



***IRISH & AMERICAN PAEDIATRIC
SOCIETY MEETING***

47th Annual Meeting

***SEPTEMBER 23-27, 2015
SHERATON MUSIC CITY
NASHVILLE, TENNESSEE USA***

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THE SOCIETY

The Irish and American Paediatric Society was conceived by Doctors Bill Kidney (Dublin), Fred Burke (Georgetown), and Thomas Cone, Jr. (Boston) at the International Congress of Pediatrics in Lisbon, Portugal in 1962. The first scientific meeting was held in Dublin, Ireland during which the Founders and Charter Members were guests in the home of Ireland's President Eamon de Valera. The aims of the Society are to promote the exchange of scientific and cultural information in the broad area of child health and life in Ireland, Northern Ireland, Canada, and the United States. Alternate meetings held in Ireland or North America on an annual basis provide a greater awareness of local color, historic and scenic vistas as well as a genuine feeling of camaraderie and warm fellowship among members and spouses.

Edmund C. Burke, M.D.



Irish and American Paediatric Society Meeting
September 23 – 27, 2015
Nashville Tennessee USA

Parnell Donahue, Jan van Eys and James O'Neill

Scientific Sessions

Thursday, September 24, 2015	8:00am-noon	Music City Mariota Hotel
Friday, September 25, 2015	8:00am-noon	Music City Mariota Hotel
Saturday, September 26, 2015	8:00am-noon	Music City Mariota Hotel

Thursday, September 24, 2015

8:00 Welcome and Opening of Meeting – President Rita Ryan

Session I – Moderator: Dean Wilcox, Emory University, Atlanta

- 8:15 D. Catherine Fuchs, MD Vanderbilt, Nashville
PEDIATRIC DELIRIUM: DEVELOPMENT OF A TEACHING TOOL
- 8:30 Brian Donahue, MD, PhD Vanderbilt, Nashville
ANESTHESIA AND NEURONAL INJURY IN INFANTS
- 8:45 Carol M. Cottrill, MD* Univ. KY, Louisville
HYPOPLASTIC LEFT HEART SYNDROME:
TWO UNUSUAL CASES AND A REVIEW
- 9:00 Karen Nora McCarthy, MB, BCh, BAO Dublin, Ireland
CASE REPORT: MOSIAC TRISOMY 21/TURNER'S SYNDROME
- 9:15 Beverly A. Powell, MD Fairfax, VA
EARLY IDENTIFICATION & INTERVENTION FOR BOYS WITH
KLINEFELTER'S SYNDROME
- 9:30 Martin Weisse, MD Bethesda, MD
EVALUATING FOR POSSIBLE LYME DISEASE: TALES FROM THE
LYME DISEASE FILES
- 9:45 Virginia Alice Miller, DO. UK Lexington, KY
SOLID PSEUDO PAPILLARY TUMOR: A CASE STUDY OF AN UNUSUAL
PANCREATIC CARCINOMA OF CHILDHOOD
- 10:00 BREAK POSTER VIEWING

10:30

Thomas Cone Founders Lecture

RAFE DONAHUE, PhD

Senior Director, Statistics BioMimetic Therapeutics, Inc.
Adjunct Associate Professor, Department of Biostatistics
Vanderbilt University

DATA STORIES AND PICTURES: DISCOVERING LESSONS AND PRINCIPLES FOR STATISTICS (AND FOR LIFE, TOO!)

Session II – Moderator: Gene Dempsey, University College, Cork Ireland

11:15

Colin ÓhAiseadha, MB, BCh, BAO Dublin, Ireland
A GEOSPATIAL INVESTIGATION OF VEROTOXIGENIC E. COLI (VTEC) O157 INFECTION IN CHILDREN AGED UNDER 5 YEARS IN THE REPUBLIC OF IRELAND, 2008–2013

11:30

Beatrice Stefanescu MD Vanderbilt Univ, Nashville
VERY LOW BIRTH WEIGHT INFANT CARE: ADHERENCE TO A NEW NUTRITION PROTOCOL IMPROVES GROWTH OUTCOMES AND REDUCES INFECTIOUS RISK

11:45

Raga Mallika Pinnamaneni, MB, BCh, BAO Rotunda Hosp. Dublin, Ireland
HEAD GROWTH AND NEURODEVELOPMENTAL OUTCOME AT 1 YEAR FOLLOWING FETAL GROWTH RESTRICTION

Friday, September 25, 2015

Session III – Moderator: James McElligott, Medical University of South Carolina

8:00

Sheena Durnin MB Bch Cork, Ireland
AUDIT OF FLUID BOLUS ADMINISTRATION IN PAEDIATRIC PATIENTS

8:15

Karen McCarthy, MB, BCh, BAO Mullingar, Ireland
TRENDS IN CYSTIC FIBROSIS HOSPITAL ADMISSIONS IN IRELAND: A NATIONAL AUDIT

8:30

Bromwyn Power, MB, BCh, BAO Cork, Ireland
THE FORGOTTEN VITAMIN – VITAMIN A DEFICIENCY IN IRISH CYSTIC FIBROSIS

8:45

Sarah A Higdon, MD Lexington, KY
RARE CYST ARISING IN THE POSTERIOR MEDIASTINUM OF A 16-YEAR OLD FEMALE: MÜLLERIAN CYST OF HATTORI

9:00

Matthew Daniel Eberly, MD Bethesda, MD
ASSOCIATION OF AZITHROMYCIN IN EARLY INFANCY WITH THE DEVELOPMENT OF PYLORIC STENOSIS

9:15

Shannon Murphy Crook, MD Lexington, KY
DISSEMINATED NEONATAL HERPES SIMPLEX VIRAL INFECTIONS WITH HEPATITIS AND CONCURRENT SEPSIS: TWO RECENT FATAL CASE REPORTS

9:30 Jsun Loong Wong, MB. BCh, BAO Galway, Ireland
CLINICAL SPECTRUM OF EARLY ONSET NEONATAL SEPSIS IN THE
NEONATAL UNIT AT GALWAY UNIVERSITY HOSPITAL FROM 2011 - 2014

9:45 Elaine Neary, MD Dublin, Ireland
COAGULATION SYSTEM IN VERY PREMATURE NEONATES

10:00 BREAK POSTER VIEWING

10:30 **William Kidney Founders Lecture**
SUSAN GUTTENTAG, MD
Julia Carell Stadler Professor of Pediatrics
Vanderbilt University School of Medicine
Director, Mildred Stahlman Division of Neonatology
Monroe Carell Jr. Children's Hospital
Nashville, Tennessee
SURFACTANT THERAPY: NOW WHAT?

SESSION IV – Moderator: Carol Cottrell, University of Kentucky

11:15 James T. McElligott, MD MUSC Charleston, SC
PEDIATRIC TELEHEALTH PROGRAMS IN THE UNITED STATES:
GROWING AND DIVERSIFYING

11:30 James A. O'Neill, MD Vanderbilt Univ, Nashville
THE NEW HUMANITARIANISM

11:45 Jan van Eys, MD, PhD Vanderbilt Univ, Nashville
THE HEALTH CARE CONUNDRUM

SESSION V – Moderator: B. Gerald Loftus, GALWAY, IRELAND

Saturday, September 26, 2015

8:00 Business Meeting

8:30 Matthew Daniel Eberly, MD Bethesda, MD
OUTBREAK! TWO UNIQUE CASES OF INFANT BOTULISM

8:45 Natalie K. Wallis, MD, PhD. U. KY, Lexington
NEONATAL ABSTINENCE SYNDROME: A KENTUCKY PERSPECTIVE

9:00 Claire Anne Murphy, M.B.,B.Ch. Donegal, Ireland
AUDIT OF THE ASSESSMENT AND INITIAL MANAGEMENT OF
FEVERISH ILLNESS IN CHILDREN UNDER FIVE YEARS BY THE
PAEDIATRIC SPECIALIST

9:15 Mohammad Omar Aziz, MB,MCh Cork Ireland
“SILENCE MEANS SNOOZE, PAUSE MEANS STOP”: NICU
HEALTHCARE PROFESSIONALS' KNOWLEDGE OF MONITOR
ALARMS

- 9:30 Rita M. Ryan, MD MUSC Charleston, SC
RESPIRATORY MEDICATION USE IN INFANTS BORN <29 WEEKS
GESTATION IN THE FIRST YEAR OF LIFE AFTER NICU DISCHARGE
- 9:45 B.M. Stefanescu, MD Vanderbilt Nashville, TN
A STUDY OF A BUBBLE NASAL CPAP BUNDLE – HOW
IMPLEMENTATION OF POTENTIALLY BETTER PRACTICES IN
RESPIRATORY CARE OF PRETERM INFANTS CAN GREATLY
IMPACT RESPIRATORY OUTCOMES
- 10:30
- 10:00 BREAK POSTER VIEWING

Fred Burke Founders Lecture

PATTI van EYS, PhD

Chief Clinical Officer, Omni Visions, Inc.
Adjunct Clinical Professor of Psychiatry and Nursing
Vanderbilt University
Nashville, Tennessee

BACK TO THE FUTURE:

HOW EARLY CHILDHOOD ADVERSITY AFFECTS HEALTH OUTCOMES

Session VI – Moderator: Rita Ryan MUSC, Charleston, SC

- 11:15 Beverly Powell, MD Fairfax, VA
ENHANCED SOCIAL BEHAVIORS IN CHILDREN WITH AUTISM
SPECTRUM DISORDERS IN THE PRESENCE OF ANIMALS IN THE
CLASSROOM
- 11:30 Rachel Powers, MB BCH BAO Limerick, Ireland
AN EVALUATION OF FATHER'S WEIGHT AND BMI AND IMPACT ON
CHILDHOOD OBESITY: A NATIONAL COHORT
- 11:45 Beverly Powell, MD Fairfax, VA
THE BENEFITS OF COMPANION ANIMALS AND PET THERAPY ON
PHYSICAL AND MENTAL HEALTH AND CHILD DEVELOPMENT

POSTER PRESENTATIONS

Posters are presented at each daily break from 10 – 10:30

ANIMAL ASSISTED INTERVENTION (AAI): THE MANY ROLES OF PET THERAPY THROUGHOUT THE LIFE SPAN, Beverly A. Powell, MD, Associate Prof. of Pediatrics, Fairfax Hospital, VA

IMPACT OF INTRANASAL MEDICATION AVAILABILITY AND USE ON THE FREQUENCY OF PROCEDURAL SEDATION AND ANALGESIA FOR FACIAL LACERATION REPAIR IN A PEDIATRIC EMERGENCY DEPARTMENT, Peter Ryan Cosgrove, MB BCh BAO, MRCPI, Matthew Wilkinson, MD, FAAP, UT Austin Dell Medical School, Dell Children's Medical Center of Central Texas

SOS: STREPTOCOCCUS AS AN OCCULT CAUSE OF SACROILIITIS IN A PAEDIATRIC PATIENT: CASE REPORT AND LITERATURE REVIEW, Bronwyn Dervyla Power, MB, BCh, BAO (hons), Children's University Hospital, Dublin, Ireland

'PASEN GAS': PERI-ANAL GROUP A STREPTOCOCCUS AS A RARE CAUSE OF ERYTHEMA NODOSUM, Rachel Florence Power MB BCh BAO(Hons), MRCPCH, MRCPI, Department of Paediatrics, University Hospital Limerick, Limerick, Ireland

REPRODUCTIVE AND SEXUAL HEALTH ISSUES IN CYSTIC FIBROSIS: AN IRISH MALE PERSPECTIVE, Rachel Florence Power, MB BCH BAO(Hons), MRCPCH, MRCPI, 1Power Rachel F., 1O'Connell Oisín J., 1Shortt Cathy, 1Fleming Clare, 1Plant Barry J., Cork Cystic Fibrosis Center, Cork University Hospital, University College Cork

ORAL PRESENTATIONS

Pediatric Delirium: Development of a Teaching Tool

D. Catherine Fuchs, MD, Professor of Psychiatry, Child and Adolescent Psychiatry
Vanderbilt University, Nashville, Tennessee, USA

Background: Delirium is a known complication of critical illness and an independent predictor of prolonged mechanical ventilation and hospitalization, long term cognitive impairment, and a higher 6 month mortality rate in adults (1,2). Delirium has been reported in up to 30% of critically ill children; the true prevalence and associated outcomes of delirium in critically ill children remains unknown (3,4).

