

Thursday 09.10-09.20

Neonatal

**EXCLUSIVE HUMAN MILK DIET (EHMD) FOR EXTREMELY PREMATURE INFANTS: NOVEL FORTIFICATION ENHANCING THE BIOACTIVITY OF FRESH, FROZEN AND PASTEURISED MILK.****RK Philip**<sup>1,2</sup>, E Romeih<sup>3</sup>, E Bailie<sup>4</sup>, M Daly<sup>5</sup>, KD McGourty<sup>6</sup>, AM Grabrucker<sup>7,8,9</sup>, CP Dunne<sup>2,10</sup>, G Walker<sup>3</sup><sup>1</sup>Division of Neonatology, Department of Paediatrics, University Maternity Hospital Limerick, Limerick,<sup>2</sup>Division of Neonatology, Department of Paediatrics, University of Limerick School of Medicine, Limerick,<sup>3</sup>Solid State Pharmaceutical Centre (SSPC), The Science Foundation Ireland, Bernal Institute, University ofLimerick, Limerick, <sup>4</sup>Human Milk Bank, Western Health & Social Care Trust, Enniskillen, Northern Ireland, <sup>5</sup>Advocacy and Policymaking, Irish Neonatal Health Alliance (INHA), Dublin, <sup>6</sup>Department of Chemical Sciences, Bernal Institute, University of Limerick, Limerick, <sup>7</sup>Department of Biological Sciences, University of Limerick, Limerick, <sup>8</sup>Health Research Institute (HRI), University of Limerick, Limerick, <sup>9</sup>Centre for Interventions in Infection, Inflammation and Immunity (4i), University of Limerick, Limerick

**Background and Aims:** Human milk (HM) fortification has been recommended for the nutritional optimisation of very low birth weight (VLBW) infants. This study analysed the bioactive components of HM and evaluated fortification choices that could accentuate or attenuate the concentration of such components, with special reference to human milk derived fortifier (HMDF) offered to extremely premature infants as an exclusive human milk diet (EHMD).

**Materials and Methods:** An observational feasibility study analysed the biochemical and immunochemical characteristics of mothers' own milk (MOM), both fresh and frozen, and pasteurised banked donor human milk (DHM), each supplemented with either HMDF or cow's milk-derived fortifier (CMDf). Gestationspecific specimens were analysed for macronutrients, pH, total solids, antioxidant activity,  $\alpha$ -lactalbumin, lactoferrin, lysozyme,  $\alpha$ - and  $\beta$ -caseins. Data were analysed for variance (ANOVA) applying general linear model (GLM) and Tukey's test for pairwise comparison. University of Limerick Hospitals' research ethics committee approved the study.

**Results:** DHM exhibited significantly low ( $P < 0.05$ ) lactoferrin and  $\alpha$ -lactalbumin concentrations than fresh and frozen MOM. HMDF reinstated lactoferrin and  $\alpha$ -lactalbumin and exhibited higher protein, fat and total solids ( $P < 0.05$ ) in comparison to unfortified and CMDf supplemented specimens. HMDF fortification offered significantly high ( $P < 0.05$ ) antioxidant activity, suggesting potential capability of HMDF to enhance oxidative scavenging.

**Conclusion:** DHM, compared to MOM, has reduced bioactive properties and CMDf conferred the least additional bioactive components. Reinstatement and further enhancement of bioactivity,

which has been attenuated through pasteurisation of DHM, is demonstrated through HMDF

supplementation. Freshly expressed MOM fortified with HMDF and given early (*3E- early, enterally, exclusively*) appears an optimal nutritional choice for extremely premature infants.

1. Chawanpaiboon S, Vogel JP, Moller AB, et al. Global, regional, and national estimates of levels of preterm birth in 2014: a systematic review and modelling analysis. *Lancet Glob Health* 2019;7(1):e37-e46. 2. Eidelman AI. Breastfeeding and the Use of Human Milk: an Analysis of the American Academy of Pediatrics 2012 Breastfeeding Policy Statement. *Breastfeeding Med* 2012;7:323c4. 3. Committee on Nutrition, Section on Breastfeeding, Committee on Fetus and Newborn: Donor Human Milk for the High-Risk Infant: Preparation, safety, and Usage Options in the United States. *Pediatrics* 2017;139(1). 4. WHO. Donor human milk for low birth-weight infants. July 22, 2019. [https://www.who.int/elena/titles/donormilk\\_infants/en/](https://www.who.int/elena/titles/donormilk_infants/en/) (accessed 14th March 2022). 5. Jensen GB, Ahlsson F, Domellöf M, et al. Nordic study on human milk fortification in extremely preterm infants: a randomised controlled trial—the N-forte trial. *BMJ Open* 2021;11:e053400. 6. Radmacher PG, Adamkin DH. Fortification of human milk for preterm infants. *Semin Fetal Neonatal Med* 2017;22(1):30-35. 7. Arslanoglu S, Boquien CY, King C, et al. Fortification of Human Milk for Preterm Infants: Update and Recommendations of the European Milk Bank Association (EMBA) Working Group of Human Milk Fortification. *Front Pediatr* 2019;22(7):76. 8. Perrin MT. Donor human Milk and fortifier use in United States level 2, 3 and 4 neonatal care hospitals. *J Pediatr Gastroenterol Nutr* 2018;66(4):664-9. 9. Hair AB, Rechtman DJ, Lee ML, et al. Beyond Necrotizing Enterocolitis: other Clinical Advantages of an Exclusive Human Milk Diet. *Breastfeed Med* 2018;13(6):408-411.

**Thursday 09.20-09.30**

**General Paediatrics**

### **A PROSPECTIVE FEASIBILITY STUDY OF THE IMPLEMENTATION OF SITTA METHOD IN THE MANAGEMENT OF ENURESIS**

**S Hoolahan**, S Abdellatif, N van der Spek

<sup>1</sup>Department of Paediatrics , Cavan General Hospital, Cavan

**Aims:** Determine efficacy of SITTA (Short and Intensive Treatment with one of Two Alarms) method in a clinic setting and inform the development of national guidelines for the management of paediatric enuresis.

**Methods:** Prospective 14 month feasibility study of SITTA protocol using body-worn or bell-and-mat alarms, in a Paediatric Continence Clinic with staff of variable expertise, overseen by an experienced practitioner. Primary outcome was Initial Success (IS) defined as 14 consecutive dry nights within 16 weeks of treatment. Secondary outcomes were time to initial success, Continued Success (CS) at 6 months and total number of clinic appointments. Clinical information was recorded in a dedicated electronic health record (EEF). Figures were compared to SITTA protocol used in a research environment by a single expert consultant <sup>1</sup>.

**Results:** 50 children with monosymptomatic or non-monosymptomatic enuresis were recruited between March 2021 and May 2022 and commenced on our SITTA protocol. Initial success (IS) was achieved in 88%. The average time to achieve IS was 8.5 weeks. 82% had Continued Success (CS). On average, patients required 6 clinic appointments.

**Conclusion:** Prior to the introduction of SITTA, a local low-intensity enuresis clinic achieved 59.5% IS. A previous large international study showed 76.8% IS<sup>2</sup>. Following development of SITTA protocol, a preliminary study in 2019 had 97.1% IS, conducted by a single experienced consultant<sup>1</sup>. The implementation of SITTA in a clinic with staff of variable expertise demonstrates similar favourable outcomes.

To date, no standardised protocol for the treatment of paediatric enuresis exists in Ireland. This study indicates that the SITTA method may be more effective than current low-intensity practices, and is feasible in a clinic setting.

Results of this study will be used to implement a national guideline to standardise the provision of alarmbased treatment methods for the management of paediatric enuresis in Ireland.

1. Hardiman D et al. SITTA: A prospective pre-RCT short and intensive enuresis alarm treatment quality improvement study. Irish Paediatric Association. Oral presentation 06/12/2019. <https://imj.ie/irish-paediatric-association-abstractsdecember-2019/> 2. Apos E et al. Enuresis Management in Children: Retrospective Clinical Audit of 2861 Cases Treated with Practitioner-Assisted Bell-and-Pad Alarm. J Pediatr. 2018 Feb;193:211-216.

**Thursday 09.30-09.40**  
**General Paediatrics**

**WORSENING CHILD HEALTH AND INEQUALITY? COVID-19 LOCKDOWNS AND THE WEIGHT, PHYSICAL ACTIVITY AND DIET OF CHILDREN LIVING IN BELFAST**

N KIRK<sup>1</sup>, A MCCLEAN<sup>1</sup>

<sup>1</sup>PAEDIATRICS, ROYAL BELFAST HOSPITAL FOR SICK CHILDREN, BELFAST, NORTHERN IRELAND

**Aims:**

1. Determine the impact of COVID-19 lockdowns (March 2020-June 2021) on the weight, physical activity and diet of children aged 5-11 living in Belfast.
2. Compare across socioeconomic groups by comparing children eligible for free school meals (FSME) with those who are not eligible (non-FSME)

**Methods:** A 20-question survey was issued to parents of children attending 2 primary schools in Belfast in June 2021. Schools were grouped according to their proportion of children receiving FSM and 2 schools selected randomly. School A < 10% children FSME and School B >60% children FSME. 480 paper questionnaires were distributed via schools. Anonymous data from completed questionnaires were collated and analysed.

