Stony Brook Children's

SCHOOL OF MEDICINE
DEPARTMENT OF PEDIATRICS

Carolyn Milana, MD
Interim Chair, Department of Pediatrics
Associate Professor of Clinical Pediatrics
Medical Director for Quality
Stony Brook Children's Hospital

May 23, 2018

Welcome to the tenth annual Pediatric Trainee Research Day. It is a pleasure to share with you the work our residents and trainees have been carrying out in children's health research and scholarship.

As we continue on our journey to creating a world class children's hospital program, we are committed to providing hope to sick children and their families by carrying out the research that will improve existing treatments and make the discoveries that will lead to new approaches to pediatric diseases.

To prepare the next generation of pediatricians to take part in this mission, each of our trainees is required to carry out a mentored project during their training. Coming from diverse backgrounds they can select from a variety of projects to best suit their career goals and meet their educational needs. Today they will have the opportunity to present their work to their faculty, peers and other colleagues.

Thank you for joining us and showing support to these young investigators. Special thanks also to Dr. Marian Evinger and this year's Organizing Committee (Drs. Fischel, Glaubach, Lane, M. Parker, R. Parker, and Woroniecki) for coordinating the day. Also, we appreciate the entire faculty who have served as mentors to provide guidance and encouragement to our trainees.

Sincerely

Carolyn Milana, MD
Associate Professor of Clinical Pediatrics
Interim Chair, Department of Pediatrics
Medical Director of Quality, Stony Brook Children's Hospital

2018 PEDIATRIC RESEARCH DAY
Wednesday, May 23rd
8 am – 1:15 pm
Charles B. Wang Center

8:00 – 8:30 **Registration and Breakfast** – Theater Lobby
8:30 **Welcome –-** Dr. Margaret Parker
Chair's Opening Remarks - Dr. Carolyn Milana– Main Theater
8:40 **Keynote Address** – Main Theater
Introduction of Keynote Speaker – Dr. Andrew Lane
8:45– 9:35 **Keynote Speaker**: **Susan Furth, MD, PhD**
Professor, University of Pennsylvania School of Medicine
Laffey-Connolly Chair in Pediatric Nephrology, CHOP

"Lessons Learned from the **Chronic Kidney Disease in Children** Cohort Study"

Platform Presentations – Main Theater

Session 1 (9:40-10:10)

9:40 **Introduction of Invited Judges –** Dr. Robert Parker

9:45 -10:10 **Residents Platform Presentations** (Chair – Dr. Robert Woroniecki)
Joseph Cafone, MD "US Adult University Rheumatologists Preferences and Knowledge Gaps Regarding the Care of Young Adults with Rheumatic Conditions"

Kathleen Murphy, DO "Rates of Blood Lead Screening and Barriers to Compliance in Stony Brook Primary Care Pediatric Offices"

10:15 – 10:30 **Coffee Bre**ak - Theater Lobby

Session 2 (10:35 – 11:20)

10:35- 10:50 **PEDsTalks** (Introduced by Dr. Marian Evinger)
Daniela Feitosa, MD "What Were We Thinking ??"
Jennifer Lutz Cifuni, DO "Dr. Patient: A Warrior"

10:55 – 11:20 **Fellows Platform Presentations** (Chair – Dr. Janet Fischel)
Aderonke Adefisayo, MD "Reception & Barriers to Long Acting Injectable Antiretrovirals (LA-ARV) for Pre-Exposure Prophylaxis (PrEP) in Adolescents"

Laura Stabin, MD "Regional Tissue Oxygenation in Term Infants with Presumed Sepsis"

11:20 – 12:15 **Poster Session** - Theater Lobby (Chair - Dr. Taly Glaubach)
Invited Judges plus Drs. Glaubach and Woroniecki

12:20 – 1:00 **Lunch** – Zodiac Gallery
Dr. Furth to discuss her academic career path
Presentation of Awards and Closing Remarks by Dr. Milana

Keynote Speaker Biography

Susan Furth, MD, PhD
Professor and Laffey-Connolly Chair in Pediatric Nephrology
Children's Hospital of Philadelphia
University of Pennsylvania

Dr. Susan Furth graduated *summa cum laude* with a degree in English from the University of Pennsylvania. She earned her medical doctorate from this institution in 1988. After contributing two years as a physician in Arizona with the Navajo Health Service, Dr. Furth completed her internship and residency in Pediatrics at Johns Hopkins University. She undertook a clinical and research fellowship in Pediatric Nephrology at Johns Hopkins, earning a doctorate in clinical investigation from Hopkins' Bloomberg School of Public Health. From 1996 to 2010, Dr. Furth was Associate Professor of Epidemiology in Public Health and Professor of Pediatrics in Medicine. In 2010, Dr. Furth was recruited by Children's Hospital of Philadelphia (CHOP) as Chief of the Division of Nephrology and named the Laffey-Connolly Professor of Pediatric Nephrology.

Dr. Furth's remarkable career in research has contributed tremendously to our understanding of pediatric kidney diseases, especially chronic kidney disease. Dr. Furth was Co-Principal Investigator of the "Teen Adherence in Kidney Transplant Effectiveness Intervention Trial (TAKE-IT)", an NIH-funded behavioral intervention to promote medication adherence in adolescents following kidney transplant. Dr. Furth is also currently Principal Investigator of NIH-funded "Chronic Kidney Disease in Children", the largest multicenter prospective cohort study of children with progressive chronic kidney disease ever conducted in North America. Dr. Furth's research has been published in *New England Journal of Medicine, Pediatrics, Journal of Pediatrics, Pediatric Nephrology, Human Molecular Genetics*, and other highly regarded journals.

Dr. Furth has actively mentored the research efforts of trainees and junior colleagues, guiding numerous individuals to establish their own research-oriented careers in nephrology, urology and rheumatology. In 2016 CHOP awarded Dr. Furth their Faculty Mentor Award. She is Associate Chair for Academic Affairs and chairs the Committee on Advancements and Promotions.

In service extending beyond her own institution, Dr. Furth has been Chair or governing Committee Member for professional organizations, including American Academy of Pediatrics, American Society of Pediatric Nephrology, and American Pediatric Society, and CDC expert panels. Notably, she was elected Councilor, Vice-President, and President of the Society of Pediatric Research. Dr. Furth also edits and reviews for numerous clinical and research journals

Abstracts

Resident Platform Presentations

Fellow Platform Presentations

Resident Posters

Interprogram Posters

NOTE: All Abstract Authors from Department of Pediatrics unless otherwise indicated.

ABSTRACT 1.

U.S. ADULT UNIVERSITY RHEUMATOLOGISTS' PREFERENCES AND KNOWLEDGE GAPS REGARDING THE CARE OF YOUNG ADULTS WITH RHEUMATIC CONDITIONS

Joseph Cafone MD[1], Catherine Messina PhD[2], Patience White MD MA[3], Julie Cherian MD[4]

[1]Internal Medicine-Pediatrics, Department of Medicine, [2]Department of Family, Population and Preventive Medicine; [3]Department of Rheumatology, The George Washington University, Washington, D.C; [4]Division of Pediatric Rheumatology, Stony Brook Medicine.

Background: Adult rheumatologists (AR) lack comfort with pediatric rheumatic disease (PRA) management due to shortcomings with formal transition of care.

Objective: Assess AR current levels of comfort in management of young adult patients (YA) with PRA and describe prevalence of formal transition program (FTP) in North American academic institutions.

Methods: AR fellowship program (ARFP) directors listed in the American Medical Association Residency and Fellowship Database were contacted via email (3 times over 3 months) for participation of an online survey. Participating ARFP were asked to distribute the survey to AR faculty and fellows. Young adults (YA) were defined as patients less than 35 years old, diagnosed with PRA. Qualtrics was used for survey distribution and data collection. Excluded surveys included non-complete response, non-consenters, and non- physicians. Statistical Package for the Social Sciences (SPSS) was used for data analysis.

Results: Of 112 rheumatology fellowship programs, 37 participated in the survey (33%), and 119 out of 753 eligible AR completed the survey (16%): 10 were excluded and 109 surveys were analyzed. Sixty-one percent of responders self-identified as female. Forty-three (40%) listed they were 30-39 years old. Six (6%) identified as Internal Medicine / Pediatric trained. Twenty-eight (26%) reported they were still in fellowship training while twenty-seven (25%) listed 1-9 years of attending experience. Sixty-one (56%) reported that $\leq$10% of their total patients are YA. Although 81 of 109 responded that a pediatric rheumatologist (PR) was employed at their institution, only 27 of 109 were familiar with FTP of YA patients. There was a significant correlation between PR and FTP, p=0.023. The most commonly selected suggestion for improving FTP was to provide a thorough patient summary (97/109). Seventy-nine of 109 participants (72%) stated they were "very comfortable" or "comfortable" when treating YA's with PRA. Fifty-three of 109 respondents (49%) answered three or more hypothetical clinical scenario questions correctly.

Conclusion: This is the first nationwide survey to target AR providers for YA with PRA. PR is associated with AR comfort of managing PRA; however this may not translate to appropriate management. Limitations of this study include self-reported responses and lack of rural areas or community hospital participation.

ABSTRACT 2.

RATES OF BLOOD LEAD SCREENING AND BARRIERS TO COMPLIANCE IN STONY BROOK PRIMARY CARE PEDIATRIC OFFICES

Kathleen Murphy, DO, and Latha Chandran, MD, MPH

Background: Elevated blood lead levels (BLL) can lead to permanent effects on cognitive development and behavior. New York State (NYS) remains an area with one of the highest levels of elevated BLL. The American Academy of Pediatrics (AAP) recommends universal screening for high risk children and following local guidelines. NYS Department of Health requires screening all children at 12 and 24 months.

