32nd Annual
THOMAS F. BOAT
DAY OF SCHOLARSHIP
April 20th, 2017
The Department of Pediatrics gratefully acknowledges the support for the Evening of Scholarship from the members of the Pediatric Faculty and the Floyd W. Denny Society.
The Thomas F. Boat Evening 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 Evening of Scholarship

The primary intent of the Thomas F. Boat Evening 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 Evening 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 Evening of Scholarship. New for 2012 is the naming of the Best Resident Award 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)

*Reducing the Power of Empiric Acyclovir in Low Risk Neonates Admitted for Sepsis Evaluation*
Lauren Bradford, MD (Pediatrics Resident)
Faculty Sponsor: Ashley Sutton, MD

*“Picture of the Day” Curriculum for Medical Students in the Newborn Nursery*
Ali Rittenberg, MD (Pediatrics Resident)
Faculty Sponsor: Eric Zwemer, MD

*Community Health Worker Case-Detection of Asthma in a Resource-Poor Community in Nicaragua*
Mary Crocker, MD (Pulmonology Fellow)
Faculty Sponsor: Sylvia Becker-Dreps, MD

*A short course of gamma tocopherol mitigates endotoxin-induced inflammation in humans ex vivo*
Allison Burbank, MD (Allergy & Immunology Fellow)
Faculty Sponsor: Michelle Hernandez, MD

12:00pm  Noon Conference Presented by Scott P. Commins, MD, PhD
Curnen Denny Conference Room
Lunch Provided

4:00pm  Formal Poster Session
Dental School Lobby
Refreshments provided

5:15pm  Award Ceremony
Kirkland Auditorium
# List of Abstracts

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Anti CD4 and Anti CD8 Antibody Co-Therapy Induces Remission of Diabetes in New Onset NOD Mice via Localized Induction of T Cell Egress from the Pancreas

MARK HENIN, MD
ENDOCRINOLOGY FELLOW
MENTOR: ROLAND TISCH, PHD

Additional Authors; Division/Institution: Matthew Clark, Ph.D.; UNC Department of Microbiology and Immunology, Post-Doctoral Fellow

Background/Introduction: Previous studies have shown that anti-CD4/anti-CD8 antibody treatment produced indefinite diabetes remission in new onset non-obese diabetic mice. Histologic studies of the mice reveal reduced T cell pancreatic islet infiltration. How the antibodies induce pancreatic T cell egress is unknown. A recently completed study showed that IFN-γ injections caused diabetes recurrence in mice with anti-CD4/anti-CD8 induced remission. It is hypothesized that anti-CD4/anti-CD8 antibody treatment decreases T cell secretion of IFN-γ and IL-2, blunting the local inflammatory response, ultimately facilitating T cell egress.

Methods: Female NOD mice with confirmed new onset diabetes (BG > 250 x2 days) were injected intraperitoneally with rat anti-mouse CD4 and CD8 or rat anti-mouse 2A3 control antibody once daily for two days. Treatment doses were serially diluted from 160 to 0.625μg/mL. Seventy two hours after antibody injection, mice were sacrificed for analyses of sera and organ T cell content. Single-cell suspensions were prepared from tissues. Leukocytes were removed from islets via an enzyme-free dissociation buffer with cytokine levels measured via ELISA and T cell counts calculated via flow cytometry.

Results: Diabetes was reversed in 79% (n= 24) of anti-CD4/anti-CD8 treated NOD mice but persisted in the control group. Both IFN-γ and IL-2 levels and total pancreatic T cell counts decreased exponentially with increasing concentrations of anti-CD4/anti-CD8 antibody while no change was seen in the control group. There was no decrease in splenic T cell counts of the treatment or control groups.

Conclusions: The results are consistent with previous studies showing anti-CD4/anti-CD8 antibody treatment reversing diabetes in new onset NOD mice. Furthermore, anti-CD4/anti-CD8 treatment causes a dose dependent decrease in concentrations of IFN-γ and IL-2 with an exponential decrease of T cell counts in the pancreas. Normal splenic T cell counts indicate the effects are localized and not due to systemic immunosuppression. Further studies to elucidate a physiologic mechanism for the regulation of T cell egress are underway. Currently the studies are focused on the transcription factor FOXO1. It is theorized that FOXO1 moves into the nucleus of unstimulated T cells, causing downregulation of CD69, an early T cell activation cofactor, as well as upregulation of S1P, a membrane protein thought to promote T cell egress from tissue.
SERUM IgE SPECIFIC FOR ALPHA-GAL SUGAR MOIETY CAN BIND GLYCOLIPID
POSTER #2

ONYINYE IWEALA, MD, PHD
ALLERGY/IMMUNOLOGY FELLOW
MENTOR: SCOTT COMMINS, MD, PHD

Additional Authors; Division/Institution:
Onyinye Iweala, MD PhD¹, Patrick J. Brennan, MD PhD², and Scott P. Commins, MD, PhD¹

(1) University of North Carolina, Chapel Hill, NC,
(2) Brigham and Women’s Hospital / Harvard Medical School, Boston, MA

Rationale: Alpha-gal meat allergy, characterized by anaphylaxis to mammalian meats like beef, pork or lamb three or more hours after consumption, has been associated with specific IgE (sIgE) antibodies against the sugar moiety galactose-alpha-1,3-galactose (alpha-gal) lining the surface of non-primate mammalian tissues. Meat fat content appears to impact reaction severity in alpha-gal-allergic patients. Because antigenic lipids are presented in complex with CD1 antigen presenting molecules, we hypothesized that delayed anaphylaxis in alpha-gal allergy may be explained by alpha-gal sIgE binding to glycolipids complexed with CD1, since CD1-based presentation of glycolipids requires additional time for assembly and loading. Because CD1d presents the canonical invariant NK T cell glycolipid agonist alpha-galactosylceramide, a molecule structurally similar to alpha-gal, we measured sIgE-binding to this isoform.

Methods: Sera from alpha-gal allergic subjects containing alpha-gal sIgE (n=5) were incubated with biotinylated human CD1d monomers unloaded or loaded with either alpha-gal containing glycosphingolipid isogloboside 3 (iGb3) or galactose-alpha-1,4-containing globotriaosylceramide (GB3) coupled with streptavidin attached to the solid phase of a sandwich immunoassay.

Results: CD1d monomers loaded with alpha-gal containing iGb3 bound IgE (2.6 ± 0.7 IU/mL), whereas unloaded CD1d did not. Less IgE binding (0.49 ± 0.2 IU/mL) was present in GB3 loaded monomers. In contrast, serum from a subject without alpha-gal sIgE was negative for iGb3 and GB3 binding.

Conclusions: Alpha-gal sIgE from mammalian meat-allergic subjects binds glycolipid complexed with human CD1d and does so with increased specificity to glycolipids containing the alpha-1,3 linkage. Thus, antigen presentation of dietary lipid through CD1 molecules may represent a mechanism of delayed food allergy.
GAMMA TOCOPHERAL INHIBITS OZONE-INDUCED INFLAMMATORY GENE EXPRESSION IN AIRWAY EPITHELIAL CELLS
POSTER #3

CHARITY DURAN, POSTDOCTORAL CANDIDATE
MENTOR: MICHELLE HERNANDEZ, MD

Background/Introduction: Allergic airway inflammation can be further exacerbated by exposure to environmental pollutants such as ozone (O3). Gamma tocopherol (gT), the primary dietary form of Vitamin E, possesses an array of anti-inflammatory properties; because airway epithelial cells are direct targets of O3-induced injury, we sought to investigate the potential anti-inflammatory effects of gT and one of its major metabolites, g-CEHC, on airway epithelial cells using an in vitro O3 exposure model.

Methods: Immortalized human bronchial epithelial cells (16HBEs), were grown at air liquid interface and exposed to 0.4ppm O3 for 4 hours with a 1 hour recovery. Cells were incubated with DMSO or gT overnight, or g-CEHC 1 hour prior to exposure. Fold changes in inflammatory gene expression were quantified using qRT-PCR. Comparisons between cytokine production and gene expression were conducted using two-sample t-tests or Wilcoxon signed-rank tests depending on whether the normality assumption was met.

Results: Treatment with gT significantly decreased O3-induced changes in expression of IL-6 (3.44 ± 1.28 vs. 1.20 ± 1.09) and IL-8 (3.20 ± 1.14 vs. 0.8146 ± 1.041) for DMSO vs gT treatment respectively. G-CEHC also modestly reduced expression of inflammatory genes, although these differences did not reach significance. In contrast, there were no significant changes in production of IL-6 or IL-8 in response to O3 exposure.

Conclusions: This study is the first to demonstrate that gT can abrogate inflammatory gene expression in airway epithelial cells in response to O3 exposure. Epithelial-derived IL-8 is integral for recruitment of neutrophils in response to airway injury, suggesting that gT is a candidate therapeutic for reduction neutrophilic airway inflammation.
GRANULOMATOUS LYMPHOCYTIC INTERSTITIAL LUNG DISEASE (GLILD) IN 22Q11.2 DELETION SYNDROME WITH COMBINED IMMUNE DEFICIENCY (CID)

POSTER #4

AMIKA SOOD, MD; ALLERGY/IMMUNOLOGY FELLOW
MENTOR: EVELINE WU, MD

Additional Authors; Division/Institution: William Funkhouser, MD, PhD (Department of Pathology and Laboratory Medicine, University of North Carolina)
Brian Handly, MD (Department of Radiology, University of North Carolina)

Background/Introduction: Immune dysregulation and autoimmunity are well-known complications of 22q11.2 deletion syndrome (22q11.2DS). GLILD has classically been associated with common variable immune deficiency (CVID) and has not generally been reported in other immunodeficiencies.

Methods: We describe the clinicopathological and radiological features of GLILD based on positron emission tomography (PET), computed tomography (CT), and lung biopsy in an adolescent male with 22q11.2DS and CID.

Results: The patient had 22q11.2DS and CID as defined by low naïve T-cell numbers, decreased lymphoproliferative responses to mitogens and antigens, hypogammaglobulinemia, and suboptimal vaccine responses. He received monthly intravenous immunoglobulin and inhaled pentamidine. Routine cardiac imaging incidentally discovered bulky mediastinal lymphadenopathy, splenomegaly, and pulmonary nodules as confirmed by PET and CT. Scattered groundglass nodular opacities and central bronchiectasis were also seen. Lung biopsy histopathology and immunophenotyping showed patchy unencapsulated lymphohistiocytic nodules containing poorly formed non-necrotizing granulomata with a predominance of T-cells and follicular bronchiolitis. Following exclusion of infectious and neoplastic processes, the patient received rituximab with marked improvement. His GLILD progressed 5 months later, incidentally with repopulation of B-cells. He was re-treated with rituximab followed by mercaptopurine with good response.

Conclusions: We believe this is only the second reported case of GLILD in a patient with 22q11.2DS. GLILD may not be unique to CVID and should be considered as a potential cause of interstitial lung disease in other immunodeficiencies. GLILD is associated with poorer outcomes in CVID. Recognition of GLILD as a rare phenotype of 22q11.2DS and other immunodeficiencies may therefore have important therapeutic and prognostic implications.
MULTIORGAN SYSTEM FAILURE WITH HEMOPTYSIS COMPLICATING ACUTE CHEST SYNDROME
POSTER #5

MATTHEW BRUEHL, MD; PULMONOLOGY FELLOW
MENTOR: TIM VECE, MD

Background/Introduction: Granulomatosis with polyangiitis (GPA) is rare with an annual incidence of less than 1 per 1 million children. Sickle cell disease affects 1 out of every 365 African-American births. This report details the only described case of acute chest syndrome with multiple severe complications leading to a diagnosis of GPA.

Case Description: A 14-year-old African-American young man with a history of sickle cell disease (HbSS) presented to the emergency department with fever, cough, and right-sided chest pain. Initial evaluation revealed tachypnea, decreased air movement on lung auscultation, and a right middle lobe infiltrate on chest radiograph, leading to a diagnosis of acute chest syndrome. He was admitted to the hospital and treated initially with intravenous antibiotics, fluids, analgesics, and a red blood cell transfusion.

He subsequently developed hepatomegaly, coagulopathy, worsening bilateral pleural effusions, and acute kidney injury. He developed frank hemoptysis that initially resolved with correction of his coagulopathy. A computed tomography (CT) scan of his chest showed a small area of cavitation in the right middle lobe concerning for a necrotic pneumonia. He developed a prolonged nosebleed with recurrent hemoptysis several days later. His renal function continued to decline, requiring hemodialysis. Due to continued multi-organ dysfunction with an extreme inflammatory state, hemophagocytic lymphohistiocytosis (HLH) was considered as an etiology; this was supported by an elevated serum ferritin concentration (1,610 ng/mL) and soluble interleukin 2 (IL-2) (2,490 U/mL) (normal 45-1105 U/mL). Therapy for HLH was initiated with plasma exchange and methylprednisolone. He improved on this regimen.

He was discharged home after 57 days, but was readmitted 9 days later with hypertension, abdominal pain, seizures, and altered mental status, for which he was intubated. A CT scan of his abdomen showed incidental progression of the right middle lobe cavitary lesion. Anti-neutrophilic cytoplasmic antibody (ANCA) testing was performed and returned positive in a cytoplasmic pattern with a quantitative proteinase 3 (PR3) of >200.0 U/mL (normal <20 U/mL). He was diagnosed with GPA complicated by macrophage activation syndrome. A lung biopsy revealed fibrous tissue, necrosis, and chronic inflammation. Kidney biopsy showed pauci-immune, C-ANCA-associated sclerosing glomerulonephritis with 100% global glomerulosclerosis. He was started on methylprednisolone, cyclophosphamide, and plasma exchange and improved on this regimen.

Discussion: In this case, the presence of a common condition skewed the differential diagnosis to conditions common in sickle cell disease. In critically ill children it is important to continuously seek a unifying explanation for clinical signs and symptoms.
ALTERED MENTAL STATUS IN A HEALTHY CHILD: THE CAT’S OUT OF THE BAG!
POSTER #6

JOANNA HALES, MD
DENISE JONES, MD
MARY TERRELL, MD
PEDIATRICS RESIDENTS
MENTOR: MICHAEL CINOMAN, MD

Case Presentation: A 6-year-old boy presented to the ED with altered mental status. He was found unconscious at home after returning from his soccer game. Parents denied trauma, ingestions and fevers. Initial vital signs were stable. He was intubated for GCS of 6. Physical exam revealed a 3cm inguinal lymph node for which his pediatrician had started Keflex 7 days prior for lymphadenitis. LFTs, CBC, lactate, blood gas, urine drug screen and chemistries were unremarkable. CT scan showed mild cerebral edema. LP was performed with an elevated opening pressure but CSF studies were otherwise normal. Parents did endorse exposure to a cat and Bartonella henselae IgG from CSF was sent. An EEG was unremarkable. A follow-up MRI was normal. He returned to baseline within 12 hours and was extubated. He was discharged with neurology follow-up; however, was readmitted within 24 hours for seizures. He was managed with multiple antiepileptic drugs. CSF Bartonella henselae IgG later resulted positive (1:640) consistent with cat-scratch disease encephalopathy (CSDE).

Discussion: Cat scratch disease (CSD) is caused by Bartonella henselae, a gram-negative bacilli. Cats are the major carrier. Close to 90% of patients with CSD have a history of contact with cats. Lymphadenopathy occurs 1-3 weeks after inoculation and is usually asymmetric involving a single node. Most patients have uncomplicated lymphadenitis at presentation, but the spectrum of CSD includes: lymphadenopathy, fever of unknown origin, eye disorders, endocarditis, CSDE and status epilepticus. CSDE occurs in approximately 2% of patients, 46-80% of whom develop seizures. CSF changes are often minimal with little or no pleocytosis and occasionally elevated protein. Brain imaging is often normal and EEG will typically show early changes of diffuse slowing. Diagnosis is made with serologic testing with positive IgG to Bartonella henselae. There is little evidence that antimicrobials help in CSDE, but options include azithromycin, rifampin and doxycycline. Prognosis is excellent in patients with CSDE with >90% achieving complete recovery.

Conclusion: Most clinicians consider CSD in the differential diagnosis of the wide variety of presentations associated with lymphadenopathy, especially if there is known exposure to cats. This case highlights the importance of considering CSDE when that presentation also includes altered mental status and/or seizures. Diagnosis is made by history in conjunction with a positive Bartonella henselae IgG. Early consideration of CSDE in these cases may help direct diagnostic testing, treatment and prognosis.
IDIOPATHIC, REFRACTORY SWEET’S SYNDROME ASSOCIATED WITH COMMON VARIABLE IMMUNODEFICIENCY

POSTER #7

QUINDELYN COOK, MD; ALLERGY/IMMUNOLOGY FELLOW; MENTOR: EVELINE WU, MD

Rationale: Manifestations of immune dysregulation in common variable immunodeficiency (CVID) include granulomatous disease, autoimmunity, chronic lung disease, enteropathy, among others. Sweet’s syndrome, or acute febrile neutrophilic dermatosis, is a hypersensitivity reaction characterized by fever, neutrophilia, and cutaneous eruptions. Few reports exist describing Sweet’s syndrome in association with primary immunodeficiencies.

Methods: We herein present a child with idiopathic, refractory Sweet’s syndrome with extracutaneous manifestations and a subsequent diagnosis of CVID.

