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<td>TRANSFUSION OF PREMATURE INFANTS WITH BIOTINYLATED RED BLOOD CELLS (BIORBCS) DOES NOT ELICIT AN ANTIBODY RESPONSE</td>
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Dr Kevin Dunne, President (IPA); Prof. Eugene Dempsey, President (IAPS), Dr Anne-Marie Murphy, Secretary & President-Elect (IPA), Dr Turlough Boiger, Treasurer (IPA); Dr Rita Ryan, President-Elect (IAPS) |
| 08.40-10.20| **SESSION 1: GENERAL & SUB-SPECIALITY PAEDIATRICS/HEALTHCARE MANAGEMENT/EDUCATION**  
Chairs: Dr Kevin Dunne, Dr Jerry Evans, Dr Paul Gallagher & Dr Lola Ihidero |
| 08.40-08.50| A COMPARATIVE STUDY OF TWO MEDICATION GROUPS OF INFANTS WITH MODERATE BRONCHIOLITIS  
F Yasin, ZS Afridi, R Khan, ¹Paediatric, Kerry General Hospital, Tralee, Ireland |
| 08.50-09.00| TWO WEEKS OF CARE – FIRST BED UTILISATION STUDY OF AN IRISH REGIONAL PAEDIATRIC UNIT, UNIVERSITY HOSPITAL LIMERICK, 2013  
CO OhAiseadh1,2, M Mannix2, J Saunders3, R Philip4, ¹Department of Public Health, Health Service Executive, Dublin, ²Department of Public Health, Health Service Executive, Limerick, ³Statistical Consulting Unit / CSTAR @ UL, University of Limerick, Limerick, ⁴Regional Paediatric Unit, University Hospital Limerick, Limerick, Ireland |
| 09.00-09.10| GROUP B STREPTOCOCCAL MENINGITIS AND VENTRICULOOPERITONEAL SHUNTS  
MR Crealey1, T Mandiwanza2, D Crimmins2, A Foran2, JF Murphy2  
¹Neonatology, Childrens University hospital, Temple st., Dublin, ²Paediatric Neurosurgery, Childrens University hospital, Temple st., Dublin, ³Neonatology, The Rotunda Hospital, Dublin, Ireland |
| 09.10-09.20| CONDENSED REFERRAL FORMS IMPROVE DATA CAPTURE IN THE NORTHERN IRELAND PAEDIATRIC INTENSIVE CARE UNIT  
P Donnelly1, J McCabe1, A Keaney1, ¹Paediatric Intensive Care Unit, Royal Belfast Hospital for Sick Children, Belfast, Northern Ireland |
| 09.20-09.30| PINE NUT ALLERGY IN CHILDREN PRESENTING TO A NEWLY ESTABLISHED ALLERGY CLINIC IN IRELAND: A CASE SERIES  
C O’Carroll1, A Byrne1,2, JA1, ¹Allergy, Our Lady's Children's Hospital Crumlin, Dublin, ²Allergy, AMNCH, Tallaght, Dublin, Ireland |
| 09.30-09.40| THE RSV PREMI PROSPECTIVE STUDY: PRETERM (32-36 WEEKS GESTATIONAL AGE) RISK ESTIMATION MEASURE FOR RSV HOSPITALIZATION IN IRELAND  
M Sheridan-Pereira1,2, J Murphy3, G Crispino3, D Corcoran3, J Murphy3, G Dempsey4, B Elnazir5, P Gavin5, LJ Bont5, B Paes10, ¹Paediatrics, Coombe Women and Infants University Hospital &Trinity College Dublin, Dublin, ²Paediatrics, Trinity College Dublin, Dublin, ³Children's Research Centre, Our Lady's Hospital for Children Crumlin, Dublin, ⁴Paediatrics & Neonatology, Rotunda Hospital, Dublin, ⁵Paediatrics & Neonatology, National Maternity Hospital, Dublin, ⁶Paediatrics & Neonatology, Cork University Maternity Hospital, Cork, ⁷Paediatrics & Respiratory Medicine, AMNCH Tallaght Hospital, Dublin, ⁸Paediatric Infectious Disease, Our Lady's Hospital for Children Crumlin, Dublin, ⁹Paediatric Infectious Disease, Wilhelmina Children's Hospital , University Medical Center Utrecht, Utrecht, The Netherlands, ¹⁰Paediatrics & Neonatology, Mc Master University , Edmonton, Canada |
| 09.40-09.50| FOOD AND EXERCISE CUES IN CHILDREN TELEVISION PROGRAMMES: THE INFLUENCE OF CHARACTER GENDER  
P Scully1, O Reid1, A Macken1,2,4, D Leddin3, W Cullen4, C Dunne4, C O’Gorman1,3,4, ¹The Children's Ark, , University Hospital., Limerick, ²National Children's Research Centre, OLCHC, Dublin, ³Dalhousie University, Halifax, , Canada ⁴Centre for Interventions in Infection, Inflammation & Immunity, Graduate Medical Entry School, University of Limerick, Ireland |
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| 09.50-10.00  | **VISUAL THINKING STRATEGIES FOR MEDICAL STUDENTS – THE TEACHER EXPERIENCE**  
Maher B,1 Magner R,2 Malone A,3 Bennett D,1 O’Flynn S,1 Ryan A,3  
1 School of Medicine, UCC,  2 Medical Student, University College Cork,  3 Department of Paediatrics and Neonatology, Cork University Hospital |
| 10.00-10.10  | **A COMPARISON OF EXCLUSIVE ENTERAL NUTRITION (EEN) AND CORTICOSTEROIDS (CS) IN THE INDUCTION OF REMISSION IN PAEDIATRIC CROHN’S DISEASE (CD)**  
Lafferty1,2, A Carey1,3, M Tuohy1,2, S Sugrue2, B Bourke1, A Broderick1, S Quinn1, S Hussey1  
1 Department of Gastroenterology, Hepatology and Nutrition, Our Lady's Children's Hospital, Crumlin, Dublin; 2 Dublin Institute of Technology, Kevin Street, Dublin; 3 National Children’s Research Centre, Our Lady's Children's Hospital, Crumlin, Dublin, Ireland |
| 10.10-10.20  | **VASCULAR AND METABOLIC STUDIES IN CHILDREN WHOSE FATHERS HAD PREMATURE CARIOVASCULAR DISEASE**  
Alan P Macken1,2,4, Michael O’Neill1, Catriona Aherne1, Ann Breen1, Ophelia Blake1, Walter Cullen4, Colum Dunne4, Clodagh S O’ Gorman1,2,4  
1 University Hospital Limerick; 2 National Children's Research Centre, Dublin; 3 Mayo General Hospital, Castlebar, Mayo; 4 Centre for Interventions in Infection, Inflammation & Immunity, Graduate Entry Medical School, Limerick, Ireland |
| 10.20-10.50  | **COFFEE/MINGLING/POSTER & EXHIBITION STAND VIEWING** |
| 10.50-11.25  | **BILL KIDNEY GUEST LECTURE – Introduction: Dr Roy Philip and Dr Turlough Bolger**  
"AUTOINFLAMMATORY SYNDROMES“  
Dr Grainne O'Regan, Consultant Paediatric Dermatologist, Our Lady's Children's Hospital, Dublin |
| 11.25-13.00  | **SESSION 2: PLENARY SESSION**  
Chairs: Prof. Eugene Dempsey, Dr Patrick Pierse, Dr Turlough Bolger & Dr John Kelleher |
| 11.25-11.40  | **PULMONARY EMBOLISM IN TEENS AND YOUNG ADULTS AND POSSIBLE LINKS TO INCREASING OBESITY RATES**  
Natalie K. Wallis, MD, PhD and Vlad C. Rudulescu, MD, Department of Pediatrics, Division of Hematology and Oncology, University of Kentucky, Lexington, Kentucky |
| 11.40-11.55  | **UNEXPLAINED SEVERE EARLY ONSET EPILEPSY – THE CLINICAL AND RESEARCH UTILITY OF WHOLE EXOME SEQUENCING IN AN IRISH COHORT**  
NM Allen1, J Conroy1, A Shahwan1, S Ennis2, B Lynch1, D McCreary1, SA Lynch1,3, MD King1,2  
1 Department of Paediatric Neurology, Children's University Hospital, Temple St., Dublin; 2 Academic Centre on Rare Diseases, School of Medicine and Medical Science, University College Dublin; 3 Department of Clinical Genetics, Children's University Hospital, Temple St. Dublin, Ireland |
| 11.55-12.10  | **MILRINONE USE FOR HEMODYNAMIC STABILITY IN PDA LIGATION**  
Matt Halliday, MD; Mino Kavarana, MD; Myla Ebeling; James Kiger, MD, Medical University of South Carolina, Charleston, SC, USA |
| 12.10-12.25  | **A 4 YEAR AUDIT OF THERAPEUTIC HYPOTHERMIA IN HYPOXIC ISCHAEMIC ENCEPHALOPATHY IN THE COOMBE HOSPITAL**  
Durnin, S & O’Connor, P., Department of Paediatrics and Newborn Medicine, Coombe Women & Infants University Hospital, Dublin |
| 12.25-12.40  | **MECONIUM ILEUS: MUCIN SECRETION IN THE CF-ILEUM**  
Koryse Sadari Woodrooffe, MD1, ChangSuk Moon Ph.D.1, Chandrima Sinha Ph.D.1, Kavisha Arora Ph. D.1, Sunita Yarlagadda M.S.1, Mehmet Kesimer Ph.D.2, Lubna Abdullah Ph.D.2, Jeffrey Whitsett M.D.1, Anjaparavanda P Naren Ph.D.1,11 Cincinnati Children’s Hospital Medical Center Cincinnati, OH 1 University of North Carolina at Chapel Hill Chapel Hill, NC, USA |
| 12.40-12.55  | **DOWN’S ARTHROPATHY – CLINICAL AND RADIOLOGICAL FEATURES**  
C Foley, EJ MacDermott, OG Killeen, 1 The National Centre for Paediatric Rheumatology (NCPR) |
| 13.00-14.00  | **LUNCH/POSTER WALK 1 AND ADJUDICATION (Poster numbers 1-33)**  
Dr Frances Enright & Dr Con Sreenan, Dr Elizabeth O’Mahony & Dr Orla Coyle, Dr John Twomey & Dr Rizwan Khan, Dr Kevin Dunne & Dr Roy Philip |
| 14.00-14.35  | **GUEST LECTURE – Introduction: Dr Michael Mahony and Dr Paul Gallagher**  
"HAEMOLYTIC URAEMIC SYNDROME“  
Dr Atif Awan, Consultant Paediatric Nephrologist, The Children’s University Hospital, Dublin |
### JOINT IPA/IAPS ANNUAL MEETING