Objective: To identify rates of lead screening of Stony Brook (SB) Children's primary care clinics, factors associated with, and practitioner barriers to compliance.

Methods: Charts of 200 children 12-36 months who had a well exam at any SB clinic in academic year 2016-2017 were randomly reviewed. An anonymous survey was distributed to providers at SB clinics. Data were analyzed with descriptive statistics. Associations were calculated for 12 month data.

Results: The rate of ordering lead screening at least once was 96.5% (95% CI 94.0-99.0). The overall rate of partial compliance (at least one lead screening result) was 74.5% (95% CI 68.5-80.5). The rate of ordering was higher ($p < 0.00001$) at 12 months (96.4%, 95% CI 93.7-99) than at 24 months (67.9%, 95% CI 55.4-80.5). Compliance was also higher at 12 months (76.8%, 95% CI 70.7-82.8) than at 24 months (56.8%, 95% CI 40.6-72.9) (p 0.01). Public insurance was associated with higher rates of compliance ($p < 0.001$). The Islip clinic site was associated with a lower rate of compliance (p 0.02). Compliance was higher in children seen by residents $\leq$ 25% of overall visits than in children seen by residents >25% of visits (p 0.003).

Most providers reported being aware of AAP guidelines (88%). Fewer were aware of NYS laws (71%). More reported ordering lead levels at 12 months (98%) than at 24 months (78%), corresponding with calculated rates. The only factor identified as a significant barrier to lead screening by most respondents was patient non-compliance (56%).

Conclusion: The overall rate of ordering lead screening was very high. The rate of compliance was lower, but higher than reported in previous studies, although Islip had lower compliance than other sites. Rates of both ordering and compliance were lower at 24 month visits compared to 12 month visits, which agreed with provider self-reporting.

ABSTRACT 3.

RECEPTION & BARRIERS to LONG ACTING INJECTABLE ANTIRETROVIRALS (LA-ARV) for PRE-EXPOSURE PROPHYLAXIS (PrEP) in ADOLESCENTS

Aderonke Adefisayo, MD and Sharon Nachman, MD

Division of Pediatric Infectious Disease, Stony Brook Medicine

Background: There are an estimated 50,000 new HIV infections every year in the USA: 26% of these are in youth 13-24 years. Oral pre-exposure prophylaxis (PrEP) has been shown to decrease the risk of HIV transmission, but the major drawback is the need for daily adherence. New LA ARVs are proposed to be useful as PrEP, but data are lacking on whether and how adolescents would use these therapies.

Objective: Assess acceptance of long-acting (LA) forms of prevention for HIV in an injectable form vs. daily oral formulations in college-attending youth

Methods: A self-administered Qualtrics Survey, evaluating risk of HIV acquisition and partiality to use LA ARVs, was distributed to adolescent clinic patients and university-attending youth at Stony Brook. Descriptive analyses of data from these surveys are provided.

Results: A total of 94 surveys were collected over a 6 month period; the median age was 19 years, and 70% of participants were female and 1 % Transgender. Over 50 % were Caucasian. All participants had at least a high school diploma. Eighty percent (80%) endorsed sexual activity in the preceding 3 months and had between 1-5 sexual partners. Only 1% reported more than 6 sexual partners. Only a third of participants reported consistent condom use, and 25% reported rarely or never using condoms. Despite this inconsistent condom use, over 70% had not been tested for HIV or other STIs. Of the 30% that had been tested, 3% reported being positive for Chlamydia and Syphilis.

Despite inconsistent condom use, over 70% reported not being very afraid of getting HIV but were more likely to consider PrEP with LA ARVs if they reported being very afraid. Reasons cited for lack of willingness to use LA ARVs as PrEP (despite high risk behaviors) included the need for frequent doctor visits, pain associated with injections, and a strong perception that they were not at risk.

Conclusion: LA-ARV's would be a helpful modality in the prevention of HIV, but there will be challenges in acceptability in college-age youth. The dual challenge of perception of risk in college-aged youth, along with the unacceptability of bimonthly injections, will prevent routine uptake of LA ARVs in prevention of HIV acquisition in this population.

ABSTRACT 4.

REGIONAL TISSUE OXYGENATION IN TERM INFANTS WITH PRESUMED SEPSIS

Laura Stabin, MD[1], Catherine Messina, PhD[2], & Jonathan Mintzer, MD[1]

[1]Pediatrics / Neonatology; [2]Family, Population and Preventive Medicine, Stony Brook Medicine.

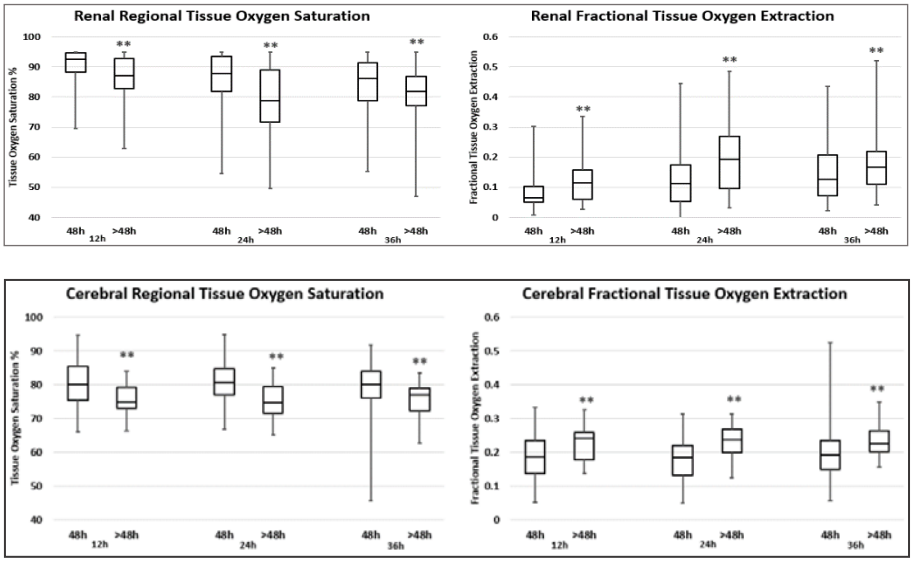
Background: Many asymptomatic term infants are admitted to the NICU for antibiotics due to maternal chorioamnionitis. Currently, antibiotic duration is determined using blood cultures and inflammatory markers. Adult studies indicate that tissue oxygen extraction, derived by near-infrared spectroscopy (NIRS), correlates with sepsis severity.

Objective: We hypothesized that infants with abnormal inflammatory markers would demonstrate elevated cerebral and renal oxygen extraction.

Methods: In this prospective observational study, we calculated cerebral and renal oxygen extraction among well term infants receiving antibiotics due to chorioamnionitis. Cerebral and renal NIRS oxygenation (rSO_2) data were collected for one-hour intervals at 12, 24, and 36 hours of life. Blood cultures were collected at birth, CBCs at 0 and 24 hours, and CRPs at 12 and 24 hours. Antibiotic duration was determined by attending neonatologists unaware of NIRS data. Infants were grouped using length of treatment. Cerebral and renal fractional tissue oxygen extraction (FTOE) were compared between treatment groups at each time point.

Results: Between June, 2016, and January, 2018, 343 term infants admitted to the Stony Brook Children's Hospital NICU due to chorioamnionitis were assessed and 44 were enrolled, of which 36 (82%) received 48 hours of antibiotics (group 1) and 8 (18%) received prolonged courses (group 2). Maternal fever was higher in group 2 (38.9 °C vs. 38.4 °C, p=0.002), as were the immature/total white blood cell ratios (0.21 vs. 0.1, p=0.001) and CRPs (1.7 mg/dL vs. 0.1 mg/dL, p<0.001). Blood cultures were negative in all subjects. Group 2 had a higher incidence of funisitis on placental pathology (75% vs. 32%, p=0.045). At all comparison points, cerebral and renal rSO_2 were reduced in group 2 (see figure; p<0.001). Consistent with our hypothesis, cerebral and renal FTOE were elevated in group 2 at all time points (figure, p<0.001).

Conclusion: Cerebral and renal NIRS have potential utility as screening modalities for clinical sepsis in asymptomatic neonates exposed to chorioamnionitis. Further analysis is needed to determine the sensitivity and specificity of NIRS in guiding these evaluations.



ABSTRACT 5.

STRONGYLOIDES INFESTATION IN RHEUMATIC DISEASES

Joseph Cafone, MD[1], Swosty Tuladhar, MD[2], Asha Patnaik, MD[2], Ayse Bag Ozbek, MD[2], Qingping Yao, MD PhD[2]

[1]Internal Medicine-Pediatrics, Departments of Medicine and Pediatrics, [2]Division of Rheumatology, Allergy, and Immunology, Department of Medicine, Stony Brook University Medicine, Stony Brook, NY

Background: Immunosuppressive agents used for treatment of rheumatoid arthritis (RA) and systemic lupus erythematosus (SLE) may result in increased risk for opportunistic infections. *Strongyloides stercoralis* infestation can present from an asymptomatic eosinophilia to disseminated multisystem hyperinfection syndrome in an immunocompromised host. There are no management guidelines for coexisting strongyloides infestation and rheumatic diseases (RD).

Objective: To present a case series of patients with RD on immunosuppressants with concomitant stronglyoides infestation and its effect on outcomes.

Methods: We present a case report of a RA patient diagnosed with strongyloides infection coupled with case review. A PubMed database search was conducted using the keywords: strongyloides and arthritis or RA or RD or SLE. Only full text articles in English published between 1997 and 2017 were included. Of those, patients with both Stongyloides positive serology and pre-existing rheumatism were included.