**Results:** 220 questionnaires were returned. 66 children (30%) were FSME. 44 (20%) parents reported their child's weight had excessively increased since the start of the pandemic. This applied to 26% in the FSME group compared to 17% in the non-FSME group. 108 (49%) reported their child as less active during COVID19 lockdowns. More children in the non-FSME group (55%) were reported as less active compared to the FSME group (36%). 89 (41%) parents across the groups indicated treat food consumption by their child had increased; 43% of non-FSME group and 36% of the FSME group.

**Conclusions:** It is concerning that 1 in 5 parents reported their child's weight increased excessively during restrictions. Using free school meal eligibility as a marker for socio-economic disadvantage, the results suggest children from poorer backgrounds have been most impacted by increased weight. Despite this, more parents in the non-FSME group reported decreased physical activity and increased treats perhaps suggesting worsening future health in this group. These findings are pertinent as we emerge from the COVID-19 pandemic into a cost-of-living crisis. We need to ensure all children have access to healthy foods and opportunities for physical activity.

Thursday 09.40-09.50

General Paediatrics

***SURF2HEAL: AN INVESTIGATION INTO THE IMPACT OF OUTDOOR ACTIVITY ON THE SOCIAL BEHAVIOURS OF THOSE WITH AUTISM SPECTRUM DISORDER.***

**SNíB Ní Bhrúdair<sup>1</sup>, AD Dumitrescu<sup>1,2</sup>**

<sup>1</sup>Medicine, UCC, Cork, <sup>2</sup>Paediatrics, CUH, Cork

**Aim:** Autism Spectrum Disorder is a highly prevalent, complex, neurobehavioural condition. Those with autism have impaired social interactions, reduced communication skills, difficulties in language development, and rigid, repetitive behavior. The volume and variety of resources available to those with ASD in Ireland is lacking, particularly given the considerable population with ASD. Surf2Heal is a voluntary camp providing those with ASD the opportunity to participate in ocean-based activities. This study was carried out with the aim of comparing the social behaviours of those with ASD before and after a surf camp tailored specifically towards those with ASD.

**Methods:** The parents of participants in the camps across Ireland filled out two copies of the Social Responsiveness Scale-2 questionnaire [SRS-2]. The first was based on participants behaviour prior to the camp, and the second based on participants behaviour immediately following the camp. Results were compared using paired t-tests to ascertain if there was an impact on social behaviors, examining overall impact and five treatment variables. The variables examined were Awareness, Cognition, Communication, Motivation, and, Restricted Interests and Repetitive Behaviours.

**Results:** Statistical analysis was done at the significance level of  $p < 0.05$  of scores obtained before and after the surf camp. Of the 27 participants, 96.4% demonstrated an overall improvement. Results showed a statistically significant improvement in the Total score and in all variables measured. Within each variable between 55.6%-92.6% of participants demonstrated improvements following analysis of the results. Analysis demonstrated that all variables had a moderate to very large effect size.

**Conclusions:** The study indicated that the surf intervention had a positive effect on the social behaviors of those with ASD in all examined variables. These results highlight how valuable further development of resources, such as Surf2Heal, are in the management of ASD, advocating for

further research and investment in this area.

Thursday 09.50-10.00

General Paediatrics

**BAAP GUIDELINES FOR CHILDREN WITH HEARING LOSS – WHAT ABOUT GENETIC PANELS?**

**T Arunan**<sup>1</sup>, S Carroll<sup>1</sup>, N van der Spek<sup>1</sup>

<sup>1</sup>Department of Paediatrics and Community Child Health, Cavan General Hospital, Co. Cavan, Ireland

**Aims:** To evaluate the outcome and cost of genetic investigations in children with non-syndromic bilateral permanent childhood hearing loss (PCHL) using the 2015 British Association of Audiovestibular Physicians (BAAP) recommendations<sup>1,2</sup>. BAAP recommends two-step testing: first for Connexin, then specific single gene mutations testing if indicated. We discuss if a one-step non-syndromic genetic panel might increase diagnostic yield and reduce costs as well as sampling.

**Method:** Retrospective analysis of the outcome and cost of genetic testing of children with non-syndromic bilateral PCHL, in the Deaf and Hard of Hearing (DHH) clinic in Cavan General Hospital (CGH), between 2017-2022. Hospital charts were used to obtain genetic blood test results and hospital accounts for the costs.

**Results:** 20 children were tested for Connexin 26 and 30, as the first-step. 40.0% (8/20) had Connexin 26 mutation explaining their hearing loss. Not following the BAAP guideline, 90.0% (18/20) were also tested for mitochondrial mutations and one for Pendrin gene, as a first line genetic test, which were all negative. One child had a non-syndromic panel as the first step, because of Cookie-bite hearing loss, and was identified with Connexin 26 mutation. As a second step, one child had a panel identifying TMPRSS3. The cumulative cost over 6 years of the two step BAAP approach in the 19 patients was €45,450 (plus non-BAAP mitochondrial and Pendrin costs)



and 21 sampling. If a genetic panel was used first line, the cost would have been €29,000, saving approximately €2,700 per year with less sampling, faster results and probable higher yield.

**Conclusion:** Using a one-step non-syndromic genetic panel instead of the BAAP recommended two-step approach of genetic testing in children with non-syndromic bilateral PCHL, would result in significant savings, faster final genetic results, less sampling and probable higher yield.

1. British Association of Audiovestibular Physicians (BAAP April 2015). Guidelines for aetiological investigation into mild to moderate bilateral permanent childhood hearing impairment. [https://www.baap.org.uk/uploads/1/1/9/7/119752718/guidelines\\_for\\_aetiological\\_investigation\\_into\\_mild\\_to\\_moderate\\_bilateral\\_permanent\\_childhood\\_hearing\\_impairment.pdf](https://www.baap.org.uk/uploads/1/1/9/7/119752718/guidelines_for_aetiological_investigation_into_mild_to_moderate_bilateral_permanent_childhood_hearing_impairment.pdf) 2. British Association of Audiovestibular Physicians (BAAP April 2015). Guidelines for aetiological investigation into severe to profound bilateral permanent childhood hearing impairment. [https://www.baap.org.uk/uploads/1/1/9/7/119752718/guidelines\\_for\\_aetiological\\_investigation\\_into\\_severe\\_to\\_profound\\_bilateral\\_permanent\\_childhood\\_hearing\\_impairment.pdf](https://www.baap.org.uk/uploads/1/1/9/7/119752718/guidelines_for_aetiological_investigation_into_severe_to_profound_bilateral_permanent_childhood_hearing_impairment.pdf)

**Thursday 10.00-10.10**  
**General Paediatrics**

**INFANTS WITH HIGHER BASELINE SPECIFIC IgE TO COW'S MILK MAKE LESS PROGRESS ON LADDER-BASED TOLERANCE INDUCTION PROGRAMS.**

**J Hayden**<sup>1</sup>, YM d'Art<sup>2,3</sup>, R van Ree<sup>4</sup>, AM Byrne<sup>1,5</sup>, JO'B Hourihane<sup>1,2,3</sup>

<sup>1</sup>Children's Health Ireland, Dublin, <sup>2</sup>Paediatrics and Child Health, University College Cork, <sup>3</sup>INFANT Centre,

University College Cork, <sup>4</sup>Amsterdam University Medical Centers, AMC, Amsterdam, <sup>5</sup>Trinity College Dublin, <sup>6</sup>Royal College of Surgeons in Ireland, Dublin

**Rationale:** Cow's milk protein allergy (CMPA) is one of the major food allergens in the first year of life. Most immunoglobulin E (IgE) mediated CMPA patients become tolerant within the first few years of life. Infants with higher milk IgE levels are known to be less likely to tolerate baked milk. Cow's milk contains over 40 proteins, with caseins (nBos d 8), beta-lactoglobulin (nBos d 5) and alpha-lactalbumin (nBos d 4) being the major allergens. These components may help predict the tolerance outcome for CMPA.

Previous work by our group has demonstrated the safety and efficacy of milk ladder use in IgE-mediated CMPA in 60 infants less than 1 year of age at recruitment. We have examined the relationship of baseline sIgE to milk and milk components with progress up the 12 step iMAP milk ladder (MLP-milk ladder position) over 6 months.

**Methods:** We conducted a secondary analysis on data on 47 serum samples taken at recruitment. Samples were tested for sIgE to cow's milk and components using UNICAP (ThermoFisher Uppsala Sweden), according to standard operating procedures. SPSS Spearman's correlation testing was performed.