**FRIDAY 26TH SEPTEMBER 2014**

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<td>POTASSIUM CHANNEL-RELATED EPILEPSY: DIFFERENT PHENOTYPES, DIFFERENT GENOTYPES, DIFFERENT OUTCOMES!</td>
<td>Dr Kevin Dunne, Prof. Tom Clarke, Dr Rizwan Khan &amp; Dr Peter Ihidero</td>
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<td>14.35-14.45</td>
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<td>Department of Paediatric Neurology, Children's University Hospital, Temple St. Dublin 1</td>
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<td>14.35-14.45</td>
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<td>Academic Centre on Rare Diseases, School of Medicine and Medical Science, University College Dublin</td>
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<td>14.35-14.45</td>
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<td>Department of Clinical Genetics, Temple St. Children's Hospital, Dublin</td>
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<td>PAEDIATRIC TONSILLOTOMY – AN IRISH PERSPECTIVE ON POTENTIAL EVOLVING INDICATIONS</td>
<td>CWR Fitzgerald², JC Oosthuizen¹, M Colreavy¹</td>
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<td>14.55-15.05</td>
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<td>Department of Paediatric Otolaryngology, Head and Neck Surgery, Children's University Hospital, Temple Street, Dublin 1</td>
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<td>SELF BOUGIENAGE IN CHILDREN WITH OESOPHAEGAL STRICURE, CASE SERIES</td>
<td>T I Y Hassan¹, E Stenke¹, S Paran², B Bourke¹</td>
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<td>15.05-15.15</td>
<td></td>
<td>Gastroenterology, Hepatology and Nutrition, Our lady's children Hospital, Crumlin, Dublin, ²Department of Paediatric Otolaryngology, Head and Neck Surgery, Children's University Hospital, Temple Street, Dublin 1</td>
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<td>RETROSPECTIVE ANALYSIS OF PAEDIATRIC APPENDECTOMIES</td>
<td>PJ Carroll¹, D Healy¹, C Travers¹, A Merrigan¹, PA Grace¹, A Murphy¹</td>
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<td>Department of Surgery, University Hospital Limerick, Limerick, ²Department of Paediatrics, University Hospital Limerick, Limerick, Ireland</td>
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<td>15.25-15.35</td>
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<td>FUMARASE DEFICIENCY: Clinical spectrum and diagnostic challenges</td>
<td>O N Oketh¹, I Knerr¹, E Crushell¹, PD Mayne², J Hughes¹, AA Monavari¹</td>
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<td>15.25-15.35</td>
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<td>National Centre for Inherited Metabolic Disorders (NCIMD), , Temple Street, Dublin, ²Department of Clinical Biochemistry, , Temple Street, Dublin</td>
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<td>15.45-15.45</td>
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<td>FRIEDREICH ATAXIA IN CLASSICAL GALACTOSAEMIA</td>
<td>SAM Neville¹, B Sweeney¹, B Lynch¹, I Knerr¹, D Hanrahann¹, SA Lynch¹, S O'Sullivan¹, E Crushell¹</td>
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<td>15.45-15.45</td>
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<td>National Centre for Inherited Metabolic Disorders, Temple Street Children’s University Hospital, Dublin, ²Department of Metabolic Paediatrics, The Royal Hospital for Sick Children, Belfast, N.Ireland, ³Department of Paediatric Neurology, Temple Street Children's University Hospital, Dublin, ²Department of Paediatric Neurology, The Royal Hospital for Sick Children, Belfast, Northern Ireland</td>
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<td>15.45-15.45</td>
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<td>Department of Clinical Genetics, Temple Street Children's University Hospital, Dublin, Ireland</td>
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<td>CONSANGUINITY IN IRELAND – ESTIMATING THE PREVALENCE AND CONSIDERATIONS FOR PUBLIC HEALTH</td>
<td>Peter Michael Barrett, MB BCh BAO, MSc, MPhil, Department of Public Health and Primary Care, University of Cambridge, UK</td>
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<td>“BONE HEALTH: THE SKELETON IN THE CLOSET”</td>
<td>Dr Ciara McDonnell, Consultant in Paediatric Endocrinology &amp; Diabetes, National Children’s Hospital Tallaght &amp; Children’s University Hospital, Temple Street, Dublin</td>
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<td>Session 4: SHORT PAPER PRESENTATIONS(Miscellaneous)</td>
<td>AN AUDIT OF THE EMERGENCY PAEDIATRIC DENTAL SERVICE IN TEMPLE STREET CHILDREN'S UNIVERSITY HOSPITAL (TSCUH)</td>
<td>Dr Paul Gallagher, Dr Michael Mahony, Dr Roy Philip, Dr Alan Macken</td>
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<td>16.40-16.45</td>
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<td>SM McGlacken-Byrne¹, E McGovern¹, R McNamara¹ Emergency Department , Temple St Children’s University Hospital, Dublin, Ireland</td>
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<td>REDUCTION IN ACUTE PAEDIATRIC ADMISSIONS FOLLOWING INTRODUCTION OF EMERGENCY REVIEW CLINIC IN MRH MULLINGAR</td>