Results: After an initial search of 64 articles, 16 articles were included in our case series, resulting in 19 cases inclusive of the patient presented from our institution. Eighty-four percent (16/19) of reported cases were from Asia and South America, which included 13 SLE and 6 RA patients. These patients were on immunosuppressive agents, including long-term glucocorticoids in 95% (18/19) of patients. Ninety-five percent (18/19) presented with gastrointestinal or pulmonary symptoms. All of the patients diagnosed with hyperinfection syndrome were chronic steroid users. The seven patients who required a higher level of care were chronic steroid users. Within this group, 3 were undiagnosed and therefore untreated, which resulted in 100% mortality.

Discussion: Immunosuppressed patients from endemic areas such as Asia and South America may be at increased risk for parasitic infestation. Strongyloides infestation should be considered in patients on immunosuppressive therapy with unexplained gastrointestinal or pulmonary symptoms. Long-term steroids may be a major risk factor for the development of hyperinfection. Delayed treatment of infestations may result in poor outcomes.

Conclusion: In patients with RD, it is important to identify risk factors for Strongyloides infestation prior to initiating immunosuppressive medications. We believe this case series will aid in future guideline development in this select group of patients.

ABSTRACT 6.

PHENOTYPIC DISCORDANCE IN TWO SIBLINGS WITH PHOSPHOGLYCERATE KINASE DEFICIENCY.

Tova Chein, DO [1], Jody Weiss-Burns, MSGC[2], and Patricia Galvin-Parton, MD[1, 2]

[1]Department of Pediatrics, [2]Division of Pediatric Genetics, Stony Brook Children's Hospital, SUNY –SB School of Medicine, Stony Brook, NY

Background: Elevations of CPK can be seen in a variety of clinical situations. Chronic, recurring presentations may signal a metabolic myopathy. Elevated CPK levels when accompanied by weakness, myalgia and tea-colored urine are the main clinical manifestations of rhabdomyolysis. Rhabdomyolysis is a clinical emergency that has the potential to cause acute kidney injury. The metabolic myopathies are a clinically diverse group of disorders. Making a specific diagnosis can be challenging.

Case: An unusual presentation of a rare metabolic myopathy in 2 siblings with PGK1 deficiency with a phenotypic discordance. Older brother presented with muscle cramps and dark urine after a bicycle accident. CPK levels reached a max of 300,000. Younger brother presented with ASD diagnoses and history of recurrent myalgia. The work-up included a combination of biochemical and genetic tests on blood, urine and muscle biopsy specimens. Specific Genetic Panels were sent and revealed carrier status but no definitive diagnosis. Whole Exome Sequencing revealed a c.639C>T mutation in the PGK1 gene responsible for a diagnosis of PKG1, which is a rare metabolic myopathy.

Discussion: PGKD affects the body's ability to utilize glucose as source of energy. There are 2 major forms of PGKD. A hemolytic form leads to chronic hemolytic anemia. There is also a rare myopathic form that leads to rhabdomyolysis and renal failure. Only one form tends to run in an affected family. An unexplained feature of the disease includes neurological manifestations and is usually seen in the hemolytic form. The family presented here is unique due to the fact that 2 brothers are affected with the same genetic defect, but the phenotypic manifestations are very different. Brother #1 has normal intelligence and severe episodes of rhabdomyolysis. Brother #2 has severe neurological manifestations with mild myopathy.

Conclusion: Svaasand (2007) proposed that the exon 6 mutation alters PGK1 at the level of transcription, causing decreased activity of PGK1. This explains the clinical manifestation of the enzyme deficiency. No explanation exists for the discrepant phenotype due to the same gene mutation. This is the first report of two brothers with the same genotype but with different phenotypes in PGK1D.

ABSTRACT 7.

IMPROVEMENT IN I-PASS HANDOFF COMPLIANCE ON THE PEDIATRIC UNIT

Alex Choi, MD[1], and Ilana Harwayne-Gidansky, MD[2]
[1]Internal Medicine-Pediatrics, Department of Medicine; [2]Department of Pediatrics, Stony Brook Medicine

Background: The importance of handoff is gaining more emphasis in clinical medicine. The I-PASS handoff is one of the leading tools developed for this purpose and has been shown in multiple studies to significantly reduce medical errors. Since its introduction to the Stony Brook Children's Hospital (SBCH) general pediatrics unit in 2015, there have been multiple efforts to improve compliance of I-PASS through an iterative quality improvement process.

Objective: To increase the rate of compliance with I-PASS handoff components to 80% between July, 2017 and June, 2018.

Methods: Pre-intervention data were collected from May 2017. Data were grouped by month, and three months were designated as one PDSA cycle. Handoff sessions on the SBCH general pediatrics floor were directly observed using a customized observation tool, which includes I-PASS components, whether the floor team was primary service, and common I-PASS deviations. A total of four interventions were planned: workshop, lecture, grand rounds lecture, and individual meeting with interns. Microsoft Excel was used for statistical analyses.

Results: Prior to intervention, two I-PASS components were above goal compliance: Illness Severity (81%) and Patient Summary (96%). After two PDSA cycles, three I-PASS components improved and were above goal compliance: Illness Severity (90%), Patient summary (100%), and Action items (100%). Situation Awareness and Synthesis by receiver showed an initial compliance of 30% and 0%. Both had intermittent increases in compliance, up to 88% and 74%, but decreased during the second PDSA cycle. In terms of I-PASS deviations, the frequency of excessive summary decreased (11 to 5), whereas that of interruptions remained consistent (7 or 8), of which one out of 22 episodes was deemed as emergent. T-test analyses showed significant difference of handoff time between floor team primary vs non-primary services (0.69 vs 2.13 min, $p<0.05$)

Conclusion: We demonstrated a partial increase in compliance with I-PASS components. We also observed a change in IPASS deviations, specifically decreasing excessive patient summary. The next phase will include interventions to increase Situational Awareness and Synthesis compliance.

ABSTRACT 8.

IN UTERO TRANSMISSION OF STAPHYLOCCOCUS EPIDERMIDIS RESULTING IN INFECTIVE ENDOCARDITIS IN A NEONATE

Alex Choi MD[1], Tom Holowka MS4[2], Saul R Hymes MD[3], Aruna J Parekh MD[4]
[1]Department of Medicine, [2]Stony Brook University School of Medicine, [3]Department of Pediatrics

Background: Endocarditis is a rare and likely underreported occurrence in the neonatal population. Risk factors include central line associated bloodstream infection (CLABSI), congenital heart diseases, and invasive procedures or surgery. Although preterm infants are at increased risk for infections, transplacental transmission of staphylococcal infection is rarely described.

Case: We report a preterm male infant with GA of 31 3/7 weeks, delivered by cesarean section to a mother who presented for fever and reduced fetal movements. Maternal history was significant for IV drug use, specifically opiate abuse and recurrent infections. Her placental cultures were positive for *S. epidermidis*.

Upon delivery, the infant was admitted to the neonatal intensive care unit for prematurity, respiratory distress and clinical sepsis. He was treated with antibiotics for 5 days. Initial blood culture was negative. An umbilical venous (UV) catheter was placed for IV nutrition and antibiotic administration. This UV line was noted to be in the right atrium and remained in place for 8 days. On day of life 11, he developed signs and symptoms of sepsis with positive cultures for *S. epidermidis*. Due to persistent positive blood cultures, a cardiac echo was performed which confirmed the findings of hyperechoic masses in the right atrium and right ventricular outflow tract. This patient was diagnosed with endocarditis and treated with vancomycin and gentamicin, with resolution of infection upon discharge.

Conclusion: This case represents a potential occurrence of an *in utero* transmission of a community-acquired infection and subsequent disease following iatrogenic injury in the hospital setting. It also raises concerns for community acquired infections, such as *S. epidermidis* occurring primarily in IV drug users, invading the hospitalized patients as noted in our case with vertical transmission from mother to her fetus. With recent rise in opiate addiction and IV drug usage, practitioners need to be aware of yet another complication.

ABSTRACT 9.

EFFECTS OF A TRAINING VIDEO ON PEDIATRIC RESUSCITATION TEAM PERFORMANCE USING A VALIDATED ASSESSMENT TOOL

Jennifer Lutz Cifuni DO, Rahul Panesar MD

Background: Pediatric residents (PR) receive insufficient training to manage acute pediatric emergencies. Simulation-based medical education (SBME) improves skills and provides training opportunities outlined by the American College of Graduate Medical Education competencies.

Objective: To describe effects of a custom-made instructional video (iV) that contrasted suboptimal and optimal resuscitation techniques on simulated pediatric cardiac arrest team performance (SCAP) as measured by a modified and validated Tool for Resuscitation Assessment Using Computerized Simulation (TRACS).

Methods: SCAP was compared during two time periods: before (07/2014 – 09/2015) and after (10/2015 – 06/2017) viewing of iV. Teams consisted of different groups of PR and nurses selected based upon schedule availability, and the team leader was a senior resident. iV viewing was unsupervised and accessible only to PR. Each simulation was tabulated on the TRACS and divided into four categories: Basics (B), Airway and Breathing (AB), Circulation and Rhythm (CR), Competency and Behavior (CB). Results were analyzed using t-tests, repeated measures Analysis of Covariance, and Chi-Square tests. All tests of significance are two-tailed and evaluated at the level of $p < 0.05$.