**Results:** Mean baseline sIgE to cow's milk was 19.1 (standard deviation 60.4). There was a negative correlation between milk IgE and milk ladder position (MLP) ( $r(8)=-0.367$ ,  $p=.01$ ) and a similar negative correlation between nBos d 8 (casein) and MLP ( $r(8)=-0.358$ ,  $p=.01$ ). There were no significant correlations between MLP and baseline nBos d 5 (beta-lactoglobulin) ( $p=.09$ ) or nBos d 4 (alpha-lactalbumin) levels ( $p=.95$ ).

**Conclusions:** Infants with higher sIgE to cow's milk make poorer progress on milk ladder-based tolerance induction programmes than infants with lower sIgE to cow's milk. Cow's milk component testing does not add additional value to this finding.

1. d'Art, YM, Forristal, L, Byrne, AM, et al. Single low-dose exposure to cow's milk at diagnosis accelerates cow's milk allergic infants' progress on a milk ladder programme. *Allergy*. 2022; 00:1–12.
2. Calamelli E, Liotti L, Beghetti I, Piccinno V, Serra L, Bottau P. Component-Resolved Diagnosis in Food Allergies. *Medicina (Kaunas)*. 2019;55(8):498.
3. Martorell-Aragonés A, Echeverría-Zudaire L, Alonso-Lebrero E, Boné-Calvo J, Martín-Muñoz MF, Nevot-Falcó S, et al. Position document: IgE-mediated cow's milk allergy. *Allergol Immunopathol (Madr)*. 2015;43(5):507-26.
4. Bartuzi Z, Cocco R, Muraro A, Nowak-Węgrzyn A. Contribution of Molecular Allergen Analysis in Diagnosis of Milk Allergy. *Current Allergy and Asthma Reports*. 2017;17.
5. Cingolani A, Di Pillo S, Cerasa M, Rapino D, Consilvio NP, Attanasi M, et al. Usefulness of nBos d 4, 5 and nBos d 8 Specific IgE Antibodies in Cow's Milk Allergic Children. *Allergy Asthma Immunol Res*. 2014;6(2):121-5.

Thursday 10.10-10.20  
General Paediatrics

**EFFECT OF FLUID TYPE AND PARENTAL PAPER DOCUMENTATION ON PAEDIATRIC ORAL FLUID CHALLENGE SUCCESS RATE : A RANDOMIZED CLINICAL TRIAL.**

**S Malik<sup>1</sup>, N Naeem<sup>1</sup>, F Khan<sup>1</sup>, J Saeed<sup>1</sup>**

<sup>1</sup>Paediatrics Department, University Hospital Limerick, Limerick, Ireland

**Aims:** Gastroenteritis, one of the most common paediatric ED presentations, requires trial of Oral Rehydration to minimize the hospital admissions burden. This study was designed to evaluate options to improve Oral Fluid

Challenge success rate;

- To determine the effectiveness of diluted apple juice (50:50 with water) versus Diorolyte.
- To determine the effectiveness of parental Paper Documentation of oral rehydration.

**Methods:** Randomized, single-blind non-inferiority Prospective trial of children aged 6 months to 14 years presenting with Acute Gastroenteritis and Mild to moderate Dehydration in Paediatric ED of University Hospital Limerick, a regional Model 3 Hospital from August 2022 till September 2022. Children who were Sick/ Suspected Sepsis or severely dehydrated were excluded . 4 groups were made on the basis of fluid choice and documentation. Participants were randomly assigned to half-strength apple juice or diorolyte and sub-randomised to whether or not they were given a leaflet enabling fluid rehydration documentation (n=70), following a departmental written guideline for oral rehydration. Children who had 2 or more vomits , not tolerating 50% of oral fluids over 1 hr or deteriorating condition were labelled as failed and admitted for IV fluids.

**Results:** Out of 70, 58 children had mild, while 12 had moderate dehydration. 25 children given dioralyte (35.71%), Apple Juice 41 (58.57%), 4 Milk (5.71%). 37(52.86%) had paper documentation and 33 Children

(47.14) did not. Overall 59 (84.29%) success rate and 11(15.71%) failure. Highest success Rate seen with

Apple Juice and Paper Documentation- 92.41%. Diorolyte and Paper Documentation had 90% Success Rate (P value>0.05)

**Conclusions:** Despite small sample size, half diluted apple juice and paper documentation by parents resulted in fewer treatment failures. So, this study recommends apple juice as a suitable rehydration fluid along with paper documentation for better outcome and further study with bigger sample size for more significant results.

1. E Umana, A Rana, K Maduemem, E Moylett; Introduction of an Oral Fluid Challenge Protocol in the Management of Children with Acute Gastroenteritis: A Regional Hospital Experience. Irish Medical Journal. Available from:<https://imj.ie/introduction-of-an-oral-fluid-challenge-protocol-in-the-management-of-children-with-acute-gastroenteritis-a-regional-hospital-experience/> 2. Stephen B. Freedman, Andrew R. Willan, Kathy Boutis, Suzanne Schuh; Effect of Dilute Apple Juice and Preferred Fluids vs Electrolyte Maintenance Solution on Treatment Failure Among Children with Mild Gastroenteritis: A randomized Clinical Trial. Pubmed. Available at:<https://pubmed.ncbi.nlm.nih.gov/27131100/> 3. D'Sa Saskia, Hussain Ahad, Hussain Mushtaq, Afridi Zahir, Twomey John, Ahmed Irfan; An audit on the implementation of administering Ondansetron in children with acute gastroenteritis and its effect on admission rate. Available at:<https://www.heighpubs.org/japch/japch-aid1025.php> 4. Gastroenteritis Acute Management. Available at:<https://www.olchc.ie/healthcare-professionals/clinicalguidelines/gastroenteritis.pdf>

**THE CORAL STUDY: DEVELOPMENTAL OUTCOME OF BABIES BORN DURING THE COVID-19 PANDEMIC**

**S Byrne**<sup>1,2,3</sup>, E O'Flaherty<sup>1</sup>, H Sledge<sup>2</sup>, S Lenehan<sup>4,5</sup>, N Jordan<sup>1</sup>, F Boland<sup>6</sup>, R Franklin<sup>1</sup>, S Hurley<sup>1,2</sup>, J McHugh<sup>1</sup>, D Murray<sup>5</sup>, J Hourihane<sup>1,2,5</sup>

<sup>1</sup>Paediatrics, Children's Health Ireland, Dublin, <sup>2</sup>Paediatrics and Child Health, Royal College of Surgeons Ireland, Dublin, <sup>3</sup>FutureNeuro SFI Research Centre, Royal College of Surgeons Ireland, Dublin, <sup>4</sup>Paediatrics and Child Health, University College Cork, Cork, <sup>5</sup>INFANT Research Centre, University College Cork, Cork, <sup>6</sup>Data Science Centre, Royal College of Surgeons Ireland, Dublin

**Introduction:** Early in the Covid-19 pandemic there was speculation about the impact of mass isolation and mask-wearing on childrens' development.

**Aims:** To look at developmental outcomes in a pandemic birth-cohort.

**Methods:** The CORAL study(Impact of **Corona** Virus Pandemic on **Allergic** and Autoimmune Dysregulation in Infants Born During Lockdown) is a longitudinal prospective observational study of Irish infants born into the pandemic(1-3). At 12- and 24-months of age, parents completed developmental questionnaires [developmental milestones, Ages-and-Stages(ASQ), and Language Development Survey (LDS)]. Results were compared to the BASELINE birth cohort(4). A subset of infants were assessed at 18-months using eyetracking technology(Tobii Pro/X3-120), to look at gaze preference in masked and unmasked adults.

**Results:** 365 infants born between March and May 2020 were recruited to the CORAL study. At 12-months we compared milestones for CORAL and BASELINE cohorts; in logistic analyses CORAL cohort had significant differences in having one definite and meaningful word, pointing and crawling compared to the BASELINE cohort. At 24-months of age infants in the CORAL and BASELINE cohort had similar developmental outcomes in all domains on ASQ (gross motor, fine motor, problem solving, personal and social) except communication where children from the CORAL cohort were more likely to fall under standardised cut-offs. In addition, children born in the pandemic cohort had on average 140 words on the LDS compared to an average of 177-184 in previously reported birth cohorts(5, 6). In the eye-tracking task when parents or strangers wore a mask, infants demonstrated less interest in the mouth.

**Conclusion:** Fewer Pandemic-born infants have achieved communication milestones at 12- and 24-months compared to the BASELINE cohort. 18-month-old infants paid little interest in the mouth region when looking at masked adults. Further work will determine if mask wearing is linked to language attainment. Universal developmental screening is extremely important for pandemic-born infants.