<td>F McCartan¹, B Rai¹, F Sharif¹, I Lambert¹ ²Paediatrics, Midlands Regional Hospital, Mullingar, Co. Westmeath, Ireland</td>
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<td>16.50-16.55</td>
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<td>AUDIT OF LIVER BIOPSIES CONDUCTED OVER A TWO YEAR PERIOD IN THE NATIONAL CENTRE FOR PAEDIATRIC GASTROENTEROLOGY, HEPATOLOGY AND NUTRITION.</td>
<td>E. Crowley¹, K. Fejer¹, E. Lowe¹, D. Connor¹, B. McDermott¹, AM. Broderick¹, B. Bourke¹, S. Hussey¹</td>
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<td>16.50-16.55</td>
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<td>National Centre for Paediatric Gastroenterology, Hepatology and Nutrition, Our Lady's Children's Hospital,Crumlin,, Dublin 12</td>
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<td>16.55-17.00</td>
<td><strong>AUDIT OF KARYOTYPE REQUEST AT UNIVERSITY HOSPITAL LIMERICK</strong></td>
<td>N Ahmed, H Adam, D O'Sullivan, AM Murphy</td>
<td>Paediatrics, UHL, Limerick, Neonatology, UHL, Limerick, Ireland</td>
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<td><strong>PRIMARY CARE ROLE OF BLOOD PRESSURE MEASUREMENT IN PAEDIATRICS POPULATION</strong></td>
<td>RA Ahmed, DK Kadar, RK Khan, AK Khan</td>
<td>Paediatrics, Kerry General Hospital, Tralee, Ireland</td>
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<td><strong>TONGUE-TIE BREASTFEEDING DILEMMA: THE GREAT DEBATE</strong></td>
<td>RF Power</td>
<td>Neonatology, National Maternity Hospital, Dublin, Ireland</td>
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<td>17.15-17.20</td>
<td><strong>REFERRAL PROCESS TO THE NATIONAL CENTRE FOR PAEDIATRIC RHEUMATOLOGY - IS IT UP TO SCRATCH?</strong></td>
<td>DM O'Leary, BP Treston, EJ MacDermott, OG Killeen</td>
<td>National Centre for Paediatric Rheumatology, Our Lady's Children's Hospital, Dublin, Ireland</td>
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<td>17.20-17.25</td>
<td><strong>HOW WELL ARE WE CONTROLLING CHILDREN'S SUGARS?</strong></td>
<td>PJ O'Reilly, W Khan, A Finan</td>
<td>Paediatrics Department, Cavan General Hospital, Cavan Town, Ireland</td>
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<td>17.25-17.30</td>
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<td>Closing Remarks and IPA AGM – Chairs: Kevin Dunne, Anne-Marie Murphy, Turlough Bolger</td>
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<td>ADJUDICATION AND DRINKS RECEPTION</td>
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<td>Dr Kevin Dunne &amp; Dr Roy Philip &amp; Dr John Twomey &amp; Dr Turlough Bolger, Prof. Eleanor Molloy &amp; Dr Michael Mahony, Dr John Kelleher &amp; Dr Liam O'Connell, Dr Ciara McDonnell &amp; Dr Paul Gallagher, Prof. Eugene Dempsey &amp; Dr Con Sreenan, Dr Anne-Marie Murphy &amp; Dr Mathew Thomas, Dr Frances Neenan and Dr Sharon Condon</td>
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<td>Sinead Cassidy (Administration &amp; Queries)</td>
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<td><strong>DINNER WELCOME AND GRACE – Dr Kevin Dunne</strong></td>
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<td><strong>After Dinner Speaker – Prof. Tony Ryan</strong></td>
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**Thank you to our sponsors!!**

This meeting has been supported by Abbvie, Alexion, Chiesi, Genzyme, GSK, Mead Johnston Nutrition, Nutricia, Pfizer, Shire and SMA through the purchase of exhibition trade space.
Joint IPA/IAPS - Annual Meeting (CPD Pending)

SATURDAY 27TH SEPTEMBER 2014

07.30-08.30 REGISTRATION – TEA/COFFEE & PASTRIES

08.40 OPENING ADDRESS
Dr Kevin Dunne, President (IPA), Dr Anne-Marie Murphy, Secretary (IPA), Prof. Eugene Dempsey (IAPS) President, Dr Liam O’Connell, Dr Barry Scanlan

08.50-10.30 SESSION 4: ACCIDENT & EMERGENCY AND AMBULATORY PAEDIATRICS
Chairpersons: Dr Ward Rice, Dr Raymond Harre, Prof. Eugene Dempsey and Dr Turlough Bolger

08.50-09.00 THE RIGHT PLACE AT THE RIGHT TIME - PROVIDING AMBULATORY FOLLOW UP TO CHILDREN WITH HENOCH SHONLEIN PURPURA (HSP)
C Martin, T Bolger, S Koe, S Moran, 1Paediatric Emergency Dept, Tallaght Hospital, Dublin

09.00-09.10 THE PATTERN OF ATTENDANCE OF CHILDREN PRESENTING TO THE PAEDIATRIC MEDICAL EMERGENCY DEPARTMENT IN THE UNIVERSITY HOSPITAL LIMERICK
O. Ihidero, P. Ihidero, J. Twomey, UCHL, Limerick

09.10-09.20 AUDIT POST EDUCATIONAL INTERVENTION TO IMPROVE THE DIAGNOSIS AND MANAGEMENT OF GROUP A STREPTOCOCCUS PHARYNGITIS IN THE EMERGENCY DEPARTMENT
M Dominguez1, J Lucey2, I Okafor3, S Deiratany3, R Mc Namara3, R Rush4, R Cunney5, K Butler6,7, P Gavin6,7.
1Medical Science, University College Dublin, Dublin, 2Medical Science, Trinity College Dublin, Dublin, 3Emergency Department, Children University Hospital Temple Street, Dublin, 4Surveillance Scientist, Our Lady’s Children Hospital Crumlin, Dublin, 5Microbiology, Children University Hospital Temple Street, Dublin, 6Infectious Diseases, Our Lady’s Children Hospital Crumlin, Dublin, 7Infectious Diseases, Children University Hospital Temple Street, Dublin, Ireland