Results: Thirty pre-video and 21 post-video groups were scored, each comprised of three to seven individuals. There was no statistically significant improvement in performance means (with standard deviations and standard errors, SE): 5.3 ± 1.40 (SE 0.26) and 5.2 ± 1.04 (SE 0.23) (B), 5.5 ± 2.11 (SE 0.39) and 5.9 ± 1.92 (SE 0.42) (AB), 6.7 ± 4.69 (SE 0.87) and 7.2 ± 4.83 (SE 1.05) (CR), 3.3 ± 1.09 (SE 0.2) and 3.8 ± 1.17 (SE 0.25) (CB)). Controlling for presence of senior residents and nurses showed significant improvement in the group score for AB (p=0.019) compared to uncontrolled presence (p=0.525). Analysis of the CB subcategories (Figure 1) showed significant improvement in "Feedback communication" from the pre-iV groups performing the task at 40% to the post-iV groups at 76% (p=0.011).

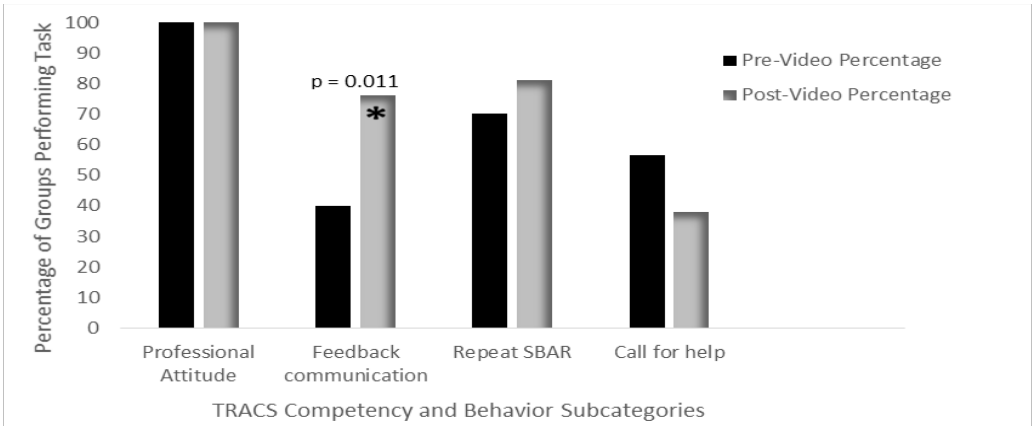


Figure 1. Competency and Behavior Performance Before and After Instructional Video

Conclusion: The effects of the iV on SCAP suggests a trend towards improvement with significant improvement in feedback communication, as calculated by a modified TRACS tool. Limitations included the inability to control for repeated or no intervention exposure by individual residents, small sample size, and nurses' inability to view the video. Training with an iV may help identify areas of acute pediatric management skills needing improvement during training.

ABSTRACT 10.

EARLY CHANGES IN GROWTH TRAJECTORY Z-SCORES PREDICT GROWTH OUTCOMES IN THE VERY LOW BIRTHWEIGHT POPULATION

Jared Dunahay DO, Susan Mathieson RD CSP, Shanthy Sridhar MD, Jennifer Pynn MD

Background: Nutritional management and achievement of adequate extra-uterine growth is challenging for very low birth weight (VLBW) neonates with physiologic weight loss of 10-20% anticipated in the first weeks of life. Postnatal growth trajectories as early as 21days of life in healthy preterm neonates have been shown to predict growth outcomes at discharge from the NICU. Studies have demonstrated the direct association in optimizing early enteral and parenteral nutrition with postnatal growth thereby improving neurodevelopmental outcomes at 22 months of age. The effects of growth at the time of maximal weight loss and at the time to regain birthweight with respect to discharge weight outcomes remain to be determined.

Objective: To determine if postnatal growth trajectories can predict discharge weight (z-scores) in both healthy and sick VLBW neonates after maximal weight loss is reached and birthweight regained.

Methods: This is a retrospective chart review of 134 VLBW neonates born in 2012 and 2015 with BW<1,500 grams. Data collected include growth parameters at birth, 35weeks post-menstrual age (PMA) and at discharge. Daily weights for the first 14days of life, time in days to maximal weight loss and days to regain BW were also recorded. Parameters were plotted on the Fenton growth curve and z-scores were calculated. We performed a multiple regression analysis with a nonparametric bootstrapping optimization algorithm to predict z-scores for weight at 35weeks PMA and at discharge.

Results: Data were stratified into 2 groups based on gestational age (GA): Group 1: <30weeks (n=86); mean GA: 27.7±1.6 weeks and mean BW: 1036±254 grams; Group 2: ≥30weeks (n=48); mean GA: 31.6±1.3 weeks and mean BW: 1279±165 grams. Maximal weight loss was achieved later (5±2days vs 4±2days; p=0.02) with a longer time to regain BW (13±4days vs 10±4days; p=0.01) in neonates with GA< 30 weeks. See table below for Multiple Regression Model results.

	<30 weeks		**≥30 weeks**	
Z-scores	**Coefficient**	**p value**	**Coefficient**	**p value**
Maximal Weight Loss Reached Compared to:				
PMA-35weeks	1.159	0.01	0.144	0.19
Discharge	1.246	0.01	0.153	0.22
Birthweight Regained compared to:				
PMA-35weeks	-0.006	0.54	1.026	0.01
Discharge	0.007	0.53	1.093	0.01

Conclusion: Postnatal growth trajectories at maximal weight loss from birth and at the time to regain birthweight predict long-term growth in the VLBW population. Larger, prospective studies are needed to demonstrate if interventions at these time points can improve growth outcomes.

ABSTRACT 11.

RIBOFLAVIN TRANSPORTER DEFICIENCY: A RARE, TREATABLE DISORDER THAT HAS POTENTIAL TO BE DETECTED ON THE NEWBORN SCREEN.

Daniela Feitosa, MD[1], Charlene Fox, MSRD[1], Jody Weiss-Burns, MSGC[2], and Patricia Galvin-Parton, MD[1,2]

[1]Department of Pediatrics, [2]Division of Pediatric Genetics, Stony Brook Children's Hospital, SUNY-SB School of Medicine, Stony Brook, NY

Background: The Newborn Screening Program in 2008 included a core of 29 metabolic disorders and an additional 25 secondary targets. These secondary targets were identified by expanding the differential diagnosis for the core disorders. MADD (Multiple Acyl-CoA Dehydrogenase Deficiency, also known as Glutaric Acidemia II; GA2) is listed as a secondary target. Riboflavin Transporter Deficiency (RTD) is caused by mutations in the SLC52A2 or SLC52A3 gene and causes impaired ability to transport riboflavin. This disorder results in sensorineural hearing loss and pontocerebral palsy. Laboratory findings include abnormal urine organic acids and an abnormal acylcarnitine profile similar to MADD.

Case Report: A 1 week old male, ex 37 week infant was evaluated for an abnormal newborn screen positive for Medium-Chain Acyl-CoA Dehydrogenase Deficiency (MCAD). The initial acylcarnitine panel showed elevations of short, medium, and long-chain acylcarnitine species and could not exclude a diagnosis of MADD. Supplementation with riboflavin and carnitine was started pending DNA results. DNA studies failed to reveal mutations consistent with MADD. The patient remained asymptomatic and riboflavin supplementation was continued. DNA studies for RTD revealed a homozygous variant in the SLC52A2 gene confirming this diagnosis. At 5 months, this infant is cooing, smiling, and social. He can push his head and chest above the plane of the exam table while prone. He has normal reflexes.

Discussion: The Newborn Screening Program has provided early detection, diagnosis, and treatment of certain genetic, metabolic and infectious congenital disorders leading to significant reductions in death, disease, and/or disabilities. RTD has been associated with mutations in the SLC52A2 gene responsible for instructions on making a riboflavin transporter protein called RFVT2 that is found in high levels in the brain and spinal cord. RTD is a rare, but treatable disorder that may look similar to the NBS acylcarnitine profile of MCAD or MADD. All forms can be life-threatening without treatment.

Conclusion: Riboflavin Transporter Defect should be considered as a secondary target for the newborn screen. Early detection and treatment with riboflavin may be lifesaving and able to change the current natural history of the disorder.

ABSTRACT 12.

DOCUMENTATION OF CHILDHOOD OBESITY IDENTIFICATION AND MANAGEMENT IN THE OUTPATIENT SETTING.

Sarah Heathcote, DO, MPH, Rosa Cataldo, DO, MPH, & Catherine Messina, PhD

Background: Childhood obesity is now estimated to affect as many as 107.7 million children worldwide. Nearly 32% of children in the United States are overweight or obese, increasing their life-time risk of complications including cardiovascular disease, diabetes, asthma, and depression. Despite a growing population of obese youth and the release of Expert Committee management guidelines in 2007, evidence demonstrates under-diagnosis and overestimation of documented obesity screening by pediatricians. To our knowledge, no prior studies have analyzed the consistency of obesity management practices as compared to Expert Committee recommendations.

Objective: To determine if overweight and obese children are appropriately identified and managed by outpatient pediatric primary care providers during annual preventive well-child visits (WCV).

Methods: We conducted a retrospective electronic medical chart review of WCV in children 2-21 years seen between 2014-2015 at 4 Stony Brook Children's outpatient sites. Subjects were stratified by body mass index (BMI) as obese (Ob), overweight (Ov), or healthy weight (H). Multiple logistical regression and cross-tabular analyses were used to examine weight category (Ob, Ov, H), presence of physician billing ICD9 weight status diagnosis (i.e. Ob, Ov, BMI percentile), and physician-assigned management: establishing diet and exercise counseling, ordering screening laboratory tests, specialist referral, and close follow-up.

Results: Among 240 WCV visits, 33 (41%) of 80 Ob and 62 (78%) of 80 Ov patients were diagnosed incorrectly. Management strategies were documented with significantly increasing frequency as BMI class increased with respect to dietary counseling, activity counseling, laboratory screening, specialist referral, and close follow-up duration (Table 1). These management strategies were applied to a low percentage of cases even in the event of accurate weight status diagnosis. Results of multiple regression analyses indicate that documented dietary goals, activity goals, and laboratory screening were significantly associated with greater odds of appropriate diagnosis (adjusted ORs range from 4.1-3.5, all p's < 0.01).

