



Stony Brook Children's

STONY BROOK LONG ISLAND CHILDREN'S HOSPITAL



**Stony Brook
Medicine**

Pediatric Department Research Day

May 9, 2012

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Welcome to the fourth annual Pediatric Trainee Research Day. It is a pleasure to share with you today 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 spectrum 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 Dr. Janet Fischel for coordinating the day, Dr. Shane McAllister who helped the presenters with their posters, members of the Pediatric Scholarship Oversight Committee, and all of the faculty who have served as mentors and provided guidance and encouragement to our trainees.

Sincerely,

Margaret M. McGovern, MD, PhD

Professor and Chair of Pediatrics

AGENDA

- 8:00 – 8:20 Sign-in: Wang Center Lobby
Breakfast and poster set up: Theater Lobby
- 8:20 – 8:30 Opening remarks: Dean Kaushansky – Main Theater
- 8:35 – 9:30 Keynote Address: Martin Pollak, MD, Beth Israel Deaconess Medical Center, Boston
“Why Do African Americans Get So Much Kidney Disease?”
- 9:30 – 10:30 Residents Platform Presentations
- Joyce Hui-Yuen, MD** *“STAT3 Mediates a Novel Mechanism to Bypass DNA Replication Checkpoint Signaling in Oncogene-Driven Transformation”*
- Sushma Krishna, MD** *“Single-course Antenatal Betamethasone Alters Lung Morphometry in Late Preterm Lambs”*
- Shweta Shah, MD** *“Multicystic Dysplastic Kidney: Serial Ultrasound versus VCUG for Diagnosing Contralateral VUR”*
- 10:30 – 10:45 Coffee Break – Theater Lobby
- 10:45 – 11:15 Fellows Platform Presentations – Main Theater
- Surabhi Jain, MD** *“Intra-nasal Lipopolysaccharide (LPS) Alters Lung Cellular Differentiation (CD)14 and Toll-Like Receptor (TLR) 2 Levels in Three-Week Old Rats Exposed to Birth Hyperoxia without Affecting CD4 and CD8 positive T cells or TLR 4 levels”*
- Shane McAllister, MD, PhD** *“A Restricted Pattern of Serum IgA Reactivity to EBV Early Antigen-Diffuse in Hodgkin Lymphoma”*
- 11:30 – 12:30 Poster Session – Theater Lobby
- 12:45 – 1:30 Lunch and Closing – Zodiac Gallery (Lower Level)

Abstracts

Resident Platform Presentations

1. Joyce Hui Yuen
STAT3 mediates a novel mechanism to bypass DNA replication checkpoint signaling in oncogene-driven transformation
2. Sushma Krishna
Single-Course Antenatal Betamethasone Alters Lung Morphometry in Late Preterm Lambs.
3. Shweta Shah
Multicystic Dysplastic Kidney: Serial ultrasound versus VCUG for diagnosing contralateral VUR.

Fellow Platform Presentations

4. Surabhi Jain, Neonatology
Intra-nasal Lipopolysaccharide (LPS) Alters Lung Cellular Differentiation (CD)14 and Toll-Like Receptor (TLR) 2 Levels in Three-Week Old Rats Exposed to Birth Hyperoxia without Affecting CD4 and CD8 positive T cells or TLR 4 levels
5. Shane McAllister, Infectious Diseases
A restricted pattern of serum IgA reactivity to EBV Early Antigen-Diffuse in Hodgkin Lymphoma

Fellow/Faculty Posters

6. RebeccaAbell, Gastroenterology
Magnets Attract Trouble in Teen
7. SeharEjaz, Endocrinology
Congenital hypothyroidism and anal atresia in a baby with chromosome 10p13 deletion : a case report
8. Janice John, Pediatrics
Do positive parenting strategies decrease externalizing behaviors? Evidence from the panel study of income dynamics.
9. Julie Khlevner, Gastroenterology
Establishing Early Enteral Nutrition with the Use of Self Advancing Postpyloric Feeding Tube in Critically ill Children

10. Sameer Lapsia, Gastroenterology
Rectal Lipoma in a Pediatric Patient with Inflammatory Bowel Disease
11. Shane McAllister, Infectious Diseases
Delayed host cell death during lytic replication of Epstein-Barr virus
12. NiyatiSkaria, Endocrinology
Accuracy of Continuous Glucose Monitoring in Pediatric Patients with Diabetic Ketoacidosis
13. Kim Tafuri, Endocrinology
Effect of Pioglitazone on the course of new onset type 1 diabetes
14. Yury Yakubchyk, Infectious Diseases
Exposure to Nanoparticles Increases the rate of Leishmaniasis and Staphylococcus aureus Infection

Resident/Student Posters

15. HeelaAzizi
Keeping Families Healthy Program: A Look at the Demographic Profile
16. ErjolaBalliu
Prevalence of undiagnosed diabetes among hospitalized patients with hperglycemia
17. Sandy Cheung
Normal Anion Gap Metabolic Acidosis: A Case Report
18. Steven Dell'Amore
The Keeping Families Healthy Program: Parent Identification and Management of Overweight and Obese Children
19. MehvishHadi
Epstein-Barr Virus-Associated HemophagocyticLymphohistiocytosis (HLH) presenting with Renal findings: Case Report
20. Stephanie Hom
Asthma, Obesity and Quality of Life
21. Zak Jacobs
A Case Report of Severe Mitral Regurgitation Associated with Refeeding Syndrome in an Adolescent with Severe Anorexia Nervosa
22. Parampreet Kathuria
Nasopharyngeal Teratoma and Imperforate Anus in a Neonate: Case Report
23. Sushma Krishna
Pain-Reducing Interventions During Pediatric Venipuncture and IV Insertion Leads to Increased Parent Satisfaction

24. Loren Murphy

Systematic survey of intubation success and opportunity in a duty-hour limited pediatric and neonatal training program

25. Lee Polikoff

Characteristics and outcomes of pediatric rapid response teams before and after automation by a pediatric early warning score (PEWS) system

26. Sheena Sharma

Successful Remission of Dense Deposit Disease (DDD) with Plasmapheresis: A Case Report

27. Mike Stern

The Stony Brook Parent Handbook: From Birth to 5 Years

ABSTRACT 1

STAT3 mediates a novel mechanism to bypass DNA replication checkpoint signaling in oncogene-driven transformation

Hui-Yuen JS, Koganti S, de la Paz A, Freeman A, Bhaduri-McIntosh S.

Background

Mechanisms that overcome the intra-S phase DNA replication checkpoint during oncogene-driven proliferation, allowing successful tumorigenesis, are not well understood. We have previously shown that *ex vivo* infection of primary human B cells with EBV was associated with activation and increased expression of signal transducer and activator of transcription-3 (STAT3), a pro-proliferative protein. STAT3 is constitutively active in the majority of human tumors, including EBV-related B cell lymphomas. We used Epstein-Barr virus (EBV), a known carcinogen, as a tool to identify mechanisms that facilitate such bypass of DNA replication checkpoint signaling.

Methods

Peripheral blood was drawn from EBV-seronegative subjects after obtaining informed consent. Peripheral mononuclear cells were isolated, CD3-positive T cells were depleted, and cells enriched for B cells were cultured for four days in media alone, with EBV, or with EBV plus a chemical inhibitor of STAT3 phosphorylation. Cells were then harvested and prepared for Western blot and flow cytometry.

Results

We confirmed that *ex vivo* infection of primary human B cells with EBV was associated with activation and increased expression of STAT3. When phosphorylation of STAT3 was chemically inhibited during EBV infection or when B cells from patients bearing naturally-occurring mutations in *STAT3* gene were infected with EBV, proliferation of cells expressing the oncogene Latent Membrane Protein-1 was halted in S phase of the cell cycle. Moreover, there was absent or delayed emergence of growth-transformed cell lines. Compared to un-infected cells, by day four after exposure to EBV, primary B cells demonstrated an 2600-fold increase in levels of phospho (activated)-ATR, the primary regulator of the DNA replication checkpoint pathway. When activation of STAT3 was chemically inhibited, a similar increase in pATR was observed along with phosphorylation of Chk1 (pChk1), its downstream target. In contrast, there was minimal to no pChk1 when B cells were infected in the presence of functional STAT3. Total Chk1 was relatively constant regardless of STAT3 function.