Results: A 9-year-old male developed fever, neutrophilia, mucocutaneous lesions, and tracheobronchial inflammation. Skin biopsy histopathology revealed a dense dermal neutrophilic infiltrate, and bronchoalveolar lavage fluid had a predominance of neutrophils. He was diagnosed with idiopathic Sweet’s syndrome, ultimately well-controlled on tocilizumab and lenalidomide. His immune evaluation was unremarkable at diagnosis. Over 15 months, he had declining immunoglobulin levels and protective vaccine responses. IgG declined from 536 to 290 mg/dL, IgM from 36 to <25 mg/dL, and IgA from 108 to 16 mg/dL. He received pneumococcal conjugate and polysaccharide vaccines and had unsustained responses. He had normal B-cell number and cellular immune evaluation. He clinically developed upper and lower respiratory tract infections. With these findings in mind, the patient was diagnosed with CVID and started on immunoglobulin replacement therapy.

Conclusion: Immune dysregulation is common in CVID. Sweet’s syndrome is an inflammatory skin condition generally associated with autoimmune disease and malignancies. This is one of few cases reporting Sweet’s syndrome in the setting of CVID. This case should encourage providers to screen Sweet’s syndrome patients for underlying primary and acquired immunodeficiencies.
A RARE CAUSE OF RECURRENT HEMOPTYSIS
POSTER #8

YOLANDA YU, MD; PULMONOLOGY FELLOW
MENTOR: CEILA LOUGHLIN, MD

Introduction:
Thoracic endometriosis syndrome (TES) describes the clinical manifestations stemming from the presence of ectopic endometrial tissue in the thoracic cavity. It entails several clinical entities including catamenial pneumothorax, catamenial hemothorax, catamenial hemoptysis, and lung nodules. Here we report a unique case of TES presenting with catamenial hemoptysis.

Case Report:
A 16 year old female with a past medical history significant for recurrent hemoptysis associated with menstruation presented with worsening hemoptysis in the setting of an acute upper respiratory tract infection with antibiotic use during her menses. She had previous workup for her recurrent hemoptysis at an outside hospital including evaluations by Gastroenterology, Gynecology and Otolaryngology, as well as autoimmune and coagulopathy workup. She had also been on long-term hormonal suppression therapy. Physical examination was unremarkable, though an episode of hemoptysis during the exam was observed. Chest computed tomography (CT) revealed no evidence of intrathoracic endometriosis. She was subsequently admitted and an oral contraceptive taper was initiated. Her hemoptysis consequently improved. Flexible bronchoscopy done at that time demonstrated no bleeding source and no airway lesions resembling endometrial tissue. She was discharged home on hormonal therapy. Follow up three weeks after discharge revealed no recurrence of hemoptysis.

Discussion:
Catamenial hemoptysis is a rare manifestation of TES. It presents as cyclical pulmonary hemorrhage associated with menstruation as the result of endometrial implants located on the pulmonary parenchyma or airway. Diagnosis of TES remains challenging. It is often diagnosed clinically after the exclusion of other pulmonary diseases, though imaging studies, bronchoscopy and video-assisted thorascopy can aid diagnosis. Histological confirmation of endometrial tissue is required for definitive diagnosis of TES, however this is not always achievable. Hormonal therapy is often first-line treatment aimed at suppressing ectopic endometrial tissue. Surgical options, such as video-assisted thorascopic surgery, have also been used for treatment.

This case report identifies a rare presentation of TES, and illustrates the challenges associated with diagnosis.
PAIN TO THE BONE: REFRACTORY THIGH PAIN IN AN ACTIVE YOUNG MAN
POSTER #9

ALEX FLORENCE, MD: PEDIATRICS RESIDENT
MENTOR: JOSHUA BERKOWITZ, MD

Case Description: 26-year-old male with well-controlled psoriasis presents with 3.5 months of left anterior thigh pain. Pain started as a jolt while exercising without specific injury. He has not had full resolution despite rest from jogging and weight lifting. He describes vague, mid and proximal thigh pain that sometimes awakens him from sleep. He thinks he has lost some weight. He notes slight worsening with exercise but without specific exacerbating movements. He denies weakness, numbness, or tingling in the left leg, fevers, night sweats, or fatigue. On exam, well-appearing young adult male with no visible abnormality of the left hip or anterior thigh. No tenderness to palpation over the hip flexors, quadriceps or greater trochanter. Full, painless ROM of the hip with symmetrically limited internal rotation bilaterally. No pain and normal strength with resisted hip flexion. Negative labral provocative maneuvers. No sensory deficits.

Radiographs of the femur show no evidence of bony abnormality, normal joint space. Initially had some relief with diclofenac but the pain returned after 3 days. MRI hip with arthrogram demonstrates a small anterosuperior labral tear and an area of increased signal on STIR sequence within the anterior mid-femoral neck. Diagnostic intra-articular hip injection yielded temporary but complete relief of pain. MRI tumor protocol obtained 2 weeks later showed interval improvement of bone marrow signal in the femoral neck. Differential included osteoid osteoma, stress reaction, labral tear, Ewing sarcoma, and impingement. Patient ultimately diagnosed with femoroacetabular impingement with anterosuperior labral tear and underwent successful arthroscopic chondroplasty and osteoplasty.

Discussion: FAI is a cause of hip/groin pain implicated in more labral injuries than previously understood. The incidence of FAI in the general population is 10-15% in most studies, but may be closer to 24% in athletes, especially in sports that involve extremes of flexion and internal rotation. FAI is due to abnormal interaction of the femoral neck and acetabulum, classically related to cam or pincer type deformity, or a combination as with our patient. It is more common in young adults, especially men, often presenting with insidious onset of pain. Few unbiased studies exist to evaluate clinical tests in accurately diagnosing FAI, and patients often require further diagnostic imaging such as MRA. There appears to be a relationship between femoral neck stress reactions and FAI, which may reconcile the MRI findings and clinical picture for our patient. Currently arthroscopic surgical management yields good outcomes as with our patient.
STUCK IN THE MIDDLE WITH MU-CIN: A 5 YEAR OLD MALE WITH OPHTHALMOPEGIA AND FEVER
POSTER #11

ALLISON RITTENBERG, MD; THIRD YEAR INTERNAL MEDICINE/PEDIATRICS RESIDENT
MENTOR: ERIC ZWEMER, MD

Case Presentation: A 5-year-old male with allergic rhinitis presented to the ER with acute onset of right cranial nerve VI palsy and afferent pupillary defect (APD). Mother reported a 5 day history of subjective fever, nasal congestion, and cough. Vital signs were T 39.4°C, HR 84, BP 121/71. Exam showed an uncomfortable-appearing boy with right-sided proptosis, orbital edema, visual field deficit, absent right eye abduction, and a sluggishly reactive right pupil measuring 3-4 mm. CT showed an expansile right-sided sphenoid mass with evidence of bony erosion. Differential diagnosis included bacterial sinusitis with orbital cellulitis, fungal infection, mucocele, and tumor. In the OR, a large mass with “peanut butter” consistency and purulent fluid was found in the right sphenoid sinus. Pathology demonstrated septate fungal hyphae and eosinophilic mucus, most consistent with allergic fungal sinusitis. Patient received systemic steroids and antibiotics for superimposed bacterial infection. At discharge, he continued to have a right cranial nerve VI palsy, visual field deficit, and improved but persistent APD.

Discussion: Allergic fungal sinusitis (AFS) is a form of chronic sinusitis due to hypersensitivity response to fungi (typically Aspergillus) in the paranasal sinuses. Atopy is a hallmark of the disease, with the majority of patients reporting a history of allergic rhinitis and/or asthma. The majority of patients are not immunocompromised. Presenting symptoms include nasal obstruction, discharge (at times purulent or dark-colored), headache, and proptosis. If pain is a presenting symptom, it often indicates a superimposed bacterial infection. Cases of visual loss and cranial nerve deficits reported in the literature are rare. The diagnosis of AFS is made based on a detailed history of atopy and symptoms of sinusitis, characteristic CT findings (opacified sinuses and bony erosion), and histologic evidence of eosinophilic mucus with non-invasive fungus. Treatment involves urgent endoscopic resection for orbital decompression and systemic steroids. There are no convincing controlled trials that demonstrate efficacy of antifungals to treat AFS. A high rate of reoccurrence necessitates close follow-up.

Conclusions: AFS occurs in immunocompetent patients and should be on the differential for any pediatric patient with a history of atopy presenting with refractory sinusitis. In rare cases, AFS can present with cranial nerve deficits and vision loss which should prompt immediate imaging and subsequent surgical decompression of the orbit. AFS does not require treatment with antifungal agents, but patients should be treated with steroids to help reduce the risk of recurrence.
LIKE FATHER LIKE SON: A CASE OF CONGENITAL LYMPHATIC DYSPLASIA DIAGNOSED WITH LYMPHANGIOGRAPHY

POSTER #12

ANNA KOWALCZYK-KIM, MD; PEDIATRICS RESIDENT
MENTOR: STEVE LICHTMAN, MD

Additional Authors; Division/Institution: J Kim, E Munns; Pediatrics

Introduction: Fetal ascites is typically a symptom found in hydrops fetalis, which is characterized by the presence of two or more of the following: ascites, pleural effusion, pericardial effusion, and skin edema. Fetal ascites can also occur independently without accumulation of fluid in other serosal cavities or subcutaneous tissue. Although not as common or as well understood as hydrops fetalis, the causes of isolated fetal ascites are due to cardiovascular disorders, idiopathic causes, chromosomal imbalances, hematologic abnormalities, infections, intra-thoracic masses, lymphatic dysplasias, twin-to-twin transfusion syndrome, placental causes, urinary tract malformations, inborn errors of metabolism, extra-thoracic tumors, and gastrointestinal disorders. Less commonly, isolated fetal ascites is caused by congenital lymphatic dysplasia or abnormal lymphatic drainage. We report the case of a child diagnosed prenatally with isolated fetal ascites, and, postnatally, developed protein-losing enteropathy likely due to congenital lymphatic dysplasia found on lymphangiography.

Case Presentation: A premature male was delivered at our hospital, and found to have isolated fetal ascites. Despite serial paracenteses not being diagnostic for chyle, he was started on a polymeric diet of medium-chain triglycerides (MCT). However, he continued to have accumulation of ascites. Ultimately, a lymphangiogram was obtained and showed an abnormal lymphatic system with numerous collateral pathways with no filling of the cisterna chyli or thoracic duct. Given the father’s history of congenital ascites, which spontaneously resolved with no surgical intervention, it was determined that our patient would undergo conservative management. A Jackson-Pratt (JP) catheter was placed within the peritoneum for additional peritoneal fluid drainage. By discharge, the JP drain was removed and Pregestimil formula was started with no further accumulation of ascites during follow up.

Discussion and Conclusions: Fetal ascites found antenatally is uncommon with an overall mortality rate as high as 67% when associated with hydrops, and 48% when isolated. Initially, it was thought our patient had primary intestinal lymphangiectasia (PIL) since his father was also diagnosed with PIL as an infant. Further diagnostic workup revealed abnormal lymphatics within the neck, thoracic cavity, retroperitoneal space, and pelvic cavity with proliferation of collateral pathways, indicating our patient did not have PIL, and instead has a diffuse central conducting lymphatic abnormality. The use of lymphangiography allowed us to determine the diagnosis and prevented our patient from undergoing more invasive testing.
PLEVA with Fevah can be a Deceivah
POSTER #13

B HAMMOND, MD; PEDIATRICS RESIDENT
MENTOR: ERIC ZWEMER, MD

Additional Authors; Division/Institution: A Kowalczyk-Kim; Pediatrics

Case Presentation: A 6 yo male with ADHD presented with 5 weeks of diffuse body rash. The rash began with erythematous, papular lesions on the neck that spread centrifugally. He was started on lisdexamfetamine (Vyvanse) the same day as the onset of rash. Initial concern was for pityriasis rosea and watchful waiting was advised. With the rash persisting, topical steroid and acyclovir were trialed without improvement. After 3 weeks, a skin biopsy was performed, followed by intravenous and oral steroids. He initially improved, but became lethargic and febrile to 101.4° F. He was admitted for IV antibiotics and IV steroids. Skin biopsy results showed pityriasis lichenoides et varioliformis acuta (PLEVA). Erythromycin, the therapy for PLEVA, was started, but the patient had recurrent fever and worsening of rash. Concern was raised for a severe variant of PLEVA called febrile ulceronecrotic Mucha-Habermann disease (FUMHD), and the patient was transferred for Dermatology consultation. Dermatology recommended starting methotrexate. By discharge, his rash had improved and he was sent home on a steroid taper.

Discussion: PLEVA is a rare, cutaneous inflammatory disorder that is typically self-limiting and classically affects young adults. Pathogenesis is thought to involve benign lymphoproliferation, or hypersensitivity response to infection. PLEVA presents as an acute eruption of inflammatory papules and papulovesicles that rapidly develop hemorrhagic or necrotic crusts. Diagnosis is made by biopsy. Based upon retrospective studies and case reports, first-line therapy for extensive disease includes use of tetracyclines or erythromycin and phototherapy for 2-3 months. Febrile ulceronecrotic Mucha-Habermann disease (FUMHD) is a severe presentation of PLEVA characterized by systemic illness, fever, and large ulcerated skin lesions. Methotrexate is used as therapy for FUMHD. Systemic glucocorticoids have been utilized in many reported cases, but whether they are truly beneficial remains unclear. Other agents that have been utilized for FUMHD include intravenous immunoglobulin, acyclovir, dapsone, cyclosporine, and phototherapy. Inpatient management is usually required for these patients.

Conclusions: Although the exact pathogenesis of PLEVA and FUHMD is poorly understood, a systemic reaction to medications may play a role.
Case presentation: A 14 yo male presented to the ER with fever, cough, a streaking erythematous rash extending upward from his left fifth toe, and an erythrodermic rash on his chest, after stubbing his toe one week prior. He had seen his pediatrician, who prescribed trimethoprim-sulfamethoxazole (TMP-SMX) without relief.

In the ER, he was febrile to 40°C, tachypenic, and ill appearing. CBC demonstrated leukopenia (1.8x10^9/L), thrombocytopenia (92x10^9/L) and blood chemistry tests revealed hyponatremia (132mmol/L. CRP was elevated to 68mg/L. PT and aPTT were prolonged to 15.1 and 46 seconds. Radiographs of the chest showed a unilateral pleural effusion. He was admitted to the Burn ICU due to concern for TMP-SMX induced drug reaction.

His presentation was judged to be most consistent with Streptococcal Toxic Shock Syndrome (TSS). Clindamycin was started and he became afebrile within the first 24 hours of therapy. After three days of IV therapy, he was discharged to complete seven further days of PO Clindamycin.

Discussion: TSS is a toxin mediated disease process, commonly caused by staphylococcus and streptococcus exotoxins. Streptococcal pyrogenic exotoxins A and B, and cell wall ‘M proteins’ M1 and M3, are the most commonly associated with TSS. Exotoxins and M proteins act as super-antigens by circumventing the normal activation of T-cells by binding directly to MHC-II receptors on antigen presenting cells, and to T-cell receptors. Through this mechanism, 5-30% of a patient’s total T-cell population will be activated, compared to the normal activation process, which activates 0.01-0.1% of a patient’s T-cells. The result is a massive release of cytokines, TNF-a, IL-6, IL-2, and IF-y, which progress to fever, rash, hypotension, ARDS, and multi-organ failure.

Unlike staphylococcal TSS, 80% of streptococcal TSS originates from skin infections and it frequently develops after negligible trauma. A combination of a β-lactam plus clindamycin is the recommended treatment for streptococcal TSS. Due to its mechanism of inhibiting protein synthesis, clindamycin not only inhibits bacterial replication, but additionally inhibits toxin production. Our patient’s progression from a simple toe infection to TSS was likely due to TMP-SMX’s limited coverage of streptococcal species and TMP-SMX’s tendency in some studies to upregulate toxin production.

Conclusion: A patient’s likelihood to progress to TSS depends on the mechanism of infection, local virulence patterns, and protective patient characteristics such as antibodies to M-protein and certain HLA-II haplotypes. Prompt identification of strep TSS and appropriate treatment with a β-lactam plus clindamycin is key for decreasing mortality.
ACUTE PANCREATITIS: A RARE COMPLICATION OF LUPUS
POSTER #15

ELIZABETH SIBRACK, MD; PEDIATRICS RESIDENT
MENTOR: ERIC ZWEMER, MD AND KATHY BRADFORD, MD

Hospital Course: A 10 y/o F with 7 years of episodic cervical lymphadenopathy and fever, presented with typical episode, new abdominal pain and emesis. ROS positive for anorexia, weight loss, night sweats, diarrhea; she denied rash, cough, sore throat and arthralgia. History notable for many prior antibiotics, supplement shake intake, and travel to Mexico 7 years ago. Past CT neck reported lymphadenitis and parotid enlargement. Admission exam found matted cervical LAD, prominent parotids, dental caries, no oral lesions, mild abdominal tenderness, no mass or organomegaly, normal strength and no joint involvement. Labs significant for WBC 3.4, Hgb 9.7, ESR >140, AST 620, ALT 614, Tbili 4.0, Dbili 3.5, lipase 82. Extensive infectious workup was negative including viral, fungal and mycobacterial studies. ANA positive 1:1280 speckled, but anti-dsDNA negative, ENA panel pending. Anti-LKM and smooth muscle antibody, ceruloplasmin, A1AT, acetaminophen and tox screen were negative. Lymph node biopsy showed reactive hyperplasia. Liver biopsy suggested biliary pattern of injury, drug-induced vs. obstruction. Recent amoxicillin/clavulanate use was excluded and herbal shakes felt unlikely cause. MRCP unexpectedly found acute pancreatitis and repeat lipase was 9452! IV methylprednisolone had been started 3 days prior, post-biopsies, with 4 criteria met for systemic lupus erythematosus (leukopenia, direct coombs, low complement, ANA). Finally, anti-Sm returned positive confirming diagnosis of SLE, also with positive anti-RNP, Ro and La suggesting MCTD and Sjogren’s overlap.