Table 1. Physician-Assigned Management by Weight Category

	Diet Counseling	Exercise Counseling	Laboratory Screening	Referral	Follow-up <5 months
Healthy, *n=80*	17(21%)	6(8%)	11(14%)	2(3%)	11(14%)
Overweight, *n=80*	28(35%)	20(25%)	18(23%)	2(3%)	14(18%)
Obese, *n=80*	53(66%)	36(45%)	26(33%)	9(11%)	33(41%)
Chi-Square, *p*	35.2, <0.00001	29.4, <0.00001	7.9, <0.02	7.9, <0.02	19.6, <0.0006

Conclusion: Physicians under-recognize and under-manage Ob and Ov subjects during WCV. Accurate Ob and/or Ov diagnosis is associated with increased provision of expert-recommended management strategies and thus should be a focus of quality improvement measures targeting the treatment of childhood obesity.

ABSTRACT 13.

MATERNAL STRESS, RECALL AND PERCEPTION OF THEIR NEONATAL INTENSIVE CARE UNIT (NICU) GRADUATE CHILD'S NEEDS

Sara Hyatt, DO[1], Catherine Messina PhD[2,] Aruna Parekh MD[1].

[1]Department of Pediatrics, Stony Brook Children's Hospital, [2]Department of Family, Population and Preventive Medicine, Stony Brook University, Stony Brook NY.

Background: Advances in neonatal intensive care have led to a decrease in neonatal mortality rates. However, complexity of care and co-morbidities, such as developmental delays, requiring close follow-up have increased. It is unclear if maternal stress influences maternal recall and perception of their child's prognosis and follow-up needs.

Objective: To determine the effect of maternal stress on recall and perception of their infant's prognosis and follow-up needs after discharge from the NICU.

Methods: At the time of NICU discharge, consent was obtained and mothers filled out 2 surveys: 1) the validated Stanford Acute Stress Reaction Questionnaire (SASRQ) and 2) our maternal recall and diagnosis/es perception/follow-up needs questionnaire. Mother's recall and their child's electronic medical record (EMR) events were coded dichotomously as present vs. not present. Mother's recall of a clinical event was then compared to their child's EMR information documenting that event using cross-tabular analyses and the McNemar test for paired measures.

Results: We enrolled 40 mother-infant pairs with a mean maternal age of 31 ± 10 years and newborn gestational age of 33 ± 7 weeks. Sixty-eight percent of deliveries (n=27) were cesarean sections. Ninety percent of infants (n=36) required supplemental oxygen, 71% (n=28) required TPN, and 20% (n=8) had intra-ventricular hemorrhage. When compared with the EMR, mothers' responses were between 65-85% in agreement regarding their newborn diagnoses. However, when mothers were asked about the need for follow-up with early intervention (EI) services, there was only agreement with EMR 50% of the time. Nineteen percent of mothers (n=6) were diagnosed with acute stress disorder. We found no correlation between maternal stress and their recall or their child's health perception/needs.

Conclusion: Our pilot study suggests that mothers were able to recall significant medical events during the NICU course. However, some mothers were not aware that their infant would require early intervention despite knowing the infant's diagnosis and presence of co-morbid conditions. A larger sample size is needed to determine true correlations. Addressing maternal stress and examining ways of optimal communication of EI needs may aid in long term infant's prognosis.

ABSTRACT 14.

SEVERE HYPOTONIA OBSERVED IN NOONAN-LIKE SYNDROME WITH A SHOC2 MUTATION: STILL DEFINING THE PHENOTYPE.

Caitlin Keane, DO[1], Jody Weiss-Burns, MSGC[2], and Patricia Galvin-Parton, MD [1,2]

[1]Department of Pediatrics, [2]Division of Pediatric Genetics, Stony Brook Children's Hospital, SUNY-SB School of Medicine, Stony Brook, NY

Background: In 2003, Mazzanti *et al*. presented three children with short stature, similar facial dysmorphism, macrocephaly, enlarged cerebral spinal fluid spaces, short neck with redundant skin, severe GH deficiency, mild psychomotor delay with ADHD, mild dilation of the pulmonary root, and a unique combination of ectodermal abnormalities. This became known as Noonan-like Syndrome with loose anagen hair.

Case: We describe a 5 year old boy with severe hypotonia originally felt to have an underlying neuromuscular disorder. At 6 months, he was sent to Genetics, Pulmonary and Neurology for evaluation. He was noted to have global hypotonia, poor head control, and mild ptosis. There were no reports of aspiration. Chromosomes including FISH for PWS, chromosome microarray, SMA testing, CPK, carnitine profile, and screening for fatty acid oxidation defects were all normal. At 10 months, he had persistent generalized hypotonia and was unable to sit without support. His head circumference was in the 95th percentile, while height and weight trended in the 15th – 20th range. MRI and CT scans were normal. A DNA panel for neuromuscular disorders was unrevealing. At 3 years, he started to walk, was language-delayed, and developed obstructive sleep apnea, as well as dysmorphic features (macrocephaly, ptosis, excessively long eyelashes, and sparse black hair). He did not have a low posterior hairline or chest wall defect. Echocardiogram revealed a mild hypertrophic cardiomyopathy without septal or valve defects. Although short, endocrine evaluations determined appropriate growth velocity and GH level.

Discussion: Whole exome sequencing revealed a *de novo* c.4A>G missense mutation in SHOC2. This recurrent missense change is the only mutation associated with Noonan-like Syndrome with loose anagen hair that shares c characteristics with cardiofacial cutaneous syndrome. The S2G variant accounts for 5% of cases with a Noonan-like phenotype. Noonan-like Syndrome with loose anagen hair is a relatively newly described disorder. The final phenotype and natural history is still emerging. This patient's findings - hypotonia, long eyelashes, and nonclassical features - may contribute to this emerging phenotype. Continued reporting on the phenotypic variability in rare syndromes will help develop a better understanding of when to suspect and further characterize these conditions.

ABSTRACT 15.

IMPROVING HIGH-RISK CLINIC COMPLIANCE OF NICU GRADUATES

Kevin Louie, DO[1], Dianne Lee, DO[1], Aruna Parekh, MD[2]

[1]Dept. of Pediatrics, and [2]Division of Neonatal-Perinatal Medicine, Stony Brook Children's Hospital,

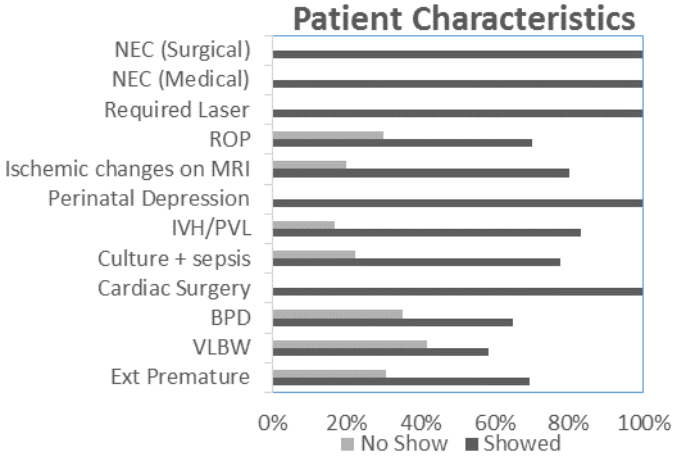
Background: Advances in antenatal and neonatal intensive care have increased survival rates in preterm infants, however incidence of neurodevelopmental disability remains high. The American Academy of Pediatrics released a statement in 2008 reaffirming "high-risk infants should be enrolled in follow-up clinics specializing in the neurodevelopmental assessment of high-risk infants". Despite the importance of high-risk clinic, national follow-up rates range from 21-40%. Reasons for low rates of follow-up are multifactorial including: socioeconomic factors such as parental demographics, lack of insurance, lack of knowledge for need for follow-up, numerous multispecialty appointments or availability of clinic dates. Association of morbidities and their impact on follow-up is less well understood.

Objective: To determine the impact of neonatal comorbidities on high-risk clinic follow-up rates.

Methods: We reviewed 1,043 patient charts of neonatal intensive care unit (NICU) graduates from April 2017 through December 2017. One hundred and four infants were referred to the Stony Brook High-Risk Clinic. Comorbidity risk factors known to be associated with altered neurodevelopmental outcomes including gestational age, birth weight, bronchopulmonary dysplasia (BPD), cardiac surgery, culture proven sepsis, intraventricular hemorrhage/periventricular leukomalacia (IVH/PVL), perinatal depression, ischemic changes on MRI, retinopathy of prematurity (ROP), and necrotizing enterocolitis (NEC) were collected for these patients.

Results: Of 104 referred patients, 50/90 (55%) patients kept their appointment, 40/90 (44%) did not, and 14 patients have appointments in future, thus not included in the analysis. Our data shows all patients referred with NEC (3/3), ROP requiring laser (3/3), perinatal depression (3/3), and cardiac surgery (3/3) followed up. The average gestational age and birthweight for those who followed up were 32 4/7 weeks gestation and 2,045 grams compared to 34 1/6 weeks and 2,238 grams who did not.

	Total # Referred	% out of 90
Ext Premature	13	14%
VLBW	24	27%
BPD	20	22%
Cardiac Surgery	3	3%
Culture + sepsis	9	10%
IVH/PVL	6	7%
Perinatal Depression	3	3%
Ischemic changes on MRI	5	6%
ROP	10	11%
Required Laser	3	3%
NEC (Medical)	2	2%
NEC (Surgical)	1	1%



CONCLUSION: Preliminary data with limited numbers to derive statistical significance show a trend towards higher follow-up rates in infants with lower gestational age, birthweight and patients with NEC, ROP requiring laser, perinatal depression, and cardiac surgery. A larger sample size and post-discharge surveys or phone calls may help answer why the remainder of patients fail to follow up.