Conclusions

EBV upregulates STAT3 to suppress levels of pChk1 in response to activated ATR, providing a mechanism by which DNA replication checkpoint signaling may be bypassed during oncogene-driven transformation.

ABSTRACT 2

Single-Course Antenatal Betamethasone Alters Lung Morphometry in Late Preterm Lambs.

Sushma Krishna MD¹, Shetal I. Shah MD¹, Sylvia Gugino MA², SatyanLakshminrusimha MD²

- 1) *Department of Pediatrics, Division of Neonatology, Stony Brook University School of Medicine*
- 2) *Department of Pediatrics, Division of Neonatology, State University of New York at Buffalo*

Background

Late preterm infants (34 0/7-36 6/7 weeks) exhibit significant rates of respiratory distress syndrome, but the use of antenatal steroids at these gestations has not been fully evaluated. The purpose of this study is to determine the effect of antenatal betamethasone on lung histology in the late preterm fetal lamb.

Methods

Time-dated pregnant ewes were injected with betamethasone (15mg/day) or placebo on day of life (DOL) 132 & 133 (term -147 days) (N=12 each). C-section was performed on DOL 134.

Lungs from 6 lambs from each group were harvested prior to first breath. Six remaining lambs were ventilated for 6 hours. Harvested lungs were fixed-inflated and morphometric assessments were performed blind on 7 μ m cross sections stained with Hematoxylineosin.

A linear 56-point-counting grid was superimposed onto each digitized image. Air space, alveolar tissue, parenchyma, air-tissue intercepts, sampled radial alveolar count, number of alveoli-per-linear field, and mean alveolar width were counted. Alveolar density and total alveolar surface area were calculated. Full-term, day-old newborn lambs were studied for comparison. One-way ANOVA testing with Bonferroni correction was used in analysis.

Results

Both full-term and steroid-treated lambs exhibited lower radial alveolar counts and decreased alveoli-per-linear field measurements compared to control ($p < 0.001$, $p < 0.01$ respectively). Mean alveolar width was increased in steroid-treated animals compared to controls ($p < 0.05$) but reduced as compared to full term sheep ($p < 0.001$). Total alveolar surface area was significantly increased in the full term sheep compared to either the control or steroid-treated groups ($p < 0.001$ for both). No differences in the percentage of air space, parenchyma, vascularity, air space/parenchyma ratio, alveolar density or alveolar surface area were observed.

Conclusions

Exposure to antenatal betamethasone in the late-preterm period increases the size, but reduces the number of alveoli, resulting in the same percentage of air space and unchanged alveolar surface area. We speculate the improvement in short-term lung function seen in steroid-treated animals is not associated with major structural changes in the lung.

ABSTRACT 3

Multicystic Dysplastic Kidney: Serial ultrasound versus VCUG for diagnosing contralateral VUR

Shweta Shah, MD¹, Margaret Parker, MD¹, Catherine Messina, PhD¹
Department of Pediatrics, Stony Brook University School of Medicine, Stony Brook, NY.

Background: Multicystic dysplastic kidney (MCDK) is a congenital anomaly of the kidney with an incidence of approximately 1 in 4300 live births and one of the most common anomalies detected by prenatal ultrasound. The natural history of these kidneys involves compensatory hypertrophy of the normal kidney, an almost complete involution of the affected kidney and preservation of renal function by the solitary kidney. Vesico-uretral reflux (VUR) has been found in upto 30% (range 5% to 43%) of MCDK cases and Voiding Cystourethrogram (VCUG) has been traditionally used to diagnose the reflux. Recent studies have questioned the utility of routinely performing a VCUG, a test that is invasive and involves ionizing radiation, for the diagnosis of the contralateral VUR. The MCDK registry data states that the vast majority of the reflux is mild to moderate with no difference in rates of UTI or renal scarring between those with and without VUR. A renal US obtained at birth, 1 month and at 2 years, 5 years and 10 years, is reasonable to rule out, initially, findings suggestive of VUR and contralateral kidney abnormalities and then, subsequently, to document MCDK involution and evaluate the growth of the contralateral kidney. Routine VCUG should be deferred unless abnormal ultrasound features are present in the contralateral kidney or ureter.

Objective: To determine if doing serial ultrasound in patients with unilateral MCDK yielded same or better results than VCUG in diagnosing abnormalities of contralateral kidney/ureter.

Materials and Method: Our study is a retrospective chart review of all patients with MCDK who were seen in Pediatric Nephrology clinic at Stony Brook Medical Center from year March 1995 to March 2012. Those patients who had hereditary cystic disease, solitary renal cyst or who did not have a documentation of a VCUG result were excluded from the review. An approval for Exemption Review was obtained from Stony Brook University Human Subjects Committee (IRB) prior to obtaining the data. In addition to results of serial ultrasound and VCUG, data was also obtained for estimating the GFR, degree and presence of proteinuria, hypertension, Wilm's tumor and need for prophylactic nephrectomy. Data was analyzed by using standard statistical tests.

Results: 32 patients were eligible for the analysis. The number of females were slightly higher than males with the M:F ratio of 0.6 to 1. There were equal number of patients with right and left MCDK. The Glomerular filtration rate ranged from 56-150 ml/min. A prenatal diagnosis of MCDK was made in 81% of the cases. 81% had complete involution of their dysplastic kidney. All but 1 patient had compensatory hypertrophy of the normal kidney. 12% of the patients had other congenital anomalies of the kidney and urinary tract, primarily duplication of the collecting system. 2 patients had hypertension that was attributed to their underlying obesity. 2 patients had prophylactic nephrectomy of the dysplastic kidney. None of them developed Wilms tumor. Only 1 patient progressed to CKD, patient had Gr II reflux into the dysplastic kidney and presence of duplicating collecting system on normal side. 19% of the patient had hydronephrosis diagnosed on US. 31% had reflux diagnosed by VCUG, with majority being low grades reflux. Out the total number of patients who did not have reflux on VCUG, majority did not have US evidence of reflux. 1 patient had hydronephrosis but no reflux. 19% had reflux diagnosed by VCUG but had no evidence of reflux on US. This was low grade reflux and had self-resolved in subsequent VCUG. None of the high grade reflux on VCUG was missed by US. The data were analysed by McNemar's test which suggests that reflux is identified similarly by both tests because the p value is much greater than 0.05.

Conclusion: US is as reliable as VCUG in detecting high grade reflux in the contralateral kidney in patients with MCDK. Low grade reflux may be missed by US, but the majority is self-resolving. Routine VCUG can be deferred unless abnormal ultrasound features are present in the contralateral kidney or ureter.

ABSTRACT 4

Intra-nasal Lipopolysaccharide (LPS) Alters Lung Cellular Differentiation (CD)14 and Toll-Like Receptor (TLR) 2 Levels in Three-Week Old Rats Exposed to Birth Hyperoxia without Affecting CD4 and CD8 positive T cells or TLR 4 levels

SURABHI JAIN, MD¹, SHETAL SHAH, MD¹, AVINASH CHANDER, PhD¹ and PAVAN VASA¹

¹Department of Pediatrics, Stony Brook University School of Medicine, Stony Brook, NY.

Background

Hyperoxia induces endothelial cell injury predisposing newborns to lung inflammation, which contributes to bronchopulmonary dysplasia. Premature infants exposed to hyperoxia at birth exhibit increased mortality because of late-onset respiratory pathogens or systemic bacteremia, possibly through impaired T cell recruitment into the lung. TLR 4 is a membrane receptor which plays a role in CD4⁺ and CD8⁺ T Cell recruitment in response to LPS and in anti-oxidant defense. Birth hyperoxia reduces lung TLR4 levels and CD4⁺ T cells immediately post-exposure. Intra-nasal LPS increases the acute systemic cytokine response following birth hyperoxia. However, the lung immune response after late-onset infection following birth hyperoxia is not known.

Objective

The purpose of this study was to determine the long-term effect of birth hyperoxia on lung cells expressing TLR4, TLR2, CD4, CD8, CD3, CD14 and CD45 proteins.

Design/Methods

Newborn rat pups were exposed for 24 hours to 100% O₂ (hyperoxia) or room air. Animals were returned to normoxia for 3 weeks, when all animals received intra-nasal LPS (10ug) or saline (Control). Lungs were harvested 24 hours later and analyzed by immunohistochemistry for indicated proteins. Quantitative microscopy was performed from 30-40 images in each case. Results were normalized for cell number. Statistical analysis was performed by ANOVA or Student's t test. P<0.05 was considered significant.