Pediatric delirium is a complex syndrome with core features of disturbances in attention and cognition. The epidemiology of delirium in children has been understudied due to the limited availability of validated tools. Currently available tools include the Pediatric Anesthesia Emergence Delirium Scale (PAED), the Cornell Assessment of Pediatric Delirium (CAP-D), and the Pediatric Confusion Assessment Method for the Intensive Care Unit (pCAM-ICU) for children age 5 years and older. Our research team recently developed and prospectively validated an IRB approved study for the development of the PreSchool Assessment Method for the Intensive Care Unit (psCAM-ICU) (manuscript under review). The reference raters for this study developed a form to standardize the assessments; this form has been adapted as a teaching tool for both Child and Adolescent Psychiatry (CAP) consultation liaison services and Pediatric Intensive Care (PICU) services.

Methods: Our pediatric delirium research team identified the importance of a consistent language to discuss delirium and the critical need for a method to screen for pediatric delirium. We developed a tool anchored in the core features of delirium to include both observation and clinical interaction with the child in order to effectively distinguish cognitive changes from pain or anxiety. The CAP team served as the reference raters using DSM V parameters (5). The CAP team developed a form to standardize the reference rater assessment. We designed the form to factor in the unique challenges of recognizing the core features of delirium in the younger age group with differences in their language development or capacity to speak. We focused on key tasks that demonstrate a level of attention, arousal, and cognition that can be universally tested in this age population. We identified the minimum skills a preschool aged child should demonstrate in order to be regarded as not delirious even if developmentally delayed.

Results: Use of specific tasks to demonstrate attention and cognition at each evaluation minimizes subjectivity among trainees when determining presence or absence of delirium. By making explicit the need to consider perception, language, sleep, and different aspects of attention, the trainee is forced to consider the developmental aspects involved. The minimum tasks necessary to assess attention and basic cognition can be done across developmental levels. As a result this tool can assess the presence or absence of delirium in children with developmental delays and in a medically fragile population with sufficient level of alertness.

Conclusion: Use of a structured teaching form facilitates consistent assessment of mental status in a critically ill child from infancy through adolescence. The form provides a consistent approach to the teaching of both CAP and PICU trainees in the assessment of pediatric delirium.

References available upon request

ANESTHESIA AND NEURONAL INJURY IN INFANTS

Brian S. Donahue, MD, PhD, Professor of Pediatrics and Anesthesia, Vanderbilt University, Nashville, Tennessee

The infant brain relies on synaptic activity for growth and development. Studies in animal models have indicated that suppression of this activity with anesthetic agents in the critical neonatal period can produce neuronal apoptosis and neurobehavioral changes. Extension of these findings to humans have relied on retrospective comparisons, with their inherent confounders. Current study in this field focuses on defining the vulnerable period for anesthetic exposure, more clearly describing the neurodevelopmental phenotype, characterizing molecular pathways responsible for neuronal injury, and identifying agents and practices which may provide neuronal protection. The current state of research remains unclear on the question of whether anesthetic exposure inflicts clinically significant damage in infants, although current investigations may soon provide some clarity.

HYPOPLASTIC LEFT HEART SYNDROME: Two unusual cases and a review.

Carol M. Cottrill, MD*, William N. O'Connor, MD*, Lisa Coleman, RN, RDCS* and Edward Bove, MD**

*Departments of Pediatrics and Pathology, University of Kentucky, Lexington, KY and

**Department of Cardiac Surgery, University of Michigan, Ann Arbor, MI

Hypoplastic Left Heart Syndrome (HLHS) is one of the biggest surgical challenges among the types of complex congenital heart diseases presenting in the neonatal period. Until 1980, it was considered an inoperable defect, and compassionate care was offered to these infants. With the advent of the Norwood operation (Stage 1) and its acceptance as a palliative measure for these babies, the outlook changed, results improved and today in several institutions the mortality rate for Stage 1 in a term, non-syndromic baby without other congenital anomalies is 90%. Of course, this is only the first of three staged procedures to fully palliate such children. The Norwood operation is usually done in the first week of life; the Glenn or hemi-Fontan operation (Stage 2) at about 6 months of age, and the Fontan (Stage 3) is completed sometime after the child's second birthday. Theories about the etiology of HLHS include disruption of flow, either forward or backward, failure of the induction of normal left ventricular (LV) growth, and programmed disruption of growth. Regardless of the etiology, HLHS is usually a fairly late-gestational event, the LV being normal until 12-15 weeks gestation.

We present two male infants with HLHS who also had Truncus Arteriosus(TA) and partial anomalous pulmonary venous return (PAPVR). These two cases are unusual, and the timing of the cardiac disruption for TA and PAPVR (5-7 weeks) is completely out of scope for the usual cases of HLHS. In addition, these babies presented unique surgical challenges, since Stage 1 and Stage 2 operations had to be drastically modified to fit the existing anatomy. Both children did well with all 3 stages, and now enjoy normal growth and development at ages 2 ½ and 3 years.

CASE REPORT: MOSIAC TRISOMY 21/TURNER'S SYNDROME

Presenters: Karen Nora McCarthy MB Bch BAO, Angharad Sian Griffiths MB Bch BAO

Authors: KN McCarthy, AS Griffiths, N McCallion. Rotunda Hospital, Dublin 1, Ireland

Background

We present the case of a female infant with mosaic trisomy 21/Turners syndrome (45 X0, 47 XX +21). Phenotypic features at birth were consistent with Trisomy 21. Her postnatal course was complicated by transient abnormal myelopoiesis which has resolved on follow up. This case presents an interesting challenge in the management and follow up of mosaic Trisomy 21/Turners including the dual endocrine issues and developmental follow up.

Case presentation

A female infant was born at 37+3 weeks gestation following an spontaneous vaginal delivery. Examination was significant for upslanting palpebral fissures, epicanthic folds and low set ears. A third fontanelle was also present. Examination of the peripheries revealed a single palmar crease on the right hand and a bilateral sandal gap. There was hypotonia on ventral and vertical suspension with mild head lag.

Demonstrated was an abnormal female karyotype of 45X, 47XX + 21 with presence of 2 cell lines. Twenty one metaphases had 45x while 19 had 47xx+21. A buccal smear revealed two abnormal cell lines - 65% trisomy 21 and 34% XO disomy 21. Also significant in post-natal course was thrombocytopaenia and haematology opinion was sought. Investigations displayed a population of immature myeloid cells (CD7 positive), thrombocytopenia on blood film and atypical mononuclear cells with circulating myeloblasts. A diagnosis of transient abnormal myelopoiesis was made. This has resolved with follow up.

We were pleased to see her growing and thriving appropriately at six week follow up. Developmentally, she had normal tone but had yet to display a social smile.

Discussion

The incidence of Trisomy 21 in Ireland is the highest in Europe at 1 in 546 live births whereas the incidence of Turner's syndrome is 1 in 2500 live female births. Double aneuploidy of both autosomal and sex chromosomes, is, however a rare occurrence. It presents an interesting challenge in interpreting clinical features and in follow up, investigation and management of associated co-morbidities. Patients with mosaic Trisomy 21 typically demonstrate a milder phenotype than non-mosaic individuals. This individual socially likely to demonstrate both features of Turner's syndrome and Trisomy 21 although previous reports on Turner/Trisomy 21 polysyndrome mosaicism suggest that phenotypic features of Down Syndrome predominates.

References:

MM Villaverde et al. Turner-mongolism polysyndrome. A review of the first eight known cases. JAMA. 1975;234(8):844-7.

Down Syndrome Medical Interest Group. Medical management of children & adolescents with Down syndrome in Ireland. University of Dublin, Trinity College, The National Childrens Hospital, AMNCH, Tallaght: Approved Guidelines; 2001

EARLY IDENTIFICATION & INTERVENTION FOR BOYS WITH KLINEFELTER'S SYNDROME

BEVERLY A. POWELL, MD, Assoc. Prof. of Pediatrics, Fairfax Hospital, VA & GWU

INTRODUCTION: Klinefelter's Syndrome is the most common sex chromosomal disorder and affects approximately one in 660 boys. It is characterized by varying of cognitive, social, learning and behavioral disorders. The phenotype varies from "near-normal" appearance & development to significant abnormalities, including delays in speech/language and tall stature for age and small testes -- often not identified until after puberty.

Decreased awareness of this syndrome among health professionals and lack of knowledge of early symptoms in affected males with 47 XXY results in lack of diagnosis prior to puberty, estimated to be 10%. As many of 75 % of non-mosaic males remain undetected until puberty or later, when their condition is identified in the course of a fertility work-up or abnormal endocrine findings.

Early identification and treatment of KS is recommended in order to offer treatment of the medical endocrine and developmental aspects of this syndrome. For example, speech delays can lead to behavioral and social maladjustment in the preschool period which can be avoided through early intervention programs and therapy. Special education services are effective for non-verbal learning disabilities and AD/HD of the Inattentive type. Males with tetrasomies (XXYY) have more severe neurodevelopmental disabilities and require more intensive interventions and support as adults.

In addition to treating infertility, regular endocrine screening and interventions can be provided to prevent osteoporosis, metabolic syndrome, Type II DM, hyperlipidemia & other medical conditions associated with hypogonadism. Testosterone supplementation can reduce the impact and severity of learning and psychosocial problems & enhance an individual's sense of well-being. Current assistive reproduction techniques make it possible for couples affected by KS to bear healthy children but there is an increased risk of XXY and other trisomies.

Recent studies have shown that KS is a multi-system disorder affecting the immune system, causing an increased incidence of lupus, RA and cancers (lung, non-Hodgkin's lymphoma & breast). The mechanism for this is unknown and further research is underway through the NCI.

Conclusions: Children with 47 XXY, Klinefelter Syndrome, have a broad range of phenotypes which can result in a variety of clinical findings. These include developmental delays, learning and behavioral problems, hypogonadism, tall stature and endocrine/metabolic disorders. Through early Identification and intervention, these boys can receive special education and therapy in addition to medical treatment to reduce the secondary clinical (e.g. metabolic) disorders.

Recommendations: When a preschool old boy presents with speech/language delay & has an early spurt in height it is important to check a chromosome analysis to determine whether he has 47 XXY or Fragile X syndrome. This enables the family to pursue genetic counseling, start developmental intervention and obtain medical follow-up for metabolic disorders in the future.

EVALUATING FOR POSSIBLE LYME DISEASE: TALES FROM THE LYME DISEASE FILES

Martin E Weisse, M.D.
Walter Reed National Military Medical Center
Bethesda, Maryland, USA

Background

Lyme Disease is a tick-borne disease with protean manifestations that is endemic in some areas of the United States and is increasingly reported in Ireland as well. While many cases of Lyme Disease are treated by General Practitioners, Pediatric Infectious Diseases specialists are consulted for the several complications and many questions related to the aftermath of a Lyme Disease diagnosis. Patients that present with early disease frequently have negative serology, and with treatment, anti-Borrelia antibodies may remain negative, causing confusion in patients and physicians alike. A review of the patients referred to one Infectious Disease Clinic in an endemic area is presented.

Methods

The handwritten log of all patient encounters in our Pediatric Infectious Diseases clinic since January 2013 was reviewed, looking for the keywords “Lyme” or “Borreliosis”. In addition, the electronic clinic log of “Reason for visit” was reviewed and words or phrases such as Bell’s palsy, arthralgia, arthritis, and E. migrans prompted review of those charts as well.

Clinical presentation, history, physical examination and laboratory values were reviewed. Assessment and disposition including treatment were noted.

Results

Over the previous 32 months, there have been 52 unique patients seen in our clinic for questions of Lyme Disease. Of these, 36 were deemed to be Lyme Disease, and an alternate diagnosis or no diagnosis was given for 16 patients.

<i>Lyme Diagnosis?</i>	<i>N</i>	<i>Diagnosis</i>		
Yes	36	Early localized	Early disseminated	Late disseminated
		10	13	13
		<i>Complaint</i>		
No	16	Fatigue	Tick bite	Other
		5	6	5

Conclusions

Lyme Disease is not an infrequent reason for referral to pediatric infectious disease specialists. Counseling of those patients with concerns for recurrent or chronic Lyme Disease is an essential skill for Infectious Disease specialists. Familiarity with the various manifestations of early localized, early disseminated and late disseminated Lyme disease is also essential and will be discussed.

SOLID PSEUDOPAPILLARY TUMOR: A CASE STUDY OF AN UNUSUAL PANCREATIC CARCINOMA OF CHILDHOOD

Virginia Alice Miller, DO.

William O'Connor MD. and Ana Ruzic MD.