ABSTRACT 16.

ASSESSING KNOWLEDGE OF CAREGIVERS ON SAFE SLEEP PRACTICES IN A LEVEL III NEONATAL INTENSIVE CARE UNIT

Denise Nunez, MD, Susan Katz, DNP, Shanthy Sridhar, MD, Aruna Parekh, MD

Department of Pediatrics, Division of Neonatology, Stony Brook Children's Hospital

Background: Premature infants are at higher risk for Sudden Infant Death Syndrome (SIDs) and Sudden Unexpected Infant Death (SUID). In 2016, Safe Sleep Recommendations (SSR) were endorsed by the American Academy of Pediatrics to minimize the risk of SIDS and SUID. Neonatal Intensive Care Unit (NICU) providers play a critical role in modeling preventive measures to be adopted by families post-discharge: positioning the baby while in NICU may significantly impact parental practice at home. However, studies show that up to 50% of NICU providers fail to communicate current SSRs to parents.

Objectives: To assess knowledge of NICU providers about SSRs and to implement an educational strategy to improve it.

Methods: Quality Improvement (QI) design using PDSA (Plan, Do, Study, Act) cycles was used to assess knowledge gaps in SSR among all NICU Providers (physicians, residents, nurse practitioners, and nurses) using an anonymous survey (S1). An educational module (EM) was then developed and distributed to NICU providers, with an anonymous Survey (S2) distributed post-intervention to assess improvement scores and knowledge retention. Student's t test was used for statistical analysis.

Results: Surveys were distributed to 192 providers using Survey Monkey. Eighty-nine of 192 (42%) providers responded to S1 and 103 of 192 (53%) responded to S2, p=0.74. Responses to questions about SUIDs, Preterm vs Full term SSRs, and influence of ethnicity are compared pre- and post-EM delivery (Table 1).

Conclusion: Consistent with national data, our results demonstrate that knowledge gaps in SSR continue to persist among NICU providers. More emphasis on education about SSR for premature infants must be adopted by all NICU providers.

Table 1.	S1 survey (n=89)	S2 survey (n=103)	P Value
Overall Improvement score after EM (mean, ± standard deviation)	6.28±1.49	7.19±1.25	0.39
Correct Answers to SUIDs Questions (%)	75.6	90.3	0.07
Correct Answers to SSR (preterm vs term newborn) Questions (%)	40.7	66	**0.01**
Correct Answers to Impact of Ethnicity on High Risk Infant Outcomes Questions (%)	70.8	81.6	0.79

ABSTRACT 17.

FACTORS OF ATTRITION IN A PEDIATRIC WEIGHT MANAGEMENT CENTER: THE PERSPECTIVE OF PARENTS

Nan Shin, DO, Rosa Cataldo, DO, MPH, Catherine Messina, PhD, and Josette Bianchi, MD

Background: More than 1/3 of children in the U.S. are overweight/obese. Pediatric weight management clinics and short-term programs have shown clinical improvement in children's health. However, high clinic attrition rates are one of the most frequently cited challenges. Multiple studies have looked at various patient factors as predictors for attrition. Previous studies by our group found that clinic attrition was associated with male gender and distance from clinic site.

Objective: To evaluate patient and familial factors associated with attrition at Stony Brook Children's Hospital Healthy Weight and Wellness Center (HWWC). To assess attitudes, perceptions, beliefs of caregivers/young adults seen at HWWC through follow-up survey on issues related to retention and attrition.

Methods:

1) We performed a retrospective analysis of 223 patients (ages 5-22 years) enrolled into the HWWC from October 1, 2013 through October 1, 2017. Using propensity score approach to estimate the impact estimate i.e. the likelihood of attrition, we examined the association between participant and familial health factors on likelihood of attrition.
2) We then administered telephone surveys to the same population (n=223) to assess parental attitudes, perceptions beliefs regarding their experience with weight management clinic, comparing attrition vs retained groups.

Results: Characteristics of Retention Group vs Attrition Group are presented in Table 1.

1) Family history of diabetes was associated with increased likelihood of retention in the clinic (Impact Estimate 1.399, 95%CI 0.197 to 2.601, p=0.023, Std. Error 0.613,) while interestingly, family history of heart problems was associated with decreased likelihood of retention (Impact Estimate -1.639, 95%CI -2.74 - -0.539, p=0.004, Std Error 0.562).
2) Our telephone survey had a response rate of 11.6% (26/223). 'Clinic characteristics' were the most commonly cited reason (69%) to not follow-up. This did not change when comparing families who dropped out after the first visit versus families who continued.

Table 1.		Retention n=90 (%)	Attrition n=132 (%)	P value
Gender	Female	53 (23.9%)	70 (31.5%)	
	Male	37 (16.7%)	62 (27.9%)	0.644
Race	White	45 (36.6%)	78 (63.4%)	0.482
	African American	11 (42.3%)	15 (57.7%)	0.679
	Other	10 (50.0%)	10 (50.0%)	0.107
	More than 1 race	8 (53.3%)	7 (46.7%)	
Family History	Type 2 DM	56 (42.4%)	76 (57.6%)	0.023
	Heart Disease	35 (35.7%)	63 (64.3%)	0.004
	High Blood Pressure	55 (39.6%)	84 (60.4%)	0.911
	Overweight/Obesity	63 (39.6%)	96 (60.4%)	0.370

Conclusions: Results suggest that most patient/familial health factors are not associated with attrition in our weight management center. However, family history characteristics were associated with retention (diabetes) and attrition (heart disease). Parental perception of the center was associated with attrition and requires a future, more detailed needs assessment to improve retention outcomes.

ABSTRACT 18.

CUMULATIVE DOSE OF METHOTREXATE AND THE RISK OF DEVELOPING OSTEONECROSIS IN CHILDREN WITH ONCOLOGIC DISEASES

Lisa Smith, MD, MPH, Catherine Messina, PhD, and Laura Hogan, MD

Background: As survival rates improve among pediatric oncology patients, they face an increasing number of late complications of cytotoxic agents, such as osteonecrosis (ON). Osteonecrosis is a potentially debilitating, long-term complication of high-dose chemotherapy in children treated for hematologic malignancy. While the association between ON and corticosteroid has been recognized for decades, ON has also been reported in association with high dose methotrexate. However, the data are lacking on studies specifically evaluating the role played by methotrexate in the development of ON.

Objective: To present a case series of pediatric oncology patients with ON and to examine the association between the cumulative dose of methotrexate and ON.

Methods: We conducted a retrospective chart review of 182 children who received chemotherapy at Stony Brook Children's Hospital from 2000-2016. Patients with ON were identified by MRI results. Cumulative chemotherapy doses per m^2 were acquired from chemotherapy roadmaps and orders sheets.

Results: ON was diagnosed in 10 out of 182 children (5.5%) with a mean age of 13 years, after a mean of 27 months from initiation of chemotherapy (table below). Underlying malignancies of the children who developed ON were Non-Hodgkin Lymphoma (NHL), B-cell Acute Lymphoblastic Leukemia (ALL), and Acute Myelogenous Leukemia (AML). There was a significant association between receiving methotrexate and developing osteonecrosis. (p=0.003). However, the cumulative methotrexate dose (mg/m2) did not differ significantly between the patients with ON (18,405mg) and without ON (17,142mg, p = 0.917). Patients with ON underwent a longer treatment course at time of their diagnosis (27 months), which differed significantly from patients without ON (17 months, p = 0.035).

Conclusion: ON as determined by MRI was found to be a rare complication in children who received chemotherapy at Stony Brook Children's Hospital. There is a significant association between developing osteonecrosis and receiving methotrexate as well as undergoing a longer chemotherapy treatment. While not statistically significant, patients who developed osteonecrosis are likely to be diagnosed with ALL, AML, or NHL, older at age of diagnosis, and have received a higher cumulative dose of methotrexate.

Table 1: Characteristics of the 10 Patients Who Developed Osteonecrosis

Case ID	Age at Diagnosis (years)	Gender	Race	Diagnosis	BSA (m^2)	Months of treatment	Cumulative MTX dose (mg)	Cumulative Corticosteroid dose (mg)
60	9.12	Male	White	ALL	1.0	39.0	38700.0	8920.0
62	4.39	Female	White	ALL	0.8	27.0	21050.0	2844.0
63	16.60	Female	White	ALL	1.64	33.0	62400.0	2987.0
156	10.89	Female	White	ALL	1.4	27.0	33932.0	5671.0
623	16.65	Female	White	ALL	1.61	28.0	43538.4	6993.0
1507	15.26	Male	White	AML	2.01	13.0	0	0
6941	12.5	Female	White	NHL	1.57	27.0	974	5903
8459	11.8	Female	Black	NHL	1.25	26.0	27200	9194
8153	12.0	Male	Other	ALL	1.46	28.0	51069	11898
9228	16.9	Female	White	ALL	1.87	26.0	40699	13510
63	16.60	Female	White	ALL	1.64	33.0	62400.0	2987.0

ABSTRACT 19.

ECHOCARDIOGRAPHIC ASSESSMENT OF RIGHT VENTRICULAR WALL THICKNESS AT THE INFERIOR WALL IN NEONATES

Terrence Tsui, DO[1], Laurie E. Panesar, MD[1], and James C. Nielsen, MD[2]

[1]Department of Pediatrics, Stony Brook University, Stony Brook, NY; [2]Department of Pediatrics, NYU Langone Health, NY, NY.