Results

TLR2 levels were lower in the LPS plus hyperoxia in comparison to the LPS group (p<0.01). LPS increased CD14 level as compared to the control saline (p<0.05). The levels of TLR4, CD4, CD8, CD3 or CD45 cells were similar in all experimental groups.

Conclusions Birth hyperoxia does not significantly alter lung levels of TLR4, CD4 and CD8 cells in 3-week old rats, suggesting that the previously reported short-term effect of hyperoxia on cellular immunity improves over time. We speculate the observed hyperoxia-induced serum inflammatory responses following late onset infection are not solely dependent on TLR4 levels.

ABSTRACT 5

A restricted pattern of serum IgA reactivity to EBV Early Antigen-Diffuse in Hodgkin Lymphoma

Shane McAllister¹, Marisa Rotenberg², Duane Shedd³, Nancy Mueller⁴, Ellen Chang⁵, George Miller⁶, and Sumita Bhaduri-McIntosh¹

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Background

Hodgkin lymphoma (HL) is associated with elevated serum antibodies to Epstein-Barr virus (EBV) lytic antigens. Evidence for EBV lytic reactivation during HL, likely in the oropharyngeal mucosal compartment, led us to hypothesize that sera from HL patients contain IgA to early-lytic antigens, essential for viral DNA replication. Furthermore, we asked if IgA to EBV early-lytic antigens can differentiate between sera from patients with HL and acute infectious mononucleosis (IM).

Methods

Convenience serum samples from 42 previously described patients with HL, 17 IM, and 15 healthy EBV-positive controls were obtained via IRB-approved protocols. Lytic replication was induced with sodium butyrate +/- phosphonoacetic acid and lytic antigen expression was detected by flow cytometry. Total serum IgA was detected by ELISA. The specificity of serum IgA was determined by immunoblotting recombinant EBV proteins.

Results

We found that 95.2% and 71.4% of HL patients, half with EBV-positive tumors, contained IgA directed against cells expressing EBV total- and early-lytic antigens, respectively. IgA in healthy EBV-seropositive sera did not recognize total-lytic antigen expressing cells; however, 20% healthy sera weakly detected early-lytic antigen expressing cells. The immunoblot pattern of IgA specific for EBV Early Antigen-Diffuse reliably differentiated between patients with IM and HL irrespective of EBV in tumors. The sensitivity of this pattern for distinguishing HL from IM was 92.9%, specificity 100%, positive predictive value 100%, and negative predictive value 83.3%.

Conclusions

The diagnostic immunoblot pattern described here may distinguish acute IM from HL, two EBV-related diseases that can present similarly. The robust performance characteristics of our test should be evaluated further to ascertain its utility as a predictive biomarker for development of HL and response to chemotherapy.

ABSTRACT 6

Magnets Attract Trouble in Teen

RebeccaAbell, DO, Thomas Lee, MD, Richard Scriven, MD, George Angelos, MD, and Jeffrey Morganstern, MD

Introduction

Foreign body ingestion is extremely common in the pediatric population. Of all foreign bodies ingested, 80% will pass spontaneously, 10-20% will require endoscopic removal, and 1% will need surgical intervention. With the increasing number of magnetic pieces in toys and household items, the number of magnetic ingestions is escalating, although still rare overall. These become difficult cases to manage because the history is often unclear. Magnetic ingestion can cause significant morbidity and mortality if not managed in a timely manner. Any magnetic ingestion involving more than one magnet or a magnet and a metallic object should prompt immediate management.

Case

A 14 year old female presented with acute abdominal pain. Imaging of her abdomen demonstrated 2 circular foreign bodies in the right upper quadrant and 3 similar foreign bodies in the right lower quadrant. The patient reported accidentally swallowing magnets after trying to create the illusion of a tongue ring. After 5 days without progression, pediatric gastroenterology was consulted. Esophagogastroduodenoscopy and colonoscopy were performed. No magnets were visualized, but upon removal of the upper endoscope, 2 magnets were found on the shaft of the instrument. Post-procedure, the patient presented with signs of perforation. She was taken to surgery, where jejunal perforation and 3 magnets embedded in the cecal wall were found. She had resection and anastomosis of both areas. The patient did well post-operatively.

Conclusions

To our knowledge, this is the first reported case of a magnetic object being attracted to the endoscope prompting its removal. It is well known that the ingestion of multiple magnets may be associated with significant morbidity due to perforation. Early intervention by endoscopy or surgery may be necessary, depending on the number of magnets ingested, time since ingestion, and anatomical location. If endoscopy is undertaken, the gastroenterologist should be aware of the ferromagnetic properties of the endoscope.

ABSTRACT 7

Congenital hypothyroidism and anal atresia in a baby with chromosome 10p13 deletion : a case report

SeharEjaz, MD¹, Kimberly Tafuri, MD¹, Andrew Lane, MD¹, Patricia Galvin-Parton, MD¹ and Thomas A Wilson, MD¹.

¹Department of Pediatrics, Stony Brook University School of Medicine, Stony Brook, NY, United States.

Background

22q11DS (DiGeorge syndrome, MIM 188400) is a developmental defect of the 3rd and 4th pharyngeal pouches associated with facial dysmorphism, conotruncal heart defects and absent /hypoplastic thymus and parathyroid. Deletion of chromosome 10p14-13 presents with similar defects, hence characterized as DiGeorge syndrome 2 (DGS2-MIM 601362). Congenital hypothyroidism has been reported with 22q11 deletion secondary to T-box transcription factor TBX1 deletion (MIM# 602054). To our knowledge, this is the first case of congenital hypothyroidism reported in DGS2.

Case

A full term Caucasian baby boy was born with anal atresia after an uneventful pregnancy to a 33 yrs old mother. Birth weight was 2.3 kg and length 48 cm. The initial newborn screening suggested hypothyroidism and decreased T cell subsets. Hypothyroidism was confirmed (TSH 635 mcu/ml, total T4 2.1 mcg/dl). Hypocalcemia (Ca 7.7 mg/dl, iCa 3.9 mg/dl) developed on day 2 of life. Baseline EKG showed right ventricular hypertrophy with non specific depolarization anomaly. No structural cardiac anomalies were detected on echocardiogram. Karyotype was 46, XY, del(10)(p13). Levo-thyroxine and calcium carbonate corrected the hypothyroidism and hypocalcemia. Sensorineural hearing loss was confirmed with BAER after he failed an initial hearing screen.

Discussion

DGS2 is usually associated with hypoparathyroidism, deafness and renal dysplasia (HDR/Barakat syndrome) due to associated GATA3 mutations/deletions. Although congenital hypothyroidism and anal atresia in our patient might be coincidental, it may also indicate the role of yet unknown genetic mechanisms responsible for the association of CH with DiGeorge syndrome 2. TBX1 gene deletion is thought to be responsible for thyroid dysgenesis with 22q11 deletion; however, whether TBX1 plays a role in the hypothyroidism of this patient with DGS2 is unclear.

Interestingly, the mother carried a diagnosis of Brugada syndrome and has an internal pacemaker. She has not had genetic testing to evaluate the mutations associated with Brugada syndrome. We speculate that the mother might have Brugada subtype 4 secondary to CACNB2 deletion located on 10p12.33-10p12.31 due to close proximity to GATA3 and DGS2 gene loci; however, the genetic results from parents are not available at this point.

ABSTRACT 8

DO POSITIVE PARENTING STRATEGIES DECREASE EXTERNALIZING BEHAVIORS? EVIDENCE FROM THE PANEL STUDY OF INCOME DYNAMICS

¹Janice Thomas John DO, MS, MPH, ¹Susmita Pati, MD, MPH, and ²Tia M. Palermo, PhD

¹Department of Pediatrics and ²Department of Preventive Medicine, Stony Brook University School of Medicine

Introduction

Externalizing behavioral problems in childhood have potentially severe, long term consequences. Over the last decade, there has been a significant rise in the prevalence of these behaviors. Previous studies evaluating the relationship between parenting strategies and externalizing behaviors are limited in that they are dated, limited in geographic scope and do not provide longitudinal data. This study examines the relationship between parenting strategies and externalizing behaviors in children.