1. Lawler M, Franklin R, McCallion N, Byrne AM, Fitzsimons J, Kinoshita M, et al. The impact of COVID-19 lockdown on infants' coronavirus exposure and routine healthcare access in Ireland: The CORAL birth cohort study at 6 months. *Pediatr Allergy Immunol.* 2021;32(8):1876-9. 2. Hurley S, Franklin R, McCallion N, Byrne A, Fitzsimons J, Byrne S, et al. Allergy-related outcomes at 12 months in the CORAL birth cohort of Irish children born during the first COVID 19 lockdown. *Pediatric Allergy and Immunology.* 2022;33. 3. Sledge H, Lawler M, Hourihane J, Franklin R, Boland F, Dunne S, et al. Parenting a newborn baby during the COVID-19 pandemic: a qualitative survey. *BMJ Paediatrics Open.* 2022;6(1):e001348. 4. O'Donovan SM, Murray DM, Hourihane JO, Kenny LC, Irvine AD, Kiely M. Cohort profile: The Cork BASELINE Birth Cohort Study: Babies after SCOPE: Evaluating the Longitudinal Impact on Neurological and Nutritional Endpoints. *Int J Epidemiol.* 2015;44(3):764-75. 5. Rescorla L, Alley A. Validation of the language development survey (LDS): a parent report tool for identifying language delay in toddlers. *J Speech Lang Hear Res.* 2001;44(2):434-45. 6. Klee T, Carson DK, Gavin WJ, Hall L, Kent A, Reece S. Concurrent and predictive validity of an early language screening program. *J Speech Lang Hear Res.* 1998;41(3):627-41.

**Oral Presentation – Thursday 10.30-10.40**  
**General Paediatrics**

**PAEDIATRIC STABBINGS – AN EMERGING PUBLIC HEALTH THREAT IN IRELAND.  
DEMOGRAPHICS, MANAGEMENT AND OUTCOMES OF DELIBERATE STABBINGS IN CHILDRENS  
HEALTH IRELAND: 2012-2022**

**F Guinness<sup>1</sup>, J Hickson<sup>1</sup>, N Beirne<sup>1</sup>, D McCollum<sup>1</sup>, D Hall<sup>2</sup>, K Synnott<sup>3</sup>, N Quinn<sup>1</sup>**

<sup>1</sup>Department of Paediatric Emergency Medicine, Children's Health Ireland at Temple Street, Dublin,

<sup>2</sup>Department of Paediatric Emergency Medicine, Children's Health Ireland at Crumlin, Dublin,

<sup>3</sup>National Office for Trauma Services, HSE, Dublin

**Aims:** To analyse deliberate stabbing incidents managed in Children's Health Ireland (CHI) over the past 10 years. We aim to highlight the changing demographics of stab wounds and report on the pre-hospital management, trauma reception and resuscitation and where available, other trauma management outcomes in this cohort.

**Methods:** A retrospective case analysis on all patients who sustained stab injuries over a 10-year period between 2012 and 2022. 'Symphony', the CHI-wide Paediatric Emergency Department (ED) system was used to identify all patients who presented with stab wounds to the emergency department (incorporating CHI at Crumlin, Temple Street and Tallaght). All patients between 2012-2022 who sustained deliberate stab wounds and were managed in CHI were included.

**Results:** 16 children sustained deliberate penetrating stab wounds requiring management in CHI. The prevalence of stab wounds has dramatically increased with 94% (n=15) of stab wounds occurring in the past

2 years, between 2020 and 2022. 60% (n=9) of the stabbing incidents occurred in the first 9 months of 2022. The age ranged from 8-18 years with an mean age of 14. 81% (n=13) were male. The thorax was most commonly affected (68.7%) followed by the abdomen (31%). 31% (n=5) required blood products in the Emergency Department. 38% (n=6) required thoracostomy/chest drains, half of which were bilateral. 6% (n=1) underwent Emergency Resuscitative Thoracotomy in the ED. 81% (n=13) of patients survived at point of discharge from the ED. There was 19% (n=3) mortality, all such patients arrived to the emergency department in Traumatic Cardiac Arrest.

**Conclusion:** Paediatric stabbing incidents have shown a marked and concerning rise in incidence over the past 2 years in particular. In addition to this becoming an emerging public health threat in Ireland, it also highlights the need for, and importance of education and training in paediatric penetrating trauma management.

**Thursday 11.40-11.50**  
**Sub-Specialty**

**REDUCING INAPPROPRIATE REFERRALS USING A STANDARDISED REFERRAL AND  
PHOTOGRAPHIC**

**ASSESSMENT AT THE NATIONAL PAEDIATRIC CRANIOFACIAL CENTRE**

MC Casey, **S Bracken**, E Holohan, L Comey, N O'Mahony, DJ Murray

<sup>1</sup>National Paediatric Craniofacial Centre, Children's Health Ireland at Temple Street, Dublin, Ireland

**Introduction:** The National Paediatric Craniofacial Centre (NPCC) provides treatment for a myriad of craniofacial conditions. Each year, approximately 700 patients are seen, including 230 new patients. An audit in 2016<sup>1</sup> showed that most referrals received queried craniosynostosis and that 40% of patients were discharged upon first review, with no craniofacial-related abnormality identified. To address this, a designated referral form was introduced. Despite this and other interventions, we receive inconsistent referral information and see increasing numbers of children presenting with head shapes that do not require craniofacial expert care. On the 1st of October 2019, a quality improvement (QI) initiative was undertaken whereby the NPCC referral form and accompanying photographs were required for all head shape referrals.

**Aim:** To evaluate the impact of standardising referrals.

**Methods:** A retrospective audit of practice was conducted, examining a year before the introduction of the QI initiative from October 2018 to September 2019 and the subsequent year, October 2019 to September 2020. Rate of discharge upon the first review, patient pathology, patient re-referral and time to review were primarily assessed.

**Results:** In the 24 months, 293 new patients were reviewed at clinic with an "abnormal" head shape. The rate of discharge upon first review reduced from 65% (n=119/182) pre- standardisation to 56% (n=62/111) post. A significant reduction of 50% was found in the number of patients reviewed in the two periods with "normal" head shapes, from 28% (n=51/182) to 14% (15/111). This group represents the inappropriate referrals seen. Following standardisation, 105 referrals were also rejected as they had no craniofacialrelated abnormality identified. Four were re-referred and subsequently reviewed with no change to the clinical diagnosis.

**Conclusion:** The introduction of a standardised referral pathway significantly impacted our clinical practice, reducing unnecessary patient review and time to review and enabling a more efficient service.

Curran, T. A., McGillivray, A., & Murray, D. (2016). A Review of Craniofacial Referrals to the NPCC and Introduction of a New Referral Pathway. *Irish medical journal*, 109(1), pp. 325.



12.00

Sub-Specialty

**ASSESSING KNOWLEDGE OF ANAPHYLAXIS MANAGEMENT AND AAI ADMINISTRATION AMONG CAREGIVERS OF CHILDREN WITH FOOD ALLERGIES: A CROSS-SECTIONAL STUDY**

C Cronin<sup>1</sup>, C O'Kelly<sup>2</sup>, L Flores Villarta<sup>1</sup>, A Gallagher<sup>1, 2</sup>, H Keohane<sup>1</sup>, E O'Rourke<sup>1</sup>, C Tobin<sup>2</sup>, J Trujillo Wurttele<sup>1, 2</sup>

<sup>1</sup>Paediatrics and Child Health , University College Cork, Cork, <sup>2</sup>Department of Paediatrics, Cork University Hospital, Cork

**Aim:** Intramuscular adrenaline with an adrenaline auto injector (AAI) is the first-line therapy for anaphylaxis (1). However, many patients and caregivers are often unable to demonstrate correct administration of the AAI, and knowledge of anaphylaxis management has been shown to be suboptimal (2). The aim of this study is to assess the knowledge of anaphylaxis management and adrenaline auto injector use among caregivers of children with food allergies.

**Methods:** This was a cross sectional study which took place in the paediatric allergy clinic at Cork University Hospital. Parents of children with food allergies who were prescribed an AAI completed an online questionnaire regarding anaphylaxis management and were assessed in the use of their child's AAI via a video call.

**Results:** 179 parents completed the online questionnaire. 61.5% had not received AAI administration training in the previous 12 months. Parents had good overall knowledge of the definition of anaphylaxis (91%), the management of allergic reactions involving the skin (92.1%) and the management of anaphylaxis with respiratory symptoms (86.5%). However, parents were less informed about the management of cardiovascular and gastrointestinal symptoms of anaphylaxis (50.2%). 111 parents were assessed in their ability to use Epipen. 20 (18%) parents correctly explained all 5 steps essential for successful AAI administration, this was largely due to failing to identify to push in and hear a 'click' from the device (65.7%).

**Conclusion:** This is the first study in Ireland to assess caregiver knowledge of anaphylaxis management and AAI administration. Parents had a good overall knowledge of anaphylaxis management, however, the ability to explain the use of an AAI was suboptimal. Further study should be carried out to identify the best educational interventions for caregivers of children with food allergies.