09.20-09.30 ASSESSING THE MANAGEMENT OF ACUTE TONSILLITIS IN A PAEDIATRIC EMERGENCY DEPARTMENT
SC Richardson1, DC Mc Collum1, D Coglan1, J Fennell2, C Martin1
1Department of Paediatrics, National Children’s Hospital, Tallaght, Dublin, 2Department of Clinical Microbiology, National Children’s Hospital, Tallaght, Dublin, 3Paediatric Emergency Department, National Children’s Hospital, Tallaght, Dublin, Ireland

09.30-09.40 TO REVIEW CURRENT PRACTICE AND COMPARE TO BEST INTERNATIONAL STANDARDS REGARDING FIRST FEBRILE CONVULSIONS. TO CREATE A CLINICAL GUIDELINE
R McGovern1,2, C Power1,2, C Martin1, Dr S Koe1, Dr T Bolger1 1Paediatric Emergency Department, AMNCH, Tallaght, Dublin

09.40-09.50 COMPLICATIONS OF HOSPITAL ADMISSION FOR INFANTS WITH NEONATAL ABSTINENCE SYNDROME (NAS) OVER AN EIGHT YEAR PERIOD.
JA Jones, D Staunton, P Curran
1Paediatric Department, Portiuncula Hospital, Ballinasloe, Galway, Ireland

09.50-10.00 INCIDENCE OF FRAGILE X SYNDROME IN IRELAND - AN ALL IRELAND STUDY
JJ O’Byrne1, M Sweeney1, D Donnelly2, D Lambert1,3, D Beattie2, C Gervin2, CA Graham2, DE Barton1, SA Lynch1 1National Centre for Medical Genetics, Our Lady’s Children’s Hospital Crumlin, Dublin, 2Northern Ireland Regional Genetics Centre, Belfast Health and Social Care Trust/City Hospital, Belfast, Northern Ireland, 3The Children’s University Hospital, Temple Street, Dublin, Ireland

10.00-10.10 EARLY LIFE TRANSEPIDERMAL WATER LOSS (TEWL) VALUES AS A PREDICTOR OF FOOD ALLERGY AND SENSITISATION AT 2 YEARS: RESULTS FROM THE BASELINE STUDY
M Kelleher1, C Cullinane1, A Dunn Galvin1, D M Murray1,2, J O’B Hourihane1, BASELINE Team1
1Dept of Paediatrics & Child Health, University College Cork, Cork, 2National Children’s Research Centre, OLCHC, Dublin, Ireland

10.10-10.20 DETERMINING THE LUNG CLEARANCE INDEX OF CHILDREN WITH CYSTIC FIBROSIS, AGED 4-16 YEARS, VERSUS HEALTHY SUBJECTS IN AN IRISH SETTING
Keane L1, Linnane B2, Saunders J3,4, Zeugolis D5 1Centre for Adult Learning and Professional Development, 2University Hospital Limerick, 3Statistical Consulting Unit, University of Limerick, 4Centre for Support and Training in Analysis and Research CSTAR Limerick, 5Network of Excellence for Functional Biomaterials, National University of Ireland

* Additional presentations from other sessions
SADAY 27TH SEPTEMBER 2014

10.20-10.45  COFFEE/MINGLING/POSTER & EXHIBITION STAND VIEWING

10.45-11.20  FRED BURKE GUEST LECTURE: Introduction: Dr John Twomey
“NEBROWN SCREENING FOR CF”
Dr Barry Linnane, Paediatric Respiratory Consultant, University Hospital Limerick (UHL)

11.20-12.50  SESSION 5: PLENARY SESSION
Chairpersons: Dr Willie O’Connor, Dr Tania Condurache, Dr Frances Enright, Dr Barry Linnane

11.20-11.35  HAVING A CHECKLIST IS ASSOCIATED WITH IMPROVED NEWBORN SCREEN SPECIMEN COLLECTION
Shelly- Ann Patricia Williams MD, James Kiger MD, Frances Koch MD
Medical University of South Carolina, Charleston SC, USA

11.35-11.50  “THE FIRST 300 THROUGH THE DOOR”: A REVIEW OF THE NEW ALLERGY CLINIC AT A DUBLIN PAEDIATRIC HOSPITAL
M Iatan1, C O’Carroll2, A Byrne3 1School of Medicine, Trinity College Dublin, Dublin, 2Allergy Department, Our Lady's Children's Hospital Crumlin, Dublin 12, Ireland

11.50-12.05  CARDIAC ASSOCIATIONS WITH ADAMS-OLIVER SYNDROME
Sarah Marie Maines, MD; Anjana Pettigrew, MD; Cristin Rolf, MD; Vanessa Smith, MD; William O’Connor, MD, University of Kentucky, Lexington, Kentucky, United States of America

12.05-12.20  ASSESSING THE IMPACT OF VACCINE REFUSAL AND DELAY ON MEASLES VACCINATION RATES
Joy Checa, MD and Jimmy McElligott, MD (Mentor), Medical University of South Carolina

12.20-12.35  EXCLUSIVE ENTERAL NUTRITION IN THE TREATMENT OF CROHN’S DISEASE IN AN IRISH PAEDIATRIC HOSPITAL OVER A TEN-YEAR PERIOD
M Tuohy1,2, A Carey1,3, L Lafferty1,2, S Sugrue1,2, B Bourke1,3, AM Broderick1, S Quinn1, S Hussey1,3
1Department of Gastroenterology, Hepatology and Nutrition, Our Lady’s Children’s Hospital Crumlin, Dublin 12, 2School of Biological Sciences, Dublin Institute of Technology, Kevin Street, Dublin 8, 3National Children’s Research Centre, Our Lady’s Children’s Hospital Crumlin, Ireland

12.35-12.50  CLINICAL OUTCOMES FOR PATIENTS WITH HIGH RISK NEUROBLASTOMA TREATED IN IRELAND BETWEEN 2003-2012
A Fox1, A Ryan1, J Pears2, M Capra2, C Owens2,1Graduate Entry Medical School, University of Limerick, Limerick, 2National Paediatric Haematology & Oncology Centre (NPHOC), Our Lady’s Children’s Hospital Crumlin (OLCHC), Dublin, Ireland

13.00-14.00  LUNCH/POSTER WALK 3 AND ADJUDICATION (Poster Numbers 71-96)
Dr John Twomey & Prof. Tony Ryan, Dr Matthew Thomas & Dr Anne-Marie Murphy, Dr Kevin Dunne & Prof. Eugene Dempsey, Dr Orla Coyle & Dr Con Sreenan, Prof. Clodagh O’Gorman & Dr Frances Neenan

14.00-15.25  SESSION 6: NEONATAL SESSION
Chairpersons: Prof Eleanor Molloy, Dr Roy Philip, Prof. Tony Ryan and Dr Saulius Sitas

14.00-14.35  GUEST LECTURE – Introduction: Prof. Eugene Dempsey
“INVESTIGATIVE APPROACH TO STILLBIRTH”
Dr Brendan Fitzgerald, Consultant Histopathologist, CUH, Cork