Discussion: History of lymphadenopathy and fever, common pediatric complaints, guided this acute presentation. The initial differential diagnosis was broad, including infections, malignancy and rheumatologic illness. The history, physical, and diagnostic studies led to the correct diagnosis of lupus as the etiology of this patient’s complicated course with hepatitis and pancreatitis. SLE is a multisystem disease, often with gastrointestinal involvement, but rarely acute pancreatitis. In several reports, as in ours, gastritis secondary to steroids was first assumed and delayed diagnosis. While steroids have been implicated in cases of pancreatitis in the general population, among studied cases of patients with SLE, this association is not seen and instead steroids demonstrate mortality benefit.

Conclusions: Providers should keep non-infectious etiologies, including SLE in mind when evaluating patients with recurrent LAD and fever. Diagnosis of SLE should be made on clinical suspicion, aided by published clinical and immunologic classification criteria. Pancreatitis is a rare, dangerous complication of SLE to recognize. Its diagnosis and treatment with immunosuppression is critical, but should first exclude more common mechanical and toxic-metabolic etiologies.
Clinical Case Description: A 5 week-old-female presented with a two-week history of decreased mobility of her left arm without inciting trauma and with no associated erythema or edema. The infant was born at term by precipitous vaginal delivery to a mother of advanced maternal age. The mother gave birth in a wheelchair in triage and the infant fell on the floor, but otherwise pregnancy and delivery were unremarkable. On presentation, the infant was well appearing with normal vital signs. There was no spontaneous movement of the left arm with some spontaneous movement of the left hand and fingers. There was no erythema, swelling or warmth of the arm. She cried when left arm was passively moved or upon palpation of the upper arm. Neurologic exam revealed intact rooting, suck, plantar and palmar grasp reflexes. Moro reflex was difficult to assess due to pain with motion of the left arm. Discussion: This case of an infant with decreased arm movement provides an opportunity to review musculoskeletal complaints in infancy. It also provides a review of neonatal osteomyelitis, especially in the context of a less common manifestation of late onset neonatal group B streptococcal disease. It also highlights the limitations of current research in decisions about optimum outpatient therapy.
MULTIPLE SYMPTOMS IN A TEENAGE MALE IMMIGRANT
POSTER #17

KRISHNA ALURI, MD; PEDIATRICS RESIDENT
MENTOR: KENYA MCNEAL-TRICE, MD

A 16-year-old Hispanic male presents with 4 months of weight loss, hair loss, generalized weakness, pain and swelling of his fingers, abdominal pain, and rash. The patient emigrated from Guatemala 9 months prior. His history is significant for a 9-month hospitalization as an infant, which his father reports was due to a respiratory illness. He also had an appendectomy in Guatemala. On admission, vital signs are normal and temperature is 36.9°C. The patient has a BMI of 16.58 kg/m². He is in mild discomfort. He has two prominent circular patches of hair loss over the right parietal and occipital scalp. The patient also has multiple 0.5 to 1cm lymph nodes in the submandibular, right supraclavicular, axillary, and inguinal regions. None of those nodes is tender to palpation. The patient’s abdomen is soft and bowel sounds are present, but there is significant tenderness to light palpation that is worse on the left. No organomegaly is identified. There is swelling and effusion present around each proximal and distal interphalangeal joint bilaterally. There is an erythematous rash with central scales over the first four metacarpophalangeal joints. Strength is 4/5 in multiple movements at the shoulder, hip, and knees. Infectious investigations reveal negative or normal studies for hepatitis A, B, C, CMV, EBV, HIV, HHV-6, syphilis, tuberculosis, and fungal infection. No concerning findings are found on oncological work-up. CT of the abdomen and pelvis shows lymphadenopathy but no other abnormalities. MRCP shows evidence of superior mesenteric artery syndrome. Rheumatologic investigation reveals negative antinuclear antibody and negative double-stranded DNA antibodies. ENA is positive at 23, and SS-A/Ro is positive. Anti-U1 RNP is also weakly positive. Dermatopathology from cutaneous biopsy of the patient’s rash shows vacuolar interface dermatitis. EMG testing is consistent with inflammatory myositis. CK is elevated to 307 U/L. These findings and the patient’s symptoms are considered to be consistent with juvenile dermatomyositis and mixed connective tissue disease. Juvenile dermatomyositis is largely diagnosed based on clinical findings, and supportive diagnostic findings in this case includes symmetric weakness, presence of Grotton papules, elevated CK, and myositis on EMG. Additionally, MCTD is characterized by an elevation of anti-ENA and anti-U1 RNP antibodies, along with overlapping clinical features of systemic autoimmune diseases. Juvenile dermatomyositis is commonly treated with a combination of glucocorticoids and methotrexate, with adjunctive therapy including IVIG and hydroxychloroquine. All of these therapies are being used in this patient, with significant improvement in symptoms.
“GANG”ING UP ON THE SKIN: AN 11-MONTH-OLD WITH ULCERATIVE SKIN LESIONS
POSTER #18

LAURA CANNON, MD; PEDIATRICS RESIDENT
MENTOR: ERIC ZWEMER, MD

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Case Presentation: A previously healthy 11-month-old female was admitted to the hospital with new onset of skin lesions. She was healthy until one month prior to admission, when her parents noticed “bumps on her bottom.” They initially thought these bumps were secondary to insect bites, and took her to an urgent care, where she was prescribed topical antibiotic ointment. The lesions persisted, and she developed new papules on her arms and legs, and fever to 101°F. At this time, her pediatrician evaluated her and diagnosed hand, foot, and mouth disease. She defervesced the next day, but the lesions persisted with development of purulent drainage, crusting, and eventual ulceration and pain. Repeat evaluation by her pediatrician resulted in prescription of cephalexin for presumed impetigo. With failure to improve, she also completed a course of sulfamethoxazole/trimethoprim as well as topical and oral acyclovir without improvement. Bacterial and viral cultures of the lesions were negative. Given persistent pain and severity of the lesions, she was admitted to the hospital.

On admission, her exam revealed diffuse well-circumscribed erythematosus papules and pustules of varying sizes on her face, legs, arms, hands, and feet intermixed with well circumscribed erosions with central crusting and a violaceous border (Figures 1-3). The initial differential diagnosis included atypical mycobacteria infection, nodular vasculitis, T-cell cutaneous lymphoma, and infantile pyoderma gangrenosum (IPG). Labs were significant for a leukocytosis with neutrophilia; LDH and uric acid were within normal limits. Intravenous antibiotics and antiviral medication were started on admission but discontinued when repeat cultures were negative. Dermatology performed a skin biopsy which was consistent with IPG. After cultures were negative, antimicrobials were stopped. Oral steroids were started with overall improvement and she was discharged home on oral steroids.

Two weeks after discharge, she experienced worsening pain and new skin lesions. She was admitted to the hospital for IV steroids given oral steroids had been ineffective. She was later started on immunomodulatory therapy with Infliximab after discussion with the pediatric rheumatology and dermatology teams. Given potential for associated diseases, she was evaluated for underlying systemic disorders with normal immunoglobulin levels, blood smear, diphtheria and tetanus antibody titers, leukocyte adhesion deficiency panel, and neutrophil function test. At this time, no other comorbid conditions have been diagnosed in this patient. She was discharged home on oral steroids and the plan for monthly Infliximab infusions.

Discussion: IPG is very rare with less than 20 cases reported in the literature. It is a neutrophilic dermatosis that presents as ulcerations of the skin with inflammation. Infants diagnosed with IPG warrant further work-up for associated systemic diseases including inflammatory bowel disease, Takayasu’s arteritis, leukocyte adhesion deficiency, and chronic granulomatous disease. Most cases, however, are idiopathic—though it can be difficult to rule out an associated condition, given skin findings may precede other systemic symptoms by several years. At this time, standardizing treatment is challenging given the rarity of IPG, but includes immunomodulation with steroids and/or biologic agents.

Conclusions: Though rare in infants, IPG should be considered in the differential diagnosis for an infant with diffuse ulcerative skin lesions.
Case Presentation: A 2-week-old female presented to the ED with tachypnea worsening over the past 3 days. No fevers, rhinorrhea, cough, or difficulty feeding. She was full term with normal prenatal ultrasounds, pulse oximetry screening, and postnatal weight gain. She had been evaluated twice by her pediatrician for tachypnea and started on reflux medication. She was admitted with concern for bronchiolitis. On exam she looked overall well but had tachypnea and supraclavicular retractions. Lungs were clear. She also had a II/VI systolic ejection murmur at the LSB and 2+ femoral pulses. Right upper extremity blood pressure was 113/68, and right lower extremity was 70/58. Corresponding O2 saturations were 100% and 99%. ECHO showed VSD and juxtaductal coarctation of the aorta with mildly to moderately decreased left ventricular systolic function and no patent ductus arteriosis (PDA). She underwent immediate surgical repair and was discharged home 3 days later doing well.

Discussion: Neonatal coarctation of the aorta (CoA) is the fifth most common congenital heart lesion. CoA typically does not cause problems in utero due to bypass of the coarctation by the PDA. Newborns with CoA may initially be asymptomatic but will start to manifest symptoms at 2-5 days of life with PDA closure. Initial manifestations include murmur, decreased/absent femoral pulses, tachypnea, and feeding difficulties. Newborns present with varying degrees of heart failure related to the severity of the coarctation, and patients can quickly deteriorate with shock and circulatory collapse. Untreated CoA has significant early mortality, but timely diagnosis and surgical repair has excellent outcomes. Early recognition is key, and screening methods include prenatal ultrasound, newborn physical exam, and pulse oximetry screening. Pulse oximetry has been utilized as a screening method in the newborn nursery for congenital heart disease. Pulse oximetry screening, however, is most likely to miss duct-dependent lesions as it is typically performed before the duct has closed, and neonates with CoA are at the highest risk of being missed. Studies show over half of neonates with CoA are discharged undiagnosed from the nursery.

Conclusions: CoA is the cardiac lesion most likely to be missed by initial newborn exam and pulse oximetry screening. Careful physical exam including cardiac exam, palpation of femoral pulses, and blood pressure measurement remain fundamental in diagnosis. CoA should be considered in infants 1-2 weeks of age who present with murmur or tachypnea and not just more overt signs of heart failure.
SEVERE RESPIRATORY DISEASE AND EVIDENCE OF INTERSTITIAL LUNG DISEASE IN A PATIENT WITH RUBINSTEIN-TAYBI SYNDROME
POSTER #20

LAUREN BRADFORD, MD; PEDIATRICS RESIDENT
MENTOR: TIMOTHY VECE, MD

Additional Authors; Division/Institution: Will Stoudemire, Timothy Vece, Celia Loughlin

Introduction: Rubinstein-Taybi syndrome (RTS) is a rare genetic disorder characterized by broad thumbs and halluces, short stature, intellectual disability, and distinctive facial features. It is caused by a 16p13.3 microdeletion with mutations in the CREBBP gene. Patients with RTS are at risk for upper airway obstruction and obstructive sleep apnea, however interstitial lung disease is not a known complication of RTS. We describe a 4 year-old patient with RTS with progressive respiratory insufficiency and evidence of interstitial lung disease.

Case Description: TY is a 4 year old with a previous pulmonary history of obstructive sleep apnea and laryngotrachomalacia without other known lung disease. Six months prior to admission, he developed increased intermittent supplemental O2 requirement. Over the course of three weeks he had progressive respiratory insufficiency characterized by desaturations and increased work of breathing and increased supplemental oxygen requirement eventually requiring intubation and high ventilator support. An extensive infectious, respiratory, and cardiac workup was initiated, but no specific etiologies were identified. Chest CT revealed diffuse ground glass opacities, septal thickening, peripheral cysts and peri hilar adenopathy. He clinically improved after the initiation of systemic corticosteroids. He continues to require intermittent increased supplemental oxygen. A repeat chest CT demonstrated continued changes consistent with interstitial lung disease.

Discussion: We describe a 4-year-old male with RTS who developed acute respiratory failure over the course of a week after months of slow respiratory insufficiency. No infectious or other definitive etiology of his respiratory symptoms was identified. Worsening hypoxemia, diffuse changes on chest CT, and improvement with systemic corticosteroids therapy are strongly suggestive of interstitial lung disease. While interstitial lung disease is not a known complication of RTS, there are reports of suspected severe interstitial lung disease in patients with RTS in the literature. The reports describe a similar pattern on CT imaging as was seen in our patient. Lung tissue samples were not obtained in those patients so it is difficult to know what exact process is occurring. This case represents a third reported case of severe interstitial lung disease in a patient with RTS. While likely a rare complication, interstitial lung disease should be considered in the differential diagnosis of a patient with RTS and increasing respiratory insufficiency.
WHY THE LONG FACE? A CASE OF NEONATAL HYPOTONIA
POSTER #21

LAUREN FRAZER, MD AND ALEX FLORENCE, MD; PEDIATRICS RESIDENTS
MENTOR: DIANE WARNER, MD

Background: The infant in this case was an ex-39 week AGA male born via C-section to a 36 y/o G4P3. He was transferred on DOL 1 due to hypotonia/weakness and respiratory failure of unknown etiology. Mother reported decreased fetal movement, and polyhydramnios was noted on ultrasound. The infant required compressions shortly after birth due to apnea and bradycardia unresponsive to bagging. He stabilized after intubation and was transferred. On physical exam, he had a prominent forehead, bitemporal narrowing, micrognathia with retrognathia, bifid uvula, elongated fingers/toes, and cryptorchidism. He also had pronounced weakness with minimal spontaneous movements of his extremities. Deep tendon reflexes were severely diminished. A workup for neonatal hypotonia/weakness was initiated.

The infant was extubated on DOL 3 but required HFNC and was unable to manage his secretions. He was relatively stable until he had a cardiac arrest event associated with respiratory failure. He then developed status epilepticus, was cooled, and seizures were controlled with AEDs. After recovery from this event, he remained mechanically ventilated. A G-tube and trach were placed, and he was discharged home.

The differential diagnosis for neonatal weakness/hypotonia is broad. Consults included endocrinology, neurology, pulmonology, ENT, ophthalmology, and genetics. The differential diagnosis was notable for Prader-Willi, Smith-Lemli-Opitz, glycosylation disorders, inborn errors of metabolism, SMA, congenital myotonic dystrophy, peroxisomal or mitochondrial disorders, and other congenital myopathies.

Methods: Diagnostic studies included a karyotype, microarray, Prader-Willi/Angelman methylation study, plasma amino acids, urine organic acids, carnitine/acylcarnitine profile, long chain fatty acids, 7-dehydrocholesterol, 185 gene panel for Muscular Dystrophy/Myopathy, MRI brain, airway evaluation, and EMG.

Results: Next generation sequencing revealed that the infant had a previously undescribed mutation in the MTM 1 gene, which encodes for the phosphatase myotubulin. The mutation affects the AG acceptor splice site for exon 9. The infant was diagnosed with X-linked myotubular myopathy.

Conclusions: This case represents a multidisciplinary approach to diagnosis and treatment of an infant with a rare genetic condition. After an extensive workup, the diagnosis was made using next generation sequencing technology. The mutation identified in this case has not been previously described. X-linked myotubular myopathy affects about 1:50,000 newborns worldwide. This patient likely has a severe form of the disease, which will results in ventilator and g-tube dependence, inability to ambulate, and severely delayed milestones. The prognosis for this infant is more complex given his history of a prolonged cardiac arrest event. Treatment is supportive.
Case: 12-year-old girl presented with altered mental status, hypothermia, and bradycardia following one week of abdominal pain, fatigue and flattened affect. Family denied ingestion, fever, cardiorespiratory symptoms, headaches or unusual exposures. Exam significant for sluggishly reactive pupils, disconjugate gaze, ability to intermittently follow commands. Laboratory studies revealed hyponatremia, hypokalemia and negative infectious work-up. CSF analysis had a persistent lymphocytic pleocytosis and oligoclonal bands. MRI Brain showed a non-enhancing, ill-defined lesion involving the hypothalamus and diencephalon, eventually with scattered grey-matter lesions which showed hypercellularity on biopsy. NMO/AQP4 titer positive in the CSF, but not the serum. Diagnosed with probable Neuromyelitis optica (NMO) autoimmune encephalitis. IVIG had no effect. Steroid treatment provided minimal improvement. Rituximab resulted in near-resolution of symptoms.

Discussion: Autoimmune encephalitis (AE) is an increasingly recognized etiology of encephalitis. It remains challenging to diagnose due to its varied presentation and limited testing. Common presentations include delirium, seizures, behavioral changes, autonomic instability, respiratory failure, and coma. NMO typically presents with optic neuritis or transverse myelitis but can present as AE. About 65% of children with suspicion for NMO have positive AQP4-IgG in the serum. Seronegativity should not exclude the diagnosis as AQP4-IgG may appear as late as 4-5 years after initial symptoms. The absence of antibodies should not delay treatment. Steroids, plasmapheresis, and IVIG are first line with advancement to rituximab or cyclophosphamide if no clinical improvement is seen.
A MIND NUMBING SENSATION
POSTER #23

LOLA OWOLABI, MD; PEDIATRICS RESIDENT
MENTOR: KENYA MCNEAL-TRICE, MD

Case Presentation: 13 y.o. male presents for evaluation of 2 weeks of left hand paresthesia, “funny sensation” bilaterally in lower extremities, and 1 week of shoulder and neck pain. Physical examination was notable for decreased left hand grip strength. A head CT angiogram was negative but MRI of the cervical spine showed T2 hyperintensity and expansion of the cord from C1-C7. A lumbar puncture was performed and CSF was significant for elevated protein, IgG, and albumin. The remainder of infectious/inflammatory disease workup was negative. He was presumptively diagnosed with transverse myelitis, treated with IVIG, and given 5-day course of steroids with notable improvement in symptoms.