1. Brown, J et al (2013). A randomized maternal evaluation of epinephrine autoinjection devices. *Pediatr Allergy Immunol* 24:173–177. 2. Trujillo J, Cronin C. Benefit of educational intervention on

Autoinjector Technique for caregivers and paediatric patients with food allergies: Allergol Immunopathol (Madr). 2022 Sep 1;50(5):100–13

**Thursday 12.00-12.10**  
**Sub-Specialty**

### **ACUTE SEVERE HEPATITIS OF UNKNOWN AETIOLOGY IN CHILDREN**

**T Fallon Verbruggen**<sup>1</sup>, R Egan<sup>1</sup>, R Mannion<sup>1</sup>, E Crothers<sup>1</sup>, C Ring<sup>1</sup>, J Hayden<sup>1</sup>, L O'Halloran<sup>1</sup>, R Hayden<sup>2</sup>, E

Liddy<sup>3</sup>, C De Gassun<sup>4</sup>, S Hussey<sup>1</sup>, B Bourke<sup>1</sup>, A Broderick<sup>1</sup>, S Quinn<sup>1</sup>, C Dunne<sup>1</sup>, E Fitzpatrick<sup>1</sup>

<sup>1</sup>National Paediatric Liver Centre, CHI at Crumlin, Dublin, Ireland

**Introduction:** Acute severe hepatitis of unknown aetiology, also known as non-A to E hepatitis or seronegative hepatitis is a common cause of paediatric acute liver failure (PALF).<sup>1</sup> While true incidence of this condition is unknown, a dramatic increase in cases was first reported in Scotland on April 5<sup>th</sup> 2022.<sup>2</sup> As of September 29<sup>th</sup>, 555 cases have been reported in Europe with a total of more than 1000 world-wide.<sup>3</sup>

**Methods:** This is a retrospective review of children referred to the National Paediatric Liver Service in Ireland between January and August 2022. Children were less than 10 years of age at presentation and had a new acute hepatitis (AST and/or ALT of >500 IU/L).

**Results:** Twenty seven children (15 females) met the case definition. The majority were previously healthy children with a median age of 3 years and 11 months (interquartile range 2 years 8 months to 5 years 2 months). The most common features at presentation were jaundice (85%), lethargy (67%), abdominal pain (62%) and acholic stools or dark urine (55%). Median peak AST was 2464 IU/L (interquartile range 1597 – 4315 IU/L). 21 (78%) children were PCR positive for either adenovirus or Human Herpes Virus-7 (HHV-7) in blood at presentation. HLA subtyping in 22

children tested to date revealed an allele frequency of 34.1% for DRB1\*04:01 (Irish blood donor allele frequency is 20%) and 4.5% for DRB1\*04:07 (control 2.4%). 5 (19%) children met criteria for PALF of whom 2 required liver transplantation.

**Conclusions:** Ireland experienced a significant and unprecedented epidemic of acute severe hepatitis in children this year. The majority had either adenovirus or HHV-7 though at low levels at the time of presentation. HLA typing suggested a possible predisposition to autoimmunity in 14/22 (60%). Further studies into the pathogenesis and aetiology of this outbreak are ongoing.

1. Squires RH Jr, Shneider BL, Bucuvalas J et al. Acute liver failure in children: the first 348 patients in the paediatric acute liver failure study group. *J Pediatr* 2006; 148:652-8. 2. World Health Organisation. Acute hepatitis of unknown aetiology – the United Kingdom of Great Britain and Northern Ireland. April 15, 2022 (<https://www.who.int/emergencies/disease-outbreak-news/item/2022-DON368>). 3. World Health Organisation. Severe acute hepatitis of unknown aetiology in children - Multi-country, 524 (2022).

12.20

Oral Presentation Thursday 12.10-

Sub-Specialty

**A REVIEW OF NATIONAL REFERRALS OF INFANTS WITH CRITICAL CONGENITAL HEART DISEASE TO**

**CHILDREN'S HEALTH IRELAND AT CRUMLIN BETWEEN 2020-2021**

**ED Dunne<sup>1</sup>, AS Smith<sup>2</sup>, OF Franklin<sup>1,3</sup>**

<sup>1</sup>Paediatric Cardiology, CHI at Crumlin, Dublin, <sup>2</sup>Neonatology, Rotunda Hospital, Dublin,

<sup>3</sup>Department of Paediatrics, Trinity College Dublin, Dublin,

**Objective:** Infants born with critical congenital heart disease (CHD) require timely identification and onward referral to national cardiology services. The aim of this study was to evaluate the overall management of infants with critical CHD and to identify factors leading to paediatric intensive care unit (PICU) admission pre-intervention.

**Methods:** This was a retrospective review of infants born with critical CHD. Critical CHD was defined as a structural lesion requiring prostaglandin or urgent postnatal review. Data was obtained from patient charts and from the electronic echocardiographic database.

**Results:** Ninety-nine patients were initially identified with 73 eligible for inclusion. The mean gestational age and birthweight was 37.99 weeks and 3.054 Kg respectively. Sixty-three (86%) had an antenatal CHD diagnosis. Of those antenatally diagnosed thirteen (21%) patients were born outside of Dublin with two of these due to be delivered in Dublin. Twelve (19%) of those antenatally diagnosed were born on a Friday, Saturday or Sunday. Of those antenatally diagnosed, where data was available, thirty-one (56%) arrived to CHI within normal working hours (08.00 - 17.00). Fifty-six (77%) infants were on prostaglandin prior to transfer. Sixty-one (84%) had a cardiac procedure during their first admission. Fifteen (20.5%) deaths were identified; 3 pre-intervention and 12 post-intervention from 12 days to 16 months. Twenty-four (33%) patients were admitted to the PICU pre-procedurally. Indications for PICU admission were multifactorial with the most frequent being requirement for intubation or non-invasive ventilation, apnoea or desaturation, requirement for central intravenous access, rising lactate or pulmonary over-circulation. Eight (33%) of these infants subsequently died though not all during the initial admission.

**Conclusion:** Infants with critical CHD have a high rate of PICU admission pre-procedurally. Our data emphasises the importance of delivery planning, prompt transfer to CHI at Crumlin and close monitoring for clinical deterioration pre-procedurally in this high risk cohort.

**Thursday 12.20-12.30**  
**Sub-Specialty**

### **EFFICACY OF GLUCAGON-LIKE PEPTIDE 1 RECEPTOR AGONISTS IN CHILDREN WITH OBESITY: A SINGLE CENTRE EXPERIENCE**

**L Corley<sup>1</sup>, J Heckmann<sup>1</sup>, N McGrath<sup>1</sup>**

<sup>1</sup>Department of Paediatrics, University Hospital Galway

**Background:** Obesity is a chronic disease of increasing prevalence in children and adolescents<sup>[1]</sup>. Ireland has one of the highest rates worldwide, with 25% of Irish children classified as obese or over-weight<sup>[2]</sup>. Historically, treatments for obesity were limited, with lifestyle therapy alone yielding poor outcomes<sup>[3]</sup>. Recent evidence supports the use of GLP1-agonists as adjuncts in achieving a reduction in BMI in adolescents with obesity<sup>[4]</sup>.

**Methods:** We conducted a retrospective chart review of all patients commenced on GLP1-agonists in our centre over a twelve month period(September 2021-September 2022). Treatment was commenced in patients with BMI >30 who failed to respond to lifestyle intervention.

**Results:** Data was available on nine patients who had commenced GLP1 treatment. Three patients(33%) were treated with daily liraglutide and six patients(66%) with weekly semaglutide. The mean age was 15 years old (range 13-18 years). Within the cohort, two patients had Type 1 Diabetes(22%) and three patients had Type 2 Diabetes(33%). The mean pre-treatment weight and BMI were 117.5kg and 40.7 respectively. Follow-up data was available at three and six months on five patients. The remaining four patients had commenced GLP1 treatment within the preceding three months. At six month follow-up the mean weight loss was 9.6% of total body weight (range 2.4%–12%). Two patients discontinued treatment due to gastrointestinal side-effects. Both had deviated from the protocol with rapid up-titration and have since recommenced treatment. Of the patients with diabetes, HbA1c pre-treatment and 6 months post-treatment was available for four patients. All patients demonstrated improvement in HbA1c with a mean HbA1c of 66.5mmol/mol(8.2%) pre-treatment and 55mmol/mol(7.2%) six months post-treatment.

**Conclusion:** Early results from our cohort suggest favourable response to GLP1-agonists as reflected by a mean total weight loss of 9.6% of body weight at 6 month follow-up and improved mean HbA1c in patients with diabetes. The medication is well tolerated by our patient cohort. In adolescents with obesity who are poor responders to lifestyle intervention, adjunctive use of GLP1-agonists should be considered.

1.The GBD 2015 Obesity Collaborators. Health effects of overweight and obesity in 195 countries over 25 years. N Engl J Med 2017;377:13-27. 2.Millar, S. R., Harding, M. (2021). Exploring 12-Year trends in childhood obesity prevalence for the Republic of Ireland - a national study using survey data from 2002 and 2014. HRB open research 3.Styne DM, Arslanian SA, Connor EL, et al. Pediatric obesity — assessment, treatment, and prevention: an Endocrine Society clinical practice guideline. J Clin Endocrinol Metab 2017;102:709-757. 4.Kelly, A., Auerbach, P., Barrientos-Perez, M. and Gies, I., 2020. A Randomized, Controlled Trial of Liraglutide for Adolescents with Obesity. New England Journal of Medicine, 382(22), pp.2117-2128.