14.35-14.40  EARLY CRP PREDICTS CHORIOAMNIONITIS IN VERY LOW BIRTH WEIGHT PRETERM INFANTS
E Ryan1, P Jayadev Menon1, D Eves2, S Alnafisee2, E Mooney3, P Downey3, EJ Molloy1
1Neonatology, National Maternity Hospital, Dublin, 2Paediatrics, Royal College of Surgeons, Dublin, 3Pathology, National Maternity Hospital, Dublin, Ireland

14.40-14.45  1,25-DIHYDROXY VITAMIN D3 INDUCES ANTIBACTERIAL ACTION IN PRETERM INFANTS BY ENHANCING NEUTROPHILS RESPIRATORY BURST IN VITRO
C Onwuneme1,2,3, F Martin1, A Blanco2, A O’Neill2, RWG Watson2, EJ Molloy1,4Neonatology, National Maternity Hospital, Dublin, 2UCD School of Medicine and Medical Sciences, University College Dublin, Dublin, 3Paediatrics, Children’s University Hospital, Dublin, 4Paediatrics, TCD, Trinity College, Dublin

14.45-14.55  INTER-OBSERVER AGREEMENT IN NEONATAL SEIZURE DETECTION BASED ON THE VISUAL INTERPRETATION OF THE ELECTROCEPHALOGRAM
NJ Stevenson1, JM Rennie1, WP Marnane1, GB Boylan1 1Irish Centre for Fetal and Neonatal Translational Research, University College Cork, Cork, Ireland

14.50-14.55  REDUCING NEONATAL MORTALITY IN CENTRAL KENYA
C Duncan, 1Paediatric Department, Altnagelvin Area Hospital, Derry, UK, 2Global Links Programme, Royal College of Paediatrics & Child Health, London, 3Nanyuki Teaching & Referral Hospital, Ministry of Health, Nanyuki, Kenya
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| 14.55-15.00 | CEREBRAL OXYGENATION IN DELIVERY ROOM STABILIZATION OF VERY PRETERM INFANTS REQUIRING LOWER VERSUS HIGHER FRACTION OF INSPIRED OXYGEN  
1 Kenosi M, 2 O'Toole JM, 3 Hawkes GA, 4 Ryan CA, 5 Dempsey EM  
1 Department of Paediatrics and Child Health, Neonatal Intensive Care Unit, Wilton, Cork,  
2 Irish Centre for Fetal and Neonatal Translational Research (INFANT), University College Cork, Cork |
| 15.00-15.05 | "BRIGHT HORIZONS" A NOVEL, MULTI-DISCIPLINARY DEVELOPMENTAL CLINIC FOR HIGH RISK NEONATES BORN IN THE UNIVERSITY MATERNITY HOSPITAL, LIMERICK.  
HA Deeny 5, S Gallagher 1, J Kelleher 1, 2  
1 University Hospital Limerick, Limerick, 2 University Maternity Hospital Limerick, Limerick |
| 15.05-15.10 | PREVENTION OF EARLY ONSET GROUP B STREPTOCOCCAL DISEASE: AN AUDIT OF PROTOCOL CHANGE  
Peter A Ihidero, C Quinn, J Kelleher, R Philip, 1 Neonatology, University Maternity Hospital Limerick |
| 15.10-15.00 | QUESTIONS                                                                                   |
| 15.20-15.40 | COFFEE/MINGLING                                                                             |
| 15.45-16.45 | SESSION 7: SHORT PAPER PRESENTATIONS (Miscellaneous)  
Chairpersons: Dr Kevin Dunne, Dr John Kelleher, Dr Turlough Bolger & Prof. Eugene Dempsey |
| 15.45-15.50 | AUDIT OF CLINICAL UTILITY OF CRP IN THE MANAGEMENT OF CHILDREN PRESENTING WITH FEBRILE NEUTROPENIA  
Maria Dominguez 1, 5, L Storey 2, F Clinton 3, R Rush 4, M Capra 3, A O'Marcaigh 2, C Owens 3, O Smith 2, J Pears 3, J K O'Brien 6, R Leahy 5, P Gavin 6, K Butler 5, 1 Medical Science, University College Dublin, Dublin,  
2 Haematology, Our Lady's Children Hospital, Crumlin, Dublin,  
3 Oncology, Our Lady's Children Hospital, Crumlin, Dublin,  
4 Microbiology, Our Lady's Children Hospital, Crumlin, Dublin,  
5 Infectious Diseases, Our Lady's Children Hospital, Crumlin, Dublin,  
6 Statistica Medica, Dublin, Ireland |
| 15.50-15.55 | TRANSITION SERVICES FOR ADOLESCENTS WITH TYPE 1 DIABETES: NEW GUIDELINES, ONGOING CHALLENGES AND HOW WE ARE DOING  
L Heavey 1, R Geoghegan 1  
1 Department of Paediatrics, Galway University Hospital |
| 15.55-16.00 | 5 YEAR REVIEW OF FIRST TIME COLONISATIONS OF PSEUDOMONAS AERUGINOSA IN CHILDREN WITH CYSTIC FIBROSIS: TREATMENT AND OUTCOMES  
D Finn 1, G Leen 1, B Elnazir 1  
1 Cystic Fibrosis Unit, National Children's Hospital, Tallaght, Dublin, Ireland |
| 16.00-16.05 | TOUCH-SCREEN TECHNOLOGY USAGE IN TODDLERS: IS THERE AN APP FOR THAT?  
C Ahearne 1, Sinead Dilworth 1, R Rollings 1, S Murray 2, DM Murray 1  
1 Department of Paediatrics and Child Health, University College Cork, Cork, Ireland  
2 Hello Games, Guildford, UK |
| 16.05-16.10 | AN AUDIT OF THE USE OF FECAL CALPROTECTIN TO RULE OUT INFLAMMATORY DISEASE COLITIS IN A PAEDIATRIC POPULATION  
Barry Scanlan 1, S Lewis 2, M Mahony 3  
1 Department of Paediatrics, University Hospital Limerick, Limerick |
| 16.10-16.15 | THE POTENTIAL IMPACT OF SUSPECTED VIRAL HAEMORRHAGIC FEVER ON EMERGENCY DEPARTMENT WORKLOAD  
Dr Octav Cristiu, ED Registrar, Dr Elizabeth Little, 3rd year BST Trainee, Mr Niall O'Connor, Consultant in Emergency Medicine, Dr Mathew Varghese Consultant Paediatrician, Our Lady of Lourdes Hospital, Drogheda, Co Louth |
| 16.15-16.30 | QUESTIONS                                                                                   |
| 16.30-16.45 | CLOSING REMARKS AND AWARDING OF PRIZES/RAFFLE * (*not sponsored by Pharma)  
Dr. Kevin Dunne, Prof. Eugene Dempsey, Dr Anne-Marie Murphy, Dr Sarah Lewis and Dr Aine Lynch |

Thank you to our sponsors!!

This meeting has been supported by Abbvie, Alexion, Chiesi, Genzyme, GSK, Mead Johnston Nutrition, Nutricia, Pfizer, Shire and SMA through the purchase of exhibition trade space
TO VAPE OR NOT TO VAPE? THE EXPERIENCE OF ONE POISON CENTER WITH A WORLDWIDE PROBLEM – THE E-CIGARETTE
George C. Rodgers Jr, MD, PhD, Webb AN, Condurache, CT, University of Louisville School of Medicine, Louisville, Kentucky, USA

Background: Over the last decade e-cigarettes have become increasingly popular as a substitute for traditional cigarettes. E-cigarettes pose several new problems both from the public health perspective and from the toxicologic perspective. The technology of e-cigarettes and the current epidemiology and impact of their use will be reviewed.

