA. His hospital course was complicated by an increase in the size of his cavitary lesion, though he clinically improved on micafungin and voriconazole.

His immediate post hospitalization course was complicated by a right pneumothorax that required readmission and chest tube placement, with prednisone started to treat ABPA. The pneumothorax resolved, and over the subsequent 8 years he has been maintained on intermittent courses of prednisone or omalizumab with posaconazole as an antifungal. He has had stable lung function and IgE generally <1000. The initial cystic lesion has decreased in size, though other lesions have grown.

Based on history of retained primary teeth, elevated IgE level, eosinophilia, and pneumatocele, the patient scored a 33 on the NIH scoring system for hyper-IgE syndrome (HIES). However, his STAT3 genetic testing was negative for mutations associated with classic HIES. Further investigation did not reveal a specific immunodeficiency.

Discussion:
Aspergillus lung disease has many manifestations, including invasive disease with cavitary lesions and ABPA, which is characterized by hypersensitivity to allergens of *Aspergillus* and elevated IgE. This patient's course was most consistent with a combination of invasive disease and ABPA, and he responded to treatment including corticosteroids, antifungals, and more recently omalizumab. Although invasive *Aspergillus* is typically associated with immunodeficiency, and HIES was suspected, no immune defect was identified.

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