Lexington Kentucky, University of Kentucky Pathology, 800 Rose St., Lexington KY, 40506.

Background: Solid pseudopapillary tumor (SPT) of the pancreas is an uncommon neoplasm of low-malignant potential occurring mostly in young adult females, and very rarely in childhood.

Case Study: a previously healthy 12 yr old female presented with bilateral abdominal pain for 2-3 days. CT of the abdomen revealed notable constipation as well as a circumscribed, solid appearing oval mass in the pancreatic body, measuring 3.6cm in greatest dimension. Further work up showed negative CEA, CA19-9 and AFP levels, but considering the size and likelihood for malignant potential/transformation, pre-operative biopsy was not attempted and a subtotal distal pancreatectomy was performed. On surgical resection, the mass involved the surgical neck and splenic vessels and closely approached the splenic vein. Gross evaluation revealed a friable, solid red-tan mass with focal necrosis and little cystic architecture. Histologically, the lesion was comprised of uniform rounded cells with a solid and pseudopapillary growth pattern with small groups of foam cells. Tumor involved peripancreatic fat in the posterior soft tissue margin with perineural and venous invasion. Additionally, one of thirteen peri-pancreatic nodes was found to be positive, though likely via direct extension, and the tumor was staged as a pT4. On follow-up the patient is progressing well with unremarkable imaging and no digestive complications.

Discussion: Diagnosis in this case is difficult for a number of reasons. SPT's comprise around 6% of exocrine pancreatic tumors overall, and account for 8 – 16% of childhood pancreatic cancer (which overall is quite rare). Though most SPT's occur between ages of 20 and 30 (90 % + in females), reports range between age 2 and 85 yrs. On radiology, SPT's present as solid and cystic masses (non-specific features which encompass a broad differential of teratomas, cystadenocarcinomas and hemangiomas). SPT in our patient appeared solid, adding differentials such as primary pancreaticoblastoma, islet cell neoplasm (the latter may present as an endocrinopathy), or, secondary pancreatic involvement by lymphoma or neuroblastoma.

Fortunately, SPT's have a good prognosis despite the tendency for local extension and/or invasion, which is seen in roughly 20% of cases. Among these, liver, portal vein and splenic involvement are the most common. Recurrence is rare (6%) and generally involves the liver or lymph nodes. Based on the data available, overall survival with resection is promising with five-year estimates at 95% and long-term survival close to 93%. Currently, the cell of origin, acinar versus endocrine, remains unclear.

Diagnosis in this case was confounded by young patient age, unusual radiologic presentation, and lack of symptoms. Although her abdominal pain may relate to constipation, pain and mass are the most frequently reported presenting symptoms for SPT, while 15% of reported cases may be symptomatic.

<https://www.dropbox.com/s/n37kxcus047a86t/Data%20Stories%20and%20Pictures%20abstract.docx?dl=0>

DATA STORIES AND PICTURES: DISCOVERING LESSONS AND PRINCIPLES FOR STATISTICS (AND FOR LIFE, TOO!)

Rafe Donahue, PhD
Senior Director, Statistics BioMimetic Therapeutics, Inc.
Adjunct Associate Professor
Department of Biostatistics
Vanderbilt University

Our world is full of data and of analyses of these data. Why are we doing to our data what we are doing to our data? Are there better ways to make inference from the deluge of data swamping us daily? Are there lessons and principles that can be applied to increase our understanding of our data and our world?

The answer is “Yes”; there are better ways. But those better ways require us to slow down, ask serious questions, take responsibility for our inferences, and, most of all, THINK about what we are doing.

We will wander through a series of data stories and pictures derived from them, along the way learning lessons and picking up principles. I promise no formulas; this talk is family-friendly. But you will be challenged to think.

A GEOSPATIAL INVESTIGATION OF VEROTOXIGENIC E. COLI (VTEC) O157 INFECTION IN CHILDREN AGED UNDER 5 YEARS IN THE REPUBLIC OF IRELAND, 2008–2013

C ÓhAiseadha¹, P Hynds², J O'Dwyer³

Department of Public Health, Health Service Executive, Dublin, Ireland

School of Civil and Structural Engineering, Dublin Institute of Technology, Dublin, Ireland

Department of Life Sciences, University of Limerick, Limerick, Ireland

Background: Verotoxigenic *Escherichia coli* (VTEC) are so called due to their ability to produce verotoxins similar to AB5-type Shiga toxins. The most frequently encountered serotype in the Republic of Ireland is VTEC O157, whose clinical presentation ranges from mild diarrhoea to hemolytic uraemic syndrome (HUS). Over the past decade, the Republic of Ireland has repeatedly reported the highest incidence of symptomatic VTEC infection in the European Union, with national incidence rates increasing ten-fold during this period. HUS is associated with approximately 4–7% of confirmed cases, particularly among younger sub-populations. The primary reservoirs of VTEC infection in Ireland are considered to be agricultural animals and untreated groundwater consumption. Accordingly, the current study examined the environmental epidemiology of pediatric VTEC infection in the Republic of Ireland through statistical analyses of geo-coded primary infections with the spatial distribution of agricultural and infrastructural risk factors.

Methods: All laboratory-confirmed, domestically acquired sporadic or outbreak index case of VTEC infection, aged <5 years, reported January 1st 2008–December 31st 2013, were georeferenced by census enumeration area, or “Small Area” (50–200 dwellings). A geographic information system (GIS) was used to assign potential explanatory variables to each area, including human population density (km⁻²), livestock (cattle and sheep) density (km⁻²), private well reliance (wells per population), septic tank density (km⁻²) and socioeconomic status. Bivariate and multivariate analyses were conducted to identify geo-statistical associations between infection occurrence and potential risk factors.

Results: Overall, 521 cases of VTEC O157 infection were geo-referenced, with 407 cases (78.1%) involving children <5 years old (annual mean incidence rate 19.7/100,000). Children <5 years currently comprise 7.8% of the national Irish population. Pediatric VTEC O157 infection in the Republic of Ireland is associated with areas characterized by low human population density ($p < 0.001$), high cattle density ($p < 0.001$), high private groundwater usage ($p < 0.001$), and marginally lower socioeconomic status ($p < 0.001$). Lower septic tank density was also observed in association with pediatric infection. Spatially derived population density and septic tank density were significantly correlated ($r_{sp} = 0.736$). A multivariate logit model shows that variables associated with infection were cattle density (OR = 1.001, 95% CI 1.001–1.003) and private well usage (OR = 4.026, 95% CI 1.167–13.89); the interaction term well usage* cattle density was found to be highly predictive (OR = 1.013, 95% CI 1.006–1.020).

Conclusions: Results indicate that pediatric VTEC O157 infection in the Republic of Ireland is a characteristically rural disease, with cattle and unregulated groundwater wells constituting the primary pathogen source and pathway, respectively. The authors consider that population density (inverse) may act as a surrogate variable for the distribution of cattle and groundwater reliance, while septic tank density may represent a proxy for periurban infrastructural development. Overall, it is concluded that a health inequality regarding pediatric VTEC O157 infection, based upon residential classification or “place”, currently exists in the Republic of Ireland.

HEAD GROWTH AND NEURODEVELOPMENTAL OUTCOME AT 1 YEAR FOLLOWING FETAL GROWTH RESTRICTION

Michael Boyle 1, 2, 4, RagaMallika Pinnamaneni^{2, 3}, Fergal Malone 2, 4, Julia Unterscheider 2, 4, Naomi McCallion^{2,3,4}, Adrienne Foran^{2, 3, 4}

1. Rosie Hospital, Cambridge University Hospitals NHS Foundation Trust, UK
2. Rotunda Hospital, Dublin, Ireland
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INTRODUCTION:

Intrauterine growth restriction (IUGR) can be defined as babies whose birth weight lie below the 10th percentile for that gestational age and affects up to 10% of all pregnancies. Approximately 5 to 10% of pregnancies complicated by IUGR will result in either stillbirth or neonatal death¹⁻². The effects of IUGR continue beyond the neonatal period and may have a profound impact on child growth and development. In one study of 1116 fetuses (PORTO study), abnormal Doppler was significantly associated with adverse obstetric and neonatal outcomes, regardless of estimated fetal weight³. We looked at a subgroup of these infants and monitored their head growth and neurodevelopment.

MATERIALS AND METHODS:

Participants were recruited from the PORTO (Prospective Observational Trial to Optimize paediatric health in IUGR infants). From this cohort, 74 infants were recruited into the StOOPS (Short-term surrogate Outcome Of infants in the PORTO) study.

Informed consent was obtained; background data and birth anthropometry measurements were recorded. All 74 infants had a detailed 3T MRI brain at term corrected gestational age (37-44 weeks). At 1 year corrected age participants parents were asked to complete a 12 month Ages and Stages Questionnaire (ASQ-3), a questionnaire assessing communication, fine motor, gross motor, social-emotional and problem solving abilities. An occipito-frontal circumference (OFC) was measured by the participants General Practitioner at 1 year corrected age.

RESULTS: Of the 74 infants, 63 had an EFW <10th centile (IUGR) and 11 infants were term infants (controls) whose weight was appropriate for their gestational age (AGA). Of the 63 SGA infants, 34 had abnormal antenatal ultrasound Doppler's and 29 had normal Doppler's. The results of the 12-month ASQ-3 showed that IUGR infants scored lower in communication when compared to term controls (see table). The difference between groups OFC was statistically significant at delivery (p-value < 0.0001) but not at one year of age (p-value=0.8074, see graph)

CONCLUSIONS: The difference between groups OFC was statistically significant at delivery; the IUGR groups had lower OFC's compared to the AGA term controls; however by 1 year this difference had normalised, displaying catch up head growth. The IUGR infants scored lower in the communication component of the ASQ-3 compared to the controls; this difference was not significant when comparing between the groups with normal and abnormal Dopplers.

Charts, graphs and reference are available upon request.

VERY LOW BIRTH WEIGHT INFANT CARE: ADHERENCE TO A NEW NUTRITION PROTOCOL IMPROVES GROWTH OUTCOMES AND REDUCES INFECTIOUS RISK

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Background: Very low birth weight (VLBW) infants are at risk for postnatal growth restriction due to inadequate nutrient delivery and concomitant illness. Integrated clinical pathways or protocols can improve growth outcomes by decreasing practice variability.

Aims: We hypothesized that a standardized nutrition bundle would result in a significant decrease in very low birth weight (VLBW) infants with postnatal growth restriction at 36 weeks postmenstrual age (PMA) and discharge. The primary aim was to determine if the intervention improved weight, length, and head circumference delta-z scores at 36 weeks PMA and decreased the percentage with severe (<3rd) or moderate (<10th) growth restriction at discharge. Secondary aims included time to first and full enteral feedings, central-line days, and rates of necrotizing enterocolitis (NEC) and sepsis/sepsis-like episodes.

Methods: A comprehensive nutrition bundle comprising standardized recommendations for initiating, advancing, and fortifying enteral feedings, and timely discontinuation of central lines was implemented in July 2012. Eligible were infants with birthweight <1500 grams and <34 weeks gestation who were born over a 1-year period pre- and post-intervention, respectively.

Results: Of the total 299 infants who were included, 156 received the proposed intervention (Nutrition bundle group), and 143 received non-standardized nutrition practices (Conventional group). Eight infants in each group died and were excluded from the growth analyses. Median delta-z scores for length (-1.2 versus -1.71; $p=0.01$) and head circumference (-0.73 versus -1.21; $p=0.03$) but not weight at 36 weeks PMA (-1.41 versus -1.58; $p=0.74$) were significantly higher in the Nutrition bundle as compared to Conventional group. Fewer infants in the intervention group had severe growth restriction (20% in the Conventional versus 11% in the Intervention Group, $p=0.036$). Compared with the Conventional group, fewer infants had moderate head growth restriction (17% versus 36%, $p<0.001$), and severe head growth restriction at discharge (18% versus 6%; $p<0.01$). The median time to first feed during the intervention decreased to 57 from 96 hours in the Conventional group (log rank test statistics $p<0.001$). Median time to full enteral feeding was also significantly reduced with intervention from 22 to 15 days (log rank test statistics $p<0.001$). Central line-days (13 versus 18 days; $p=0.004$), the incidence of NEC (2% versus 6%; $p=0.05$), sepsis/sepsis-like episodes (49% versus 64%; $p=0.01$) and chronic lung disease (25% versus 36%; $p=0.045$) decreased with the intervention.