Methods

We analyze data on 1,176 children in Waves II and III (2002 and 2007) of the Child Development Supplement of the Panel Survey of Income Dynamics. Externalizing behavior problems were operationalized using the Behavior Problem Index. Independent variables included parental warmth, television exposure, discussing household rules and limits, disciplining styles and other covariates. We performed pooled and longitudinal analyses using multivariate linear regressions.

Results

Children who experienced more parental warmth had lower externalizing behavior problem subscale scores in the subsequent wave ($p < 0.05$). The pooled analysis indicated that increased parental warmth ($p < 0.001$), decreased television exposure ($p < 0.001$), and nonaggressive disciplining styles ($p < 0.05$) were significantly associated with lower externalizing behaviors. Discussing rules and limits more frequently was associated with higher externalizing behaviors in both the pooled ($p < 0.05$) and longitudinal analysis ($p < 0.01$). Other covariates such as household income, primary care giver education, ethnic background and family structure also may have a relationship with the exhibition of externalizing behavior problems in children.

Conclusions

This study suggests that positive parenting strategies may help reduce externalizing behavior problems in children. While many risk factors are difficult to modify, limiting television exposure, parental warmth and disciplining strategies are ones that care givers can control. Policymakers and clinicians should partner to promote targeted evidence-based interventions to increase positive parenting approaches such as limiting and restricting screen

time, increasing parental warmth, teaching healthy disciplining styles and other beneficial parenting strategies.

ABSTRACT 9

Establishing Early Enteral Nutrition with the Use of Self Advancing Postpyloric Feeding Tube in Critically ill Children

Julie Khlevner, MD¹, Janice Antino, RD, MS, CSP², Rahul Panesar, MD³, and Anupama Chawla, MD¹

¹Division of Pediatric Gastroenterology, Hepatology and Nutrition, Stony Brook University School of Medicine; ²Division of Dietary Services, Stony Brook University School of Medicine; ³Division of Pediatric Critical Care Medicine, Stony Brook University School of Medicine

Introduction

Early nutrition support is an integral part of the care of critically ill children. Early enteral nutrition (EN) improves nitrogen balance and prevents bacterial translocation and gut mucosal atrophy. Adequate EN is often not achieved as gastric feeds are not tolerated and placing postpyloric feeding tubes can be difficult. Spontaneous transpyloric passage of standard feeding tubes without endoscopic intervention or use of anesthesia can range from 30% to 80%. We report on our experience with a 14Fr polyurethane self-advancing jejunal feeding tube in a pediatric population. These tubes have been used in the adult population with success but to our knowledge there have been no reports of its use in the pediatric age group.

Case Series

We present 7 critically ill patients 8-19 years old, admitted to pediatric intensive care unit, in whom prolonged recovery, inability to tolerate gastric feeds and dependence on ventilator was predicted at the outset. The jejunal feeding tube was successfully placed on first attempt at the bedside in all of the 7 patients within the first 24hrs without the use of a promotility agent or endoscopic intervention. Nutritional goal achieved within 48hrs of feeding tube placement was reported for each patient. Our case series demonstrates that children fed via the small bowel reached their nutritional goal earlier and did not require parenteral nutrition.

Conclusions

The self-advancing jejunal feeding tube can be used effectively to establish early EN in critically ill children.

ABSTRACT 10

Rectal Lipoma in a Pediatric Patient with Inflammatory Bowel Disease

Sameer Lapsia, MD, Julie Khlevner, MD, Jeffrey Morganstern, MD, and Anupama Chawla, MD

Pediatric Gastroenterology, Stony Brook University School of Medicine, Stony Brook, NY.

Background

Gastrointestinal manifestations of inflammatory bowel disease, especially Crohn's disease, include inflammation, edema, pseudopolyps, strictures, and stenosis.

Objective

We present a case of a rectal lipoma in a patient with inflammatory bowel disease and one of very few cases reported in children.

Design/Method

Patient is a 16 year old female with Crohn's disease complicated with perineal fistulas, joint pain, episcleritis, and erythema nodosum. Due to worsening disease despite being on maximized medical therapy such as Humira, a TNF• antagonist, the patient underwent a colonoscopy to assess the full extent of her disease and determine if surgical intervention would be an option. At approximately 10 cm from the anal opening, a smooth, polypoid mass 1.5 cm in diameter with a 1 cm base was noted protruding into the lumen. Due to concerns for a possible cancerous lesion, the polypoid mass was resected. Pathology diagnosed the mass to be a lipoma. No malignant characteristics were noted.

Results

Colonic lipomas are soft tissue tumors originating from mature adipocytes. Although rare, they are the second most common benign tumor of the colon after adenomatous polyps with an incidence ranging between 0.2% and 4.4% with no cases reported in patients with inflammatory bowel disease. Resection of these masses is essential due to complications arising from lesions larger than 2 cm such as abdominal pain, change in bowel pattern, massive bleeding, obstruction, intussusception, or perforation. Cases of these lipomas metamorphosing into liposarcomas have been reported making removal of this lesion a necessity. The large base of the mass in our patient warranted a 2-step procedure, firstly achieving hemostasis with the endoloop followed by snare cauterization of the polypoid mass.

Conclusions

Lipomas must be considered as part of the differential diagnosis when a colonic polypoid lesion is encountered in patients with inflammatory bowel disease.

ABSTRACT 11

Delayed host cell death during lytic replication of Epstein-Barr virus

Shane McAllister¹ and Sumita Bhaduri-McIntosh^{1,2}

¹Department of Pediatrics, Stony Brook Long Island Children's Hospital and ²Department of Molecular Genetics and Microbiology, Stony Brook University, Stony Brook, NY.

Background

Periodic reactivation of the tumorigenic herpesvirus Epstein-Barr virus (EBV) from latency results in the release of progeny virus. This lytic reactivation is important in the pathogenesis of EBV-mediated diseases such as lymphomas and lymphoproliferative diseases that arise during immunocompromise. Detection of lytic viral DNA and proteins is an important component of host innate and adaptive defense mechanisms. While on the one hand, attempts are made to stall production of infectious virus and pro-apoptotic signals are delivered within the infected cell, EBV is able to stave off apoptotic death of the host cell by expressing anti-apoptotic viral genes and inducing expression of anti-apoptotic and pro-survival cellular genes. Delay of host cell death allows for replication and release of progeny virus. The mechanisms contributing towards such delayed cell death in lytically infected cells has received little attention.

Methods

LCLs were established by infecting peripheral blood mononuclear cells with EBV in the presence of Tacrolimus to prevent T cell-mediated killing of EBV-infected B cells. DNA content of LCL was determined by propidium iodide staining followed by flow cytometry. DNA content of lytically-infected LCL was correlated with early or late lytic cycle by co-staining for Early Antigen-Diffuse (early lytic cycle) or gp350 (late lytic cycle). Autophagy was identified in LCL by immunofluorescent detection of LC3. Nuclear morphology of lytically-infected LCL was determined by immunofluorescent staining for either EA-D or gp350 with counterstaining of host-cell DNA with DAPI.

Results

LCL expressing surface gp350 had condensed nuclei consistent with cells undergoing the nuclear reactions of apoptosis. Notably, many of these intact cells appeared to have fractional nuclei when examined by immunofluorescence. Flow cytometry-based examination of DNA content confirmed that while all cells expressing EA-D contained 2N to 4N DNA, approximately 25% of live, gp350-positive late lytic cells contained less than 2N DNA. Simultaneous staining with Annexin V revealed a progressive increase in the fraction of Annexin V-positive lytic cells in early, intermediate lytic, and late lytic stage cells. Autophagic vesicles were identified in lytically-infected LCL.

Conclusions

Together, these results indicate that signs of cell stress are demonstrable upon lytic reactivation and worsen as lytic replication proceeds. Further studies in our laboratory are aimed at characterizing the cell death pathways activated by lytic replication of EBV and delineating viral mechanisms that are necessary to delay host cell death.

ABSTRACT 12

Accuracy of Continuous Glucose Monitoring in Pediatric Patients with Diabetic Ketoacidosis

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Background

Continuous glucose monitors (CGM) are increasingly used in the outpatient setting. Patients may make clinical decisions based on CGM readings, including during development of diabetic ketoacidosis (DKA). Because CGM devices rely on measurement of generated hydrogen ions, it is unknown whether DKA affects CGM accuracy.

Objective

We aim to determine whether a commercially available CGM (Dexcom) measures blood glucose measurements accurately compared to a standard of care bedside glucometer in the setting of DKA.