The patient was readmitted within days of completing the steroid course with worsening symptoms. Repeat MRI of the cervical spine revealed diffuse expansion of the cervical spinal cord. Lumbar puncture was repeated and CSF cytology was negative. Ultimately, he underwent cervical spinal biopsy of the lesion and pathology showed a highgrade diffuse infiltrating glioma consistent with diffuse midline glioma of the spinal cord.

Discussion: Intramedullary spinal tumors are primary spinal cord lesions that arise from glial cells in the spinal cord. Spinal cord tumors are extremely rare in children, representing only 1% of all CNS tumors. Approximately one-third of these spinal tumors are intramedullary. Of those that are intramedullary, high grade gliomas only account for 1-3%. Despite advancements in therapy high grade diffuse gliomas are a challenging diagnosis with ultimately poor prognosis.

Conclusions: Although primary spinal tumors are extremely rare in pediatric populations, patients who present with multiple neurological complaints including motor and sensory deficits should raise a high index of suspicion for a CNS tumor. Clinicians should be aware that the differential also includes inflammatory or autoimmune processes including multiple sclerosis, neuromyelitis optica, and transverse myelitis, in addition to vascular lesions. When symptoms don’t improve, continued re-evaluation is always necessary and should include a multidisciplinary team of pediatric hospitalists, neurologists, hematologist/oncologists, and neurosurgical specialists. A family-centered approach is also crucial to ensuring the patient and family understand the differential diagnosis and the plan for diagnostic evaluation. Pediatric palliative and supportive care consultation was integrated early into the care management plan, and proved invaluable as the poor prognosis for the confirmed diagnosis became evident.
AN EYE OPENER: KLEBSIELLA NEONATAL CONJUNCTIVITIS IN A TERM INFANT  
POSTER #24

MARY TERRELL, MD; PEDIATRICS RESIDENT  
MENTOR: JACOB LOHR, MD

Introduction: Neonatal conjunctivitis (NC) is a common neonatal condition. In addition to ophthalmologic complications including vision loss, systemic infection can occur. Neonates with infectious NC are thought to acquire the pathogens vertically regardless of mode of delivery. Ocular prophylaxis at birth has reduced Neisseria gonorrhea NC worldwide, but Chlamydia trachomatis remains prevalent. Other pathogens implicated in NC, though less studied, abound and appear to vary geographically.

Case Presentation: A two-day-old term male in the newborn nursery presented with unilateral purulent eye discharge. Vaginal delivery of the infant was uncomplicated without prolonged rupture of membranes, and erythromycin was applied to eyes. Mother received adequate prophylaxis for GBS+ status. Standard prenatal labs were otherwise unremarkable. Notably, gonorrhea and chlamydia were negative in both the first and third trimesters and there was no history of HSV. The neonate was vigorous on exam with normal vital signs for age. The left eye was edematous without spontaneous opening and profuse green discharge was projectile on palpation. The left conjunctiva was markedly injected. An eye swab was sent for bacterial, AFB and fungal cultures as well as N. gonorrhea, C. trachomatis, and HSV PCR. The culture resulted positive for Klebsiella pneumoniae resistant to ampicillin; remaining studies were negative. Blood and urine cultures resulted negative. IV cefotaxime was administered prior to discharge on cefdinir and topical tobramycin. Complete resolution was noted at follow up.

Discussion: This case illustrates an unusual pathogen for neonatal conjunctivitis (NC) in a healthy term infant in the newborn nursery. Klebsiella pneumoniae are Gram negative bacteria found in GI and vaginal flora, and are known surface water contaminants. Though rarely reported in healthy newborns, they are a common cause of hospital-acquired conjunctivitis in the NICU. B-lactam and carbapenem resistance in isolates is an increasing problem and susceptibility testing is important for guiding treatment.

This case reinforces the need to consider Klebsiella pneumoniae and other Gram-negative organisms whenever NC is suspected, especially when the maternal history does not suggest Neisseria gonorrhea or Chlamydia trachomatis. While history and timeline of symptoms are helpful in diagnosis, sending a culture with susceptibilities and PCR when available is imperative. It remains important to include empiric gonococcal and chlamydial coverage while awaiting cultures. Given potential for systemic infection, IV antibiotics, at least initially, should be considered for this high-risk population.

To our knowledge, this is the second reported case of Klebsiella NC presenting in a healthy newborn. The first case occurred locally, which prompts consideration of a geographical predilection.

References:


Case Presentation: A male infant born at 34 weeks was noted on newborn exam to have significant abdominal distension, hepatosplenomegaly, and a diffuse maculopapular rash with petechiae (figure 1). Birth history was notable for normal routine prenatal ultrasounds, maternal exposure to ParvoB19 in the 3rd trimester, and induction of labor due to decreased fetal movements. Initial CBC showed anemia (Hgb 6.3) and thrombocytopenia (PLT 4), followed by leukopenia (WBC 6.8 on DOL2). Abdominal ultrasound revealed marked hepatosplenomegaly. Differential diagnosis included TORCH infections, Parvovirus, neuroblastoma, and Langerhans cell histiocytosis. He subsequently developed ascites and worsening liver function with direct bilirubin of 18.7. All viral testing (including HSV, EBV, CMV, HHV 6, ParvoB19, enterovirus, adenovirus) and toxoplasma were negative. Skin biopsy of an arm lesion revealed dense, histiocyte infiltrates most consistent with hemophagocytic lymphohistiocytosis (HLH). Additional labs consistent with HLH included elevated ferritin (3670) and elevated soluble interleukin-2 receptors.

Discussion: Hemophagocytic lymphohistiocytosis is a disorder of impaired T-cell and natural killer cell activity characterized by cytopenias, hepatosplenomegaly, and persistent fevers. When presenting in the first days of life, symptoms can significantly overlap with more common neonatal disease processes, particularly congenital infections. This patient had hepatosplenomegaly and a rash that was initially thought to be the classic maculopapular “blueberry muffin” rash associated with TORCH infections. However, as opposed to congenital infections that typically present with isolated thrombocytopenia, patients with HLH have at least 2, if not all 3, cell lineages affected. The clinical picture can further be confused as neonatal HLH can also be triggered by viral infections, particularly HSV, CMV, and enterovirus. HLH should be considered in the differential if TORCH infection is suspected and treatment is initiated, but the infant continues to demonstrate clinical decline. Ferritin can serve as an initial screening test, and levels above 500 should prompt further evaluation.

Conclusion: The clinical presentation of neonatal onset HLH considerably overlaps with that of TORCH infections, including cytopenias, maculopapular rash, and hepatosplenomegaly. Although rare, clinicians should consider HLH in the differential diagnosis in the following circumstances: when 2 or more cell lineages are affected or there is continued clinical decline despite appropriate treatment of the suspected infectious etiology. Ferritin can be used as a screening test with levels above 500 prompting further testing and involvement of an HLH expert.
THE CLINICAL OUTCOMES WITH EARLY INITIATION OF TREATMENT IN A PATIENT WITH INCIDENTALLY DIAGNOSED INFANTILE HYPOPHOSPHATASIA
POSTER #26

SARA DUFFUS, MD; PEDIATRICS RESIDENT
MENTOR: ALI CALIKOGLU, MD

Additional Authors; Division/Institution: Bradley Thrasher, DO, fellow UNC Department of Pediatric Endocrinology

Objectives: To describe an unusual presentation of hypophosphatasia and the outcomes of treatment when initiated early in the disease course

Results: A 4-week-old, term male was found to have a hip click bilaterally on exam by his primary care provider. Hip films were interpreted as having an abnormal appearance of the proximal femurs concerning for non-accidental trauma. Subsequent skeletal survey revealed cupping and metaphyseal irregularities in the majority of the long bones, in addition to rachitic rosary most consistent with rickets. The patient had normal calcium (9.7 mg/dL), normal parathyroid hormone (25 pg/mL), only mildly elevated phosphorous (5.8 mg/dL), and significantly low alkaline phosphatase level (33 units/L). Additional testing was consistent with hypophosphatasia, including elevated urine phosphoethanolamine (2228 nmol/mg creatinine, normal 0 – 372) and elevated vitamin B6 (1030 mcg/L, normal range 5 - 50). The patient was started on asfotase alfa 2 mg/kg three times weekly at 7 months of age. After 1 month of therapy, his laboratory findings had normalized. Repeat imaging revealed grossly normal long bone appearance with no evidence of low bone mineralization or metaphyseal flaring or widening. At 11 months of age, four months following initiation of therapy, he continued to have no apparent bony abnormalities or respiratory insufficiency. His only notable clinical symptom was premature loss of two primary teeth.

Conclusions: Infantile hypophosphatasia is a rare disorder that is life-threatening and frequently diagnosed late in the clinical course. Without treatment, mortality is greater than 50% by 9 months of age. This case report describes a unique presentation of infantile hypophosphatasia in which the diagnosis was incidentally made following evaluation to rule out congenital hip dysplasia, prior to the development of any clinically evident symptoms. Treatment with enzyme replacement therapy was initiated early, after which significant radiographic and laboratory improvement was demonstrated. Findings from this case emphasize the importance of early diagnosis and argue for timely implementation of enzyme replacement therapy.
17 year-old male with a history of anti-NMDAr encephalitis three years prior presented with two days of confusion, agitation and insomnia. Associated symptoms included anorexia, blunted affect, slow movements and, though awake, refusal to respond or engage with others. Family also reported apparent hallucinations and associated aggression including biting. Neurologic examination with non-verbal patient, intermittent ability to attend and follow commands, hypertonic extremities, 3+ patellar reflexes and ankle clonus bilaterally. MRI Brain was normal and CSF analysis confirmed anti-NMDAr antibodies. EEG showed diffuse slowing. Paraneoplastic work-up was negative. Prior presentation in Mexico was associated with seizure and protracted impairment which resolved after treatment with steroids and IVIG. Initial management with pulsedose steroids and IVIG resulted in minimal improvement, thus the patient was treated with Rituximab. Patient received monthly infusions and after three months normalized. Anti-NMDAr autoimmune encephalitis is a well-described syndrome and one of the most common etiologies of non-infectious encephalitis. Though possible, anti-NMDAr encephalitis less likely to be paraneoplastic in children and males. Presentation classically includes neuropsychiatric symptoms such as agitation, psychosis, behavior changes as well as memory loss, chorea, and coma. Studies may be normal other than CSF anti-NMDAr antibody detection, though can reveal CSF lymphocytic pleocytosis, elevated protein, and MRI abnormalities. Up to 25% of patients experience a relapse of symptoms, often months or years after initial presentation. Relapse may be more likely to occur in patients who do not receive immunotherapy at first presentation and who do not have an associated tumor. Relapsed symptoms may differ from prior and classical presentation. Early diagnosis and initiation of immunotherapy leads to decreased levels of anti-NMDAr antibodies. This aids in reducing the risk of relapse; relapses have been reported in 12-25% of patients. Risk for relapse includes lack of immunotherapy and may also be related to absence of tumor, although there is limited information regarding risk for relapses overall. Clinical presentations of relapses often differ from initial or classic symptoms of anti-NMDAr encephalitis. Relapses have been described as occurring up to 15 years later with significant recovery between presentations. Early treatment, milder disease, and malignancy resection come with a more favorable prognosis. This case is unique as relapse is less often described in pediatrics and the patient did receive immunomodulator therapy with return to baseline and thus demonstrates the need for clinical suspicion for relapse in cases where patients with a history of anti-NMDAr encephalitis present with concerning neurologic symptoms.
A 3-year-old boy presented to the Emergency Department with intermittent abdominal pain for 6 months. Additionally, he has had episodes of non-bloody non-bilious vomiting along with this pain, usually after meals. History is significant for recent travel to Africa 8 months ago. The patient appeared well, and his physical exam and vitals were completely benign. He was previously seen twice for similar complaints and was diagnosed with viral gastroenteritis both times. Abdominal radiography was obtained due to concern for constipation, and x-ray revealed a non-obstructive bowel gas pattern. However, the haustra looked thickened on x-ray, prompting a CT for further investigation (Image 1). The CT revealed a massive burden of tubular opacities consistent with Ascaris lumbricoides (Image 2). The patient was also discovered to be harboring trichuriasis. After a course of Albendazole, the patient recovered completely.
CEREBRAL VENOUS SINUS THROMBOSIS IN THE SETTING OF ACUTE VIRAL INFECTION AND IRON-DEFICIENCY ANEMIA
POSTER #30

ZACHARY SMITH, MD; PEDIATRICS RESIDENT
MENTOR: KENYA MC-NEAL TRICE, MD

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Background: Iron-deficiency is a common cause of anemia in infants and young children that is easily corrected in most cases with dietary supplementation, but that can be associated with serious sequelae. Cerebral venous sinus thrombosis (CVT) is one of the most serious complications of IDA, but the mechanisms surrounding this association remain unclear. Previous reports have proposed that a reactive thrombocytosis or the development of a hypercoagulable state may contribute to CVT in patients with iron-deficiency anemia (IDA), but no studies to date have evaluated these claims. In addition to IDA, other well-documented independent risk factors for CVT in pediatric patients include infection and dehydration. This is consistent with previous reports of CVT as a complication of IDA where patients initially presented with complaints associated with acute viral illnesses and dehydration. A number of treatment modalities are described in the setting of CVT, including thrombolysis and mechanical removal of thrombus, but the vast majority of patients in the literature are managed with long-term systemic anticoagulation. Outcomes also vary widely, ranging from full-recovery within several months to severe neurological impairment.

Case: We present a case of a 15 month old exclusively-milk-fed male with IDA who subsequently developed CVT and left thalamic stroke. Patient initially presented to a community pediatric emergency department complaining of two days of subjective fever and emesis consistent with an acute viral illness, and was subsequently diagnosed with IDA and baseline hemoglobin of 4.4 g/dL. He was discharged to home following one pRBC transfusion, but presented again within 24 hours with right-sided weakness and evidence of left-sided thalamic stroke on head CT. Following transfer to and further evaluation in a Pediatric ICU at a tertiary care facility, the patient was discharged to complete several months of systemic anticoagulation therapy. By two month follow-up, he made a near full recovery.

Conclusion: While IDA remains a common diagnosis in community and inpatient settings, the morbidity and mortality associated with certain outcomes like CVT requires practitioners to maintain a reasonable level of suspicion when managing such patients. As seen in previous cases, our patient’s CVT was also preceded by an acute viral illness, thereby underscoring the importance of also understanding other independent risk factors for this condition. Prompt recognition and management are key in the assessment of CVT and as seen here, may assist in leading to favorable long-term neurologic outcomes.
OFF-LABEL SURFACTANT USE IN PREMATURE INFANTS  
POSTER #30.5

GENNY TAYLOR, MD; NEONATAL-PERINATAL FELLOW  
MENTOR: MATT LAUGHON, MD

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Background: Surfactant is indicated for respiratory distress syndrome (RDS) in premature infants within certain gestational age (GA) or birth weight (BW) ranges, during mechanical ventilation. We hypothesize many infants receive surfactant “off-label” outside the GA or BW ranges, or without obligatory mechanical ventilation.

Objective: Characterize off-label surfactant administration in premature infants.

Design/Methods: We included all inborn infants <37 weeks gestational age who received poractant alfa, calfactant, and beractant and were discharged from Pediatrix Medical Group neonatal intensive care units between 2005 and 2013. Off-label surfactant use was defined according to FDA labeling: 1) use of any surfactant without mechanical ventilation; or 2) beractant use GA < 23 weeks or > 29 weeks, BW < 600 g or > 1750 g, or > 48 hours after birth; poractant alfa use BW < 600 g or > 2000 g, or use > 48 hours after birth; or calfactant use GA > 29 weeks, BW > 1251 g, or use > 72 hours after birth. We examined prenatal characteristics, demographics, reasons for off-label use, and clinical outcomes during the birth hospitalization.

Results: Of 91,546 premature infants that received surfactant, 56,689 (62%) received surfactant off-label. These infants were more likely to be older GA (median 32 weeks, IQR 30 - 34 weeks) and higher birth weight (median 1810 g, IQR 1335 - 2347 g) than infants who received surfactant on-label (GA 27 weeks, IQR 25 - 29; BW 945 g, IQR 750 - 1185 g). Among infants who received surfactant off-label, mortality was 5%, bronchopulmonary dysplasia (BPD) 9%, BPD or death 14%, air leak 7%, and pulmonary hemorrhage 1%.

Conclusion(s): The majority of surfactant in premature infants is used off-label. The primary reasons are higher GA and BW, followed by administration without obligatory mechanical ventilation.
A SHORT COURSE OF GAMMA TOCOPHEROL MITIGATES ENDOTOXIN-INDUCED INFLAMMATION IN HUMANS EX VIVO
POSTER #32

ALLISON BURBANK, MD; ALLERGY/IMMUNOLOGY FELLOW
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Background/Introduction: We have shown that 1 week of daily oral supplementation with 1200 mg γT attenuated endotoxin-induced airway inflammation. However, rapid onset anti-inflammatories are greatly needed to treat respiratory exacerbations. The goal of this proof-of-concept study is to determine if an abbreviated course of γT can attenuate endotoxin (LPS)-induced inflammation.