Conclusions: Our study showed that a nutrition bundle can lead to an improvement in growth parameters at 36 weeks PMA and reduction in postnatal growth restriction at discharge in VLBW infants. In addition, this nutrition intervention resulted in a reduction of NEC, sepsis, central line-days, and CLD. This was achieved by addressing multiple aspects already known to individually affect change: breast milk, early feedings, and careful fortification with protein. Future studies should focus on higher enteral protein fortification of breast milk and the neurocognitive development of these infants in infancy and childhood. Our group is prospectively collecting results of developmental testing in toddlerhood in our cohort of patients and will present these in a future publication.

AUDIT OF FLUID BOLUS ADMINISTRATION IN PAEDIATRIC PATIENTS

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Introduction: The British Advanced Life Support Group and the National Institute for Health and Clinical Excellence (NICE) recommends an immediate 20 ml/kg bolus of 0.9% sodium chloride (NaCl) for the resuscitation of acutely unwell paediatric patients in hypovolaemic shock. Incorrectly prescribed or administered fluids are potentially very dangerous. This audit aimed to assess the documentation of dehydration status and the appropriate use of fluid boluses in line with these guidelines in the Emergency Department (ED) and patients admitted directly to the paediatric ward.

Methods: A retrospective chart review of 50 patients was conducted in Cork University Hospital. Inclusion criteria were patients under 16 years of age who received a 0.9% NaCl bolus on admission from October 2014 until March 2015. Cardiac, surgical, diabetic and trauma patients were excluded. A fluid bolus was a defined volume of 0.9% NaCl administered over a defined time period (maximum 1 hour). The assessment of dehydration was based on the NICE clinical guideline on diarrhoea and vomiting in children and included fifteen attributes of signs and symptoms of clinical dehydration and shock.

Results: Fifty patients were recruited (age range 0.03-15.8 years, 54% male). The diagnosis was gastroenteritis in 64%, sepsis in 14%, respiratory tract infection in 14% and mesenteric adenitis in 8%. The fluid bolus was administered in the ED in 58% of cases and on the paediatric ward in 42%. Fluid boluses prescribed were 10 ml/kg in 50% of patients and 44% were prescribed 20 ml/kg. A blood gas performed in 20% of cases.

The mean time interval from finishing the bolus to starting maintenance IV fluids was 73 minutes (range 0-555 minutes). Thirty two percent waited over one hour to commence the prescribed maintenance IV fluid. A deficit volume of fluid was prescribed in 32%. The median number of clinical dehydration and shock attributes documented in the assessment of dehydration was 9.8 (range 4-14). Attributes of fever, tachycardia and tachypnoea were documented in all cases but there was a significant paucity of documentation (50% or more of cases) for pulse volume, urine output, blood pressure, mucous membranes, periphery temperature, skin turgor and the presence of sunken eyes.

The gastroenteritis proforma was used in 38% of the gastroenteritis cases and the median number of attributes documented was 11.5. In the other cases of gastroenteritis where the proforma was not used, the median number of attributes was 8.4. The use of gastroenteritis proforma enhanced the documentation (p-value 0.002 by Mann-Whitney U test).

Conclusion: The audit has highlighted that our practice is not in keeping with the guidelines. A delay has been highlighted between bolus administration and commencing maintenance IV fluids. Deficits in the documentation of the clinical assessment of dehydration and shock were noted. The use of gastroenteritis proforma should be encouraged to improve documentation of the assessment of dehydration if appropriate. Education of trainees in the documentation of dehydration assessment and the appropriate prescription and use of fluid boluses was implemented. This data will be re-audited in 6 months.

TRENDS IN CYSTIC FIBROSIS HOSPITAL ADMISSIONS IN IRELAND: A NATIONAL AUDIT

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Background: Cystic Fibrosis is the most commonly occurring life threatening inherited disease in Ireland with an incidence of approximately 1 in 1461 and a carrier rate of 1 in 19 among the Irish population. We sought to evaluate the trends in hospital admissions for patients with any listed diagnosis of cystic fibrosis using the Hospital In-Patient Enquiry (HIPE) system data.

Methods: The HIPE system was evaluated from 57 Irish hospitals from 2009-2013 for patients admitted with any listed diagnosis of cystic fibrosis. Data recorded included diagnosis, average age, gender, principle procedure, and average length of stay. Results are shown as totals and mean (Standard deviation). Statistical analysis was carried out using Prism 6 software. Trends were examined by logistic regression analysis. T test was used for length of stay comparison. Kruskal wallis test was used for comparison of 5 year age groups. P value <0.05 was considered statistically significant.

Results: The number of patients aged 0-14 years admitted with a principal diagnosis of CF significantly increased annually from 659 to 809 in 2013 ($r^2=0.8$, $p=0.04$). CF was most frequent admitting diagnosis in the 0-14 year age group with $n=3763$, followed by bacterial pneumonia with a total of 193 and an annual average of 39 (sd 11) admissions when patients had an average age of 10 yrs (SD 4). The average age of all patients admitted for CF significantly increased from 2009-13 rising from 24yrs (7) to 29yrs (9), ($r^2=0.9$, $p=0.02$). CF was most frequent admitting diagnosis with 7419 in those older than 15. Admission for management of an implanted device ($n=451$) was the predominant reason in those >15 years with an average annual admission rate of 90 (SD 6) when patients had an average age of 28 yrs (SD 6). Average length of stay was significantly greater for those 15 and over than those <14 (mean 8 v 15, $p<0.01$ – IQR unavailable). Death data was available for 4 years from 2009-2012 inclusive during which 46 in-hospital deaths occurred in CF patients. No in-hospital deaths were recorded in patients <14 and the average annual incidence of death was 12 patients (SD 3) with a mean age of death of 27 yrs (SD 7).

Conclusion: The number of paediatric cystic fibrosis patients admitted to hospital has increased from 2009-2013. This may reflect increased early diagnosis following the introduction of national newborn screening for CF in 2011. The most commonly occurring diagnoses at discharge differ between the paediatric and adult population with the latter being more frequently admitted for management of implanted devices. The average length of stay for paediatric patients is significantly shorter than the adult population, perhaps due to reduced comorbidities in the paediatric population. This study has several limitations however. Each HIPE discharge record represents one episode of care. Patients may be admitted to hospital more than once in any given time period with the same or different diagnoses. In the absence of a unique health identifier, therefore, the data reported to HIPE facilitate analysis of hospital discharge activity, but do not permit analysis of discharges at individual patient level. Provision of means and medians only limited statistical analyses.

THE FORGOTTEN VITAMIN – VITAMIN A DEFICIENCY IN IRISH CYSTIC FIBROSIS

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Background: Vitamin A deficiency (VAD) due to pancreatic insufficiency and hepatobiliary disease has been reported in CF since the earliest descriptions of the disease. A Cochrane Review (2012) suggested that there was insufficient data to draw any conclusions about the benefits of vitamin A supplementation in CF. To date, only one paediatric case report has addressed VAD in an Irish CF population.

Aim: To determine the prevalence and clinical implications of VAD in Irish patients with CF.

Methods: This quantitative study was a combination of a retrospective medical record review from January 2010 to December 2012, and a cross-sectional questionnaire-based study of CF patients in Ireland. VAD was defined, in accordance with the WHO, as serum retinol $<0.7\mu\text{mol/L}$, with severe deficiency defined as serum retinol $<0.35\mu\text{mol/L}$.

Results: 147 patients were screened for study entry resulting in a total of 100 patients meeting the inclusion criteria. In 2012, 8% ($n=8$) were VAD (serum retinol $<0.7\mu\text{mol/L}$) and 17% ($n=17$) were insufficient in vitamin A (serum retinol $0.7\text{--}1.05\mu\text{mol/L}$). In 2011 and 2010, 8% ($n=8$) and 9% ($n=9$) were VAD respectively. There was no association between VAD and zinc deficiency, liver disease, retinol-binding protein deficiency or markers of CF clinical phenotype. 83% ($n=83$) were taking multi-vitamin supplementation, including vitamin A. The questionnaire response rate was 51% ($n=51$); none of these were VAD. 23.5% ($n=12$) reported ocular symptoms; of those, three patients were insufficient in vitamin A. 5.9% ($n=3$) reported night blindness. 17.64% ($n=9$) had mild/moderate conjunctival xerosis.

Discussion and Conclusions: The prevalence of VAD in Irish adult CF patients is low when compared to international studies. No association was identified between VAD and previously reported risk factors in the literature including zinc deficiency, liver disease, retinol-binding protein deficiency and markers of CF clinical phenotype. Our results question the role of measuring zinc and RBP levels routinely in VAD in patients with CF. Based on ocular questionnaire results, there is no clear relationship between ocular symptoms and VAD in CF. These results validate the multi-disciplinary approach to CF care, specifically the role of the Clinical Nutritionist in vitamin supplementation. This has been the first Irish study to investigate the prevalence, associated factors and clinical implications of VAD in CF.

References available upon request.

RARE CYST ARISING IN THE POSTERIOR MEDIASTINUM OF A 16-YEAR OLD FEMALE: MÜLLERIAN CYST OF HATTORI

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The differential diagnosis of cysts in the mediastinum include: foregut (bronchogenic), gastroenteric (contain alimentary tract mucosa), neurenteric (contain enteric and neural tissue), mesothelial, and thoracic duct cysts (contain lymphatic channels)². Teratomas and thymic cysts should also be considered⁵. In 2005, the first mediastinal cyst with müllerian differentiation was described in an 18-year-old female³. Extra-genitourinary cysts of müllerian origin are rare with only 18 histologically and immunohistochemically proven reported cases. However, two retrospective studies show that the müllerian cysts may not be so rare and/or are misdiagnosed^{4,7}. They are commonly mistaken for enterogenous and bronchogenic cysts, which are also lined by ciliated, columnar epithelium².

We report a posterior mediastinal cyst with müllerian differentiation in the youngest patient to date. A 16-year-old female presented to the University of Kentucky pediatric surgery department with a 3 cm T5/6 paraspinal mass, visualized by chest x-ray and computer tomography. Prior, she had a one-month history of cough and back pain that radiated to her right chest, for which she had completed several courses of antibiotics without resolution of symptoms. Resection of the mass was completed by video-assisted right-sided thoracoscopic surgery.

Grossly, the mass was pink and cystic with a wrinkled, thin wall and measured 2.0 x 2.0 x 0.6 cm. Microscopic sections revealed a cyst lined by a ciliated epithelium which stained positive for estrogen (ER) and progesterone receptor (PR) immunohistochemical stains. The lining epithelium stained negative for calretinin, caldesmon, CD10, and inhibin immunohistochemical stains. Fascicles of smooth muscle cells within the cystic wall are highlighted by caldesmon immunohistochemical stain. No cartilage or glands were found in the wall.

ER and PR receptors are known to be the best markers for this type of cyst². Additionally, immunohistochemical stains PAX8 and WT1, both markers of müllerian differentiation, can be used⁶. The etiology of müllerian cysts arising in the mediastinum is unknown. It has been proposed that these cysts are either derived from the primary müllerian apparatus or they may represent misplaced mesothelium⁶.

Müllerian cysts occur almost exclusively in women 40-60 years of age (with the exception of the first reported and present cases). They typically occur in paravertebral locations between the 3rd and 8th thoracic vertebra and present with cough and chest pain⁶. Treatment is surgical removal. Their behavior is benign and to date, there are no reports of recurrence¹. We report the youngest case of a mediastinal müllerian cyst. This case serves as a reminder to include this entity in the differential diagnosis of pediatric mediastinal cysts.

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ASSOCIATION OF AZITHROMYCIN IN EARLY INFANCY WITH THE DEVELOPMENT OF PYLORIC STENOSIS

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Objective: Administration of oral erythromycin in infants has been shown to increase the risk of developing infantile hypertrophic pyloric stenosis (IHPS). The risk associated with azithromycin remains unknown. We evaluated the association between exposure to oral azithromycin and erythromycin and subsequent development of IHPS.

Methods: A retrospective cohort study of children born between 2001 and 2012 was performed utilizing the U.S. military health system database. Infants who were <90 days of life were evaluated and those prescribed either oral erythromycin or azithromycin as outpatients in the first 90 days of life were determined. IHPS was identified using specific diagnostic and procedural codes.