Background: Echocardiographic (ECHO) reference values distinguish normal from abnormal heart morphology and function. Normative data for anterior right ventricular wall thickness (aRVWT) are published. However, obtaining aRVWT is technically difficult in neonates due to poor acoustic windows limiting its clinical utility. Quantification of inferior RVWT (iRVWT) may be a more accurate and reproducible method to assess right ventricular hypertrophy (RVH) in neonates due to improved visualization.

Objectives: To explore the feasibility of iRVWT as a measure of RVH in neonates and to report normal values for iRVWT in neonates.

Methods: Retrospective ECHO data from 2012-2014 were analyzed for all full-term neonates ≤ 72hours old. ECHO indications were: murmur, family history, arrhythmia, abnormal electrocardiogram. Exclusion criteria were: ECHO abnormality (except PFO or/and small PDA). Diastolic aRVWT and iRVWT measurements were performed by single observer. iRVWT was obtained from a sub-xiphoid short-axis view at mid-ventricular level. From same image, left ventricular (LV) inferior mid-ventricular wall thickness was measured. aRVWT was obtained from parasternal short-axis view at mid-ventricular level. The mean (±SD) in aRVWT and iRVWT were compared using Student T-test. Number of successful measurements of aRVWT versus iRVWT was compared using Chi-Squared test. P value < 0.05 was significant.

Results: Seventy subjects were identified and 5 were excluded due to suboptimal ECHO views. Sixty-five remaining subjects (35% were female) had median (range) weight (kg) 3.42 (2.45 - 4.68); median (range) BSA (m^2) 0.22 (0.18 - 0.27). iRVWT was thicker than aRVWT: 2.36 ± 0.46mm versus 1.89 ± 0.31mm, respectively, p <0.001. The ratio of iRVWT to iLVWT was 0.97 ± 0.18 indicating similar wall thickness. Ability to visualize iRVWT was superior to aRVWT: 86% versus 56%, respectively, p<0.05. Indexed iRVWT to BSA^1 and BSA^0.5 were 10.7±2.07 and 5.0±0.96, respectively.

Conclusion: iRVWT appears to be a superior method to assess RVH in term neonates compared to aRVWT. Normative data for iRVWT were presented unindexed, indexed to size (BSA^1 and BSA^0.5) and as a ratio of iRVWT/iLVWT. Validity of iRVWT as a measure of RVH requires further study with a larger sample size, multiple observers, and in disease states with abnormal RV afterload.

ABSTRACT 20.

CLINICAL AND MOLECULAR CHARACTERISITCS OF A RARE GENETIC DISORDER CAUSING SELF-INJURY: PRDM12 MUTATIONS ASSOCIATED WITH HEREDITARY SENSORY AND AUTONOMIC NEUROPATHY

Jessie Zhang, MD[1], Denise Nunez, MD[1], Jody Weiss-Burns, MSGC[2] , Patricia Galvin-Parton, MD[1,2]

Department of Pediatrics[1], Division of Pediatric Genetics[2], Stony Brook Children's Hospital, SUNY SB School of Medicine, Stony Brook, NY

Background: Young children with Congenital Insensitivity to Pain (CIP) may have mouth or finger wounds due to repeated self-biting. CIP is a form of peripheral neuropathy which can result in unintentional, self-mutilation behavior that can be devastating. The patient described here had an evolving clinical picture that has not been previously described. Whole Exome Sequencing (WES) identified the underlying gene defect which is responsible for a rare disorder that causes CIP.

Case: This patient was born at term via C-Section for breech presentation. He developed tachypnea and was transferred to NICU. He had generalized hypotonia and limited range of motion in elbows and knees. His cardiac echocardiogram, brain/spinal cord MRI, skeletal x-rays, NBS, and chromosome microarray were all normal. When his teeth erupted at 7 months, the patient developed lacerations of his tongue, lips and thumb from self-biting. He displayed no evidence of pain from these injuries. Studies to rule out Lesch-Nyhan were negative. Whole Exome Sequencing revealed a novel homozygous mutation in the PRDM12 gene that has been associated with autosomal recessive hereditary sensory and autonomic neuropathy type VIII (HSAN8).

Discussion: Prdm proteins are a family of epigenetic regulators that control neural specification and neurogenesis. Mutations in the PRDM12 gene family is responsible for HSAN8 which is characterized by being unable to feel acute or inflammatory pain from birth. This patient is homozygous for the c.600_606delGTGGTACinsCA pathogenic variant in the PRDM12 gene. In addition, this patient has developmental delays and limited range of motion in major joints which have not been previously described.

Conclusion: CIP is a rare disorder. The scope of investigation includes new genes that were not previously recognized. The phenotype and natural history of disorders associated with these newly described genes is still emerging. This patient's finding of self-injury behavior coupled with joint limitations will help add to the emerging phenotype. Continued reporting on phenotypic variability in rare syndromes will provide better understanding of when to suspect these rare genetic disorders and will aid in earlier therapeutic options.

ABSTRACT 21.

BASELINE DEPRESSION AND QUALITY OF LIFE IN TRANSGENDER YOUTH: ADOLESCENT AND PARENTAL PERCEPTIONS

Christal Achille, MD[1], Tenille Taggart, MA[2] , Nicholas Eaton, PhD[2] Jennifer Osipoff, MD[1], Kimberly Tafuri, DO[1], Andrew Lane, MD[1] and Thomas A. Wilson, MD[1]

[1]Division of Pediatric Endocrinology, Department of Pediatrics and [2]Department of Psychology, Stony Brook University, Stony Brook, New York

Background: Depression and suicide attempts are common among lesbian, gay, bisexual and transgender (LGBT) youth; transgender individuals are at the highest risk. Risk factors include lack of family support, hopelessness and prior suicide attempts. Parental support has been shown to protect against mental health problems in lesbian, gay, bisexual adolescents, but this has not been as closely investigated in transgender adolescents and youths.

Objective: To examine baseline depression and quality of life scales of transgender youths and parents' baseline perceptions of their children as part of a longitudinal study on the effect of endocrine intervention in transgender patients (either suppression of endogenous pubertal hormones utilizing GnRH agonists/anti-androgens/suppressors of menstruation, or addition of cross-sex hormones).

Methods: Individuals with sex chromosome abnormalities and disorders of sexual differentiation were excluded from the study. After obtaining informed consent, subjects completed the Center for Epidemiologic Studies Depression Scale, Patient Health Questionnaire Modified for Teens, and Quality of Life Enjoyment and Satisfaction Questionnaire-modified while parents were given Nisonger Child Behavior Rating Form.

Results: Ninety-five subjects were enrolled from the pediatric endocrine clinical practice: 27 (28%) male-to-female, 68 (72%) were female-to-male. Mean age was 16.3 (+/-2.3) years, ranging from nine to 21 years old. Thirty-four (36%) were on psychiatric medication at first visit. Sixty-two (65%) reported depression as defined by the depression scale, and 33 (14%) reported current suicidal thoughts. Thirty (32%) patients reported having had at least one lifetime suicidal thought. This compares to 10.5% prevalence of depression and suicidal ideation among adolescents in the general population in NY in 2017 per CDC. Parents noted that transgender children scored the highest percentage in withdrawn behaviors [15.86 of 48 points (33.04%)] and overly sensitive behaviors [3.98 of 12 points (33.15%)]. Parents scored children very low on conduct disorder problems [3.25 of 42 points (0.07%)].

Conclusion: Transgender youth demonstrate a high prevalence of depression and suicidal ideation. Parental perception matched the high depression scores and noted higher withdrawal and overly sensitive behaviors in their children. This underscores the importance of an ongoing longitudinal study to examine whether endocrine intervention has an impact on depression and suicidality.

ABSTRACT 22.

THE INFLAMMATORY MODULATOR HIGH-MOBILITY GROUP BOX PROTEIN 1 IS POST-TRANSCRIPTIONALLY REGULATED DURING THE LIFE CYCLE OF THE ONCOVIRUS EPSTEIN-BARR VIRUS

Jozan Brathwaite, MD[1], Sandeepta Burgula, PhD[2], Eric Burton, BS[3], and Sumita Bhaduri-McIntosh, MD, PhD[4]

[1]Division of Neonatology, Department of Pediatrics, Stony Brook Children's Hospital, Stony Brook, NY; [2]Department of Microbiology, College of Science, Osmania University, Hyderabad, India; [3]Department of Molecular Genetics and Microbiology, Stony Brook University, Stony Brook, NY; and [4]Division of Infectious Diseases, Department of Pediatrics and Molecular Genetics & Microbiology, University of Florida and Shands Children's Hospital, Gainesville, FL, USA

Background: High-mobility group box protein 1 (HMGB1) is a ubiquitous, highly-conserved, protein with multiple functions depending on its subcellular location. In the nucleus, it is a non-histone DNA chaperone involved in transcriptional regulation while in the cytosol, it is a key modulator of the inflammatory response. HMGB1 is elevated in multiple medical conditions and has been shown to promote tumor metastasis. In the perinatal period, it is elevated in tracheal aspirates of neonates with bronchopulmonary dysplasia and in amniotic fluid of mothers in preterm labor. Despite its well-described role in mediating inflammation, the regulation of HMGB1 itself is poorly understood.

Objective: To determine whether HMGB1 is regulated during the transition from latency to the pro-inflammatory lytic phase of Epstein-Barr virus (EBV).

Methods: We compared HMGB1 levels among pure populations of cultured human B lymphocytes latently-infected to those lytically-infected with EBV via western blot, qRT PCR and flow cytometry. Bromouridine sequencing was then utilized to determine if HMGB1 was transcriptionally regulated.