Methods: Ten healthy volunteers received 1200 mg γT orally every 12 hours for 3 doses. Serum levels of γT and active metabolite γ-CEHC were obtained at baseline, 24 hours, and 30 hours after dose #1. Peripheral blood mononuclear cells were isolated at baseline and 6 hours following dose #3 and challenged with 0 ng/mL and 0.1 ng/mL of LPS. IL-1β, IL-6, and IL-8 levels were measured following each challenge and comparisons made using a paired t-test. Pearson’s correlation coefficients were used to examine relationships between serum γT/γ-CEHC and cytokine concentrations.

Results: Three doses of γT supplementation significantly increased serum levels of γT and γ-CEHC (p<0.001 for both). γT supplementation significantly reduced LPS-induced production of IL-1β (p=0.002) and IL-6 (p=0.04) ex vivo. Both before and after supplementation, serum γT concentration negatively correlated with LPS-induced IL-1β production (r = -0.45, p = 0.04). With γT supplementation, change in γ-CEHC serum concentration negatively correlated with change in LPS-induced IL-8 production (r = -0.76, p = 0.01).

Conclusions: An abbreviated course of γT supplementation reduced inflammatory cytokine production following LPS challenge ex vivo. Moreover, γT/γ-CEHC serum concentrations negatively correlated with LPS-induced cytokine production. An abbreviated course of γT supplementation may present a feasible approach to quickly target inflammation secondary to respiratory exacerbations.
RESIDENT LEADERSHIP AND AUTONOMY AT NEONATAL DELIVERY ROOM RESUSCITATIONS
POSTER #33

ANDREW HELING, MD; NEONATAL-PERINATAL FELLOW
MENTOR: SOFIA ALIAGA, MD

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Background: A key goal of pediatrics residency training is leading multidisciplinary teams for neonatal delivery room resuscitations. Resident competency in neonatal resuscitation program (NRP) and leadership skills may be compromised by insufficient exposure to neonatal resuscitation events.

Objective: Describe patient and team characteristics of resident-attended neonatal delivery room resuscitations at a level IV NICU.

Design/Methods: We collected a longitudinal convenience sample of resident-attended neonatal delivery room resuscitations at UNC. Resuscitations were video recorded; those for which intervention was not required were excluded. Patient characteristics were identified via chart review. Team characteristics and leadership were identified via questionnaire and masked video review. Low risk deliveries were defined as: 33-36 weeks GA, minor congenital anomalies, fetal distress, meconium stained fluid, unscheduled c-section, operative vaginal delivery, shoulder dystocia or IUGR. High risk deliveries were defined as: <33 weeks GA, major congenital anomalies or emergency c-section for severe fetal distress. An existing 3-phase curriculum during each UNC NICU block encourages increasing resident autonomy at low risk deliveries. Residents receive 2 NRP-based programs each block, with each program signaling a transition between phases of resident autonomy.

Results: We analyzed 99 neonatal resuscitations (65 low risk and 34 high risk) involving NICU residents. Patient and team characteristics are described for all deliveries (Table 1) and low risk deliveries (Table 2). Residents were identified as the initial team leader in 75% of low risk and 21% of high risk deliveries. Residents maintained leadership throughout 58% of low risk and 6% of high risk deliveries. Within NICU blocks, residents increasingly maintained leadership during low risk deliveries over time (phase 1: 19%, phase 2: 62%, phase 3: 79%). Within NICU blocks, experienced supervising providers (neonatal fellows and NPs) were present at fewer low risk deliveries over time (phase 1: 100%, phase 2: 81%, phase 3: 61%).

Conclusions: Residents demonstrated increasing autonomy as evidenced by initiation and maintenance of leadership at low risk deliveries over the course of a NICU block. Residents were offered limited autonomy and leadership opportunities at high risk deliveries. To further optimize resident competency in neonatal resuscitation, supplemental learning opportunities may be needed to augment existing neonatal resuscitation exposure, particularly high risk deliveries.
TEAMWORK AND COMMUNICATION AT RESIDENT-ATTENDED NEONATAL DELIVERY ROOM RESUSCITATIONS
POSTER #34

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Background: Teamwork and communication are essential components of the neonatal resuscitation program (NRP). Multidisciplinary teamwork and communication scores during simulated neonatal resuscitations are improved following educational programs with additional emphasis on these skills. It is unknown how teamwork and communication at actual deliveries progress throughout a resident NICU rotation in the setting of a curriculum that includes educational programs that focus on these skills.

Objective: Evaluate multidisciplinary teamwork and communication at resident-attended neonatal delivery room resuscitations at a level IV NICU.

Design/Methods: Residents at UNC attend low and high risk neonatal deliveries. Low risk deliveries were defined as: 33-36 weeks GA, minor congenital anomalies, fetal distress, meconium stained fluid, unscheduled c-section, operative vaginal delivery, shoulder dystocia or IUGR. High risk deliveries were defined as: <33 weeks GA, major congenital anomalies or emergency c-section for severe fetal distress. An existing 3-phase UNC NICU curriculum offers residents 2 NRP-based educational programs with an emphasis on teamwork and communication. We collected a longitudinal convenience sample of resident-attended neonatal delivery room resuscitations at UNC. Resuscitations were video recorded. Objective assessment of teamwork and communication was assessed via a modified University of Texas Behavioral Marker Audit Form and subjective assessment of teamwork, leadership, communication and situational awareness via a modified Team Events Assessment Non-Technical Skills form. Statistical analysis was performed using one-way ANOVA and chi-squared tests.

Results: We analyzed 99 neonatal resuscitations (65 low risk and 34 high risk) involving NICU residents across 6 separate NICU blocks. Team characteristics and objective data are described for all deliveries (Table 1) and resident-led low risk deliveries (Table 2). Throughout individual NICU blocks, residents increasingly maintained leadership for all deliveries (phase 1: 23%, phase 2: 59%, phase 3: 77%). Total teamwork events did not change over time for all deliveries (phase 1: 7.0 total events/min, phase 2: 5.9, phase 3: 6.4) or for resident-led low risk deliveries (phase 1: 5.4 total events/min, phase 2: 5.6, phase 3: 5.6). Subjective assessments of teamwork, leadership and composite scores of communication and situational awareness did not change over time for all deliveries (Table 1) or for resident-led low risk deliveries (Table 2).

Conclusions: We demonstrated that resident leadership and autonomy at neonatal delivery room resuscitations within NICU blocks can be increased while maintaining both objective measurement of teamwork and communication events and subjective assessment of teamwork, leadership and communication quality, through use of a longitudinal educational curriculum stressing these skills.
THE ASSOCIATION BETWEEN ROUTINE HEAD ULTRASOUNDS AND CLINICAL INTERVENTIONS IN PREMATURE INFANTS
POSTER #35

ANDREW HELING, MD; NEONATAL-PERINATAL FELLOW
MENTORS: WAYNE PRICE, MD

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Background/Introduction: National guidelines recommend performing a routine head ultrasound (HUS) at 7-14 postnatal days in premature infants delivered at <30 weeks gestational age (GA) to screen for intraventricular hemorrhage (IVH). IVH can result in posthemorrhagic hydrocephalus (PHH) and is associated with neurodevelopmental impairment. However, there are no established treatments for IVH and therapeutic interventions for PHH typically occur only when infants become symptomatic. Premature infants who are eligible for a routine screening HUS often are scheduled for follow-up clinic visits regardless of ultrasound results.

Objective: To assess whether routine, screening head ultrasounds obtained at 7-14 postnatal days are associated with clinical interventions for infants delivered at <30 weeks GA.

Methods: This retrospective cohort study included all inborn infants delivered at <30 weeks GA between 1/1/2012 and 12/31/2015. We defined 'routine HUS' as a screening HUS performed at 7-14 postnatal days to assess for intraventricular hemorrhage (IVH). We defined 'clinical intervention' as a routine HUS followed by neurosurgical intervention before a 36-40 week postmenstrual age HUS or elective withdrawal of critical care within 30 days.

Results: We identified 337 eligible patients. Thirty-four infants were excluded; 17 infants died prior to obtaining a routine HUS, 9 had prenatally diagnosed structural CNS abnormalities and a HUS performed prior to a routine HUS, 5 never received a routine HUS, and 3 infant records were unavailable. Out of 303 included infants (Table 1), 4 (1.3%) had neurosurgical intervention prior to a 36-40 week postmenstrual age HUS; all had a diagnostic HUS performed prior to postnatal day 7 to assess for an IVH due to clinical instability. No infant had critical care electively withdrawn within 30 days of a routine HUS.

Conclusions: Clinical intervention rarely followed routine HUS studies performed at 7-14 postnatal days for inborn infants delivered at <30 weeks GA. In no case did clinical intervention occur based on findings from a routine HUS in instances where the routine HUS was the infant’s first HUS performed.
Background/Introduction: Turner Syndrome (TS) has been associated with T cell immune alterations and chronic otitis media, suggestive of a possible immune deficiency. Recently, ubiquitously transcribed tetratricopeptide repeat on the X chromosome (UTX), a histone H3 lysine 27 demethylase, was identified as a gene downregulated in TS immune cells. Mice with T cell-specific UTX deficiency (UTX-TCD) have normal clearance of acute viral infections but impaired clearance of chronic viral infection due to decreased frequencies of Tfh cells. Whether vaccine response in UTX-TCD mice and TS patients are impaired is not known.

Methods: In 15 females (7 females with TS and 8 females without TS, ages 9 to 26), total IgG response to HPV vaccine was measured via HPV 16 virus like particle (VLP) ELISA. Subjects received standard dose of HPV quadrivalent or 9-valent HPV vaccine at 0, 2 and 6 months. Serum was obtained before vaccine was administered. In 18 mice (9 UTX T cell deficient mice and 9 wild type mice), total IgG response to HPV vaccine was measured via HPV 16 VLP ELISA. Mice received 4ug per dose (1/10 of human dose) of HPV quadrivalent vaccine at 1 and 14 days. Serum was obtained two and four weeks after initial inoculation.

Results: Total IgG response to HPV 16 VLP was similar pre-vaccine and at months 2 and 6 post-vaccine in TS versus non-TS age matched controls. Total IgG response to HPV 16 VLP was similar pre-vaccine and at days 14 and 28 post-vaccine in UTX T cell deficient mice vs wild-type.

Conclusions: At these early time points, there are no significant differences in humoral response to vaccine between UTX-TCD vs wild type mice and TS subjects vs non-TS age matched controls. This may reflect a Tfh independent immune response early after vaccination. Future directions include determination of total IgG responses at 60 days post-vaccine and evaluation of IgG subsets to determine if particular subsets may be affected.
APPLICABILITY OF THE CURACAO CRITERIA IN THE DIAGNOSIS OF HEREDITARY HEMORRHAGIC TELANGIECTASIA IN CHILDREN AND YOUNG ADULTS

POSTER #36

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Background/Introduction: Hereditary hemorrhagic telangiectasia (HHT) is an inherited disorder which leads to the development of mucocutaneous telangiectasia and arteriovenous malformations (AVMs) in visceral organs. Diagnosis of HHT is either through genetic testing for the three known disease causing mutations, or clinically using the Curacao criteria. All HHT causing mutations occur in members of the transforming growth factor beta pathway, including endoglin, activin like receptor kinase I, and SMAD4. The Curacao criteria include the presence of multi-site mucocutaneous telangiectasia, recurrent epistaxis, family history, and visceral organ AVMs. Individuals whom meet three or more criteria are said to have definite HHT, two criteria as possible HHT, and one or less as unlikely HHT. The curacao criteria have been validated and are highly sensitive and specific in the adult population. Given that children with HHT are more likely to be asymptomatic when compared to adults, it is unclear if the Curacao criteria are an appropriate diagnostic tool.

Methods: We conducted a multi-center, retrospective study of patients seen at the HHT Centers at UNC, Yale, Washington University, and Cincinnati from 2002-2016. Data collected included age at evaluation, gender, Curacao criteria present, and genotype positive or negative. Patients were divided into five age groups; 0-5, 6-10, 11-15, 16-20, and 21-25 years. Sensitivity and specificity of the Curacao criteria was evaluated for each age group.

Results: A total of 196 patients were included (with data pending from Washington University and Cincinnati). 46 patients were aged 0-5, 45 patients 6-10, 59 patients 11-15, 23 patients 16-20, and 23 patients 21-25. Sensitivity was 3% for the 0-5 year age range, 15% for 6-10, 36% for 11-15, 44% for 16-20, and 70.5% for 21-25. Specificity was 100% for all age groups except for the 11-15 year olds (96%).

Conclusions: This is the largest study to date of pediatric HHT, and the first to evaluate the utility of the Curacao criteria for diagnosis of HHT in the pediatric and young adult setting. We found that the Curacao criteria had a high specificity in all age groups. Sensitivity was low in the very young subjects and improved significantly with increasing age. This is likely because of decreased frequency of disease related complication in childhood. Our findings suggest that the Curacao criteria are useful and can be applied in the pediatric setting. However, genetic testing would be preferred in asymptomatic children, particularly in the presence of a strong family history of HHT.
COMMUNITY HEALTH WORKER CASE DETECTION OF ASTHMA IN A RESOURCE-POOR COMMUNITY IN NICARAGUA
POSTER #37

MARY CROCKER, MD; PULMONOLOGY FELLOW
MENTOR: SYLVIA BECKER-DREPS, MD

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Background/Introduction: Asthma is a major cause of morbidity and mortality in children worldwide, and in Nicaragua the prevalence is estimated at 15.2%. However, many cases of asthma may remain undiagnosed, likely due in part to the overlap of symptoms with pneumonia. Community health worker (CHW) led programs have been successful in increasing case detection of childhood pneumonia, but no such programs have been validated for the detection of asthma in resource-poor settings. We hypothesize that a CHW administered questionnaire will be effective in case-detection of asthma in a semi-urban, poor Nicaraguan community.

Methods: We sought to enroll every child aged 2-17 in Vladimir Hernandez, a small community near Managua, Nicaragua. Presence or absence of asthma was established through clinical evaluation by a pediatric pulmonologist, including spirometry. Separately, a trained CHW administered a screening questionnaire based on a previously validated school-based questionnaire developed in Argentina, consisting of 11 symptom-focused questions. Sensitivity and specificity of the questionnaire were assessed, and risk factors for asthma were explored.

Results: We enrolled 199 out of 218 eligible children during a 4-week period in summer 2016. Asthma prevalence was measured to be 33%. The risk factor most closely associated with asthma diagnosis was respiratory infection in the first 3 months of life (p=0.02); parasite infection, crowding, secondhand smoke exposure, mold or animal exposure, and indoor cookstove use were not significant. Mean scores on the CHW questionnaire were 3.6 points (95% CI 2.9-4.3) for children without asthma and 11.0 points (95% CI 9.7-12.3) for children with asthma, and were significantly different (p<0.0001). Various cut-off points for scoring the questionnaire were tested. Using a cutoff of 5 points (out of 22 possible), sensitivity of the CHW questionnaire was 89% and specificity was 70%, compared to pulmonologist evaluation as a gold standard. An ROC curve was generated with AUC = 0.868, indicating a good screening test.

Conclusions: The prevalence of asthma in Vladimir Hernandez, Nicaragua determined by pediatric pulmonologist evaluation was high compared to previously determined national prevalence, possibly due to increased exposure to risk factors including low socioeconomic status, use of biomass fuel, or ambient particulate matter. The community health worker questionnaire had high sensitivity and area under the ROC curve, making it an ideal screening tool. This questionnaire could be administered by lay health workers without the need for a physician, and could greatly increase the detection of asthma, allowing for education and referral for ongoing care.
DEVELOPMENTAL AND HEALTH CORRELATES OF OBESITY AMONG TEN-YEAR-OLD CHILDREN BORN EXTREMELY PREMATURE
POSTER #38

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Background/Introduction: Infants born extremely premature (EP) exhibit growth delay during the first few postnatal months, which is often followed by accelerated growth. This “catch-up growth” is associated with better cognitive outcomes in early childhood, but might also lead to obesity, itself a known risk factor for cognitive problems and lower school achievement. Limited study has investigated school-age developmental and health correlates of obesity among children born EP.

Methods: The ELGAN study is a multi-center prospective, observational study of EP infants born in the years 2002-2004. This investigation included 871 children who were evaluated at 10 years using neurocognitive testing, parent-reported health outcome surveys, and height and weight measurements. Per standard definitions, overweight was defined as BMI centile ≥85 and <95 and obese as ≥95. For neurocognitive tests, a low score was defined as a Z score ≤ -1. To describe the strength of association between overweight/obesity and adverse outcomes at 10 years, we used logistic regression models adjusting for confounders.

Results: Among 10 year-old children born EP, the health and neurodevelopmental outcomes of overweight and obese children were similar to those of healthy weight EP peers. BMI category at 10 years of age was not associated with differences in intelligence, language, or academic achievement test scores (Figure 1). Children with BMI centile ≥85 had lower communication skills (Figure 2), and their parents more often reported poorer quality of life on social function scales. Parents’ rating of their child’s general health was more likely to be less than “good” with increasing BMI, and obese children were more likely to be diagnosed with, and prescribed medication, for asthma.