Design/Methods

Successive eligible patients ages 2 to 21 years old admitted to our hospital in DKA (venous pH <7.25) were invited to participate. Venous blood gases and chemistry panels were drawn as per standard PICU protocol. Exclusion criteria included: other known metabolic conditions, or dermatologic conditions preventing sensor placement. The CGM was placed subcutaneously at presentation. After warm-up, CGM was calibrated to the bedside glucometer initially and every 12 hours. CGM values were analyzed by comparison to bedside and serum glucose values.

Results

In this ongoing study, 91 paired reference vs. CGM values have been obtained to date from 5 patients, ages 12 to 15 years old. The mean absolute relative difference (MARD) between glucometer and CGM values is 29% (SD=25.7). The MARD between serum glucose and CGM values is 26.7% (SD=24). The MARD between mild, moderate, and severe acidosis groups was not statistically different ($p>.05$). In a Clark Error Grid Analysis- 45% of values fell in category A- desirable correlation and 44% in category B- acceptable correlation. A total of 89% fell in category A and B. Of the rest of the values, 9% fell in category C- risk for inappropriate treatment, 2% in categories D and E- high risk.

Conclusion

Other investigators studying correlations between CGM and reference values in healthy individuals with diabetes have demonstrated MARDs of 13.2 to 21.5%, and 98% of values falling within Clarke Error Grid category A and B. In contrast, our data suggest that during DKA, there is a much higher MARD between CGM and reference values, and more values fall outside Clarke categories A and B. Our results have important implications in counseling patients and caretakers regarding reliance on CGM values at times of risk for the development of diabetic ketoacidosis.

ABSTRACT 13

Effect of Pioglitazone on the course of new onset type 1 diabetes

Kimberly Tafuri, DO, MustaqGodil, MD, Andrew Lane, MD and Thomas Wilson, MD.

Background

Type 1 diabetes (T1DM) is caused by insulin deficiency resulting from progressive destruction of insulin producing beta cells. The histological hallmark of the diabetic islet is mononuclear cell infiltration or insulinitis. Thiazolidinediones (TZDs) activate PPAR γ & enhance the actions of insulin. Studies in the non obese diabetic & streptocotozin treated mouse models demonstrated that pretreatment with TZDs prevented or delayed the development of T1DM.

Objective

This study examines whether the TZD, pioglitazone (PIO), given with insulin, preserves beta cell function in patients with new onset T1DM.

Design/Methods

This is a prospective, randomized, double-blind, placebo controlled 24 week study in patients with new onset T1DM. Subjects received either PIO or placebo. A total of 15 patients were enrolled. Eligibility criteria included: T1DM, age > 6 years, & absence of liver disease. Blood sugar, HBA1C, C-peptide, & liver enzymes were measured at baseline. Boost[®] stimulated C-peptide responses were measured at baseline & 24 weeks. Patients were followed at 2 and 4 weeks and monthly thereafter. Blood sugar, insulin dose, height, weight & hepatic enzymes were monitored at each visit. HBA1C was performed every 12 weeks.

Results

Of the 15 patients, 8 received PIO and 7 placebo. 6/8 patients in the PIO group completed the study. 1 was removed from the study because of elevated transaminases (transient) and 1 withdrew. Age at entry & duration of diabetes (approximately 3 months) was similar between groups. Baseline daily insulin dose was higher in the PIO group (0.46 vs. 0.28 u/kg/d); HBA1C was slightly lower in the PIO group (6.7% vs. 7.9%). Mean peak C-peptide values were similar between groups at baseline (PIO: 1.6 vs. Placebo: 1.54 ng/ml). Mean peak C-peptide was slightly greater at 24 weeks in the PIO group compared to placebo (1.75 vs 1.5 ng/ml) which is thought to be clinically insignificant. The interaction of HBA1C and insulin dose (HBA1C* insulin/kg/day), which combines degree of diabetic control and dose of insulin required to achieve this control, showed transient improvement in the PIO group at 12 weeks but was not sustained at 24 weeks, whereas the placebo group demonstrated a progressive increase throughout the 24 weeks.

Conclusions

In this pilot study, PIO did not appear to preserve beta cell function when compared to placebo. Our negative results may be impacted by the small sample size.

ABSTRACT 14

Exposure to Nanoparticles Increases the rate of Leishmaniasis and *Staphylococcus aureus* Infection

Yury Yakubchyk, Miriam H. Rafailovich

Background

Leishmaniasis is a vector borne parasitic disease affecting millions of adults and children, mainly in underdeveloped countries. Cutaneous form of this disease is presented as chronic skin ulcerations with primary involvement of macrophages, where parasite reproduces and propagates. Often these cutaneous lesions are infected with pathogenic microflora, including *Staphylococcus aureus* (*S. aureus*) as a common commensal of skin and mucous membranes. TiO₂ and ZnO nanoparticles (NPs) due to their known anti-bacterial action are often applied to the skin in different formulations (sunscreens, ointments and creams). These NPs can also penetrate the cell membrane and alter multiple cellular functions. We studied the effect and influence of TiO₂ and ZnO NPs on *Leishmaniatropica* (*L. tropica*) parasites and on the response of J774A.1 murine macrophages to infection by *L. tropica* and *S. aureus*.

Methods

L. tropica parasites and murine J774A.1 macrophages separately were exposed to 0.01 mg/ml of TiO₂ and ZnO NPs for 1 hour with subsequent assessment of parasites and cells proliferation via light microscopy. J774A.1 cells were exposed to 0.01 mg/ml NPs for 1 hour or 24 hours and infected with *L. tropica* or opsonized *S. aureus* respectively. Infected murine macrophages cultures were incubated for 24 hours with following fixation and staining with Giemsa. The percentage of infected cells and the intensity of infection (number of parasites per 100 total host cells) were determined by light microscopy. Transmission Electron Microscopy (TEM) was performed to reveal the localization of TiO₂ and ZnO NPs in exposed to NPs and infected J774A.1 cells.

Results

We found no effect on parasite and cell proliferation after 1-hour exposure to 0.01 mg/ml TiO₂ or ZnO NPs. Intensity of infection of NP-exposed macrophages with *L. tropica* was 2 to 6 times greater than in the control group ($p < 0.05$). TEM revealed sequestration of the parasite and NPs within the same parasitophorous vacuole, yet *L. tropica* viability was not affected by the presence of NPs within the vacuole. Infection rate with opsonized *S. aureus* of NP-exposed macrophages was 40% less than of control cells.

Conclusions

These results indicate that exposure to TiO₂ and ZnO NPs decreases the ability of J774A.1 macrophages to fight bacterial infection; while at the same time increases the probability of cutaneous infection by both *S. aureus* and *L. tropica*.

ABSTRACT 15

Keeping Families Healthy Program: A Look at the Demographic Profile

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The program's study aims to identify the effectiveness of home visits in improving patient compliance and childhood immunization in a pediatric population from birth through 17 years of age. Created by pediatricians at Stony Brook Children's Hospital, *Keeping Families Healthy* is a free home visitation program designed to help families receive information and resources needed to make the best decisions regarding their child's health. The program acts as an extension of the pediatrician's office by communicating families' health questions with their child's doctor and navigating the healthcare system. The program has 3 main areas of focus; management of chronic diseases, well child/adolescent care, and newborn care. In an observational study, a target of 1300 individuals from the SBUH Tech Park Clinic is being recruited as the control group. Another 1300 individuals from the Patchogue, NY area are expected to be enrolled in the home visitation program as the intervention group. The trained community health workers tailor each family's visit to their child's specific needs using 7 program protocols: Newborn, Premature Gestation, Asthma, Diabetes, Obesity, Other Chronic Conditions, and Healthy Adolescents. While this study is ongoing, the 2 aims of my research will be (1) to determine how well the Tech Park enrollees function as a control for the intervention group, and (2) to identify the types of families likely to participate in a home visitation program, based on their demographics. These results will help us better understand what populations to target for similar home intervention programs in the future.

ABSTRACT 16

Prevalence of undiagnosed diabetes among hospitalized patients with hyperglycemia

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Introduction

Early detection of diabetes can significantly alter the disease progression. Our objective was to screen hyperglycemic patients and identify those with undiagnosed diabetes using a hemoglobin A1c test. In addition, other causes of hyperglycemia were further characterized such as pre-diabetes, stress hyperglycemia and steroid induced hyperglycemia.