Results: Steady-state levels of HMGB1 mRNA were 30-fold higher among lytic B cells at 24 hours; via western blotting, HMGB1 protein levels were also higher in lytic B cells in comparison to latent B cells. At the single cell level, more than 50% of lytic B cells had higher levels of HMGB1 compared to latently-infected cells. However, via bromouridine sequencing, >2-fold decreased levels of nascent HMGB1 mRNA were observed in lytic B cells in comparison to latent B cells. These findings represent preliminary data; further replicates need to be performed in order to prove statistical significance.

Conclusions: As expected, increased levels of HMGB1 were demonstrated among lytic populations of B cells. However, contrary to our expectation, we found via bromouridine sequencing that HMGB1 mRNA synthesis decreased when cells transited from latency to the lytic phase. This suggests that HMGB1 is regulated at the post-transcriptional level, likely through increased stabilization of the message. These findings provide insight into possible mechanisms by which HMGB1 may be regulated during inflammatory processes.

ABSTRACT 23.

CAN DEGREE OF CELIAC SEROLOGICAL MARKER ELEVATION PREDICT SEVERITY OF HISTOLOGICAL CHANGES?

Sherin Daniel, MD[1], Grace Gathungu, MD[1], Daniel Mockler, MD[2], Anupama Chawla, MD[1]

[1]Departments of [1]Pediatrics and [2]Pathology, Stony Brook Hospital

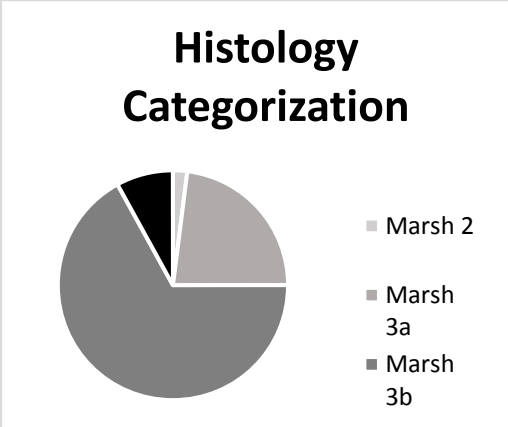
Background: Celiac disease (CD) is an autoimmune inflammatory disease of the small intestine as a result of reaction to gluten. Exclusion of dietary gluten is the mainstay of treatment that necessitates a precise diagnosis. Currently, the diagnostic gold standard for CD is duodenal biopsy. The usual approach is to perform serological screening and, if abnormal, confirm by biopsy.

Objective: To determine the association between symptoms, serology and histology among CD patients is the primary aim. The secondary aim is to compare time to normalization of serology after being gluten-free.

Methods: This is a retrospective study which includes patients with celiac serology and confirmed histology following endoscopies completed between 2006 -2016. Patient data for associated factors including abdominal pain, short stature, anemia, weight loss, diabetes and constipation were gathered. Celiac markers, including total IgA, tissue Transglutaminase (TTG) IgA/IgG, Gliadin IgA/IgG, and Endomysial IgA/IgG, data were collected. Archived histology specimens were retrieved, and the pathologist who was blinded to serological and clinical data determined a histological score using the modified Marsh classification.

Results: Histology was categorized as Marsh 2, 3a, 3b and 3c (Figure1). TTG IgA, which is the most reliable marker for CD was assessed as increase in titer fold: 1-4.9x (33%), 5-9.9x (8%) and ≥ 10x (59%). The most prevalent symptoms were abdominal pain and constipation in our sample size of 133 (Figure2). The frequency of most GI symptoms was highest among subjects with Marsh 3 scores; however, none were significantly greater. There was a significant correlation between increased TTG IgA titer fold >10x upper limit and Marsh 3c by Chi-Square analysis (p .005). Mean time for normalization of TTG IgA titer was 11 months after being gluten-free: there was no significant difference among histology groups.

Conclusion: There was a correlation between degree of elevation in TTG IgA and severity of histological changes that is suggestive of CD. However, there were a few cases where it was difficult to diagnose CD based on the serology alone. At present, biopsies should continue to be a part of the investigation for CD, and prospective studies will be needed to further explore the ability to diagnose without biopsy.



.
Figure 1

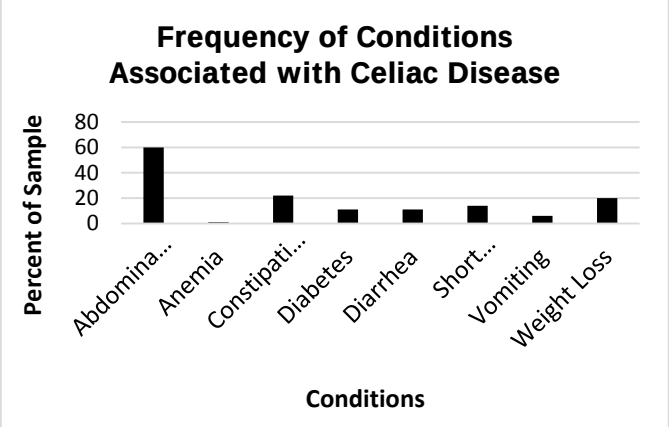


Figure 2

ABSTRACT 24.

H. PYLORI INFECTION CAUSING PERSISTENT HICCUPS

Narendra Yallanki, MD, Lisa Wilks-Gallo, MD, Jennifer Lutz Cifuni, DO, and Lesley Small-Harary**,** MD

Background: A hiccup is an involuntary, reflex-like activity that begins with contraction of the diaphragm and terminates with the abrupt closure of the glottis. Systemic disorders, including gastrointestinal (GI) pathology, can cause intractable hiccups. This case focuses on an atypical presentation of *H. pylori* infection that presented with acute persistent hiccups.

Case: An 18-year-old Hispanic male presented to the ED with acute onset of severe hiccups, nausea, and vomiting. The patient was an ill-appearing male with near continuous hiccups and frequent retching. Laboratory data were unremarkable. Over the first 5 days of admission the patient had persistent, progressively worsening symptoms with continuous hiccups and bilious emesis. Multiple therapies to abort the hiccups were attempted without sustained response. He had an extensive workup which yielded normal results until an upper endoscopy showed significant nodularity in the gastric antrum. Biopsies revealed severe chronic gastritis with evidence of rod shaped bacteria consistent with *H. pylori*. He received 3 days of IV antibiotic therapy followed by oral triple therapy. Symptoms improved within 24 hrs with complete resolution by 72 hrs.

Discussion: Causes of intractable hiccups include GI pathology, CNS lesions, toxic-metabolic disorders, drugs and psychogenic pathology. Triggers for hiccups are described as central or peripheral. GI pathology, including gastritis or GE reflux, is considered a peripheral triggering stimulus for hiccups. *H. pylori* infection has been implicated as a cause of hiccups, confirmed via successful bacterial eradication resulting in cessation of hiccups. The putative pathogenesis is that increased acid production following *H. pylori* infection stimulates esophageal mucosa irritating vagal afferents and triggering hiccups. The majority of cases of hiccups are self-limited and resolve spontaneously. Persistent hiccups refers to episodes that last for between 48 hours to 2 months duration. Episodes of hiccups lasting > 2 months are considered intractable. This case of acute persistent hiccups was presumed to be due to moderate to severe gastritis secondary to *H. pylori* infection.

Conclusion: Hiccups are usually self-limited, but with persistent hiccups GI pathology should be considered. Our case suggests an association between *H. pylori* gastritis and hiccups, suggesting that *H. pylori* should be considered early in the differential diagnosis of causes of persistent hiccups.

ABSTRACT 24.

IMPROVISATION, TEAM BUILDING, AND INTERPROFESSIONAL COMMUNICATION IN THE HEALTHCARE ORGANIZATION

Brenda MacArthur, PhD[1], Susmita Pati, MD, MPH[2], Radha Ganesan, PhD[1], and Laura Lindenfeld, PhD[1]
[1]Alan Alda Center for Communicating Science and [2]Department of Pediatrics

Background: Health care team members across disciplines often feel overworked and undervalued, leading to increased stress and eventual burnout. This phenomenon is becoming more common as the structure of the health care delivery system shifts to team-based models where a range of health care team members with different skillsets and experience levels must work together to deliver high quality patient care. Persistent challenges in interprofessional communication suggest a need for communication to be highlighted as a core outcome of training for all health care team members, regardless of profession or specialty.

Methods: At the invitation of Stony Brook Children's Pediatric Professionalism Committee, the Alan Alda Center for Communicating Science conducted 9 pilot workshops with health care providers, nurses, and allied health professionals at Stony Brook Children's Hospital (n = 83). The workshops taught effective communication skills in health care teams to create a collaborative climate and in turn, high quality patient care. Improvisation was used as a mechanism to teach participants ways to be present in the moment, listen to their colleagues, and react appropriately. Each workshop lasted approximately 1.5 hours and was co-led by an Improvisation Lecturer and Medical Facilitator.

Results: After participating in the workshop, 85.5% of participants rated the workshop "above average" or "excellent" and 90.1% would recommend it to colleagues. Participants were more likely to recommend the workshop if they rated it highly ($r = .46$, $p < .0001$) or indicated a higher interest in interprofessional communication ($r = .41$, $p < .0001$). There was a noteworthy change in interest in interprofessional communication with 84.2% of participates indicating that they were "interested" or "very interested" after the workshop, compared to 48.7% before. Those who indicated higher interest in interprofessional communication were also more likely to rate the workshop highly ($r = .67$, $p < .0001$).

Conclusion: These results offer initial support for the continued exploration of improvisation as a method for teaching communication skills in the health care setting. Improvisation-based communication training focuses on humanism in health care delivery to help team members connect to their passion for their work, and hone their communication skills to communicate clearly and efficiently with other team members.