Methods: The Kentucky Regional Poison Center (KRPC) serves the Commonwealth of Kentucky, with a population of about 4 million people, and some adjoining areas. The Center receives about 65,000 calls each year. Calls to the Center concerning e-cigarettes or the refill liquid for the period 1/1/2013-6/30/2014 analyzed after removal of identifiers.

Results: The first calls concerning e-cigarettes came in 2012 (9 calls). In 2013 the Center received 58 calls relating to exposure to either the e-cigarette itself or the liquid used to refill the cartridges. During the first half of 2014 the center has received 72 calls relating to e-cigarettes or the refill liquid. Descriptive and demographic analysis of the data from 2013-2014 was performed to further characterize these exposures. Of the 130 total calls, 71 (55%) occurred in children <6-years old, with 41 of these being <2-years old. Only 4 (3.1%) exposures occurred in children 6-17-years of age and the remainder (55 or 42%) occurred in people >17-years of age. Sixty-eight cases (52%) were symptomatic. The most common symptoms reported were: miscellaneous neurologic (sweating, flushing, jitteriness etc.) in 18%; nausea and/or vomiting in 12%; ocular irritation in 10%; miscellaneous cardiovascular (tachycardia, hypertension, palpitations) in 5%. Seventeen percent of cases were either in an emergency room or were referred to an emergency room. Only one patient required admission to hospital: an adult patient with preexisting medical problems who suffered a cardiac arrest after one day of ‘chain-vaping’ and ultimately died.

Discussion: Data from the KRPC mirror data from other sources and show an astounding increase in the use of e-cigarettes. The technology of these devices, the general lack of regulations concerning their packaging and sales, and the potential for very large nicotine exposures in young children are raising serious concerns about their safety and desirability. We will review existing data from a variety of sources to assess the toxicologic impact of these products on the pediatric population. As a pediatric community, we need to be aware of their potential for toxicity, and vigilant in advising families about the risks.

Conclusions: E-cigarettes pose a serious and growing worldwide threat to children. There is a need for urgent regulation of the sales of these products and paediatricians need to advocate for such changes.
TRANSFUSION OF PREMATURE INFANTS WITH BIOTINYLATED RED BLOOD CELLS (BIORBCS) DOES NOT ELICIT AN ANTIBODY RESPONSE

John A. Widness1, Ronald G. Strauss1,2, Robert L. Schmidt1, Demet Nalbant1, Svetlana Kiosseva3,4, Gretchen A. Cress1, and Donald M. Mock3,4

1Department of Pediatrics and 2Department of Pathology, Carver College of Medicine, University of Iowa, Iowa City, IA
3Department of Biochemistry & Molecular Biology, University of Arkansas for Medical Sciences, Little Rock, AR
4Department of Pediatrics, University of Arkansas for Medical Sciences, Little Rock, AR

Background. Allogeneic and autologous red blood cells (RBCs) may be reliably biotinylated ex vivo, transfused, and enumerated by flow cytometry for accurate determination of commonly used indicators of RBC kinetic parameters: RBC volume, 24 hr post-transfusion recovery, and RBC survival. The BioRBC method has been applied in adults and infants to concurrently study RBC kinetics in multiple distinct populations of BioRBCs in individual study subjects (Pediatr Res 74:592, 2013; Pediatr Res 74:689, 2013). This is easily achieved by merely varying the concentration of the biotinylating reagent. Although about 15% of adults receiving autologous BioRBCs develop antibodies (Transfusion 52:1596, 2012), there are no data for infants and children. Because of the immunologic immaturity, we hypothesized that infants are less likely than adults to develop BioRBC antibodies following BioRBC transfusions.

Study Design & Methods. Thirty premature infant subjects received allogeneic and/or autologous BioRBCs. Subject plasma was saved every 4 weeks for approximately 6 months for subsequent antibody testing using two indirect agglutination assays—a standard wet-phase Coombs test we developed (Cordle et al. Transfusion 39:1065, 1999) and a recently developed IgG gel card test (Ortho Clinical Diagnostics, MTS™ Anti-IgG Card). With the gel card method, subject plasma is incubated with both BioRBCs and RBCs (control). If specific BioRBC IgGs are present, they will bind to (“sensitize”) BioRBCs, but not to control RBCs. Cards are then centrifuged such that non-agglutinated RBCs pass through to the bottom while agglutinated RBCs are retained in a graded fashion in the gel matrix. Agglutination of sensitized cells is enhanced by the presence of anti-human IgG contained in the gel matrix.

Results. None of the 30 premature infants who were transfused with BioRBCs, developed anti-BioRBC antibodies when tested by the gel card method. Eleven of the 30 infants were also negative by standard Coombs testing. None of 39 premature infants without prior BioRBC exposure tested positive for anti-BioRBC antibodies by the gel card method.

Discussion & Conclusion. Inability of infants to produce antibodies against RBC surface antigens has been ascribed to immunologic/developmental immaturity and serves as the basis for AABB standards requiring only one pre-transfusion antibody screen before 4 mo of age. In the present infant study, failure to detect specific BioRBC antibodies among study subjects is consistent with these recommendations. Failure to detect BioRBC antibodies in the separate group of 39 BioRBC naïve infants suggests that the occurrence of naturally occurring anti-BioRBC antibodies is uncommon. We speculate that, from an immunogenicity safety perspective, BioRBCs are well suited for studies of RBC kinetics in infants.

Presenter: John (“Jack”) Widness, M.D., Univ of Iowa Children’s Hospital, Dept of Pediatrics, 200 Hawkins Drive, Iowa City, IA 52242-1083; (319) 356-8102  john-widness@uiowa.edu
A SURVEY OF BREAST FEEDING KNOWLEDGE, ATTITUDES, AND BELIEFS: PEDIATRIC RESIDENTS IN A TERTIARY HOSPITAL SETTING
L. McCarthy Clark, DNP, CPNP-AC/PC, M. Connolly, MD, L. Campfield, D.O
Stony Brook Children’s, Department of Pediatrics
Stony Brook University Medical Center, Stony Brook, NY

Purpose/Objective Breastfeeding is an important public health recommendation due to the multiple health benefits for both infants and mothers. (WHO, 1998; CDC, 2012; & AAP, 2012) Exclusive BF rates in the United States still do not meet Healthy People 2020 objectives, despite health care providers understanding of the benefits of breastfeeding. Prior studies have shown that maternal initiation and continuation of breastfeeding is a complex relationship with health care providers. Influencing factors identified include the attitudes and support put forth by the health care professionals who breastfeeding mothers encounter before and after delivery. Exploring breastfeeding knowledge and attitudes is important to achieve breastfeeding goals among breastfeeding mothers. The objective is to investigate the relationship between knowledge, attitudes and beliefs toward breastfeeding among pediatric residents in training.

Design/Methods: A descriptive survey was self-administered to pediatric residents PGY 1-4, at an academic tertiary-care medical center. Conducted with IRB approval, between March and June 2013. The survey was adapted with permission from "Nurses Support for Breastfeeding Questionnaire" originally designed by L. Bernaix (2000). The instrument consists of five subscales; all formatted using a 5-point Likert-type scale; Breastfeeding Experience: Confidence; Attitudes; Behavioral Beliefs; Impotence and Knowledge of Breastfeeding. Analysis of the data was performed by SPSS 2013 Version 22.0 for correlation between variables for significance of a p < 0.05 level.