Conclusions: Children born EP with higher BMI have similar neurocognitive skills as their peers at 10 years of age. As in other cohorts, obese EP children were more likely to have asthma. Overall, these findings provide reassurance that excessive catch-up growth, which may contribute to obesity, does not confer adverse effects on neurocognitive outcomes later in childhood. Further study is needed for individuals with obesity born EP to evaluate for cardiovascular and metabolic outcomes that typically manifest later in life.
SUCCESSFUL USE OF SIROLIMUS IN A 5 MONTH OLD WITH PROS DUE TO SOMATIC PIK3CA MUTATION

POSTER #39

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The phosphatidylinositol 3-kinase /AKT/mTOR pathway regulates cell metabolism, growth, proliferation, and survival. Mutations in the mTOR (mammalian or mechanistic target of rapamycin) signaling pathway are commonly identified in various tumors or cancers, and inhibitors of the pathway have been used in targeted treatment in this context. A spectrum of segmental overgrowth phenotypes including PIK3CA-Related Overgrowth Spectrum (PROS), Proteus syndrome, and Congenital Lipomatous asymmetric Overgrowth of the trunk with lymphatic, capillary, venous, and combined-type Vascular malformations, Epidermal naevi, Scoliosis/Skeletal and spinal anomalies (CLOVES) syndrome result from activation of this pathway due to mosaic mutations in affected tissue. A current clinical trial at the NIH is investigating the use of sirolimus amongst patients 3 years and older who have confirmed somatic PIK3CA mutations as a systemic therapy. The purpose of the trial is to determine whether sirolimus can reduce or prevent overgrowth and associated complications. Unfortunately, no trials exist for patients less than 3 years of age and the only currently established management for these patients is serial debulking, amputation and vascular intervention techniques. We report a successful trial of sirolimus therapy in a 5 month old who met clinical diagnostic criteria for PROS due to a somatic PIK3CA mutation. Based on our results, we suggest a protocol for obtaining baseline imaging and laboratory studies, dosing sirolimus, monitoring for adverse reactions, and clinical follow-up for patients under 3 years of age. We plan to expand treatment to other patients who could benefit from sirolimus therapy to spare them the medical, cosmetic, and social consequences of surgical therapy.
Infants who contract Hepatitis B (Hep B) have a 90% risk of developing chronic Hep B, which can lead to complications such as cirrhosis, hepatocellular carcinoma and death. The American Academy of Pediatrics (AAP) and the Advisory Committee on Immunization Practices (ACIP) strongly recommend that the 3-dose Hep B vaccine course be commenced at birth to prevent acquisition of Hep B.

This retrospective case-control series includes infants born at North Carolina Women’s Hospital between January 1st and December 31st, 2011. The “not vaccinated at birth” (nVaB) group was defined as all infants who did not receive the birth dose of Hep B by seven days of life and the “vaccinated at birth” (VaB) group consisted of a random sample of infants who did receive the birth dose of Hep B by seven days of life. Data were collected through the Carolina Data Warehouse for Health (CDW-H) and sent to the North Carolina Immunization Registry (NCIR) to determine each infant’s vaccination status. Completion of the primary vaccination series by 18 months of age was defined as follows: Hepatitis B (3 doses), Rotavirus (2 to 3 doses), Diphtheria, Tetanus & acellular Pertussis (DTaP) (4 doses), Haemophilus influenza type b (Hib) (3 to 4 doses), Pneumococcal conjugate (PCV13) (4 doses), Inactivated Poliovirus (IPV) (3 doses), Measles, Mumps and Rubella (MMR) (1 dose), and Varicella (1 dose). Incomplete vaccination was defined as failure to receive one or more of the above recommended vaccinations by 18 months of age.

1495 infants were included in the initial chart review. Vaccination data were not available for 59 infants (4% of total). Of 696 infants in the nVaB group, 163 (23%) completed the primary series by 18 months of age compared with 326 (44%) of 740 in the VaB group (p<0.001). Infants in the VaB group were more likely to complete the 3-dose Hep B series (88% v 64%, p<0.001).

Infants who miss the birth hepatitis B vaccine are at risk for incomplete vaccination at 18 months of age – both with the Hepatitis B series and the full primary vaccine series. Determining why these patients miss the birth dose and how that relates to missing subsequent doses is a vital question for future research.
IN-HOSPITAL OUTCOMES OF PREMATURE INFANTS WITH SEVERE BRONCHOPULMONARY DYSPLASIA
POSTER #41

WESLEY JACKSON, MD
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Background/Introduction: Bronchopulmonary dysplasia (BPD) is the most common pulmonary complication of prematurity. Compared to infants with mild or moderate BPD, infants with severe BPD who require positive pressure ventilation at 36 weeks postmenstrual age (PMA) are more likely to be rehospitalized for pulmonary causes/complications following discharge and have more severe neurodevelopmental impairment at 18-22 months of age. In this population, mortality, co-morbidities, and short-term outcomes are not well-defined.

Methods: We identified all inborn infants <30 weeks gestational age with severe BPD, defined as receiving positive pressure ventilation at 36 weeks PMA, discharged from 348 Pediatrix Medical Group neonatal intensive care units from 1997-2015. We examined prenatal characteristics, demographics, interventions including medications used and level of respiratory support, and clinical outcomes during hospitalization. We compared distributions of study variables across categories using Wilcoxon rank sum, Chi square, and Fisher’s exact tests where appropriate.

Results: There were 10,752 infants with severe BPD, and 549/10,752 (5%) died prior to discharge. Infants who died were more likely to be male, small for gestational age, received more medical interventions, and more frequently diagnosed with surgical necrotizing enterocolitis, culture-proven sepsis, and pulmonary hypertension following 36 weeks postmenstrual age (PMA) compared to survivors. At the time of discharge, 5855/10,203 (57%) of infants were receiving supplemental oxygen, 302/10,203 (3%) had a tracheostomy, 1019/10,203 (10%) a gastrostomy tube, and 332/10,203 (3%) were receiving mechanical ventilation. Approximately 70% of infants with severe BPD were discharged by 44 weeks PMA, and 86% were discharged by 48 weeks PMA. Of the 2525 infants receiving mechanical ventilation at 36 weeks PMA, 1860 (74%) were discharged home by 52 weeks PMA, and 336 (13%) died prior to discharge.

Conclusions: Approximately 95% of infants with severe BPD survived to discharge, and the majority of these infants were discharged home by 44 weeks PMA. A minority of infants received tracheostomy and/or gastrostomy tube prior to discharge. These findings provide valuable information for families and caregivers about the expected hospital course of infants diagnosed with severe BPD.
ASSOCIATION OF BACTERIAL INFECTION AND OBSTRUCTIVE LUNG DISEASE IN YOUNG CHILDREN WITH RECURRING WHEEZING
POSTER #42

WILLIAM STOUDMIRE, MD; PULMONOLOGY FELLOW
MENTOR: CHARLES ESTHER, MD, PHD

Background/Introduction: Protracted bacterial bronchitis (PBB) is a relatively common cause of recurrent wheezing and cough in young children, with 40-60% of these patients having positive bacterial cultures at the time of bronchoscopy. However, the pathophysiological consequences of PBB are not well understood. We examined the short and long term impact of PBB on lung function. Methods: The UNC pediatric bronchoscopy database was used to identify all children who underwent bronchoscopy with bronchoalveolar lavage as preschoolers (<5 years old) for evaluation of recurrent cough or wheeze from 2002-2009. Children with identified systemic illness were excluded. Patient characteristics, bronchoscopic findings, as well as rates of bacterial, viral, and mycobacterial infections were collected. The subset of patients who underwent infant spirometry at that time of bronchoscopy was identified, and multiple regression was utilized to analyze the relationship between lung function and PBB controlling for age, sex, and the presence of airway malacia. Long term impact of PBB was assessed in a different subset of patients who had spirometry performed in later childhood (>5 yrs old).

Results: Positive bacterial cultures were observed in 209/358 (58.1%) of preschool children who underwent bronchoscopy for recurrent cough or wheeze. Moraxella catarrhalis was the most commonly identified bacteria (31.9%), followed by Haemophilus influenzae (24.4%) and Streptococcus pneumoniae (21.3%). In patients with infant lung pulmonary function testing at time of bronchoscopy (n=29), bacterial infection was associated with significantly lower FEV0.5 (p=0.02) and FEF25-75% (p <0.01). Bacterial infection at the time of bronchoscopy was also associated with decreased lung function later in childhood (n=16), with significantly lower FEV1 (84.2% vs 103.8%, p<0.02) and lower FEF25-75% (67.3% vs. 103.1%, p<0.02) compared to those without bacterial infection.

Conclusions: Consistent with previous studies, PBB was found in 58.1% of infants and young children with recurrent wheezing or cough. Presence of bacterial infection was associated with obstructive lung disease, as patients with bacterial infection had significantly lower FEV0.5 and FEF25-75% by infant pulmonary function tests at time of bronchoscopy. Furthermore, patients with PBB at preschool ages had significantly lower percent predicted FEV1 and FEF25-75 on future spirometry. These results suggest that bacterial infection in young children may play a role in the development of airway obstruction later in life.
REPRODUCTIVE CONCERNS IN PATIENTS WITH CYSTIC FIBROSIS, A QUALITATIVE STUDY
POSTER #43

CLAIRE HAILEY, MD CANDIDATE
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Background/Introduction:
Treatment advances in cystic fibrosis (CF) have increased the median predicted survival to 40 years.[1] With increased life expectancy, more people with CF are considering becoming parents.[2] Data suggest that women with CF lack knowledge about reproductive health in the context of CF [3] and that there are deficiencies in reproductive health education for men with CF.[4] Clinical experience corroborates these deficiencies and suggests that many patients grapple with reproductive decisions or have made decisions without standardized education or resources. The objective of our research is to better understand the reproductive and parenting concerns of people with CF. We hypothesized that participants would report wanting more provider-initiated education on reproductive health topics and would endorse specific reproductive health and parenting concerns unique to their illness.

Methods:
In this ongoing study, 19 adults with CF have participated in in-depth, semi-structured interviews about reproductive health and parenting concerns, including knowledge of reproductive health in CF, psychosocial adaptation to this chronic illness in regard to fertility and parenthood, parenting concerns related to CF and its treatments, and supportive care needs around reproduction. Interviews were audiotaped, transcribed, and analyzed using thematic content analysis. We used descriptive statistics to describe participants’ demographics and health variables.

Results:
Of the 19 participants (mean age=32, range: 22-44), 10 are parents and 9 do not have children. We have enrolled 9 females and 10 males. The average FEV1 % predicted is 67 (range: 30-105). Seven participants had never discussed reproductive health with a CF care provider, and the remaining 12 reported varied depth and content of conversations. Participants described concerns about fertility, the heritability of CF, and the remaining 12 reported varied depth and content of conversations. Participants described concerns about fertility, the heritability of CF, and several concerns related to the impact of their illness on their children including emotional, financial, anticipated health decline and mortality concerns. Nearly all parents described concerns related to illness-related communication with their children. Participants provided recommendations for enhancing CF-related reproductive health education, such as who should initiate reproductive health conversations and what topics should be discussed.

Conclusions:
Results from this study can provide guidance for CF providers to address reproductive health concerns with their patients. Future research interventions to address reproduction and parenting topics may lead to improved education and support for patients with CF and their families.
PREVALENCE OF PEDIATRIC FUNCTIONAL GASTROINTESTINAL DISORDERS ACCORDING TO THE ROME IV CRITERIA
POSTER #44

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BACKGROUND: Pediatric Functional Gastrointestinal Disorders (FGID), such as Irritable Bowel Syndrome (IBS) and Cyclic Vomiting, cannot be diagnosed based on laboratory testing and therefore symptom-based criteria have been developed. Recently an update to these criteria (Rome IV) has been published which includes the addition of new diagnoses such as functional nausea. The goal of the current study is to assess the prevalence of FGIDs in children 0-18 years old according to the new Rome IV diagnostic criteria in a representative community sample.

STUDY DESIGN: Mothers (n=1255) of children aged 0-18 years old in the US were recruited to complete an online survey about their child’s gastrointestinal symptoms, quality of life, and other health conditions.

RESULTS: Based on the Rome IV criteria, 24.7% of infants and toddlers aged 0-3 years and 25.0% of children and adolescents aged 4-18 years fulfilled symptom-based criteria for an FGID. The most common FGID were Infant regurgitation among infants (24.1%), functional constipation among both toddlers (18.5%), and children and adolescents (14.1%). Quality of life was diminished in pediatric patients with FGIDs for toddlers (M= 79.41 vs. M=88.61, p<0.001) and children 4 years and older (M=66.4 vs. M=82.8, p<0.001). Children 4 years and older were more likely to qualify for an FGID if their parent qualified for an FGID (p<0.001).

CONCLUSION: This is the first study to report the prevalence of pediatric FGIDs based on Rome IV criteria. FGIDs are common in pediatric populations of all ages and are associated with decreased quality of life. These data are of importance to treatment and research of FGIDs.
RAPID DETECTION OF PEDIATRIC BACTERIURIYA USING NARROW ANGLE FORWARD LASER SCATTERING TECHNOLOGY (NAFLST) WITH BACTERIOSCAN
POSTER #45

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Introduction: Pediatric UTI is a common diagnosis. However, culture data can take up to 48 hours, delaying diagnosis and exposing pediatric patients to antibiotics unnecessarily. The sensitivity of screening urinalysis (UA) can be low, and false positives can be common, depending on collection method (i.e. clean catch). Therefore, rapid detection of bacteriuria by another means would be beneficial.

Narrow Angle Forward Laser Scattering Technology (NAFLST) with Bacterioscan can promptly identify bacterial growth in a liquid sample. Through this technique, a laser beam is shown through a liquid sample containing replicating bacteria in nutrient broth. Over time as bacteria replicate in the media, the laser beam light is refracted. Higher degrees of light refraction represent higher initial bacterial load and continued bacterial growth. The amount of optical scatter is graphed over time by the machine, allowing Bacterioscan to identify which samples are “Likely Positive” or “Likely Negative” in approximately 3 hours.

We compared Bacterioscan results to urine culture data to determine if it could be an effective screening tool to rapidly exclude bacteriuria and avoid unnecessary urine culture.

Methods: This protocol was reviewed and approved by the UNC Biomedical Institutional Review Board. A total of 169 pediatric (<18 yo) urine culture samples were collected as part of routine patient care at UNC Health Care from 1/11/2017-2/17/17. A standardized amount of an individual urine sample and 2.5mL of Sterile Tryptic Soy Broth (TSB) were pipetted into a Bacterioscan micro-curette. These results were then compared with urine culture results obtained by routine microbiologic methods. Results: 169 urine samples were analyzed. When considering the performance of NAFLST in identifying any growth on urine culture (Trial 1), 68 samples were positive, and Bacterioscan labeled 86 cultures as “Likely Positive”. 55 cultures yielded no growth, and Bacterioscan labeled 83 as “Likely Negative”. This yielded a sensitivity of 70.8%, specificity of 75.3%, PPV of 79.1%, and a NPV of 66.3%. When the results were limited to detection of only clinically relevant/pathogenic growth (Trial 2), 27 cultures were positive, and Bacterioscan identified 86 as “Likely Positive”. 83 cultures yielded no growth, and Bacterioscan identified all 83 samples as “Likely Negative”. This yielded a sensitivity of 100%, specificity of 58.4%, PPV of 31.4%, and NPV of 100%, respectively.

Conclusions: By rapidly identifying urine cultures likely to be positive, NAFLST with Bacterioscan can obviate the plating of every urine sample and reduce empiric antibiotic use while waiting for culture results.
UN-LINQED: SPONTANEOUS EXTRUSION OF NEWER GENERATION IMPLANTABLE LOOP RECORDERS
POSTER #46

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INTRODUCTION: Insertable cardiac monitors (ICMs) have been used for years for long-term monitoring of cardiac rhythm in children with or without known arrhythmias or structural heart disease. The Medtronic’s LINQ Reveal™ is a new generation wireless, automated, and patient responsive subcutaneous ECG monitoring device measuring just 1cc in size. Despite several advantages to a small size we have noted an unusually high incidence of extrusion in our center.

METHODS: We conducted a retrospective case note analysis to review all new generation Reveal LINQs extruded at our center. All devices were inserted using the provided insertion and incision tools. Patients with extruded devices were identified and details relating to the implantation, namely the site of insertion, technique utilized for insertion and wound closure, use of peri or post-operative antibiotics and follow up details were noted.

RESULTS: 81 patients underwent 85 Reveal LINQ implants at a tertiary care University Hospital referral center between March 2014-Oct 2016. The most common reason for implant was suspected arrhythmia with or without structural heart disease or unexplained syncope. All implants were performed using sterile technique in electrophysiology/catheter suites. There were 4 spontaneous extrusions of devices occurring within 7-24 days after insertion with an incidence rate of 4.7%. There was slight variability in the technique of implant noted in each case. Variation in techniques included use of subcutaneous sutures in combination with a topical skin adhesive or paper based butterfly closures or the use of a topical skin adhesive alone without subcutaneous sutures. In one case with a device extrusion the device was anchored down with absorbable suture material. One patient was on chronic anticoagulation, however, the wound was noted to be hemostatic despite the extrusion. All extrusions were reported to be pain free. There were no device or pocket infections noted in any case.