Results: A total of 2466 of 1,074,236 children in the study period developed IHPS. Azithromycin exposure in the first 14 days of life demonstrated an increased risk of IHPS (adjusted odds ratio [aOR], 8.26; 95% confidence interval [CI], 2.62-26.0); exposure between 15 to 42 days had an aOR of 2.98 (95% CI, 1.24-7.20). An association between erythromycin and IHPS was also confirmed. Exposure to erythromycin in the first 14 days of life had an aOR of 13.3 (95% CI, 6.80-25.9), and 15 to 42 days of life, aOR 4.10 (95% CI, 1.69-9.91). There was no association with either macrolide between 43-90 days of life.

Conclusions: Ingestion of oral azithromycin and erythromycin places young infants at increased risk of developing IHPS. This association is strongest if the exposure occurred in the first 2 weeks of life, but persists although to a lesser degree in children between 2 and 6 weeks of age.

DISSEMINATED NEONATAL HERPES SIMPLEX VIRAL INFECTIONS WITH HEPATITIS AND CONCURRENT SEPSIS: TWO RECENT FATAL CASE REPORTS

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Introduction: Neonatal herpes simplex virus (HSV) infection is a rare (1 in 3,200 births) but potentially preventable cause of morbidity and mortality. Acquired intrapartum during vaginal delivery, mortality is over 85% without early treatment. Sixty to eighty percent of mothers with an HSV-infected infant have no known history of infection or clinically evident genital HSV infection at the time of delivery. We report pathologic findings in two recent autopsy-confirmed neonates with disseminated HSV born to asymptomatic mothers with primary infections.

Case 1: A term female, with pregnancy complicated by chorioamnionitis, had a vacuum-assisted vaginal delivery secondary to shoulder dystocia. The mother remained hospitalized for postpartum fever with newborn until day of life (DOL) 6 when baby girl developed respiratory distress and a fever of 100.4°F. Following intubation, antibiotics and acyclovir were started. A blood sample revealed HSV by PCR. She developed bilateral pneumonia, pleural effusions, and acute liver failure, with death on DOL 12. Immediate premortem blood cultures grew *E. coli*. At autopsy, the lungs showed gross consolidation with microscopic HSV-related necrosis of the lower lobes. HSV necrosis affected adrenal glands and liver also, with some viral cytopathy, but abundant HSV in cell nuclei throughout by immunostains. The CNS was not involved.

Case 2: A term male was discharged home following an uncomplicated spontaneous vaginal delivery. The mother was re-admitted for postpartum fever and an infected episiotomy that grew *Enterococcus faecalis* and Group B *Streptococcus*. Infant stayed with mom and had an unremarkable course until DOL 7 when he developed feeding difficulty. On DOL 8, he was poorly responsive, rushed to the ER, and was found to be in respiratory distress with hypotensive shock. The infant was intubated, and antibiotics and acyclovir were started. Labs showed metabolic acidosis, elevated hepatic transaminases, and hyperammonemia. Despite treatment, the infant passed away less than 12 hours later. That night, the mother reported new-onset genital lesions consistent with herpetic vesicles (positive exposure from her husband). Pre-mortem blood and urine cultures grew Group B *Streptococcus* and *Staphylococcus aureus*. Postmortem lung microscopy showed acute HSV pneumonia with necrosis. The colon showed ulcerative HSV colitis with coexistent bacterial overgrowth. HSV viral changes were seen in the liver, adrenal gland, and spleen, confirmed by immunohistochemistry. Ultrastructure of the liver confirmed typical herpes virus features on electron micrographs. The CNS was not involved.

Discussion: Our clinicopathologic findings highlight the importance of close maternal monitoring for signs of primary HSV infection. Furthermore, a high index of suspicion is required for HSV when acute liver failure presents in the neonate. Acquisition of bacterial sepsis complicates the outcome for the highest-risk cases of neonatal HSV.

CLINICAL SPECTRUM OF EARLY ONSET NEONATAL SEPSIS IN THE NEONATAL UNIT AT GALWAY UNIVERSITY HOSPITAL FROM 2011-2014

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Background: Early-onset neonatal sepsis (EONS), defined as sepsis that occurs within the first 72 hours of birth, is a significant cause of neonatal mortality and morbidity
Objective: The purpose of this study was to identify the incidence, presentation, maternal risk factors and bacteria associated with EONS in our neonatal unit over a four year time period.

Methods: A retrospective chart review of all cases of blood culture positive early-onset neonatal sepsis between 2011 and 2014. Neonatal clinical and laboratory data, and maternal clinical risk factors were analysed (Ref 1).

Results: A total of 12,946 infants were born at our institution during the study period (2011-2014). Nine infants had EONS giving an overall incidence of 0.7/1000 Births. Four of the infants were born at term gestation while five infants were premature (< 37 Wks Gestation, Range 25 Wks to 35 Wks). Group B Streptococcus (GBS) was the commonest organism isolated accounting for 45% (4 Infants) of all cases. There were two cases (22%) of Escherichia coli sepsis and one case of each of the following: Listeria monocytogenes, Klebsiella pneumonia and Actinobacteria baumannii complex. Maternal risk factors were documented in three infants (33%). Clinical symptoms suggestive for neonatal sepsis were present in all cases, with respiratory distress being the commonest presenting symptom (60%). FBC abnormalities were uncommon and were only identified in two patients (Low WCC in 2 and low platelet count in 1). All the GBS isolates were sensitive to Penicillin but half of the Escherichia coli isolates were resistant to Amoxicillin.

Conclusion: The rate of EONS at 0.7/1000 births in our neonatal unit is slightly lower than recently reported rates in the UK (Ref 1). However, the clinical presentation and distribution of responsible organisms is similar to previous reports.

Neonatal infections in England: the NeonIN surveillance network. Stefania Vergnano, Esse Menson, Nigel Kennea, Nick Embleton, Alison Bedford Russell, Timothy Watts, Michael J Robinson, Andrew Collinson, Paul T Heath. Arch Dis Child Fetal Neonatal Ed 2011;96:F9-F14

COAGULATION SYSTEM IN VERY PREMATURE NEONATES

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Background: Very premature infants are at risk of bleeding complications and are frequently given plasma because of a perception that coagulation test results are abnormal. However, "abnormal" clotting times may simply be due to physiological immaturity. We hypothesized that prospective characterization of coagulation tests alongside assessment of thrombin generation would provide information on overall haemostatic balance in this population. **Methods.** In a prospective observational study, blood was drawn into citrated tubes from cord blood of neonates (<30/40) at delivery, day 1, day 3 and fortnightly until 30/40 corrected gestational age from non-heparinised lines. Exclusion criteria included antenatal intraventricular haemorrhage. Platelet poor plasma was obtained by centrifugation of whole blood. Prothrombin time (PT), activated partial thromboplastin time (APTT), fibrinogen, procoagulant and anticoagulant factor activity were measured and tissue factor (TF)-stimulated thrombin generation was characterized. Control plasma was obtained from cord blood of term neonates.

Results: Between 2013-2015, 137 patients <30/40 were admitted, 11 excluded and 126 recruited. Median (25th-75th) gestational age and birth weight was 27.7 (26.3-28.7) wks and 1020 (818-1221) g respectively. Median Day 1 values for PT, APTT and fibrinogen were 17.9 (12.9-17.7) s, 79.1 (48.8-131.9) s and 1.3 (0.7-3.9) g/L respectively. Correlation of cord blood results to neonatal blood for PT, APTT and fibrinogen was $r=0.8, 0.6, 0.7$ respectively ($n=38$). PT and APTT were higher on day 1 vs. matched CGA infants for all GA and attenuated with postnatal age ($p<0.001$). Endogenous thrombin potential and peak thrombin generated were comparable in preterm and term infants ($n=100, p=0.08, p=0.4$). Thrombin generation was dose dependently suppressed in presence of increasing amounts of APC in both cord samples from term and preterm neonates and in preterm neonatal samples. Alpha 2 macroglobulin activity was estimated to be lower in cord blood of preterm infants compared to cord blood of controls (12.2 (7.8-57.8) nM vs. 19.6 (12-25) nM, $p<0.001$) and increased with increasing gestational age ($n=100$). Further analysis of procoagulant and anticoagulant mechanisms was performed where sufficient plasma was available from umbilical cord samples. Vitamin K dependent factors (II, VII, IX, X) were lower in preterm cord samples vs. controls ($p=0.007, p=0.31, p=0.011, p=0.01$ respectively). Protein C ($p=0.004$), S ($p=0.07$), antithrombin ($p=0.0001$) and TFPI ($P=0.14$) had lower levels than controls ($n=9$). PT and APTT were further analysed in pooled preterm cord blood ($n=16$) with preterm factor levels and with individual factor levels corrected to that of pooled term cord blood, with no correction of APTT observed.

Conclusion: In conclusion, in the largest prospective study to date of very preterm infants born <30 weeks, we describe typical ranges for standard coagulation tests. We also demonstrate differences in both procoagulant and anticoagulant pathways, to which standard clotting tests may have limited sensitivity and correction of individual factors does not attenuate coagulation profiles. Finally we demonstrate that thrombin generation is similar in very preterm and term infants. These findings call into question the current practice of exposing infants to plasma products based on standard laboratory parameters.

SURFACTANT THERAPY: NOW WHAT?

Surfactant Therapy and Antenatal Glucocorticoids to promote lung maturation are unquestionably the two most significant advances in neonatal care of the last century, resulting in large scale improvements in morbidity and mortality in a preterm population that continues to grow. And yet there have been additional advances that now challenge us to rethink what we know about the benefits of these evidence-based therapies. This talk will review that evidence base and present the current conundrums that face the neonatologist and pediatrician responsible for managing perinatal consultation, delivery room care, NICU care and long term outpatient care of prematurely born infants.

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PEDIATRIC TELEHEALTH PROGRAMS IN THE UNITED STATES: GROWING AND DIVERSIFYING

James McElligott, MD

Background: The use of telehealth has grown rapidly in recent years, with a 400% increase in its use since 2012 to estimated 100 million e-visits performed in 2014. Pediatric services that utilize telehealth are likely to be growing, but no assessment of growth or variation in focus of these services has been done to date.

Methods: The Pediatric Special Interest Group (Peds SIG) of the American Telemedicine Association conducted a Pediatric Telehealth Census in 2011 to determine the number and diversity of various pediatric telehealth programs across the country. In 2011, the census was compiled through a survey of the members of the American Telemedicine Associations (ATA) Pediatric Special Interest Group. Each respondent indicated whether the institution for which they work offers pediatric telemedicine services for each of the Accreditation Council for Graduate Medical Education (ACGME) specialties for inpatients as well as for outpatients. The same method was used to evaluate the diversity of international pediatric telemedicine services. The survey was repeated in 2015 and expanded to include the members of the American Academy of Pediatrics' (AAP) Section on Telehealth Care.

Results: The number of responding organizations grew from 22 in 2011 to 48 in 2015. The findings for inpatient specialties indicated a 24% increase in the number of specialties offering pediatric telehealth services. The diversity of outpatient pediatric programs grew even more significantly with a 61% increase. The most notable increase was seen in the international pediatric telemedicine services being offered as evidenced by growth of 380%. The census also revealed a number of unique pediatric telemedicine programs. The inpatient unique programs identified were: hospice and palliative medicine, plastic surgery (including craniofacial surgery), and thoracic surgery (including congenital cardiac surgery). The only unique outpatient program was that of pain management. The largest growth in unique programs was in international telemedicine. The programs identified were burn surgery, general pediatrics, infectious disease, nephrology, neonatal-perinatal medicine, nuclear medicine, ophthalmology, pathology, rheumatology, and congenital cardiac surgery.

Conclusions: The increase in total number of pediatric organizations utilizing telehealth as well as a greater number of services offered by each organization indicates that the use of telehealth in pediatrics is following the national trend of growth. The development of common services across multiple institutions provides an opportunity for the development of pediatric telemedicine subgroups. By partnering with one another through the implementation of specialty specific subgroups, each telemedicine program will benefit by the experience and expertise of other programs providing similar services. This will not only improve the quality of care being provided but also allow for the development of guidelines and standards. The lack of existing evidence within the pediatric telehealth field could also be addressed by the development of pediatric specialty specific subgroups. By establishing a common set of measures and combining data across each subgroup, outcomes will be stronger and more generalizable.

THE NEW HUMANITARIANISM

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The traditional model for surgeons to be involved in humanitarian activities has been the short-term mission of 2 to 4 weeks, doing a number of cases with complexity based on local resources and limited followup. The author has been involved in such activities for over 30 years in China, South and Central America and East Africa. However, 10 years ago it became evident that a new model would be needed to answer modern day needs in Africa. Current problems in East Africa include extreme poverty, a huge burden of surgical disease, a critical shortage of surgical specialists, limited hospital and medical resources, limited technology, an inadequate medical education system, almost no research training capability, cross border violence and numerous other things.