Results: The participation rate of the pediatric residents was 72% (47/65). The survey identified the pediatric residents with more experience with breast feeding support to mothers to have rated the importance of achieving breast feeding success as very important (95.7%); extremely necessary (98%) and a positive experience (74%). Analysis of the survey identified significant support for the relationships of: experience and confidence (r: 0.406; p<0.005); and confidence and attitudes (r: 0.487;p<0.001). Knowledge and timing of last breast feeding educational program among participants yielded no significant difference as to experience; confidence; attitudes; importance; knowledge and behavioral belief score.

Conclusion/Discussion: Exploring breastfeeding knowledge, positive attitudes, and successful maternal breast feeding support is important in order to achieve breastfeeding goals among mothers who wish to breast feed. These finding may be important in future interventions to improve breastfeeding rates in hospital and exclusive breastfeeding for the first 6 months of a baby’s life. The strategies and education of breast feeding training among the pediatric residents may need to be varied with didactic, observational and hands on training to improve residence confidence and attitudes.

References:
http://www.cdc.gov/breastfeeding/data/NIS_data/index.htm
Nicholas was a 4-week-old infant when his mom brought him to the pediatrician over concern about difficulty feeding with frequent choking and noisy breathing. He was born at term and appeared to be a healthy, well-developed baby but mom noticed he was having increasing difficulty feeding. He could suck from his bottle but had to stop frequently during feeds to cough and catch his breath. At times he appeared to turn pale to dusky gray during these episodes but recovered quickly. His pediatrician noted Nicholas was stridorous on exam with mild increased work of breathing. He was quickly referred to a cardiologist and the diagnosis of a vascular ring with double aortic arch was made by CT angiography. Nicholas was an excellent surgical candidate and he was quickly taken for repair. Postsurgery his choking episodes resolved and he was left with only mild persistent stridor. Today he is more than 6 months old and his is growing and developing normally.

Vascular rings are rare defects of fetal development with only a few published case reports in the literature. Double aortic arch is one of the two most common forms of vascular ring, a class of congenital anomalies in which the trachea and esophagus are encircled by connected segments of the aortic arch and its branches. It typically presents in early infancy with symptoms such as stridor due to compression of the trachea causing upper airway obstruction. Esophageal symptoms including emesis, choking, or dysphagia are common as well.

Although the double aortic arch has various forms, the common defining feature is that both the left and right aortic arches are present. Embryologically, the aorta is formed when the ventral and dorsal aortas are connected by aortic arches. The right fourth aortic arch normally involutes and the left fourth aortic arch persists to give rise to the normal left aortic arch. The persistence of both the right and left fourth aortic arches leads to a double aortic arch. It is not uncommon for some portion of the duplicate arch to be atretic. In Nicholas’ case, a portion of left sided anterior arch was atretic and this portion was wrapped around his trachea and esophagus causing significant impingement and leading to a clear need for surgical repair.

This case of an infant with a double aortic arch is unique but highly relevant to the general pediatric audience. It is a classic presentation of a rare disorder and one that should be considered whenever an infant presents with stridor or feeding difficulty. To best illustrate this case, the audience will be shown a video of Nicholas attempting to feed from a bottle prior to his surgical repair. Furthermore, three-dimensional images of Nicholas’ CT angiogram will be shown to help clarify the anatomy of the double aortic arch.
EFFECTS OF EDUCATIONAL INTERVENTION ON FUNCTIONAL OUTCOMES OF HOSPITALIZED CHILDREN WITH ASTHMA: A RANDOMIZED-CONTROLLED STUDY

Carmen Tania Condurache, MD, MSc, Janice Sullivan, Allison Burger, Nemr Eid. University of Louisville, KY, United States.

Background: Childhood asthma is the main reason for school absenteeism and hospitalization, which could be reduced through better understanding of and compliance with the plan of care. Educational models have been published, but to our knowledge, the educational impact on functional outcomes and quality of life (QOL) has not been assessed in a randomized-controlled trial in children.

Objectives: Evaluate the impact of education on functional outcomes of children with asthma. Primary outcomes: asthma knowledge scores (AKS), i.e. proportion correct answers on asthma knowledge quiz (AKQ); and asthma knowledge retention rates (AKRR), i.e. AKS at different times compared to baseline. Functional outcomes: Quality of Life/Asthma Control Test (QOL/ACT) scores, and asthma-related missed school days. Secondary outcomes: unplanned Primary Care Physician (PCP) visits, Emergency Department (ED) visits, hospitalizations, and systemic steroid courses.

Design/methods: A randomized-controlled study assessed the AKRR, QOL, asthma-related missed school days, and unplanned medical care in children 5-12 years hospitalized for asthma. The caregivers attended a one-time education session at baseline (t₀), versus standard education at t₀ with periodic reinforcement over time. Subjects were randomized to group A (intervention) or group B (control), and the same caregiver completed the AKQ and QOL/ACT at t₀, prior to asthma education, and again by phone at 2 weeks (t₁), 1 month (t₂), and 3 months (t₃). In addition, group A received reinforced asthma education by phone at t₁, t₂, and t₃. Information on asthma-related missed school days and unplanned medical visits was collected by phone at 6 months (t₄). The PCP provided asthma-related medical records for the 6 months duration of the study.

Results: A sample size of 128 was calculated to detect a 5% effect size (difference in AKS between groups) with 80% power; 144 subjects were enrolled: 75 in group A and 69 in group B. Demographic variables were similar between groups. Repeated measures mixed effects model showed significant improvement in AKS from t₀ to t₁ in both groups (p<0.001); AKS remained above baseline at t₂ and t₃, suggesting that periodic quizzing alone may improve AKRR. The QOL scores improved significantly from t₀ to t₁ for both groups (p<0.001); the rate of improvement from t₀ to t₁ was significantly higher in the intervention group (p<0.001). The control group had a significantly higher rate of school absenteeism (HR: 1.24, 95% CI: 1.02-1.54, p=0.04) and systemic steroid use (HR: 2.25, 95% CI: 1.98-2.76, p<0.001) than the intervention group. Unplanned medical visits were not statistically different between groups.

Conclusions: 1. Education with periodic reinforcement improves parental ability to identify and avoid triggers and recognize asthma symptoms early. 2. Periodic phone calls from healthcare providers help improve caregiver compliance with the plan of care. 3. Reinforced asthma education contributes to better QOL through better control of asthma (less absenteeism and systemic steroids use in the intervention group).
AORTOPATHY (WHATEVER THAT IS) IN CHILDREN
Carol M. Cottrill,* MD  Doug Schneider, MD,*MD  and William N. O’Connor**, MD.
Departments of Pediatrics (*) and Pathology(**)
University of Kentucky, Lexington, Kentucky.

For many years, congenital disorders involving a dilated aorta were thought to have their genesis in associated abnormalities. Therefore, the aortic root with a bicuspid or stenotic aortic valve was thought to dilate secondary to shear forces as eccentric jets of blood hit the side of the aorta. The dilated aortic root in Marfan’s was explained as a poorly understood “connective tissue” disorder; in fact, 30 years ago clinicians taught that about 10 years elapsed between the first audible murmur of aortic insufficiency and dissection.