Methods

We randomly selected 611 patients admitted to the General Medicine Floors, the Coronary Care Units (CCU), and the Intensive Care Units (ICU) at Stony Brook University Hospital. Patients with random blood glucose over 200mg/dL were identified and a hemoglobin A1c test was performed on each patient. A diagnosis of undiagnosed diabetes was made if the hemoglobin A1c was greater than or equal to 6.5% and there was no prior history of diabetes. Patients with HbA1c 5.7-6.4% were identified as pre-diabetes. In addition, the prevalence of stress hyperglycemia and steroid induced diabetes was also made.

Results

148/611 (24%) patients had elevated random blood glucose levels > 200 mg/dL. Upon review of these patients' medical records, 9/148 (6.1%) were found to have an elevated HbA1c • 6.5% without a previous diagnosis of diabetes, while 10/148(6.8%) were found to have HbA1c 5.7- 6.4% and were classified as pre-diabetes. 22/148 (15%) were found to have elevated serum glucose levels with HbA1c < 5.7%. Of the latter group, 17 patients (11.5%) were on steroid therapy and 5 patients (3.4%) were categorized as stress hyperglycemia. There was an increased prevalence of undiagnosed diabetes in the ICU/CCU in comparison to the General Medicine Floors (18.8% vs. 4.9%, respectively).

Conclusion

Recent reports have documented a prevalence of undiagnosed diabetes in the inpatient population as high as 18%, our study shows a prevalence of 6.1% among hyperglycemic patients, with an increased prevalence in patients admitted to the CCU/ICU compared to the General Medicine Floors.

ABSTRACT 17

Normal Anion Gap Metabolic Acidosis: A Case Report

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Background

Milk and soy are the most common food triggers of food protein enterocolitis syndrome (FPEC) in young infants, and up to 50% of those with milk protein allergy also react to soy. The incidence of FPEC attributed to milk protein is reported between 0.35-2%. In the case of milk and soy-induced FPEC, the majority of reactions are thought to be non-IgE-mediated.

Objective

We present the case of a 6 week old African American male with diarrhea and dehydration.

Methods

Our patient presented with one-day history of projectile, non-bilious, non-bloody vomiting, non-bloody diarrhea and decreased oral intake with weight loss of 570 grams over the past week. The infant was recently switching from regular formula to lactose free formula. Laboratory studies on admission reveal a metabolic acidosis. After 24 hours of rehydration, switched to a casein hydrolysate formula, emesis has resolved, the diarrhea has lessened, however the normal anion gap metabolic persists. Evaluation for amino and organic acidemias was negative. Given the persistence of a normal anion gap metabolic acidosis in the setting of improving diarrhea, the working diagnosis is a renal tubular acidosis.

Results

Renal studies including ultrasound reveal both kidneys to be within the normal limits. Subsequent follow up reveals the true diagnosis of food protein enterocolitis syndrome. The patient was then switched to an amino acid based formula. At subsequent follow up, his diarrhea had completely resolved, as had the anion gap acidosis and hypokalemia.

Conclusions

In most of milk protein allergy, the diagnosis is made by elimination of exposure to milk protein, by limiting ingestion in breast feeding mothers, or changing to a hydrolyzed casein formula. Stool occult blood is often positive, even in the absence of grossly bloody stools. In severe cases, a more extensive laboratory work up may be warranted, especially if other conditions must be excluded. In the case of milk and soy-induced FPEC, the majority of reactions are thought to be non-IgE mediated. 50-80% of children diagnosed with FPEC tolerated milk-based foods after a year of life.

ABSTRACT 18

The *Keeping Families Healthy* Program: Parent Identification and Management of Overweight and Obese Children

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Childhood overweight and obesity are increasing in prevalence. The early diagnosis of childhood obesity is important to prevent future health problems. *Keeping Families Healthy* (KFH) is a free service provided by the Stony Brook Long Island Children's Hospital. KFH sends trained community health workers (CHWs) into the home for scheduled visits. *Keeping Families Healthy* acts as an extension of the pediatrician's office by offering enriched health education. Among other topics, CHWs focus on nutrition, activity, and weight management. KFH CHWs are trained to take the heights and weights of participants and calculate BMIs and percentiles. Once a child is identified as overweight or obese, based on their BMI percentile, the CHW asks specific questions about the child's lifestyle. We hypothesize that most parents do not identify overweight/obesity as a medical condition. We also hypothesize that the children of these parents will be less active and have less healthy diets when compared with the children whose parents identified overweight/obesity as being a medical condition. This study includes 62 children enrolled in the KFH program, ages 2 to 17 years, who have a BMI at or above the 85th percentile. Upon initially signing up for the program, parents were asked if their child has any pre-existing medical conditions. Of the 62 overweight/obese children, 21% of their parents identified their child as having an overweight/obesity medical condition. This finding supports our hypothesis that many parents have an inaccurate perception of their children's health. Furthermore, this finding demonstrates the need for early identification and education for both parents and their children so that their overweight state or obesity can be addressed by modifying nutrition, eating habits and other aspects of lifestyle. Data analysis is currently being conducted and we anticipate that the results will support our hypotheses. The results from this study are critical, as they may impact future primary care practices.

ABSTRACT 19

Epstein-Barr Virus-Associated HemophagocyticLymphohistiocytosis (HLH) presenting with Renal findings: Case Report

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Background

Hemophagocyticlymphohistiocytosis (HLH) is a potential life threatening syndrome in which the body is in a state of uncontrolled inflammation. The main features of this are impaired function of natural killer (NK) cells and cytotoxic T cells (CTL). There is uncontrolled accumulation of activated T-lymphocytes and activated histiocytes. An important aspect of HLH is cytokine dysfunction. HLH is either due to familial (primary HLH) or associated with infection, autoimmune or malignancies (secondary HLH). Criteria for diagnosis include the following signs and symptoms: fever, cytopenia in two cell lines, splenomegaly, hypertriglyceridemia and or hypofibrinogenemia, hyperferritinemia, and demonstration of hemophagocytosis. An established treatment protocol exist for patients diagnosed with HLH, this protocol has significantly reduced mortality in children diagnosed with HLH. The HLH-2004 protocol consists of therapy over an 8-week period consisting of dexamethasone, etoposide, and cyclosporine +/- intrathecal methotrexate. EBV is the most commonly identified trigger in secondary HLH.

Case

We present an unusual case of EBV associated HLH and nephrotic syndrome. To the extent of our knowledge this is the first case report of EBV associated HLH presenting with nephrotic syndrome. We describe a case of a 16 year old female that presents with 1 month history of persistent fevers and malaise that was diagnosed with infectious mononucleosis via mono spot test. She subsequently developed a diffuse maculopapular rash, jaundice, lower extremity edema, low urine output and with a hepatosplenomegaly.

Discussion

It is important to have a high index of suspicion for macrophage activation syndrome (MAS) on presentation as some authors consider it secondary HLH. MAS is a life threatening syndrome seen in patients with systemic juvenile idiopathic arthritis (systemic JIA) and systemic lupus erythematosus (SLE). Although no treatment protocol is established for MAS, standard of treatment currently consist of corticosteroids +/- cyclosporine. Recent studies have reported positive outcomes with the use of Anakinra which is a soluble IL-1 receptor antagonist. Anakinra has fewer side effects and is a highly effective treatment.

ABSTRACT 20

Asthma, Obesity and Quality of Life

S Hom, C Messina, F Qureshi, C Kier

Background

Quality of life (QOL) measures have recently been included in outcome studies, emphasizing both medical scientific and patient-related outcomes. Both asthma and obesity pose significant limitations on QOL. In addition, there is literature to support a possible link between obesity and the development of asthma. Nonetheless, there is limited data evaluating the QOL of obese asthmatic and non-asthmatic subjects with their normal weight counterparts. It is also unclear whether patients who are obese and asthmatic have poorer QOL compared to normal weight asthmatics.

A preliminary study comparing QOL differences between obese asthmatics and non-asthmatics (Kier protocol Asthma Obesity) demonstrated mean QOL scores that were lower in the obese asthmatic group; however, these results were not statistically significant. This study increases sample size by extending QOL measures from the original pilot study to other subpopulations, including normal weight asthmatics and non-asthmatics.