Beginning 10 years ago, the author and his colleagues in global health at Vanderbilt developed what might be called the university model to address some of the above problems. The model is based on having full-time faculty on site with rotating part-time faculty (the author spends 2 months a year on site and is in contact throughout the year). Training programs have been developed for African residents in general, neuro, plastic, pediatric and orthopedic surgery using criteria of both the American RRCs as well as the COSECSA African accrediting organization, incorporating R-4 Vanderbilt general surgical residents as well. A curriculum has been developed for each specialty with lectures 3 times a week in addition to twice daily bedside group rounds. Self-study resources have been developed as well. An anesthesia program is associated as a shortage of anesthesiologists exists as well. Surgery is performed at a high level. In addition, the trainees have been incorporated into clinical research projects, some Kenya-based and some associated with funded projects at Vanderbilt in order to train them in research methodology.

The results have been gratifying. A high level of complex surgery is being done and the model allows for close postoperative followup. A liaison with the United Nations refugee camps in Northern Kenya has been developed for the care of children needing surgical care. Each of the surgical specialty residency programs has produced 1 or 2 trainees per year since 2006 and all have been involved in at least 1 clinical research project a year, most with publication. They have integrated well with the Vanderbilt senior surgical residents on the team also. Thus far, all the pediatric surgical trainees have taken full-time academic positions in Kenya, Uganda, Sierra Leone and Ethiopia after completing their training. The goal is to eventually have the local African faculty take over the program completely though it will be many years before sub Saharan Africa is totally independent in this regard. This model is applicable to most developing nations with severe medical and surgical workforce shortages.

THE HEALTH CARE CONUNDRUM

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The United States has attempted multiple times over the last century to curtail health care expenses, improve outcomes of medical interventions, and to make health care more equitable and affordable. Today, health care in the US remains the most expensive in Western Societies, yet the outcome is poor by many measures. The latest attempt has been the Affordable Care Act (ACA), often referred to as Obama Care. President Obama championed a significant change in financing health care. He was not proposing the most sweeping and radical reform ever. That honor goes to President Harry S. Truman. I suggest that the conundrum of health care in the United States will never be solved until and unless we, as a nation, decide what health care really represents.

I pose four possibilities: Health care is either: 1. A Commodity; 2. A Privilege; 3. A Charity, or 4. A Right. The problems we have gotten into designing a health care system is that in any given proposal each concept is applied simultaneously to a subset of the population but not to all. A Commodity implies that health care can be purchased when needed following the rules of free enterprise commerce. A Privilege implies that health care is a reward for a certain social status or behavior. A Charity implies that health care is doled out based on the good will of those in power, be it the government, the rich to the poor, or the citizen to the non-citizen. A Right implies that health care is available as a consequence of being human and living in the United States.

In the current climate, the far right and the libertarians favor the commodity model. Most people, who have some middle class status or better, favor privilege as the best model. They reserve the charity model for those less fortunate than they are. Few people like or want to be recipients of charity. Those that favor the models of Commodity and Privilege make the assumption that health care is delivered under the free enterprise system. However, when you add up Medicare, Medicaid, (and its various variants like TennCare, Medi-Cal, etc.), Veterans Administration, Indian Health Service, Champus, health care for incarcerated persons, and charity care by local governments, somewhere between 40-50% of persons receive their health care benefits through the government.¹ Any plan that is based on the assumption of free enterprise is bound to fail if one wants to maintain current systems that work: e.g. Medicare.

1 US Government National Health Expenditure data from 2013

OUTBREAK! TWO UNIQUE CASES OF INFANT BOTULISM

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Abstract:

We describe two unique cases of infant botulism. The first is a 6 month-old previously healthy infant with poor tone, poor feeding, weak suck, and weak cry who was admitted to our institution for suspected sepsis. He progressed to flaccid paralysis with cranial nerve deficits and required mechanical ventilation for respiratory failure. A diagnosis of infant botulism due to *Clostridium botulinum* toxin type B was confirmed by the state health department's toxin neutralization assay. The child received a single dose of intravenous Botulism Immune Globulin (BIG-IV) and fully recovered. Three months later, an 8 week-old infant with a history of constipation, presented to our emergency department with progressively poor feeding, poor head control, and weak cry. Despite aggressive fluid resuscitation for suspected sepsis and dehydration, the infant remained globally hypotonic. Sluggishly reactive pupils, diminished gag reflex, and diminished deep tendon reflexes prompted a suspicion for infant botulism. BIG-IV was administered within 3 days of admission and the infant was discharged after 10 days, never requiring intubation. The state health department also confirmed this case to be due to toxin type B. An investigation found that the two children lived in the same neighborhood, on the same street, and within 100 yards of each other. Although soil sampling was ultimately not performed, it was suspected that a nearby construction area containing a disturbed dirt mound was the source of the *Clostridium* spores. With fewer than 100 cases of infant botulism in the United States each year, these cases represent an extreme for geographic and temporal clustering.

NEONATAL ABSTINENCE SYNDROME: A KENTUCKY PERSPECTIVE

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The use of addictive drugs, such as opioids, during pregnancy often causes newborn infants to suffer from addiction as well. If a baby goes through withdrawal with symptoms such as extreme irritability, tremors, seizures, vomiting, diarrhea, poor feeding, and inconsolability this is called Neonatal Abstinence Syndrome, or NAS. In the United States, rates of NAS have quadrupled since 2004 from 7 to 27 cases per 1000 NICU admissions in 2013. Incidence varied greatly by region with the highest including Kentucky, Tennessee, Mississippi and Alabama. Here, the incidence is more than 16 per 1000 total hospital births. This increase has great social and financial implications both for the families and for society as a whole. Approximately 87% of infants with NAS receive pharmacotherapy such as morphine in addition to behavioral interventions to ease withdraw symptoms. By 2009 the cost of treatment rose to approximately \$53,400 per infant with 77.6% of charges being attributed to state Medicaid programs.

Symptoms of NAS typically do not start until 48-96 hours after delivery as drug levels drop but the effects of prenatal drug exposure can linger for months to years. For example, these infants are at high risk for ongoing problems such as failure to thrive and child abuse due to their excessive irritability and crying. Also, prenatal drug exposure can affect brain development although this may not be detected at birth, as the most frequent manifestations do not result in nervous system malformations but in functional abnormalities years later. These abnormalities typically result in problems with behavior and attention as well as an increase in anxiety and depression. The good news is that outcomes can be changed. By improving psychosocial support of these children and their families they can overcome these initial challenges. Given the dramatic rise in NAS over the past decade despite aggressive anti-drug policies, we should turn our focus on efforts to find interventions that minimize the adverse effects of prenatal drug exposure.

AUDIT OF THE ASSESSMENT AND INITIAL MANAGEMENT OF FEVERISH ILLNESS IN CHILDREN UNDER FIVE YEARS BY THE PAEDIATRIC SPECIALIST

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Background: Feverish illness in young children is a cause for concern in both parents and paediatric specialists and accounts for 20% of paediatric presentations to hospital. It is important to identify children with serious illness who require urgent treatment. The assessment and management of children with febrile illnesses is a challenge and NICE issued Clinical Guideline 160 in 2013 on this subject. Our aim was to assess whether these recommendations were followed at our institution and to identify how our practice could be improved.

Method: A retrospective review of the medical notes of 30 children (<5 years) who presented to the paediatric ward in February 2015 with a documented temperature >38°C was performed. The data collected were vital signs at admission, investigations performed and the initial management. This data was input into Microsoft Excel.

Results: The age of the children ranged from 2 months to 4 years 10 months. All vital signs were recorded in 36% (n=11), heart rate in 80% (n=24), respiratory rate in 76% (n=23), temperature in 90% (n=27) and capillary refill time in 53% (n = 16).

Using the traffic light system, 5 children were assessed as high risk, 16 as intermediate risk and 9 were deemed to be at low risk of serious illness. All children in the high risk group should have a full blood count (FBC), CRP, blood culture and urine tested for infection. In this high risk group (n=5) 100% had FBC and CRP, 80% had blood cultures and 80% had urine tested for infection. 60% received intravenous antibiotics. Of the children at intermediate risk (n=16) 94% had FBC and CRP, 87% had blood cultures and 81% had urine tested for infection. Children at low risk of serious infection should be assessed for signs of pneumonia and have their urine tested for infection; they do not routinely require blood investigations. This group (n=9) 88% had FBC and CRP, 66% had blood cultures, 78% had urine tested for infection. In the total study population 33% had a chest x-ray, 96% were admitted overnight and 43% received IV antibiotics.

Conclusions: Overall there was good documentation of the initial vital signs but there is still room for improvement. A small proportion of the high risk group did not receive all of the recommended investigations and in the low risk group the blood sampling was often not in accordance with the guidelines. These findings were presented to the department and it was decided that all incoming doctors would receive a copy of Clinical Guideline 160 in their induction pack and that the "Traffic Light Table" should be clearly displayed in all clinical assessment areas. This audit will be repeated in six months' time.

References: "Feverish Illness in Children - Assessment and initial management in children younger than five years" Issued May 2013. NICE Clinical Guideline 160

“SILENCE MEANS SNOOZE, PAUSE MEANS STOP”: NICU HEALTHCARE PROFESSIONALS’ (HP) KNOWLEDGE OF MONITOR ALARMS

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Background: The alarm systems employed in the neonatal intensive care unit (NICU) are vital for patient care and safety. They give readings of many physiological variables including heart rate, respiratory rate, oximetry and the cut-offs for the alarms are set by the individual or the manufacturer. The alarms contain a Stop and Pause function.

Aim: Following the recent event, where a neonate may have been hypoxaemic for a period due to misunderstanding of monitor alarms, we decided to evaluate nurses and doctors’ understanding of the alarms in the NICU.

Methods: The biomed technician for the unit was consulted and exact parameters for Stop and Pause button as well as extreme alarm were defined. An 8 item questionnaire was developed. Doctors and nurses were approached with the structured questionnaire to test their knowledge of the unit-specific monitor.

Results: Although all HP’s, except one, had claimed to know difference between Stop and Pause button, the average score overall for the remaining 7 questions was 75%. Over 80 % answered correctly the response to the length of time the alarms are disabled when the silence button is pressed. Majority (66%) knew that silence turns off 1 parameter alarm and 83% knew that pause turns off all parameter alarms. Most HP’s (73%) understood the extreme alarm concept (e.g. desat alarm can be de-activated when oxygen saturation < 90%, but will re-activate when oxygen saturation < 80%). There was no difference overall between clinicians and nurses knowledge of alarms.

Conclusion: HP knowledge of monitor alarms was satisfactory, but not excellent, amongst doctors and nurses. For such an important patient safety action, all HP’s should be fully aware and competent in all monitor alarm functions. HP’s should be reminded that ‘Silence means Snooze (1 alarm) and Pause means Stop (all alarms)’. Continnnual education on alarm functions is necessary in the NICU setting.

Data charts and references available upon request.

RESPIRATORY MEDICATION USE IN INFANTS BORN <29 WEEKS GESTATION IN THE FIRST YEAR OF LIFE AFTER NICU DISCHARGE

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Rationale: Resource utilization due to respiratory illness is prevalent following preterm birth. The NHLBI Prematurity Respiratory Outcomes Program (PROP) [HL101794] aims to identify better predictors of post-NICU discharge respiratory statuses. The NICHD provided supplemental support under the Best Pharmaceuticals for Children Act (BPCA) to ascertain respiratory medication use in premature infants both in the NICU and in the first year of life after hospital discharge.

Methods: Preterm infants <29 weeks gestation at birth were enrolled (n=835) at 6 research centers (13 clinical sites) in the first week of life. Respiratory medication exposure was collected by parental questionnaire every 3 months after discharge, to 12 months corrected age and 656 infants have completed at least one quarterly assessment. Approximately 600 subjects were assessed at each quarter.

Results: Data are reported as number (%) of patients interviewed in each quarter with detailed data (Table) for the most common drugs used. Beclomethasone, caffeine, furosemide, hydrochlorothiazide, hydrocortisone, ipratropium, and racemic epinephrine were used in <5% of babies, and amiloride, aminophylline, bumetanide, formoterol, methylprednisolone, montelukast, sildenafil, and theophylline in <1% of subjects. In any given quarter, 66-75% of infants received no respiratory medications, with the least medication use in the first quarter after discharge.