**Oral Presentation**

**Thursday 12.30-**

**12.40**

**Sub-Specialty**

## **THE EFFECT OF COVID-19 ON FOREIGN BODY INGESTION IN CHILDREN: THE LONDON EXPERIENCE**

**P Tormey<sup>1,2</sup>, A Brown<sup>1</sup>, A Parikh<sup>1</sup>, I Morrison<sup>1</sup>**

<sup>1</sup>Paediatric Emergency , Royal London Hospital, London, <sup>2</sup>Paediatric Emergency, CHI @ Tallaght, Dublin

**Introduction:** The most common foreign body (FB) ingestion are coins (61.7%), toys (10.3%), jewellery (7%) and batteries (6.8%)<sup>1</sup>. Most FB ingestions are managed conservatively without any harm. High-risk ingestions such as button batteries and magnets can cause significant morbidity and mortality. The Covid19 pandemic has had a significant impact on injury patterns in children and presentations to Paediatric Emergency Departments.

**Aims:** Our primary objective was to determine if the societal changes relating to Covid-19 resulted in an increase in higher risk FB ingestions in children and if there was a correlating increase in the need for surgical intervention.

**Results:** We recorded 173 FB presentations in our “pre-covid” cohort, accounting for 0.005% of ED presentations. There were 138 FB presentations in the “post-lockdown” cohort, accounting for 0.007% of ED presentations. There was a decrease in coin ingestions and an increase in high risk ingestions (batteries and magnets) in the post-lockdown group. In the pre-covid group 4 were admitted for observation and discharged with no intervention. Three were admitted for observation and subsequently needed surgical intervention and 12 needed emergent surgical intervention from ED. In the post-lockdown group 7 were admitted for observation and subsequently discharged. Ten were admitted for observation and subsequently needed surgical intervention and 10 needed emergent surgical intervention.

**Discussion/Conclusion:** Covid-19 resulted in a huge drop in PED attendances. Despite this, the rate of FB ingestion increased during the period. Children were spending more time at home and their parents had to balance childcare with work and other commitments. The rise of contactless payments and the reduction of coins in circulation has contributed to a large reduction in coin ingestions, a trend which persisted 12 months after the onset of the pandemic. However, there was a worrying increase in high risk ingestions, requiring surgical treatment.

1. Orsagh-Yentis, D., McAdams, R. J., Roberts, K. J., & McKenzie, L. B. (2019). Foreign-Body Ingestions of Young Children Treated in US Emergency Departments: 1995-2015. *Pediatrics*, 143(5).  
<https://doi.org/10.1542/PEDS.20181988>

Thursday 12.40-12.50  
Sub-Specialty

**SARS-COV-2 VACCINATION INDUCES VIRUS-SPECIFIC T-CELL RESPONSES IN PERIPHERAL BLOOD BUT NOT TISSUE RESIDENT MEMORY T CELL RESPONSES IN NASAL MUCOSA**

**KN McCarthy**, KHG Mills

<sup>1</sup>Immune regulation research group, School of Immunology and Biochemistry, Trinity College Dublin, Dublin

**Aims:** Tissue resident memory T-cells (TRM) represent a population of effector memory T-cells that persist in tissues, such as the skin, lung and nasal mucosa and mediate rapid immunity against re-infection.<sup>1</sup> Severe acute respiratory syndrome-coronavirus-2 (SARS-CoV-2) infection induces antigen-specific CD4 and CD8 Tcell responses in blood, however there is limited data on the impact of infection or vaccination on T-cell responses at tissue level.<sup>2</sup> The objective of this study is to investigate the possible induction of SARS-CoV-2specific TRM cells in nasal mucosa following immunization with an mRNA vaccine.

**Methods:** We enrolled healthy volunteers between 25-35 years of age due to receive a Pfizer/BioNTech SARS-CoV-2 vaccine. Blood and nasal mucosal samples were taken prior to vaccination and two-weeks post the first and second doses. Peripheral blood and nasal mononuclear cells were isolated, stimulated with a panel of SARS-CoV-2 specific peptides and antigen-specific T-cell responses were analysed by intracellular cytokine staining and flow cytometry.

**Results:** Among an mRNA vaccinated cohort (n=8), the frequency of SARS-CoV-2 Spike (S)-specific Interferon gamma (IFN- $\gamma$ -producing CD4 and CD8 was significantly enhanced in the blood two-weeks post the second vaccination, compared with pre-vaccination (p=0.02, p<0.001). In contrast, in the nasal mucosa, S-specific IFN- $\gamma$ -producing CD4 or CD8 TRM were not enhanced following vaccination. Furthermore, nucleocapsid (N)-specific CD4 or CD8 IFN- $\gamma$ <sup>+</sup> cells or IL-17 producing antigen-specific T-cells were not detected in the blood or nasal mucosa after vaccination.

**Discussion:** The results support the findings of larger published studies showing that SARS-CoV-2 infection or vaccination induces antigen-specific cytokine-producing CD4 and CD8 T cell responses in the circulation.<sup>3</sup> Despite the limitations of a small sample size, our results suggest that parenteral administration of SARSCoV-2 mRNA vaccines do not induce antigen-specific TRM responses in the nasal mucosa. These findings may in part explain the failure of current vaccines to prevent infection of the nasal mucosa.

1. Mueller, S., Mackay, L. Tissue-resident memory T cells: local specialists in immune defence. *Nat Rev Immunol* 16, 79–89 (2016). <https://doi.org/10.1038/nri.2015.3>
2. Moss, P. The T cell immune response against SARS-CoV-2. *Nat Immunol* 23, 186–193 (2022). <https://doi.org/10.1038/s41590-021-01122-w>
3. Painter, Mark M. et al. Rapid induction of antigen-specific CD4+ T cells is associated with coordinated humoral and cellular immunity to SARS-CoV-2 mRNA vaccination. *Immunity*, Volume 54, Issue 9, 2133 - 2142.e3

**Oral Presentation**

**Thursday 12.50-**

**13.00**

**Sub-Specialty**

### **PAROXYSMAL MOVEMENT DISORDERS – MORE PREVALENT AND TREATMENT RESPONSIVE THAN PREVIOUSLY BELIEVED**

**S Harvey<sup>1</sup>, S Byrne<sup>2</sup>, B Lynch<sup>1</sup>, M O'Regan<sup>2</sup>, D O'Rourke<sup>1</sup>, D Webb<sup>2</sup>, KM Gorman<sup>1</sup>**

<sup>1</sup>Department of Neurology, Children's Health Ireland at Temple Street, Dublin, <sup>2</sup>Department of Neurology, Children's Health Ireland at Crumlin, Dublin

**Aims:** Paroxysmal movement disorders (PxMD) are clinically and genetically heterogenous disorders characterised by episodic involuntary movements (dystonia, dyskinesia, chorea and/or ataxia) divided into paroxysmal dyskinesias and episodic ataxia (EA). Both may co-exist with additional co-morbidities like epilepsy and migraine. Prevalence in the paediatric population is unknown. This study aims to determine the number of children in Ireland with a PxMD and characterise clinical features.

**Methods:** Cross sectional cohort study involving retrospective chart reviews (clinical presentation, treatment response, co-morbidities, genetics, neuroimaging, electrophysiology) and phone reviews with patients and families. Study sites were CHI at Temple Street and Crumlin.

**Results:** In total, 48 patients were identified with a PxMD (EA=22, Paroxysmal dyskinesias=24, Both=2). Prevalence is thus estimated at 1/29,00 children under 18 years of age based on 2016 census results. For paroxysmal dyskinesias, median age at symptom onset was 31 months (IQR 7-75 months) with median time to diagnosis of 11 months (IQR 2-43 months). Treatment was commenced in 69%(18/26) with 56%(10/18) improving on treatment alone. For EAs, median age



at symptom onset was 21 months (IQR 14-35 months) with median time to diagnosis of 10 months (IQR 4-27 months). Treatment was commenced in 46%(11/24) with 91%(10/11) improving on medication. Across all PxMD, 46 children were genetically investigated with a confirmed cause identified in 26%(12/46). Four other children have likely disease-causing variants. Single gene testing, in many cases targeted due to family history, had the highest yield at 27%(3/11). This was followed by whole-exome sequencing at 22%(3/14) and PxMD panels 20%(6/30).

**Conclusion:** PxMD prevalence is higher than previously reported. Despite this, PxMD remain rare conditions and so there is often prolonged time to diagnosis. Appropriate genetic investigations help to identify underlying cause thus guiding treatment. With medication, a large proportion (56-91%) of our cohort demonstrated symptom improvement.