Methods

A goal of 80 subjects between 8-17 years of age was to be recruited from Stony Brook outpatient facilities. Children were screened for eligibility by completion of an intake form, which reviewed previously-made diagnoses of asthma by a physician, current medications, and co-morbid medical conditions, as well as the child's height, weight, and calculated body mass index (BMI). Subjects and their parents were then distributed standardized PedsQL 4.0 questionnaires pertaining to general health and/or asthma, depending on the subject.

Results

Enrollment of subjects was challenging due to limited encounters with patients that met eligibility criteria, in particular the age cut-off. Statistical analysis of patient data using one-way analysis of variance (ANOVA) and Dunnett tests is still ongoing at the time of this abstract.

Conclusions

Analysis of patient data is ongoing at the time of this abstract. Continuing enrollment will enable us to further elucidate differences between obese/normal weight asthmatics from obese/normal weight non-asthmatics. Identifying these differences in QOL inventories will allow us to tailor diagnosis and management for these subpopulations of patients. Furthermore, it will enable us to target individuals at risk for poor compliance to provide them with specialized education and guidance.

ABSTRACT 21

A Case Report of Severe Mitral Regurgitation Associated with Refeeding Syndrome in an Adolescent with Severe Anorexia Nervosa

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BackgroundAnorexia Nervosa is a well-known disorder with a multitude of well-known medical complications due to starvation and extreme malnourishment. Complications are varied and commonly include electrolyte abnormalities, refeeding syndrome, amenorrhea, anemia, and cardiac abnormalities, most commonly bradycardia. Refeeding syndrome is encountered during the reintroduction of feeding after a period of starvation and is also associated with various medical complications, commonly cardiac and neurologic. Although arrhythmias and cardiac failure have been documented as complications from refeeding syndrome, there have been no documented cases of significant mitral regurgitation secondary to refeeding syndrome.

ObjectiveWe present a case of a 16 year old Caucasian female with severe anorexia nervosa who developed refeeding syndrome early in hospital treatment and subsequently developed moderate-to-severe mitral regurgitation.

MethodsOur patient, a known anorexic, presented with complaints of soreness and weakness in bilateral lower extremities, as well as difficulty ambulating for 2-3 days. She was hypotensive and bradycardic at an outside hospital and was given two normal saline boluses prior to transfer to our pediatric ICU for further management. Her initial BMI was 11.7 and was at 60% of ideal body weight. Shortly after initiating feeds she was found to have hypokalemia, hypophosphatemia and hypomagnesemia consistent with refeeding syndrome. On hospital day three, our patient was found to have a new-onset murmur, which was assessed with an echocardiogram and found to be caused by moderate-to-severe mitral regurgitation.

ResultsWhen electrolyte abnormalities were first appreciated oral supplementation was commenced with potassium phosphate. This was increased due to continued deficiencies, and magnesium was also supplemented. All electrolyte abnormalities were corrected by hospital day three. Feeding regimen was continued with modest increases in caloric intake during the first week of refeeding. The patient never developed significant symptoms from mitral regurgitation and did not sustain right heart failure because of early diagnosis, appropriate management, and careful observation.

ConclusionThis case of an adolescent female with severe malnutrition secondary to anorexia had several risk factors for the development of refeeding syndrome and its complications. However, those risk factors were accounted for during treatment and our patient still developed refeeding syndrome, likely due to the two boluses received prior to admission to the ICU. These fluids were not needed due to the compensation that is observed in patients with anorexia nervosa. Fortunately, there were no further complications and the mitral regurgitation remained asymptomatic, but had the potential to cause more significant effects, such as heart failure.

ABSTRACT 22

Nasopharyngeal Teratoma and Imperforate Anus in a Neonate: Case Report

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Background

Teratomas arising from the nasopharynx are rare occurring approximately in 1 out of every 4,000 live births. These tumors are composed of the three germinal layers - ectoderm, mesoderm, and endoderm - and are characterized by poor cell differentiation. Teratomas of the head and neck account for 2-9% of all cases of teratomas in children. Nasopharyngeal teratomas usually cause immediate dyspnea at birth requiring urgent intubation and ventilatory support. Resolution of symptoms requires surgical excision after which a low recurrence rate of nasopharyngeal tumors has been reported.

Case

We report a female neonate born at 37 weeks gestation who was found to have a large nasopharyngeal mass undiagnosed in utero, an imperforate anus, and a laryngeal web. She was born via C-section due to decreased fetal growth and significant polyhydramnios. She was born floppy and no respirations were noted. There was a significant airway obstruction and she was successfully intubated after which physical examination revealed a hard mass on the palate and a clear cystic mass on the soft palate. A CT scan of her brain and facial bones confirmed a mass extending into the nasopharyngeal region. She was taken to the OR for resection of mass and tracheostomy. Intraoperative findings with direct laryngoscopy revealed a 2.5 to 3 cm hard mass emanating off the inferior edge of the left palate. She was extubated intraoperatively and her tracheostomy was reversed at one week of life after which she developed respiratory distress. She was brought back to the OR when a bronchoscopy revealed a laryngeal web and subglottic stenosis. At this time, the laryngeal web was resected and she was retrached. On initial exam she was thought to have an anteriorly displaced anus. At 6 weeks of age she underwent an anoplasty for an imperforate anus with perineal fistula found on intraoperative examination as well as PEG tube placement. She received physical, occupational, and speech therapy while in the hospital and these services were continued after being discharged. At 7 months she appears to be doing well with no evidence of tumor recurrence.

Conclusion

Teratomas of the head and neck are rare congenital neoplasms that arise from pluripotent cells that account for less than 2% of reported cases of congenital teratomas. The recommended management of nasopharyngeal teratomas is surgical excision. Congenital anomalies have been reported in patients with nasopharyngeal teratomas such as a cleft palate, pneumothoraces, unilateral choanal atresia, and inguinal hernias. To the best of our knowledge, this is the first report of a child with a nasopharyngeal teratoma, imperforate anus requiring anoplasty, and a laryngeal web requiring resection.

ABSTRACT 23

Pain-Reducing Interventions During Pediatric Venipuncture and IV Insertion Leads to Increased Parent Satisfaction

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Background

Topical anesthesia during painful procedures leads to lower pain scores in children; however the effect on parent satisfaction has not been characterized. The purpose of this study is to determine the effect of various pain-reducing interventions during painful pediatric procedures on parent satisfaction.

Methods

Parent(s) of children undergoing either IV insertion or venipuncture on the general pediatric floor at Stony Brook Long Island Children's Hospital were included. Children received no intervention, topical anesthetic alone, distraction techniques alone, or topical anesthetic with distraction techniques. Distraction techniques included child life specialist, nursing distraction, and child life box. Patient age, gender, and type(s) of intervention used were recorded. Parent(s) reported whether the intervention(s) increased their satisfaction with their hospital stay based on a 4 – point scale (1–Strongly Disagree, 2–Disagree, 3–Agree, 4–Strongly Agree).

Results

Parents of 79 children, ages 0 to 20 years, undergoing IV insertion or venipuncture were included; 49.4% females (n=39), 45.6% males (n=36), and 5.1% (n=4) gender was not reported. Sixty patients (75.9%) received pain-reducing intervention(s) and 19 (24.1%) patients received no intervention.

Parents of children who received pain-reducing interventions were more likely to agree or strongly agree that this intervention increased satisfaction with their child's hospital stay as compared to parents of children who received no intervention (85.0% versus 52.7%, $p < 0.014$).

Topical anesthesia alone increased parent satisfaction ($p < 0.022$, OR 5.2, 95% CI 1.3-21.1), as did distraction techniques alone: child life specialist ($p < 0.003$, OR 10.8, 95% CI 2.3-51.2), nursing distraction ($p < 0.012$, OR 5.6, 95% CI 1.4-21.7) and child life box ($p < 0.010$, OR 23.7, 95% CI 2.2-260.1).

Conclusions

Our findings suggest that the use of pain-reducing interventions including topical anesthetic and distraction techniques in children during venipuncture and IV insertion is significantly related to increased parent satisfaction with their child's hospital stay. We propose that similar interventions be routinely used during pediatric hospital admissions.