In the last 10 years, there has been a paradigm shift in our understanding of dilation of the aortic root. Rather than mechanical forces, we have come to understand that aortic dilation is a primary intrinsic abnormality of the aorta itself. This shift has been mostly clinically driven, as methods of imaging have become more available and longitudinal follow-up of patients has yielded information about the changing aorta. Along with this information, identification of genetic disorders and their concomitant biochemical defects has further confirmed the aorta itself as the culprit in many of these diseases and offer drug treatment options.

This talk will try to put the various conditions associated with a dilated aorta into perspective with respect to associated findings that allow accurate diagnosis and risk stratification for dissection. Children with some diseases with dilated aortas do not dissect, while others are at risk for progressive dilation and eventual sudden death due to dissection. Understanding of the disease process and prevention of dissection is the primary goal of most of the new “Aortopathy” clinics that many large pediatric cardiac programs now have.

The role of the general pediatrician to notice subtle abnormalities that will alert one to read about or refer such patients is crucial. Pediatricians are usually the first to suspect Turner and Noonan phenotypes and refer them for work-up. Many of the genetic diseases so far found with aortic dilation are autosomal dominant and local pediatricians are in an excellent position to look at and talk about other family members. A history of premature deaths must be carefully explored. Information is often available from Forensic offices who investigate sudden early deaths. Usually one can identify a family member who is the reservoir for such information.

Only by cooperation between local health care givers, pediatric cardiologists, geneticists and other scientists will the biologic basis of aortic dilation and dissection be fully understood to the point of safety for all of our children.
INCREASED INTRACELLULAR GRANZYME B AND PERFORIN LEVELS IN TRACHEAL ASPIRATE LYMPHOCYTES OF PRETERM INFANTS WHO DEVELOP SEVERE BRONCHOPULMONARY DYSPLASIA

Rita Marie Ryan, MD, Melissa A. Micallef, MD, James R. Yawn, BA, Frances R. Koch, MD, John E. Baatz, PhD, Jennifer K. Mulligan, PhD, Medical University of South Carolina, Charleston, SC, USA

Background: Inflammation is central to the pathogenesis of bronchopulmonary dysplasia (BPD). Little attention has been given to lymphocyte responses and the chronic inflammation phase of BPD. Additionally, the role of the cytotoxic protease, granzyme B (GrB), and perforin have not been studied in this disease.

Objective: To determine what role, if any, cytotoxic lymphocytes play in the immunopathogenesis of mechanically ventilated preterm infants at risk for BPD, and determine if a relationship exists between clinical outcomes and these markers of inflammation directly in the lung.

Methods: Tracheal aspirates (TAs) of 13 mechanically ventilated preterm infants were prospectively collected after 7 days of life. Immune cell phenotyping was conducted by intracellular immunostaining with flow cytometric analysis. Total secreted GrB in TA samples was measured by enzyme-linked immunosorbent assay (ELISA).

Results: T-lymphocytes were identified easily in human preterm infants who were still mechanically ventilated at day 7-14. At day 7-14, TA GrB and total protein were correlated. A higher percentage of day 7-14 TA live cells with cytotoxic factors GrB and perforin were associated with a higher number of respiratory support (mechanical ventilation or continuous positive airway pressure (CPAP)) days) for GrB (p=0.048; for perforin p=0.037). The percentages of TA live cells positive for GrB and perforin were also positively correlated with the severity of BPD, severe vs. moderate (p=0.0286 for both GrB and perforin). There was no correlation with day 7-14 TA T-lymphocyte (%CD3), % CD4, or % CD8 and later severity of BPD. All 13 infants who were ventilated at 7-14 days and enrolled in this study developed either moderate (n=5), or severe (n=5) BPD; or died (n=3) before 36 weeks corrected gestational age. Both NK cells and CD8+T-cells were responsible for the production of GrB and perforin, with neither cell type dominating production.

Conclusion: T-lymphocytes that produce granzyme B and perforin are found in the tracheal aspirates of preterm ventilated infants. Preterm infants who develop more severe lung disease had a significantly higher percentage of TA live cells with GrB and perforin early in the course of their lung disease. These lymphokines may play a role in a possible mechanism of lung injury in premature infants who develop BPD.
CASE REPORT: LIMITED SUCCESS OF CORMATRIX IN SURGICAL REPAIR OF SEVERE TRUNCUS ARTERIOSUS
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Introduction: The clinical and post surgical course of an infant girl with complicated Truncus Arteriosus and severe truncal valve dysplasia who died unexpectedly 79 days postoperative is described.

Background: Commercially produced decellularized porcine intestinal submucosa (Cormatrix) has been successfully used as a non-calcifying alternative to harvested pericardium for patch repair in cardiac surgery. Once implanted, this artificial extracellular matrix (ECM) is designed to facilitate seeding of the patient’s stem cells with ingrowth of native capillary supply from the adjacent heart wall to form new tissue. Recently, Cormatrix has been employed to fashion right and left heart valves and conduits.

Clinical Summary: Born at 38 weeks of gestation, a fetal diagnosis of Truncus arteriosus was confirmed by postnatal echocardiogram with Type II pulmonary arterial origin, dysplastic truncal valve with regurgitation and severe stenosis, unrestricted VSD, severe biventricular hypertrophy, and a secundum atrial septal defect. Complete surgical repair was undertaken at a well respected congenital heart center at 5 days of life. This comprised GoreTex patch VSD closure, RVOT muscle resection, and right ventricle to pulmonary artery 12 mm Cormatrix valved conduit, truncal valve reconstruction with Cormatrix 9 mm conduit, and primary secundum ASD repair. The intraoperative and post operative course was challenging as expected in this high risk neonate. By postoperative day (POD) 13, decreased biventricular function, elevated right sided heart pressure, tricuspid valve regurgitation and moderate truncal valve regurgitation was present, but she improved enough for discharge on POD 27. She subsequently thrived well at home for the next six weeks. However, on POD 79, acute onset of shortness of breath over a short period was followed by bradycardia and sudden cardiac death.

Autopsy Findings: Mother kindly gave permission for limited examination with procurement of relevant cardiac tissue sampling. Grossly, the postsurgical, globoid, enlarged 72 gm heart had biventricular hypertrophy and surprisingly extensive ongoing acute myocardial infarction demonstrated by subendocardial mottling and tissue retraction. A dilated conduit from right ventricle to the pulmonary arteries was virtually devoid of valve tissue guarding the outflow. The neo-aortic, formerly truncal valve was severely incompetent due to two poorly developed foreshortened thick leaflets, while the third leaflet was 50% lacking. Microscopically, all neo-valves had limited residual collapsed porcine ECM collagen central core. Inflammation with macrophages, lymphocytes and eosinophils at the ECM interface was covered by a thick layer of overlying spindly cellular neointima. This is consistent with a recently described reaction to Cormatrix with overlying fibrous reaction seen in some cases. The LV infarction histology ranged from 2 weeks to under 24 hours.

Discussion: Pathologic evaluation is consistent with sudden cardiac death due to ongoing acute myocardial infarction. Recognizing the exposure to significantly abnormal hemodynamics, Cormatrix used surgically to form a valved conduit between right ventricle and pulmonary arteries, and new left sided semilunar valves leaflets in this case of Truncus arteriosus demonstrated a newly described pathology with future clinical implications.