CONCLUSIONS: Device migration and erosion through the skin are included as potential adverse events for this device. Skin closure and securing of this device is a variable process that has been shown to be largely physician dependent which is higher than the incidence with older generation larger devices. The authors feel that the higher incidence of the new device extrusion compared to the older generation devices is in part related to the rectangular contour of the device and the pressure effect of the head of the device. This holds especially true in children with relatively scanty subcutaneous fat and tissue. Proposed measures to avoid spontaneous extrusion in this population includes design of a longer tool in order to increase the skin to device distance.
MEASURING THE ACCURACY OF THERMOMETRY MODALITIES IN HEALTHY NEONATES
POSTER #47

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Background: Rectal thermometry (RT) is the gold standard for measuring temperature in children under age 3, according to the AAP. However, the use of temporal artery thermometry (TT) is increasing, specifically with Exergen models. Within healthy neonates, we aim to (1) determine the accuracy of mean TT and mean axillary thermometry (AT) when compared to mean RT measurements and (2) determine the reliability of each of the three modalities by comparing variations between the dual TT, AT, and RT measurements.

Methods: Newborn patients in the UNC Healthcare Children's newborn nursery were recruited based on the following criteria: newborns born between July-October 2016, gestational age ≥ 37 weeks, lack of any single documented temperature ≥100.4° F, and chronological age 12-72 hours of life. During the recording of routine vital signs on each subject, trained nurse providers measured two temperatures each via TT, AT, and RT modalities within a 15-minute interval, using the Exergen TAT-2000C for TT measurements, and Welch Allyn SureTemp Plus 690 for AT and RT measurements (this instrument has interchangeable probes to measure AT and RT). To assess accuracy, a linear mixed effects model was fit to the data to determine if the mean temperatures differed across modalities. To assess reliability, the absolute values of the residuals from the mean model were analyzed with a subsequent linear mixed model.

Results: 245 patients were approached. 205 consented and met criteria for final analysis (6 temperatures each, 1230 total measurements). For accuracy, TT differed from the gold standard mean rectal measurement, overestimating temperature by 0.25° F on average; AT and RT measurements did not significantly differ, with their means being only 0.02° F apart. For reliability, the variability of the modalities differed significantly, with RT measurements differing by 0.44° in either direction of the model’s predicted mean for each patient, compared to differences of 0.31° and 0.33° for AT and TT (respectively).

Conclusions: AT demonstrated the best combination of accuracy and reliability. It had equal accuracy when compared to the known gold standard, RT, with TT being the least accurate. AT also demonstrated great reliability in obtaining consistency between an individual subject’s two AT measurements. Our results endorse AT as the method of choice for measuring temperatures in the newborn nursery setting.
ATTITUDES, MOTIVATIONS, AND EXPERIENCES OF UNC HOSPITAL VOLUNTEERS
POSTER #48

PAULA GOMEZ, UNDERGRADUATE
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Background/Introduction: Many hospitals have a volunteer services, yet there is limited data on the characteristics, experiences, and contributions of the volunteers. This study describes characteristics of volunteers in one academic medical center and reports on their motivations for volunteering, knowledge, confidence and experiences. Further, the study compares the similarities and differences between general UNC Hospital Volunteers and those who volunteer through Carolina Conexiones, a program that specifically trains bilingual volunteers as patient navigators.

Methods: We sent a web-based survey to all volunteers at UNC’s Women’s and Children’s Hospital in fall 2016. Survey items assessed respondents’ motivations for volunteering, experiences, and self-reported knowledge and confidence with health care-related skills. Likert scales (5 points) were used to assess agreement with survey items. We used descriptive statistics to examine volunteer characteristics and experiences. Bivariate analyses were used to compare the relationship of volunteer characteristics and experiences for general Women’s and Children’s Hospital volunteers with those in the bilingual patient navigator program.

Results: We sent 417 surveys and received 145 responses (response rate of 35%). A majority (86%) of volunteers were female and 47% reported being enrolled in a graduate or undergraduate degree program. The average number of hours per month spent volunteering was 10.7 hours per month (range=0-30). Volunteers strongly agreed that interacting with patient and families (86%) and serving the community (84%) were main motivations for volunteering. 48% plan to pursue a health care career in the future. The volunteers reported that their experiences included continuously improving experience for patients (62% strongly agree). Volunteer confidence and knowledge were highest for ability to work as part of a health care team (48% and 31% strongly agree, respectively). Volunteer confidence and knowledge were lowest for taking care of limited English proficiency patients (21% and 11% strongly agree, respectively). Carolina Conexiones volunteers witnessed higher barriers to health care due to lack of English proficiency (43%) when compared to the general hospital volunteers (29%).

Conclusions: Volunteers contribute a diverse set of backgrounds and experiences to healthcare environments. Their perceived core contributions include service, and many intend to eventually join the paid healthcare workforce. Experience with working on healthcare teams provides valuable preparation for volunteers who join the permanent healthcare workforce. Opportunities exist for maximizing the skills and effectiveness of volunteers by assisting them with the areas where they feel least skilled and confident, such as in service to patients with limited English proficiency.
EVALUATION OF DISEASE SEVERITY AND QUALITY OF LIFE IN PEDIATRIC EOSINOPHILIC ESOPHAGITIS
POSTER #48.5

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Background/Introduction: Eosinophilic esophagitis (EoE) is a relatively new and increasingly common chronic inflammatory disease of children and adults. There have been a wide range of studies on health-related quality of life (HRQoL) in pediatric chronic disease, but HRQoL in pediatric EoE has been less extensively investigated. To date, a study showed that HRQoL negatively correlates with increased symptom severity. However, it is unknown if this reflects disease severity. The purpose of this study was to assess to what degree health-related quality of life is associated with disease severity in pediatric patients with eosinophilic esophagitis.

Methods: This was a prospective cross-sectional study of children age 7-18 years of age diagnosed with EoE as per consensus guidelines at a tertiary care center. Participants completed the PedsQL EoE Module (PGN, 2013, 57, 57-66) before they underwent endoscopy. Peak eosinophil count and EoE Endoscopic Reference Score (EREFS) were used to grade disease severity.

Results: We enrolled 23 children. The mean age was 12.09 (SD=3.01); 19 (82.6%) participants were male and 4 (17.4%) were female. At baseline, the mean EREFS was 1.68 (SD=1.36), the mean of the peak eosinophils was 36.74 eos/hpf (SD 41.68), and the mean QL score was 72.60 (SD=18.28). Peak eosinophil count (r=.184, p=.279) and EoE Endoscopic reference score (r=.130, p=.413) were not significantly correlated with quality of life. Quality of life was also not correlated with edema (r=.102, p=.306), rings (r=-.109, p=.306), exudates (r=.210, p=.512), furrows (r=.097, p=.341), or strictures (r=.169, p=.341) found on endoscopy. The total HRQoL scores were also not correlated with dietary restrictions or food allergies (r=-.285, p=.344). Most of these correlations are so small low power is likely not the reason for insignificance.

Conclusions: This small prospective study indicates that disease severity as measured by endoscopic findings and eosinophil count is not related to quality of life in children with EoE. Future studies should examine if other clinical factors (e.g., disease management strategies) and psychological factors impact HRQoL in pediatric EoE. This information is important for clinical treatment and development of treatment trials in pediatric EoE.
RACE, PLACE, AND DEPRESSION: ADOLESCENT DEMOGRAPHICS AND DEPRESSION IN NC
POSTER #48.75

EMILIE KADHIM
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UNC UNDERGRADUATES
MENTOR: TAMERA COYNE-BEASLEY, MD, MPH

**Background**
- Suicide is the second leading cause of death for adolescents aged 10-24 in the US, and self-harm is second for those ages 10-19
- Both suicide and self-harm are preventable and associated with depression. Depression in adolescents and young adults is underdiagnosed and undertreated
- Intersectionality in race, gender, place, and socio-economic status create multidimensional and interplaying layers of disadvantage that may impact depression

**Methods**
- Data were obtained from patients, ages 11-21 from 2012-2016
- A PHQ9 score of ≥10 indicated a positive screen for depression
- Insurance status was used as a proxy for socioeconomic status
- Sample characteristics were assessed with univariate analysis, bi-variate chi-square analyses were utilized to assess associations of depression with demographics and location

**Results**

**Results cont.**
- Depression was significantly positively correlated with being female (OR=2.36, p<0.041), Hispanic (OR=2.92, p<0.048), and on public insurance (OR=2.29, p<0.041)
- There is a statistically significant relationship between insurance status and geographic location (p<0.031)
- More adolescents screened positive for depression (score ≥10) category who are from Chapel Hill and Durham (24.49% and 26.77% respectively), than other locations (Raleigh, Hillsborough, and Burlington)

**Conclusions**
- In the sample of 939 adolescents, females, people of Hispanic origin, uninsured/publicly insured individuals, and those in Durham, NC and Chapel Hill, NC were the most likely to report depressive symptoms
- Hispanic publicly insured females were the most likely to have the highest PHQ9 scores. These findings call for an increase in attention to adolescent groups that experience multiple facets of disadvantage and depression

**Race, Place, and Depression: Adolescent Demographics and Depression in NC**
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**Positive Depression Screen by Race**

**Depression by Gender and Race**
A QUALITY IMPROVEMENT APPROACH TO DESIGNING AND IMPLEMENTING AN OUTPATIENT EDUCATION PATHWAY FOR NEWLY DIAGNOSED TYPE 1 DIABETES
POSTER #49

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Background/Introduction: The significant emotional/psychological stress associated with a new diagnosis of Type 1 Diabetes (T1DM) may be exacerbated by the historic process of hospitalization after diagnosis. As of November 1, 2016, UNC Pediatric Endocrinology transitioned to an outpatient diabetes education model for patients with newly diagnosed T1DM. Given that T1DM is a chronic illness that requires continued self-management to optimize health status, self-management strategies are essential components of diabetes education provided at diagnosis. To optimize the outpatient diabetes education model, we implemented an improvement initiative to redesign the outpatient care processes, refine patient education content, and identify ideal educational strategies. Our aims were to achieve patient self-management, reduce stress and ensure patient/family and provider satisfaction with the outpatient pathway.

Methods: A multidisciplinary team was formed to implement the improvement initiative. A 3-day outpatient diabetes education pathway, including criteria for eligible patients, was developed based on the education previously provided in both the inpatient and outpatient settings. Using formal QI methods and tools, the initiative focuses on redesigning the content and format of the pathway based on results from key measures and individual PDSA cycles. Patient and family input into content and format is integral to our PDSA cycles. Primary outcome measures include: self-efficacy, stress and satisfaction. Key process indicators include knowledge and presence of a school plan. School absences and unplanned readmissions and ED visits are monitored as balancing measures and investigated for opportunities to improve the pathway.

Results: This is a Quality Improvement initiative in the early stages. Initial baseline data for the outpatient program has been collected for parent- and adolescent patient-reported self-efficacy, stress, and satisfaction. Ongoing monitoring for unplanned ED visits and admissions identified the need for closed loop communication between the provider and CDE at all education visits. A PDSA cycle was conducted and the pathway modified. There have not been any return ER visits for other patients since that time. Formal feedback identified that the Day 1 plan was overwhelming for patients and families, because it included a full discussion about nutrition at the end. Another PDSA cycle resulted in a pathway modification, the addition of an additional education day, so that Day 1 could focus on Introduction to Diabetes and Survival Essentials, and Day 2 would focus on Nutrition and Meal Planning.

Conclusion: We report the initial stages of a quality improvement initiative aimed at reducing stress and improving patient self-management for newly diagnosed patients with Type 1 Diabetes Mellitus.
THE SIMULATED FAMILY MEETING: A STANDARDIZED APPROACH TO A COMMUNICATION CURRICULUM AND ASSESSMENT OF THE ACGME PEDIATRIC MILESTONES
POSTER #50

JENNIE HART, MD; HEMATOLOGY-ONCOLOGY FELLOW
MENTORS: ELISABETH DELLON, MD AND BENNY JOYNER, MD

Additional Authors; Division/Institution: Jennifer McEntee, Eric Zwemer, Kenya McNeal-Trice, John Hipps, Pediatric Hematology/Oncology

Background: Communication skills are an important competency in the evaluation and certification of residents. There exists difficulty in assessing subjective competencies between physician and patient. Simulation provides a platform for practicing these skills and reliably assessing residents’ performance in interpersonal communication.

Objective: To use simulation and didactic sessions to teach residents how to lead a family meeting and discuss transitions to end of life care. To use simulation to evaluate the ACGME Milestones.

Methods: Residents conducted two simulated multidisciplinary family meetings before and after a live didactic session for leading a family meeting and an online palliative care lecture. One reviewer assessed the residents’ behavior on a checklist for each simulated encounter. Residents self-evaluated their confidence in leading family meetings at the start and completion of the curriculum. Two faculty members evaluated residents’ performance on Milestones, PROF6 and ICS2, for each simulation.

Results: All thirteen residents who completed the curriculum demonstrated improvement in the number of items observed on the behavioral checklist. Of 24 possible items, an average of 12 were observed for the first simulation and 17 for the second. All residents reported greater self-confidence on a scale of 1-4 with a mean improvement in overall confidence by 0.7. Confidence increased on an average of 8 of 13 items. The most common confidence level for the first simulation was 2 and the most common for the second simulation was 3. Five residents showed improvement on PROF6, while two residents received a lower score on the second simulation. Four residents showed no change in score on ICS2, two residents improved and one resident received a lower score on the second simulation. Statistical analysis and data collection is in progress.

Conclusions: Given the improvements in observed behaviors and self-confidence at the start and completion of the curriculum, we conclude that the combination of didactic sessions and simulated experiences is an effective method for teaching residents to lead family meetings. Since the Milestones are a longitudinal evaluation, a significant change in resident performance may not be observed in a short time period. Our data suggests that an intensive curriculum may accelerate residents’ progress on some Milestones. This curriculum provides a foundation for assessing Milestones and mapping them to specific skills. As more data is collected, we expect to make a stronger conclusion about resident performance on the Milestones and the role simulation plays in teaching and evaluating communication skills.
SHORT HANDBOOK IMPROVE SEPSIS KNOWLEDGE IN PEDIATRIC HEMATOLOGY-ONCOLOGY CAREGIVERS
POSTER #51

MARK DEXTER, MD; CRITICAL CARE FELLOW
MENTORS: REBECCA SMITH, MD

Additional Authors; Division/Institution: Hillary Spangler, Sean Miller / UNC School of Medicine, Sheelah Shortell / UNC Pediatric Hematology-Onecology

Background/Introduction: Sepsis is a common, world-wide medical problem with significant morbidity and mortality. Over the last several years, there has been a push to improve quick recognition and treatment of sepsis by medical providers, but patient education about sepsis has not been studied.

Methods: A group of experts met and devised a brochure containing information about sepsis to be given to caregivers of patients in the pediatric hematology-oncology outpatient clinic. Several PDSA cycles were done to improve the delivery of these materials, while caregiver knowledge about sepsis was tracked by pre- and post-education surveys. Retention of knowledge was measured by e-mail survey approximately 30 days after initial education.

Results: Caregiver self-reported knowledge about sepsis improved by 4.98 points (95% CI: 4.44-5.51) on a 16 point summed Likert-like scale after participating in electronic presentation education in hematology-oncology clinic. Sixteen percent of caregivers completed follow-up at 30 days post-education; their knowledge of sepsis, while lower than immediately posteducation, remained 4.98 points (95% CI: 3.53-6.40) higher than baseline on a 16 point summed Likert-like scale.

Conclusions: Education delivered by brochure in the outpatient setting increases caregiver knowledge about sepsis, and this increased knowledge is sustained even 30 days post-education. Further studies are indicated to determine whether these findings can be replicated in other pediatric populations, and whether this education results in improved outcomes for pediatric sepsis patients.
USE OF UNFRACTIONATED HEPARIN LEVELS FOR HEPARIN TITRATION AND ANTICOAGULATION MANAGEMENT IN PEDIATRIC EXTRACORPOREAL LIFE SUPPORT
POSTER #52

MELISSA CROWDER, MD; CRITICAL CARE FELLOW
MENTOR: KATHERINE CLEMENT, MD

Jackie Patterson, MD, Ann Marie Castleman, MPH, Ana Williams, DDS, Laura Parajon, MD, & Carl Bose, MD

Historically, anticoagulation for all extracorporeal life support (ECLS) patients at UNC Medical Center was monitored using activated clotting time (ACT). The ACT has been the gold standard for anticoagulation monitoring for ECLS patients of all ages in all centers due to its historical use in cardio-pulmonary bypass surgeries [1]. Unfortunately over the last few years, ACT testing seems to have become a less reliable strategy for heparin titration [2]. Due to clinical observation of the inaccuracy of ACTs in our own unit, it has been difficult to standardize ACT driven anticoagulation using our existing standardized clinical pathway. Outcomes from several pediatric studies demonstrate better correlation between unfractionated heparin levels compared to the ACT [3-4]. We then initiated our own new standardized protocol using unfractionated heparin levels as a quality improvement project to reduce bleeding and clotting events, with other objectives being investigated are number of blood product transfusions, number of heparin infusion rate changes, number of circuit changes, and overall patient survival. Data was collected prospectively, and is compared to retrospective data from ECLS patients in our unit before the new protocol was initiated. We hypothesize that the new anticoagulation protocol will demonstrate an improvement in bleeding and clotting events compared to use of the prior protocol.
MEASURING MILESTONES: TRAINING PEDIATRIC SUBSPECIALISTS IN COMMUNICATION AND PROFESSIONALISM
POSTER #53

MELISSA SMITH, MD; CRITICAL CARE FELLOW
MENTOR: KIMBERLY BLASIUS, MD

Additional Authors; Division/Institution:
Benny Joyner, Rebecca Smith, Sofia Aliaga, Gene Hobbs, Robert Isaak

Background: The Accreditation Council for Graduate Medical Education (ACGME) initiated the Pediatric Subspecialty Milestones (PSM) in July 2015 to assess development of fellows in important aspects throughout training. Many of the advanced level training milestones are targeted at professionalism. Feedback for fellows can be sporadic, or overlooked, due to clinical demands and time. Several of the PSM are new and challenging to assess. The use of an objective structured clinical exam (OSCE), via simulation, is a well-developed tool in other parts of medical education, however, there is limited data on the use of OSCEs at the fellowship level and for assessing professionalism.