ABSTRACT 24

SYSTEMATIC SURVEY OF INTUBATION SUCCESS AND OPPORTUNITY IN A DUTY-HOUR LIMITED PEDIATRIC AND NEONATAL TRAINING PROGRAM

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Background

Neonatal intubation is critical in resuscitation. Duty-hour restrictions limit real-life training opportunities, increasing reliance on mid-level providers (NNP). The 2012 Neonatal Resuscitation Program (NRP) guidelines emphasize simulation training to improve resuscitation proficiency. Our trainees receive NRP certification with reinforcement during neonatal intensive care unit (NICU) rotation. Assessment of airway skills in this post-training era has not been evaluated.

Objective: To survey intubation practice patterns in a tertiary care Level III NICU.

Methods

From Mar to Oct 2011, a sample of intubations performed by NICU providers was independently evaluated. Data on trainee level, intubation success, reason for failure, location of intubation & proficiency (0-5 Likert Scale) were collected. Demographic data included post-menstrual age & weight at intubation. Data were analyzed using Fisher's Exact Test and ANOVA with Bonferroni correction for multiple comparison.

Results

282 intubations were analyzed with an overall first-attempt success rate of 52.1%. This rate was higher in the Delivery Room (DR), (62% vs. 47%, $p < 0.02$). Distribution of intubation attempts differed among provider levels (Resident 18%, Fellow 30.8%, NNP 31.9%, Attending 20.2%). NNPs, Attendings & Fellows had increased success rates and higher Likert Scale proficiency scores compared to residents ($p < 0.0005$, $p < 0.001$ & $p < 0.007$ respectively). As expected, proficiency increased with level of experience. There was no difference noted between PGY 2, 3, & 4 (means \pm SD 2.98 ± 0.77) or the PGY 6, NNP & Attendings (4.5 ± 0.85), but there was striking change at level PGY5 when compared to both groups ($p < 0.001$). Failures included airway visualization (28.8%) esophageal intubation (26.6%) & desaturation/bradycardia (14%). Intubation success did not differ based on gestational age.

Conclusions

Resident trainees have decreased intubation success rates compared to other providers despite similar opportunities. The combination of NRP & resident training does not confer competence in infant airway management. While the PGY1 level represents inexperience in intubation skill there is an increase in proficiency at the PGY5 level that reflects exposure at the PGY4 level. We speculate added focus on intubation skill and DR exposure may increase success rates in NICU.

ABSTRACT 25

Characteristics and outcomes of pediatric rapid response teams before and after automation by a pediatric early warning score (PEWS) system

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Introduction

The Pediatric Early Warning Score (PEWS) System was created to help identify unstable patients prior to a life-threatening event. Rapid Response Teams (RRTs) were developed to assist in the evaluation and management of these patients. Since December 2008, we have automated the activation of RRTs by a PEWS of 5 or greater (red score).

Hypothesis

We aim to identify differences in characteristics of RRT calls before and after automation by the PEWS System.

Methods

We compared RRT data compiled from 2008 to 2010, with 44 RRTs recorded before automation and 69 RRTs after automation by the PEWS System. Chi-Square analysis was performed; P-value <0.05 considered statistically significant.

Results

Tachycardia was identified more often as a trigger to an automated RRT, with an increase of 26.1% ($p = 0.004$). There was a 22.9% ($p = 0.009$) reduction in RRTs triggered by recognition of acute change in mental status or agitation post-automation. Post-automation, an increase of 15.1% of RRTs required no respiratory interventions and the use of supplemental oxygen via nasal cannula or face mask decreased by 24.5% ($p = 0.011$) as well as bag-mask ventilation by 11.6% ($p = 0.035$). Patients were stabilized less often by initial interventions by 19.7% ($p = 0.038$) post-automation, but there was no significant increase in transfer to the pediatric intensive care unit (PICU) or in Code Blue calls.

Conclusions

Those RRT calls automatically triggered by the PEWS system more readily identified objective than subjective physiologic changes, such as tachycardia as opposed to changes in mental status. The use of supplemental non-invasive respiratory support decreased after automation. Automation may have influenced clinical practice as clinicians knew a priori that an RRT would be triggered by a red score based on respiratory rate and therefore triaged patients to the PICU. However, automation may also have selected for borderline patients to be triaged to the floor based on their PEWS. These patients subsequently decompensated more acutely, since they were less readily stabilized after initial interventions. Automation did not affect the calls for code blue, or deaths, but did show a trend toward increased transfers to the PICU.

ABSTRACT 26

Successful Remission of Dense Deposit Disease (DDD) with Plasmapheresis: A Case Report

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Background

DDD is an extremely rare condition affecting 2-3 persons per million in the general population. The most common etiology is due to C3 nephritic factor (C3NF), an IgG autoantibody which stabilizes C3 convertase (C3bBb), resulting in uncontrolled activation of the alternative pathway of complement. Remnants of C3 are deposited in the glomerular basement membrane causing glomerular injury. Approximately 50% of patients develop end-stage renal disease within 10 years. Due to its rarity, there are no evidence based guidelines for the treatment of DDD. We present the case of a 15 year old Caucasian male with DDD secondary to C3NF who achieved remission with short-term plasmapheresis therapy.

Methods

Our patient presented with gross hematuria and proteinuria. A native kidney biopsy revealed DDD. Components of the complement cascade were measured. C3 was found to be low and C3NF was elevated. In order to remove the pathologic antibody, pheresis treatments were initiated. Treatments were performed with the Cobe Spectra Collect Flow Path using citrate dextrose containing heparin for anticoagulation and 5% albumin as replacement fluid. Our patient had a total of 18 treatments over 6 months; weekly for 3 months then bimonthly for an additional 3 months. C3 levels and proteinuria were monitored bimonthly, and C3NF was monitored prior to the initiation of treatments, mid-way through treatments, and following cessation of treatments.

Results

C3 and proteinuria normalized after 6 months of plasmapheresis. C3NF initially decreased while receiving treatments, but increased once stopped. Despite this, he remains in clinical remission defined as urine protein/creatinine < 0.5 on 1st AM void, normal estimated GFR, normotensive, and normal serum C3.

Conclusions

We have shown that plasmapheresis is a safe and effective method for inducing remission in patients with DDD secondary to C3NF. The efficacy may be due to removal of the pathologic antibody. The persistent elevation of C3NF after pheresis could possibly be explained by change in epitope or anti-idiotypic antibodies which should be explored in the future.

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The Stony Brook Parent Handbook: From Birth to 5 Years

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Background

There is currently a staggering amount of medical information of varying quality that is readily available both online and in print. There is a need for a simplified starting point for the typical first-time parent seeking accurate healthcare information.

Objectives

To create a concise, high-yield and easily understandable handbook for parents. The handbook will aim to serve as a starting point for new parents that will complement but not replace the advice of pediatricians or that found in more comprehensive parenting guides. The handbook will review vaccines, standard anticipatory guidance, treatment of common problems and basic first aid. It will also provide a listing of suggested sources for more detailed information. When complete, the handbook will be printed in an economical format and provided free of charge to any interested parent of a patient of the Stony Brook Long Island Children's Hospital.

Methods

Content and format of anticipatory guidance was synthesized from reviewing a breadth of the available literature. Major print sources include several of the American Academy of Pediatrics (AAP) topic specific handouts and comprehensive parenting books. Other sources include the "What to Expect" series by Murkoff and Mazel and "Caring for Kids: The Complete Guide to Children's Health" from the Hospital For Sick Children in Toronto, Canada. Electronic sources included parent targeted websites of the AAP (www.healthychildren.org), the DuPont Children's Hospital (www.kidshealth.org) and the Hospital For Sick Children (www.aboutkidshealth.ca). Any discordance was settled through personal communication between the author and various board-certified pediatricians. Primary assessment of readability was performed using the SMOG score for readability. This estimates the reading level required to achieve 100% comprehension of the material in question. Secondary assessment of readability and content was assessed by directly surveying physicians and laypersons. Separate surveys were prepared for attending and resident physicians and parents. Drafts of the handbook were distributed with surveys to 15 attendings, 15 residents and 30 parents.

Results

Primary assessment with SMOG score revealed that the first draft required greater than a 12th grade reading level to comprehend most of the content. After a comprehensive revision and rewrite; the second draft scores between the 6th and 7th grade reading level. At the time of abstract submission, secondary assessment via physician and parent survey has yet to yield sufficient results for statistical evaluation. Preliminary survey response from 6 Attending Physicians and 5 Resident Physicians revealed that 100% would give a revised and finalized version of this handbook to their patients. It is anticipated that continued response via returned surveys will provide valuable insights to be included in a final revision and reformatting of the book.