Drug category:	Month 3 (n = 707)	Month 6 (n = 703)	Month 9 (n = 683)	Month 12 (n = 693)	P-value (by chi-square)
No respiratory drug	531 (75.1%)	478 (68.0%)	464 (67.9%)	462 (66.7%)	<.0001
At least one	176 (24.9%)	225 (32.0%)	219 (32.1%)	231 (33.3%)	<.0001
Diuretic	76 (10.7%)	41 (5.8%)	11 (1.6%)	11 (1.6%)	<.0001
Inhaled Bronchodilator	91 (12.9%)	182 (25.9%)	196 (28.7%)	215 (31.0%)	<.0001
Inhaled Steroid	59 (8.3%)	68 (9.7%)	84 (12.3%)	97 (14.0%)	0.0053
Leukotriene receptor antagonist	0	0	2 (0.3%)	3 (0.4%)	.
Methylxanthine	17 (2.4%)	11 (1.6%)	0	0	.
Pulmonary Vasodilator	2 (0.3%)	7 (1.0%)	6 (0.9%)	6 (0.9%)	0.0045
Systemic Corticosteroid	28 (4.0%)	29 (4.1%)	39 (5.7%)	39 (5.6%)	0.1450

Conclusion: In the PROP cohort, the most frequently used respiratory medication post-discharge is the bronchodilator, albuterol. Systemic and inhaled steroids and diuretics are used in 15%-20% of the population. Despite the high use of caffeine previously described in the neonatal period in preterm infants, post-discharge use remains low. At any given time, 2/3 to 3/4 of former preterm infants are on no respiratory medications. Better understanding of current respiratory medication usage in former premature infants may inform the design of clinical trials to assess drug efficacy and safety.

A STUDY OF A BUBBLE NASAL CPAP (BNCPAP) BUNDLE – HOW IMPLEMENTATION OF POTENTIALLY BETTER PRACTICES (PBP) IN RESPIRATORY CARE OF PRETERM INFANTS CAN GREATLY IMPACT RESPIRATORY OUTCOMES

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BACKGROUND: VLBW infants are at high risk for respiratory failure after birth, which is associated with high morbidity, mortality and hospital costs. Interventions such as prophylactic intubation and mechanical ventilation can increase the risk of complications. Studies showed that many of these complications can be prevented by the use of NCPAP immediately after birth with subsequent selective surfactant administration as an alternative to routine intubation. The objective of this study is to determine the impact of a standardized BNCPAP bundle protocol on short-term respiratory outcomes of a subgroup of VLBW infants.

METHODS: Non-standardized respiratory care practices for VLBW infants admitted to our level 4 NICU were replaced by a comprehensive bundle protocol in October 2014. The bundle consisted of 3 potentially better practices (PBP) based on review of existing literature. Essential parts of the bundle included: 1) support of infant's functional residual capacity (FRC) starting with infant's first breath by placing BNCPAP prongs upon placement under the warmer in the DR, 2) strict rules for selective surfactant administration, and 3) continuation of CPAP support preferentially until 32 weeks PMA. The target population consisted of inborn infants at 26-30 weeks GA. An a priori plan was made for a 1-year review of infants' outcomes pre- (Control group) and post- (BNCPAP group) bundle implementation. Primary outcomes are: the proportion of infants requiring mechanical ventilation, days on mechanical ventilation, and the proportion of infants with O₂ dependency at discharge. Secondary outcomes include the rate of chronic lung disease (CLD) and the length of stay (LOS). Group differences in outcomes were assessed using chi square, Student's t, and log rank tests.

RESULTS: There were 62 infants in the Control group. In the BNCPAP group, 28 infants were discharged in the first 6 months following the implementation of the bundle protocol. An interim univariate analysis after 6 months following the implementation of the intervention showed no significant differences between the study groups with regard to baseline characteristics. Compared to the Control group, infants in the BNCPAP group had a significantly lower rate of intubation (50 vs 84%, $p < 0.001$), and a trend towards lower rate of O₂ dependency at discharge (7 vs 16%, $p = 0.25$). Infants in the BNCPAP group spent a significantly shorter amount of time on mechanical ventilation than infants in the Control group (median 1.4 days vs 5.4 days; log rank test $p < 0.001$). CLD rate was more favorable in the BNCPAP, although it did not reach statistical significance (23 vs 7%, $p = 0.08$). There was no significant difference with respect to LOS.

CONCLUSIONS:

Supporting the first breath of VLBW infants with BNCPAP in the DR was safe and well tolerated. Furthermore, BCPAP had excellent benefits such as reduction of time on mechanical ventilation and intubation rate in the DR. The downward trend of infants with CLD and O₂ dependency at discharge is likely due to continued FRC support up to 32 weeks PMA in the BNCPAP group.

BACK TO THE FUTURE: HOW EARLY CHILDHOOD ADVERSITY AFFECTS HEALTH OUTCOMES

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Does preverbal history matter? Does early experience significantly affect our children or do they forget it? The newborn brain lays down 700 neural synapses per second! Neural circuitry for essential functions is foundationally complete in the first months/ years of life. Knowing the facts about the early effects of toxic stress through experiences like abuse and neglect, maternal depression, and attachment disruption orients us toward effective therapeutic intervention.

This talk will deliver the current state of the literature on how both adverse and positive factors dramatically affect the developing brain and set the foundation for varying developmental outcomes. Discussion will include the current science regarding how adverse factors (e.g., toxic stress, maltreatment, disrupted attachment, maternal depression) and positive factors (e.g., nurturing caregiver interaction, solid attachment) affect neural development and overall health. Some of the sources in this overview include the Adverse Childhood Experiences (ACE) study, information from the Harvard Site on the Developing Child and the Bucharest Early Intervention Project.

ENHANCED SOCIAL BEHAVIORS IN CHILDREN WITH AUTISM SPECTRUM DISORDERS IN THE PRESENCE OF ANIMALS IN THE CLASSROOM

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BACKGROUND: Multiple studies have documented the positive effects of animals in the lives of children including the promotion of empathy, increased awareness of the needs of others & prosocial behaviors. A prominent researcher in the area of Animal Assisted Interaction (AAI) designed two unique projects looking in a rigorous, controlled manner to determine whether there is sufficient evidence to utilize AAI in educational & therapeutic settings to reduce social anxiety and enhance interaction with peers for children with ASD. **METHODS:** Following a review of 14 studies published in peer-reviewed journals that reported positive effects of companion animals upon social behaviors of children with ASD and other disabilities, Dr. O'Haire of Purdue found a lack of control groups, standardized rating scales & methodologic errors. Her team devised two rigorous studies using blinded observers & uniform measures of social skills. In their preliminary paper, published in 2/13. They documented a significant reduction in social anxiety and enhanced social skills, eye contact and verbal interaction. A follow-up was done using skin conductance as a measure of physiologic arousal (which correlates with sweat gland activity and sympathetic nervous system activity).

RESULTS: As in previous studies, children with ASD had a greater sense of well-being and positive affect in the presence of companion animals. On standardized measurements of social skills & severity of ASD they exhibited 1) more social approach behaviors to peers 2) increased verbal interaction and eye contact; 3) more positive emotional displays, i.e. smiling & laughing; 4) they received more social approach (and less negative feedback) from peers; and 5) had fewer problem behaviors, including ag-aggression and self-stimulation. Both the children with ASD and their typically-developing peers (TD) indicated increased emotional valence, i.e. sense of happiness and well-being, in the presence of animals vs. toys.

The second study, published in 4/15, added the use of skin conductance (via a wrist band) to measure physiologic arousal. A cyclic relationship is postulated between social anxiety, autonomic nervous system arousal and the autistic symptoms of social isolation, withdrawal, social skill deficits and negative interaction with peers. In this study, using blinded observers and a control group Dr. O'Haire found global improvements in all clinical parameters listed above. This occurred after 8 weeks with companion animals in the classroom compared to a wait-list control period. In addition, there was a significant decrease in physiologic arousal during 10-minute intervals when the children with ASD were playing with the guinea pigs vs. toys. It was postulated that animals are perceived as non-judgmental. A similar study at UNC showed that Oxytocin has the ability to produce calming & soothing hormones.

CONCLUSIONS: Companion animals are known to have positive effects on physical & mental health and child development. The studies by Dr. O'Haire demonstrate their ability to act as buffers against social anxiety and enhance the mood, behavior and interaction with peers for children with ASD. They have the potential of supplementing other interventions such as speech, social skills training & performance in inclusion class rooms.

AN EVALUATION OF FATHER'S WEIGHT AND BMI AND IMPACT ON CHILDHOOD OBESITY: A NATIONAL COHORT

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Aims: The obesity epidemic may lead to shorter life expectancies in the current generation of children compared to their parents. 19% of Irish children (age nine) are overweight and 7% are obese. One in four Irish males are obese. We aim to examine whether differences exist between paternal self-reported and measured height, weight and BMI classification in a population representative sample; explore paternal perceptions of their own weight status and assess relationship between paternal and child's BMI.

Methods: Population representative sample from the National Longitudinal Study of Children Growing Up in Ireland. Trained social interviewers conducted computer-assisted personal interviews with the study child and both parents/guardians (where applicable) in the home. A self-report of weight (kg), height (cm) and self-perceived weight status (categorised as very underweight, moderately underweight, slightly underweight, about the right weight, slightly overweight, moderately overweight and very overweight) was recorded for all fathers. Measures of height and weight were obtained by trained interviewers using validated methods during the household interview visit. A two-way mixed single measure interclass correlation coefficient (ICC) was used to assess the extent of agreement between father's self-reported and objectively measured height and weight.

Results: Almost three quarters of fathers (6, 263 of 8,568 study children) with a mean age of 42 years (SD 5.04) provided information on themselves and the study child. Mean height was 177cm (SD 6.57). Median weight was 86kg (first quartile=78kg, third quartile=95kg). Median BMI was 27.7 kg/m² (first quartile=25.4, third quartile=30.3kg/m²). The mean difference between self-reported and measured weight was -1.03kg (SD=4.52) indicating that weight was underestimated on average. The mean difference between BMI derived using self-report data and BMI derived from measured data was -0.58 (SD=1.70), indicating that BMI was underestimated on average using self-reported data. The underestimation of weight and BMI increases with BMI category; with obese fathers underestimating their weight by an average of 2 kg. Obese fathers were more likely to have an obese son (9.4% compared to 5.3% for the full cohort; a similar pattern was found for girls (12.4% compared to 7.7%).

Conclusions: Overweight and obesity are rapidly becoming public health issues. Our data suggests that there is a strong relationship between fathers' weights and his sons' and daughters' weights. This relationship is likely to be complex and multi-factorial but at a minimum, this data suggests that tackling overweight and obesity in the child should occur simultaneously with tackling overweight and obesity in the parents (in this study, the Fathers). Obese Dads are not good at estimating their own weight status; we need Irish data on how good they are at estimating their kids' weight status. The impacts of Father's perception of own body image can't be underestimated. Fathers need to recognise overweight and obesity before it can be brought to medical attention.

THE BENEFITS OF COMPANION ANIMALS & PET THERAPY ON PHYSICAL AND MENTAL HEALTH AND CHILD DEVELOPMENT

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BACKGROUND: The use of Animal Assisted Interventions (AAI) has been shown to reduce both the physical & mental effects of stress triggered by illness, disability and hospitalization. Measures of sympathetic nervous system arousal, eg. salivary cortisol levels decrease and Oxytonin production increase during interaction with pets and Immune function is enhanced. Use of AAI has increased to a variety of populations & therapeutic settings.

METHODS: A historical review is combined with presentation of studies looking at the impact of companion animals on physical and mental health & human development. Information will be provided on the goals of AAI, the variety of populations in which it has been used and results of intervention on cardiovascular status, mood & behavior, rehabilitation, cognitive function, socialization and quality of life.

RESULTS: The first known report of AAI comes from 1792 when rabbits were introduced into a Quaker institution for adults with mental illness to promote a sense of calm. Later (1867) in a German hospital for patients with epilepsy, animals were used for comfort and relaxation. These initial trials were in the field of mental health and 50% of clinicians currently advise patients to adopt pets to provide companionship and combat loneliness and depression.

Therapeutic riding has been documented to benefit patients with cerebral palsy through normalization of muscle tone, improved balance & equilibrium, core strength and posture. In addition, children report enhanced self-confidence and esteem. When those with PTSD participated in this activity, there was a calming effect associated with grooming the horses as well. Youngsters with autism were reported to increase their socialization with staff and peers.