Objective: Our objective was to assess the ability of an OSCE to evaluate PSM in Pediatric ICU fellows.

Design/Methods: OSCE scenarios were designed to target difficult to assess milestones. The following milestones were chosen to be assessed bi-annually: Transfer of care with seamless transitions, Work in inter-professional teams, High standards of ethical behavior. Using a checklist and global score mapped to the PSM, each fellow was assessed during the simulations by two faculty per station. Post-surveys were completed by faculty and fellows.

Results: Survey data included questions scored based on a 5-point Likert scale. Data reported as means (M). Sample faculty (6) responses: It is important to have a milestone simulation program (4.6); It is useful to observe these scenarios specifically (4.83); The case was appropriate for the objective (4.8); Ease of using the grading rubric (4.66). Sample fellow (4) responses include: These helped to understand potential communication issues (4.25); These were believable scenarios (4.5); I understood the purpose of this activity before participating (4.0). Data regarding inter-rater reliability and comparison of faculty evaluation to self-evaluation is ongoing.

Conclusions: Based on surveys, the simulation exercises were well received. Faculty agreed that it was useful to observe fellows objectively with the intention of milestone evaluations. We plan to validate the evaluation tools to ensure that the scenarios and observations are reproducible. We are in the process of adapting our scenarios and grading rubrics to other pediatric subspecialty programs as well as other institutions.

References:
Background/Introduction: In 2015, the University of North Carolina Health Care System launched a system-wide sepsis collaborative to improve the timely and appropriate screening and management of patients with sepsis. Subsequently, the Pediatric Emergency Department Code Sepsis Quality Improvement Initiative team was established. The overall aim was to improve bundle compliance, defined as the following four metrics occurring within one hour of presentation: Blood cultures drawn before antibiotics; serum lactate levels determined; appropriate antibiotics initiated; and first fluid bolus initiated.

Methods: A fourth year medical student was assigned the role of developing a fishbone diagram of factors leading to non-compliance by observing workflow and interviewing stakeholders. Based on the diagram the medical student was tasked with identifying potential areas for improvement and developing a key driver diagram for improving compliance. A survey was also used to assess provider management decisions related to key interventions.

Results: A key barrier identified during interviews was that ordering lactate levels was confusing. The PED sepsis EPIC order set was built to standardize ordering practices for both inpatients and the ED and thus contained four different options for lactate orders. A revision to the order set, a pre-check of one of the options, was implemented on March 7. Interviews also confirmed where to send the samples and whether or not samples should be placed on ice. Clarification of these processes was provided to ED staff and providers. Additionally, providers reported significant alarm fatigue from a seemingly overly-sensitive BPA. Recommendations to revise the EPIC sepsis BPA have been submitted and a team has been chartered. Furthermore, a survey designed to assess provider use of one-hour lactate has provided insights that suggest that initial one hour sepsis management is not impacted by a serum lactate result.

Conclusions: Medical students can play an important and unique role in hospital quality improvement initiatives since they can take a high-level view of processes and initiatives while simultaneously being an informed member of the team. This dynamic allows medical students to objectively assess a situation, identify areas for change, and develop appropriate plans of action.
“PICTURE OF THE DAY” CURRICULUM FOR MEDICAL STUDENTS IN THE NEWBORN NURSERY
POSTER #55

ALISON RITTENBERG, MD; PEDIATRICS RESIDENT
MENTOR: ERIC ZWEMER, MD

Additional Authors; Division/Institution: Carl Seashore, MD

Objective: The newborn nursery is a critical learning experience for third-year medical students and provides excellent teaching opportunities for residents. Restructuring of our medical school curriculum decreased the time that students spend in the newborn nursery. The One-Minute Preceptor (OMP) model has previously elicited high satisfaction from learners and educators and has been shown to improve preceptors’ ability to evaluate students. In order to improve resident-led teaching of medical students and to improve residents’ abilities to evaluate students’ clinical reasoning, we designed, implemented, and evaluated a “Picture of the Day” curriculum based on the OMP model.

Methods: Our “Picture of the Day” curriculum consists of color photos of common newborn physical exam findings, accompanied by clinical scenarios structured via the OMP model with a clear question and “take-away” points. Third-year medical students and residents who rotated in the newborn nursery were eligible to participate. Printed educational materials were placed in the newborn nursery for resident use. Anonymous pre- and post-intervention surveys regarding the amount and quality of teaching were completed by students and residents.

Results: Nine residents and 13 students completed the pre-intervention survey, and to date, 5 residents and 16 students have completed the post-intervention survey. All respondents exposed to the curriculum reported that the “Picture of the Day” materials were effective and engaging. Medical students reported significantly increased satisfaction with the amount of teaching after the intervention (p=0.03). Resident reported awareness of (p=0.006) and comfort (p=0.02) using the OMP method increased significantly post-intervention. Resident satisfaction with the amount (p=0.002) and quality (p=0.02) of teaching they provided increased significantly post-intervention. Residents also reported significantly improved ability to generally evaluate students’ fund of knowledge (p=0.04) and clinical reasoning (p=0.002).

Conclusions: Use of “Picture of the Day” materials in the newborn nursery has preliminarily been associated with increased satisfaction with resident teaching for both medical students and residents. Residents report greater abilities to evaluate their students’ fund of knowledge and clinical skills. This novel translation of the OMP model into a visually driven curriculum can facilitate increased resident-student interactions, especially in environments where dedicated teaching time is limited.
Making Decisions About Pushing Fluids When Your Patient Has Sepsis

Poster #56

Michael Baca-Atlas, MD; Family Medicine Resident
Mentors: Rebecca Wheeler, RN, William Mills, MD, MPH, and Sue Tolleson-Rinehart, PhD

Background: Adequate fluid resuscitation is a critical part of goal-directed therapy for sepsis, and the use of the fastest methods – via pressure bag or push/pull syringe – is also emphasized in guidelines. Yet recommended resuscitation guidelines are rarely achieved in clinical practice. Because sepsis affects over 100,000 US children annually, closing the gap between recommended and actual practice is important. As a first step toward understanding and eliminating this disparity, we sought to capture the decision-making processes of nurses and physicians who must initiate a bolus of fluids for a septic patient. We surveyed practitioners’ preferences for fluid delivery (push/pull, pressure bag, gravity) in the UNC Pediatric Emergency Department.

Methods: We administered a 10-question web-based survey to Emergency Department nurses and attending physicians using the Qualtrics online survey tool supported by UNC. The survey aimed to identify current practices surrounding sepsis fluid resuscitation and barriers to administering 20 mL/kg bolus x 3 over 15 minutes, a goal set by the pediatric emergency department sepsis response plan. One email reminder was sent to complete the survey 2 weeks after the initial survey was distributed. The results come from 88 completed surveys, including 75 who answered all questions about fluid delivery. Respondents were predominantly nurses (63.2%), but a substantial number of physicians (36.8%) also answered. More than 60% of respondents have worked in an ED for 5 years or more, and more than three quarters of respondents (77%) said they are comfortable caring for pediatric emergency patients. We used Stata v.14.2 to analyze survey data.

Results: The two fastest methods, pressure bag and push/pull syringe, were preferred by most respondents (34 [45%] and 27 [36%] respectively. But 8% of respondents (N=6) preferred the pump, and 10.7% (N=8) preferred other methods. For a large number of respondents (80%), the patient’s age matters to the choice of fluid delivery. Respondents reported obtaining access and an inadequate number of team members to aid with charting and delivering fluids as the two largest barriers to successful fluid delivery.

Conclusions: The fact that almost a fifth of providers prefer a method that doesn’t meet current recommendations offers a significant opportunity for improvement. Despite efforts to standardize sepsis care to bring it into conformity with the American College of Critical Care Medicine Guideline, the UNC Pediatric Emergency Department has not yet reached that goal. Rapid fluid resuscitation is essential to good outcomes, making adherence to the new guideline important. The survey results show that PDSA cycles and other quality improvement strategies must be deployed to bring practice into alignment with current guidelines.
IMPLEMENTATION OF AN EVIDENCE BASED ALGORITHM FOR MANAGEMENT OF FEBRILE INFANTS
POSTER #57

CAMERON LANG, MD; PEDIATRICS RESIDENT
MENTOR: NICOLE CHANDLER, MD

Background: Adequate fluid resuscitation is a critical part of goal-directed therapy for sepsis, and the use of the fastest methods – via pressure bag or push/pull syringe – is also emphasized in guidelines. Yet recommended resuscitation guidelines are rarely achieved in clinical practice. Because sepsis affects over 100,000 US children annually, closing the gap between recommended and actual practice is important. As a first step toward understanding and eliminating this disparity, we sought to capture the decision-making processes of nurses and physicians who must initiate a bolus of fluids for a septic patient. We surveyed practitioners’ preferences for fluid delivery (push/pull, pressure bag, gravity) in the UNC Pediatric Emergency Department.

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IMPLEMENTATION OF FOOD INSECURITY SCREENING IN THE PRIMARY CARE CLINIC
POSTER #58

CHRISTIAN LAWRENCE, MD AND JEFFREY OKONYE, MD; PEDIATRICS RESIDENTS
MENTOR: MELISSA FITGERALD, MD, COLIN ORR, MD, AND EMILY VANDER SCAAF, MD, MPH

Background: Food insecurity is a significant issue at the local, national and international level. Food insecurity is defined as a household-level economic and social condition of limited or uncertain access to adequate food. It is estimated that 1 in 4 children in the USA live in food insecure homes. North Carolina is among the hungriest states in the US, having the 5th highest level of food insecurity.

Aim Statement: Increase the percentage of provider visits that include food insecurity screening as a part of well child check (WCC) visits to >50% over a 9 week period.

Interventions: We introduced a two question screening tool as a part of well child visits. To increase the number of children screened, we added the two questions to our note templates and held conferences to discuss the importance of screening for food insecurity. The provider screens for food insecurity using this tool and identifies food insecure families. Once identified, we provided a list of resources and allowed access to a food pantry established within our clinic.

Measures: We reviewed documentation from WCC visits from 8/23/16 (week prior to intervention) until 11/5/16. We calculated the percentage of well child provider visits that included food insecurity screening in their documentation. In addition, we documented the county of residence, ethnicity and age of every patient to try to identify trends in food insecure families.

Results: We found that 0% of WCC visits included documentation of food insecurity screening during our baseline week prior to implementation. This number increased to 58.75% after our interventions. Of those patients screened, 13.5% screened positive for food insecurity.

Conclusions and Next Steps: Though we reached >50% of provider visits screening for food insecurity, this data is limited by several factors including lack of screening tool placement into every template and provider comfort using the tool. Due to >40% of WCC visits not including documentation of food insecurity screening, a large number of food insecure families were possibly missed or screened without documentation. In addition, many patients may not be comfortable disclosing this information, which would lead to data underestimating the true number of patients in food insecure homes. Food insecurity screening can help identify families in need. With an effective tool and intervention, there is a significant enough need to continue including this in our practice.
IMPROVING RESIDENT DOCUMENTATION IN A PRIMARY CARE CLINIC
POSTER #59

JOE ZAKHAR, MD; PEDIATRICS RESIDENT
MENTOR: CARL SEASHORE, MD

Introduction: This project is a resident led effort at improving visit documentation at UNC Children's Primary Care Clinic. Our Electronic Health Record (Epic) allows for the creation of standardized note templates as ‘System Lists’, which means they can be edited by their creators and the changes are instantly reflected in any new notes generated by users. This allows for continued editing and adaptation of templates to a user and group’s documentation needs.

Aims: 1) Create standardized note templates for well-child visits in 4 distinct age groups: (Birth - 12 months, 15 months - 5 years, 6 - 10 years, and 11+ years through adolescence)
2) Measure documentation of selected core measures for each age group before and after template implementation
3) Improve resident efficiency and experience when documenting well-visits

Methods: We created standardized note templates (System Lists) in Epic to be used at all well-child visits starting in July 2016 at UNC Children’s Primary Care. We reviewed prior documentation methods for well-child visits via a random selection of well-child visit progress notes. We recorded the frequency of documentation of select core measures which vary by age-group. We then compared documentation of these core measures in notes created before and after the implementation of our standardized templates. Residents were also periodically surveyed to gain insight into their experience with these templates and to obtain suggestions for improvement.

Results: Over 200 well-child visit notes were reviewed from the pre and post template eras. There was universal improvement in frequency of identified core measures over the past year. In particular, documentation of infant secondhand smoke exposure (38% vs 92%) and carseat/booster seat use (19% vs 96%) were improved post-templates. Survey respondents reported a near universal adoption of these templates, though not all residents were surveyed. Interns in particular expressed appreciation for consistent and readily available note templates with age-appropriate anticipatory guidance and developmental milestones included. In this time period we have also noted adoption of these note templates at other UNC Pediatric satellite clinics.

Conclusions: Standardization and optimization of note templates can improve frequency of documentation of important measures in pediatric primary care. They also show potential in improving resident efficiency and overall experience in documenting these visits. In future, we will involve a member from each residency class in fostering continued improvements to our templates.
REDUCING THE USE OF EMPIRIC ACYCLOVIR IN LOW RISK NEONATES ADMITTED FOR SEPSIS EVALUATION
ORAL PRESENTATION ONLY

LAUREN BRADFORD, MD; PEDIATRICS RESIDENT
MENTOR: ASHLEY SUTTON, MD

Additional Authors; Division/Institution: Elizabeth Darnell, Peyton Wilson; Christine Walsh-Kelly, Ravi Jhaveri

Background/Introduction: Herpes simplex virus (HSV) infection is a rare but potentially fatal infection in neonates. Neonatal fever, however, is a common reason for evaluation and hospitalization. A lack of consensus on the use of empiric acyclovir in neonates presenting with symptoms concerning for infection results in the potential for its overuse. Previously published baseline data from our institution confirmed variation in practice and poor stewardship. We aimed to adapt a published, evidence-based care process model (CPM) at our institution to reduce inappropriate testing and use of empiric acyclovir in neonates at low-risk for HSV infection, particularly in neonates over 42 days old. We also aimed to improve complete testing for neonates at high risk for HSV infection.

Methods: After analysis of baseline data over a 13 month period at our institution, a multi-disciplinary team was convened to implement a quality improvement (QI) project with the above aims. Included infants were the same as the baseline cohort: younger than 90 days of life and treated with IV acyclovir during hospitalization. Infants admitted to the neonatal intensive care unit were excluded. Patients were identified by weekly reports within the electronic health record. Eligible patient charts were reviewed for baseline clinical presentation and laboratory findings, in order to retrospectively classify infants as low- or high-risk for HSV infection. The QI initiative included iterative changes including the creation of a local diagnostic algorithm on risk-stratification, evaluation and treatment of neonatal HSV, evidenced-based conferences for pediatric residents and faculty on HSV management, and ongoing monitoring of algorithm adherence via real-time chart review and coaching of acyclovir prescribers. Results were analyzed using statistical process control charts interpreted by standard rules.

Results: Six months of prospective data during the ongoing improvement project demonstrated an overall decrease in the use of empiric acyclovir from 28% to 17% of all infants presenting for rule-out sepsis evaluation and an increase in the rate of complete diagnostic testing from 11% to 33% in those infants initiated on acyclovir. Acyclovir use in infants greater than 42 days of age was nearly eliminated.

Conclusions: Our QI project was successful in decreasing the use of empiric acyclovir for infants at low risk for HSV who presented for sepsis evaluation. Additionally, the rate of complete laboratory evaluation was improved for infants whose clinical presentation warranted empiric acyclovir initiation.
Understanding the Factors Contributing to the Successful Establishment of a Free School Readiness-Centered Well Child Clinic.

Kathryn Blew, Lourens du Pisanie
University of North Carolina-Chapel Hill School of Medicine

**Motivation**
- Over 5.9 million US children are uninsured.
- 1,007 free clinics operate in the United States, providing care to around 1.6 million people.
- Among these free clinics, only 30 are student-run and few of these are devoted to serving children.
- Many children miss several days of school due to lack of access to vaccinations and or providers.
- Students who miss greater than 10 days of school have a statistically significant higher risk of falling behind in their work and underperforming at their grade level.
- Student-run free clinics centered on school readiness well child visits could serve as an excellent intervention to help underserved children reach their school readiness health goals.

**Methodology**
- Partnered with a community free clinic which served as a hub for vaccinations, space, and equipment.
- Partnered with the UNC pediatric department for physician recruitment.
- Advertised in local community centers, on school websites, and with the county nursing at local schools.
- First year: 3 weekend clinics located at the community free clinic.
- Second year: 2 weekend clinics at the community free clinic, and 2 weekday clinics at elementary schools.

**School Readiness: Immunization & Health Exams**

**Results**
- Our first clinic series served 25 children and the second served 03 children.
- 151 vaccines were administered.

**Conclusions**
- Multiple variables affected the success of a school readiness-centered well child clinic including timing, advertising, and personnel.
- Clinic volume was most affected by timing; clinics held closer to the start of school or during the grace period were most popular.
- Advertising with nurses directly proved more successful than with community centers.
- Personnel were more likely to volunteer during the week.
- Clinic volume was unaffected by weekday or weekend hosting.

**References**
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