**Thursday 15.05-15.10**  
**General Paediatrics**

### **CHILD NEEDS FOR REHABILITATION THERAPY INPUT FOLLOWING NEUROLOGICAL INJURY CONTRIBUTE TO DELAYED DISCHARGE FROM ACUTE TERTIARY HOSPITALS**

**O McNerney<sup>1</sup>, J Pouch<sup>1</sup>, I Gill<sup>1,2,3</sup>**

<sup>1</sup>Acute Rehabilitation Service, CHI Temple Street, Dublin, <sup>2</sup>Rehabilitation Service, National Rehabilitation

Hospital, Dublin, <sup>3</sup>School of Medicine , University College Dublin, Dublin

**Aims:** The Rehabilitation Complexity Scale (RCS) is a measure of complexity of a patient's rehabilitation needs including basic care, level of skilled nursing and medical involvement and therapy/equipment needs. It is administered within one week of admission for patients accessing the Acute Rehabilitation Service in Children's Health Ireland at Temple Street, and fortnightly thereafter. We aimed to use the RCS to assess the healthcare needs of acute rehabilitation patients, and to identify the point at which patients admitted for 28 days or longer were suitable for discharge but remained in hospital solely to access intensive therapy input.

**Methods:** This study was conducted by retrospective review of the Acute Rehabilitation Service database which comprises clinical data recorded prospectively during patient admissions. Initial RCS scores were extracted as a measure of baseline complexity. The date at which patients were suitable for discharge from a medical/ nursing perspective was identified, as determined by an RCS 'Medical Needs' score of M0/M1 and 'Skilled Nursing Needs' score of N0/N1/N2.

**Results:** Thirteen patients were admitted for 28 days or more between June 2021 and June 2022. Total length of hospital stay ranged from 42 to 184 days with a mean average of 87 days. Length of stay after patients were fit for discharge based on RCS scores ranged from 1 to 70 days with a mean average of 28 days. Prolonged admissions accounted for a minimum of 307 extra bed days in one year.

**Conclusion:** Patients with rehabilitation therapy needs often remain admitted to CHI at Temple Street after their potential date of discharge (as determined by RCS score) solely to access therapy input at a level which cannot be provided in the community. Given the cost of inpatient admission, development of an intensive day rehabilitation service may be a cost-effective solution.

Turner-Stokes L, Scott H, Williams H, Siegert R. The rehabilitation complexity scale–extended version: detection of patients with highly complex needs. *Disabil Rehabil.* 2012;34(9):715–720.

Thursday 15.10-15.15

General Paediatrics

**PAEDIATRIC INCLUSION HEALTH: THE BEGINNINGS OF EQUITABLE HEALTHCARE FOR CHILDREN EXPERIENCING SOCIAL ADVERSITY**

**A Walsh<sup>1,2</sup>, B Joyce<sup>1</sup>, E Molloy<sup>2</sup>, C Ni Cheallaigh<sup>2,3</sup>, C Hensey<sup>1,2</sup>**

<sup>1</sup>General Paediatrics, Children's Health Ireland at Temple Street, Dublin, <sup>2</sup>Faculty of Paediatrics and Child Health, Trinity College Dublin, Dublin, <sup>3</sup>Inclusion Health, St. James Hospital, Dublin

**Background & Aims:** Paediatric Inclusion health (PIH) is model of healthcare delivery for children that seeks to overcome inequitable access to necessary services. Adverse social determinants of health (SDH) contribute to higher rates of illness and increased barriers to accessing healthcare. Our hospital began a pilot project assessing the feasibility of PIH delivery to facilitate equitable access to services and better understand SDH in clinical practice.

**Methods:** Need for PIH care was determined by the presence of adverse SDH including insecure housing, lack of secure primary care infrastructure, ethnic minorities, or vulnerable migration status. Healthcare encounters were optimised with outreach clinics, care pathways focused on specific population needs (e.g. migrant, Roma), and the inclusion of community representatives in service planning.

**Results:** Over 15 months, 125 children were referred to the PIH project, the majority from the following cohorts: homeless children, Irish Travellers, Roma and vulnerable migrants. High rates of homelessness (72%, n=90), low complete vaccination uptake (26.4%, n = 33), need for interpreters (49.6%, n=62) and less formal access to primary care (35.2%, n=44) were notable findings. Attendance at first appointment was 76.8% (n=96), 8% (n=10) cancelled due to illness. Of the 15.2% (n=19) who did not attend, 52.6% (n=10) attended a rescheduled appointment.

**Conclusion:** A PIH model for children experiencing social deprivation enabled us to better understand their individual and population needs. Tailoring healthcare provision to overcome barriers and enable equitable access has developed pathways of care that facilitate attendance and further our understanding of the most marginalised children in our society.

Thursday 15.15-15.20

General Paediatrics

**CRISIS, CONFLICT AND CHILDREN'S HEALTH: THE CHILDREN'S HEALTH IRELAND (CHI) RESPONSE TO THE UKRAINIAN CONFLICT.**

**J McGirr<sup>1</sup>, A Walsh<sup>1,2</sup>, E Molloy<sup>2</sup>, C Ní Cheallaigh<sup>3,4</sup>, I Okafor<sup>1</sup>, C Hensey<sup>1,2</sup>**

<sup>1</sup>General Paediatrics Department, CHI at Temple Street, Dublin, <sup>2</sup>Faculty of Paediatrics and Child Health,

School of Medicine, Trinity College Dublin, Dublin, <sup>3</sup>Inclusion Health, St. James's Hospital, Dublin, <sup>4</sup>Clinical Medicine, School of Medicine, Trinity College Dublin, Dublin

**Aims:** Since the 24th February 2022, Ireland has experienced an unprecedented influx of refugees. At the end of August 2022 there were an estimated 12,042 Ukrainians under 18 claiming refugee status in Ireland, many with known underlying pathologies. CHI Global Migrant Health Response (MHR) was created to support Ukrainian children and colleagues in accessing and providing healthcare respectively.

**Methods:** MHR identified additional logistical and social considerations needed to support Ukrainian children to engage with an unfamiliar health service. This was an iterative process to facilitate hospital engagement, access to community resources, obtaining necessary medications and ensuring continuity of care.

**Results:** As of 17th July 2022, 495 Ukrainian children had registered in CHI sites under the Ukrainian Temporary Protection Directive (UKTPD). 48.5% (n=227) were acute, unplanned presentations. The remainder required scheduled care, often for complex and significant pathologies. Co-operation with community health and social care professionals (HSCPs) was integral to providing comprehensive care, especially when primary health infrastructure was not established. A resource repository was compiled, including information pertaining to immunisation, prescribing, translation and psychological support. Adaptations to existing services were made involving the provision of additional outpatient appointments, protected phlebotomy slots and transport support.

**Conclusion:** This dedicated response continues to coordinate hospital care for Ukrainian refugees across CHI sites. Multi-agency collaboration led by MHR has enabled comprehensive care for those with complex pathologies. This is the first example nationally of a hospital level service for vulnerable migrant children, the tactics employed will inform the strategic development of CHI-led migrant child health care.

Thursday 15.20-15.25  
General Paediatrics

**HEEADSSS UP – REVIEW OF USE AND DOCUMENTATION OF AN ADOLESCENT PSYCHOSOCIAL RISK**

**ASSESSMENT INSTRUMENT IN AN IRISH UNIVERSITY TEACHING HOSPITAL**

**MS Smyth<sup>1</sup>, EG Gordon<sup>2</sup>**

<sup>1</sup>Department of General Paediatrics, Our Lady of Lourdes Hospital, Drogheda, Louth, <sup>2</sup>Department of General Paediatrics, Our Lady of Lourdes Hospital, Drogheda, Louth

**Background:** The HEEADSSS assessment is a psychosocial screening tool the World Health Organisation recommends should be used in all clinical adolescent encounters (1, 2). To date, there is no Irish data on the use of this screening tool. The aim of this review was to establish the compliance with HEEADSSS in an Irish context and establish the efficacy of a proforma.

**Methods:** A retrospective chart review was carried out on all adolescent medical presentations to Drogheda emergency department (ED) over a one month period. A standardised data collection form was used to capture use of the HEEADSSS assessment. Next, an anonymous survey was carried out by staff (n=30) to identify knowledge deficiencies. Following result analysis and presentation, an education session was facilitated. A blank HEEADSSS proforma was implemented in clinical notes of all adolescent patients presenting to ED, attached at time of nursing triage. One month post implementation, a retrospective review was on a sample of charts to assess adherence.

**Results:** In total, 103 charts were reviewed and 6.7% (n=7) had a complete HEEADSSS assessment documented, with 57.1% (n=4) using the recommended format. An incomplete assessment was documented in 27% (n=28). Deliberate self-harm represented 14% (n=15) of presentations and only 4 of these had partial or complete HEEADSSS assessment. There was a 63% (n=19) response rate to the anonymous survey. Of those surveyed, 84% (n=16) denied receiving any training on use of HEEADSSS and cited this as a primary obstacle. In the post intervention phase 50 charts were reviewed and 64% (n=32) of HEEADSSS were appropriately completed; an improvement of 57.3%.

**Conclusions:** The implementation of a proforma, together with staff education and training on use of the HEEADSSS assessment, led to an increase in use of a comprehensive medical review of adolescent patients. Consequently, there is greater opportunity for crucial intervention.