Cardiovascular studies have shown reduction in blood pressure, heart rate and lipid levels as well as a dramatic increase in one-year survival for MI patients with companion animals (8 times the rate of those without pets). Following the adoption of a dog, their number of health problems decreased as did mental health concerns. Exercise via walking their dogs increased significantly from baseline. In addition, their socialization was enhanced. Pets act as facilitators & conversation starters.

Especially during Senior years, whether living alone in the community or in long-term care centers, individuals suffer from loneliness, depression & social isolation – all of which impair quality of life. Having a companion animal or pet therapy helps combat these negative feelings which can impair cognitive function, health & nutrition. The presence of animals, eg. fish or birds, in nursing homes can give Residents a sense of purpose by feeding and caring for living things, a principle known as the Eden Philosophy.

Children learn a great deal from having pets in the home or at school. Studies have shown this brings out responsibility, empathy, kindness, affection & concern for other living things. When they move, pets can provide a sense of security and stability thus facilitating their adjustment. They also learn important lessons about life, eg. coping with loss and grief. [Refer to Charlotte's Web].

CONCLUSIONS: The positive bond between humans and companion animals has been shown to have multiple benefits to physical and mental health and child development. Physiologic factors have been identified & measured showing the basis for reducing stress, which interferes with healing.

POSTER PRESENTATIONS

Animal Assisted Intervention (AAI): The Many Roles of Pet Therapy throughout the Life Span

Beverly A. Powell, MD, Associate Prof. of Pediatrics, Fairfax Hospital, VA

Anderson Cooper recently did a 60 Minutes segment on the science that proves our dogs really DO feel emotions and exhibit intelligence. On a personal note, I know my Bichon developed object permanence & memory after 12 months of life -- and my ShihTzu and parrot both have phenomenal memories. I will present 5 short videos documenting the value of pets in nursing homes; the therapeutic effect of riding for kids with CP, the value of Seeing Eye dogs and other Service Animals, and the great work pets do for Vets with PTSD and physical disabilities.

IMPACT OF INTRANASAL MEDICATION AVAILABILITY AND USE ON THE FREQUENCY OF PROCEDURAL SEDATION AND ANALGESIA FOR FACIAL LACERATION REPAIR IN A PEDIATRIC EMERGENCY DEPARTMENT

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Background: Pain and anxiety are frequently under-recognized and under-treated in the pediatric emergency department (PED). The introduction of intranasal medications for pain and anxiety has revolutionized the treatment of these symptoms in pediatric patients. Multiple studies have shown intranasal fentanyl and midazolam to be at least as effective as their oral and parenteral counterparts for the relief of pain and anxiety. It is not known, however, what kind of impact the introduction of intranasal medication has had on the rates and types of procedural sedation for certain painful or difficult procedures, such as facial laceration repair. This study aimed to analyze the association between intranasal medication availability and procedural sedation and analgesia for facial laceration repair in the PED.

Methods: This was a retrospective, before and after, observational cohort study involving patients seen at a large, urban PED between July 1, 2007 and December 31, 2014. In April 2009, at the study facility, intranasal medications were introduced as an option for procedural sedation and analgesia. Patients who presented with facial lacerations before the widespread use of intranasal medications were compared with those who presented after. A randomly selected subset of patients were included if they were < 18 years of age and presented with a facial laceration that was repaired in the emergency department (ED). Patients were excluded if they were: trauma activation, arrived with an IV in place, or had another significant injury. Stepwise logistic regression was used to identify significant predictor variables for procedural sedation, IV sedation, and injectable anesthetic use. Multivariable linear regression was used to compare the groups with regard to ED length of stay (LOS). Candidate predictor variables were: cohort (pre or post); age; number, size, and location of laceration(s); triage pulse and pain score; topical anesthetic use, and repair type. Results: 776 subjects were randomized (389 per group). 49% of subjects in the post-cohort received some sort of procedural sedation and/or analgesia, versus 11% in the pre-cohort. After adjusting for confounders, children in the post cohort had a significantly higher rate of sedation and analgesia (OR=27; 95% CI: 15-48; $p<0.001$). There was no difference with respect to IV sedation (OR=1.4; 95% CI: 0.7-2.7; $p=0.34$). There was significantly less injectable anesthetic use in the post cohort (OR=0.3; 95% CI 0.2-0.5; $p<0.001$). The post cohort had a mean ED LOS 15 minutes greater than the pre-cohort (169 vs 154 minutes; $p=0.004$).

Conclusion: Our results suggest that the availability of intranasal medications was associated with an increased use of appropriate sedation and analgesia and a decreased use of injectable anesthetic for pediatric facial laceration repairs in the ED. We did not find an associated reduction in IV sedations, and there was a slightly increased length of stay in the post-cohort.

References available upon request.

SOS: STREPTOCOCCUS AS AN OCCULT CAUSE OF SACROILIITIS IN A PAEDIATRIC PATIENT: CASE REPORT AND LITERATURE REVIEW

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Background: Group A Streptococcus (GAS) is an aerobic gram-positive coccus, associated with significant morbidity and mortality rates. There is a paucity of data describing pyogenic sacro-iliitis secondary to GAS infection in the current literature. The limited data that exists is predominantly historic and focused primarily on adult obstetric cohorts. , , To date, only one case report has addressed GAS sacro-iliitis in a paediatric patient. Furthermore, a recent Cochrane review (2013) concluded that there was insufficient evidence to show clinically meaningful differences between antimicrobial therapies for Group A Beta-haemolytic Streptococcus, outlining the importance of continued research in this area.

Aim: We report the first documented case of an acute pyogenic sacro-iliitis secondary to GAS in an Irish population

Methods: Several online databases were used to identify relevant papers which could be included in this review. These included PubMed, Medline, Google Scholar, the Cochrane Library and BMJ journals online. The initial search criteria included a combination of the keywords 'sacro-iliitis', 'pyogenic sacroiliitis', 'Group A Streptococcus', 'Streptococcus pyogenes' and 'paediatrics'. The clinical data, radiological imaging and microbiological results of the index case were reviewed.

Results: We present the case of a seven year old boy who presented with a one day history of fevers, severe left hip and lower back pain. On clinical examination, he was pyrexial and rigorous. Examination of the left lower limb revealed significant pain on movement, reduced range of movement of his left hip and knee joint and an antalgic gait. His white cell count ($16 \times 10^9/L$), neutrophils ($15 \times 10^9/L$) and inflammatory markers (C-reactive Protein: 247 mg/L , ESR: 20 mm/hr) were elevated on admission. A septic work up was performed and he was commenced on empiric intravenous benzylpenicillin and clindamycin. Blood cultures isolated Group A Streptococcus. Throat swabs were negative. Plain radiographs of the left hip, and lumbo-sacral spine showed no obvious abnormality. No organisms were cultured from an aspirate of the left hip. Further imaging with a bone scan and subsequent MRI pelvis highlighted radiological features consistent with a left septic sacroiliitis. In total, he completed 14 days of intravenous antimicrobial therapy, 4 weeks of oral amoxicillin and intensive physiotherapy. He was followed up as an outpatient by the infectious disease team, and made a full clinical recovery.

Discussion and Conclusions: This case outlines a rare presentation of GAS sacroiliitis without apparent predisposing risk factors. The diagnostic procedures of choice to localise occult sites of bone inflammation are bone scanning and MR imaging. A multidisciplinary approach to patient care is emphasised in this case with infectious disease specialists, microbiologists, radiologists and physiotherapists playing a central role in the diagnosis and management of the patient. This case is the first Irish case report of GAS sacroiliitis; a treatable condition with few long-term complications if appropriate therapy is initiated promptly. In addition, this case should alert physicians to the possibility of a pyogenic sacro-iliitis in paediatric patients who present with hip pain, back pain and fever.

‘PASEN GAS’: PERI-ANAL GROUP A STREPTOCOCCUS AS A RARE CAUSE OF ERYTHEMA NODOSUM

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Aims:

Erythema nodosum (EN) is the most frequent panniculitis in childhood and has been associated with various conditions, such as infectious and autoimmune disorders, medications, and malignancies. A cause is not always found (up to 55%). Streptococcal infection is the most common underlying cause (28-48%). To highlight the importance of thorough examination for concealed causes of EN in children, we report a case of a previously well 3 year old girl with EN secondary to peri-anal group A streptococcus (GAS).

Methods:

The clinical details and radiological records of our index case were reviewed. Contemporary peer-reviewed literature was perused.

Results:

We present the case of a 3 year old girl, presenting with persistent fever despite antipyretics. A rash was noted on the anterior aspect of her lower limbs. The lesions were red, tender and nodular. Borders are poorly defined and varied between two and six cms in diameter. Her coagulation screen was normal. Inflammatory markers were elevated: CRP 47mg/L, ESR 81mm/hr. ASOT, Chlamydia, EBV and mycoplasma screens were negative, as was a quantiferon test for TB. Blood, urine, stool and throat swab were all culture negative. A chest xray was normal. Non-steroidal anti-inflammatories (NSAIDs) were commenced and she was discharged when her fever had settled.

At follow up, the rash was improving to faded yellow lesions. A peri-anal erythematous rash was noted. A peri-anal skin swab cultured Beta hemolytic GAS +++, Coliform ++, Enterococcus species ++ Specific therapy with oral erythromycin (penicillin allergic) for 10 days was initiated, with progressive clinical improvement and complete recovery within eight weeks.

Conclusions:

The prevalence of EN is 2.4 per 10,000. EN secondary to streptococcal infection is common in children; however, this is the first reported case of erythema nodosum secondary to peri-anal group A streptococcus. Other causes including TB, EBV, various forms of gastroenteritis and myoplasma pneumoniae, can be associated; all were ruled out in our case. Most cases are self-limiting; NSAIDs are useful. Infective aetiology should be treated, but antibiotics should not be given blind. Steroids are beneficial but should be used with caution. If aetiology is infectious, the lesions usually heal within eight weeks. We recommend that children presenting with EN of unknown origin and a napkin rash have a peri-anal swab sent for GAS.

REPRODUCTIVE AND SEXUAL HEALTH ISSUES IN CYSTIC FIBROSIS: AN IRISH MALE PERSPECTIVE

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Aims: 95-98% of male patients with cystic fibrosis (CF) experience infertility. An important corollary of improved survival in CF is the increased significance of developmentally sensitive complications in adolescence and adult life, such as those affecting reproductive and sexual health (RSH).

Methods: A descriptive, cross sectional study was carried out on male CF patients attending Cork University Hospital. An RSH questionnaire was developed by the CF multi-disciplinary team based on previous studies, to address knowledge of infertility, sources of information and genetic implications. A coded version of the patient database was accessed to obtain clinical data. Responses were correlated with clinical phenotype, based on FEV1%, BMI and total serum testosterone level. Independent sample t tests were used ($p < 0.05$ was considered significant). Ethical approval was obtained from the Clinical Research Ethics Committee of the Cork Teaching Hospitals.

Results: Based on 35 completed questionnaires, 34 patients (97%) were aware that fertility is affected. Sources of first hearing information included: CF healthcare professional (62.1%), parent (13.8%) and other (24%). Patient's preferred source of information was their HCP and their recommended place of discussion was the clinic. The mean (SD) age when reproductive issues were broached was 19.47 years (4.79) (median age 19, range 14-30), which is significantly later ($p < 0.001$) than the desired mean (SD) age of 15.92 years (2.57) (median age 16, range 12-20) in our population surveyed. Rate of contraceptive use varied between patients, with 34.2% ($n=12$) admitting to never using contraception. Lower FEV1 levels are associated with self-reported affected sex life ($p < 0.01$). There is an inverse relationship between self-reported impotency and testosterone levels ($p < 0.05$).

Conclusion: Our study provides a detailed description of reproductive and sexual health issues in an Irish male CF population. The findings highlight that Irish males with CF are well informed about fertility issues. The key timing identified for disclosure of such information was earlier than our current practice and the HCP was identified as the person most desirable to initiate conversations. Our findings noted a significant relationship between self-reported impotence rates and low testosterone levels. Disconcertingly, over one-third report never using contraception. These issues require counselling and education as part of standard CF care, commencing in early adolescence. There is scope for future work in this area, as no study has yet been carried out evaluating Irish parental perspective on RSH issues in male CF patients. Furthermore, there is a need to explore RSH in Irish female patients with CF, and the implications of differing contraceptive practices. This study paves the way for patient-centred clinical care for males with CF.