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**Thursday 15.25-15.30**  
**General Paediatrics**

#### **A CHILD CENTRED PHLEBOTOMY SERVICE FOR CHILDREN WITH AUTISM SPECTRUM DISORDER**

**A Gallagher<sup>1,2</sup>, J Lyons<sup>1</sup>, C Chawke<sup>1</sup>, A O'Brien<sup>1</sup>, S Richardson<sup>1</sup>, U Murtagh<sup>1</sup>**

<sup>1</sup>Children's Health Ireland, Connolly Hospital, Dublin, <sup>2</sup>Department of Paediatrics and Child Health, University College Cork

**Aims:** The aim of the service is to have equal access to phlebotomy services for children with autism spectrum disorder (ASD) in order to reduce trauma, aid diagnosis and support access to medications.

**Methods:** Referrals are collated at central referrals at Children's Health Ireland. Suitable children are contacted by the team to discuss the child's specific needs and interests. A social story and blood sampling pack (needle removed) are sent to the family. Desensitisation appointments are offered during which blood sampling can take place with or without nitrous oxide.

**Results:** 41 children attended the clinic, 2 children were excluded as they were not suitable for the clinic. All children had a diagnosis of mild to moderate autism and were unable to have successful blood sampling on previous occasions. A successful outcome is every positive step a child takes towards blood sampling. 87% had successful visits with 46% having successful blood sampling. 31% used Entonox (50/50 mix of nitrous oxide and oxygen). Parental feedback was overwhelmingly positive (94.4%).

**Conclusion:** Children with ASD have similar medical needs to their neurotypical peers however sensory sensitivities and overstimulating environments can make minor medical procedures such as phlebotomy particularly difficult. This phlebotomy service allowed children with autism to have blood sampling with minimal distress and should be continued and expanded nationwide.



**STABILITY OF APPETITIVE TRAITS AND DIET QUALITY IN CHILDREN AGED 5 AND 9-11 YEARS OLD: FINDINGS FROM THE ROLO LONGITUDINAL BIRTH COHORT STUDY.**

**A Delahunt**<sup>1</sup>, SL O'Reilly<sup>1,2</sup>, FM McAuliffe<sup>1</sup>

<sup>1</sup>UCD Perinatal Research Centre, University College Dublin, Dublin, <sup>2</sup>School of Agriculture and Food Science, University College Dublin, Dublin

**Aim:** Establishing how appetitive traits track from early to later childhood, and how this impacts diet quality has not been extensively researched. We explored change in appetitive traits and impact on diet quality in a cohort of 5 year olds and 9-11 year olds.

**Methods:** This is secondary analyses of the ROLO longitudinal birth cohort study. Data included 306 mother-child dyads from the 5 year and 224 from the 9–11-year follow-up. Children's appetitive traits were measured using the Children's Eating Behaviour Questionnaire (CEBQ). Dietary quality was measured using the Healthy Eating Index (HEI-2015). Paired t-tests and linear mixed models determined change in appetitive traits from 5 to 9-11 years old. Multiple linear regression examined associations between child appetitive traits and HEI.

**Results:** Mean (Standard Deviation) scores for 'Emotional Overeating' (1.63 (0.51) vs 1.99 (0.57),  $p < 0.001$ ) and 'Enjoyment of Food' (3.79 (0.72) vs 3.98 (0.66)  $p < 0.001$ ) increased from aged 5 to 9-11 years old. 'Desire to Drink', 'Satiety Responsiveness', 'Slowness Eating' and 'Food Fussiness' decreased from 5 years to 9-11 years old. Boys had higher mean scores for 'Food Fussiness' at 5 years old. Mean scores for 'Food Responsiveness' and 'Desire to Drink' were higher in boys and 'Satiety Responsiveness' was higher in girls at 9-11 years old. 'Food Responsiveness', 'Emotional Overeating' and 'Enjoyment of Food' were positively associated with higher quality diet and 'Desire to Drink', 'Satiety Responsiveness' and 'Food Fussiness' were negatively associated with diet quality at 5 years old. 'Enjoyment of Food' was positively and 'Desire to Drink' and 'Food Fussiness' were negatively associated with diet quality at 9-11 years old.

**Conclusion:** Aspects of food approach appetitive traits increased over time, whereas food avoidant appetitive traits decreased over time. Some food avoidant appetitive traits were associated with lower quality diet.

**AUDIT OF COMPLIANCE AND FINDINGS OF DMSA SCAN IN CHILDREN WITH URINARY TRACT INFECTIONS COMPARED TO NICE GUIDELINES**

**I Dafalla**<sup>1</sup>, T Andijani<sup>1</sup>, M Ryan<sup>2</sup>, M Nadeem<sup>1</sup>



<sup>1</sup>General Paediatrics, CHI at Tallaght, Dublin, <sup>2</sup>Radiology, CHI at Tallaght, Dublin, Ireland

**Aim:** Urinary Tract Infection (UTI) is a common cause of hospital admission in children. Follow-up imaging such as ultrasound (US), Micturating cystourethrogram, and Dimercaptosuccinic acid scintigraphy (DMSA) [1-4] was illustrated in numerous guidelines. This study aimed to examine whether the guidelines in terms of follow-up imaging were followed and to investigate the relationship between DMSA and the US results, recurrent or atypical UTI, age, or length of hospital stay.

**Methods:** A retrospective review of the imaging of all children admitted with UTI from 01/08/2021 to 01/08/2022 was conducted. UTI was defined as per the guideline [4]. Atypical and recurrent UTIs were defined per NICE guidelines.[2]

**Results:** We examined 66 patients admitted with UTI, mean (SD) age 34 (43.83) months. Mean (SD) LOS was 4.18 (2.43) days. Of 66 patients, 23 (34.85%) had recurrent UTI and 1 (18.2%) exhibited atypical UTI, 26 (39.4%) had normal US and 13 (19.7%) had abnormal DMSA.

There is a significant association between abnormal DMSA and abnormal US imaging (p-value 0.02) but not pyelonephritis or normal ultrasound. Of 13 patients with abnormal DMSA, 6 (46.2%) had underlying abnormalities shown by ultrasound, such as (Bifid, duplex kidneys, hydronephrosis, or size discrepancy). Moreover, abnormal DMSA was associated with atypical UTI (p-value 0.001) and recurrent UTI (p-value 0.02). Notably, of 13 patients with abnormal DMSA, 9 (69.23%) and 8 (61.54%) had atypical UTIs and recurrent UTIs, respectively. However, no association has been reported between abnormal DMSA and age at presentation or length of hospital stay (p-value > 0.05).

**Conclusion:** In conclusion, the current guidelines were followed in this cohort of patients with UTI. In children with UTI, follow-up DMSA is recommended particularly in children with recurrent or atypical UTIs or those with abnormal ultrasounds. These results highlight the importance of following current guidelines regarding imaging following UTI in children.

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**AUDIT OF COMPLIANCE WITH BRAIN IMAGING IN SYMPTOMATIC HEAD INJURED CHILDREN PRESENTED TO ED, COMPARED TO CHALICE AND PREDICT GUIDELINES**

**A Khaky**, I Dafalla, H Azam, I Krebit.

<sup>1</sup>Emergency Department, CHI at Tallaght, Dublin , Ireland

**Aim:** Head injuries are one of the most frequent causes for children to visit the Paediatric emergency department. Most of these head injuries are benign and brain imaging is usually unnecessary. CHALICE and PREDICT clinical guidelines were both designed to guide clinicians who care for children with head injuries. The aim of this Audit is to look at our compliance with CHALICE guidelines, and to compare the indications and findings if the PREDICT guidelines were used instead of CHALICE.

**Methods:** Data were collected from computerized patient charts reviewing the history and physical exam findings from Symphony program, and reviewing CT brain findings from NIMIS program. Patients presented with head injury requiring Computerised Tomography (CT) of the brain were included. The study period was from 01/01/2021 to 31/12/2021.

**Results:** During the study period, 1494 patients attended Paediatric ED with head injury. Of these, 87 patients underwent brain CT (6%). Male patients comprises two third, mean age of 12 years (IQR: 7-14) and female patients were third, mean age 7 years (IQR: 7-11). Almost two third of these 87 patients (63%) met the indication criteria for brain CT according to CHALICE guidelines, eleven patients of these has positive findings (12%). In the remaining 32 patients who didn't meet the criteria for brain imaging, the CT brain was reported as normal. PREDICT indication criteria were met in 49 Patients (56%). Of these, 11 patients had positive findings. The remaining 37 patients in whom brain imaging was not indicated, all of them has normal results. The positive findings were mostly skull fractures and extracranial haematomas.

**Conclusion:** Both CHALICE and PREDICT guidelines are sensitive in picking the right patients for brain imaging. Deviating from these guidelines resulted in unnecessary brain imaging in almost one-third of the patients. Therefore, strict compliance with these guidelines prevents unnecessary exposure to radiation.

[www.olchc.ie/healthcare-professionals/clinical-guidelines/mild-moderate-tbi.pdf](http://www.olchc.ie/healthcare-professionals/clinical-guidelines/mild-moderate-tbi.pdf)

[www.rch.org.au/clinicalguide/guideline\\_index/Head\\_injury/](http://www.rch.org.au/clinicalguide/guideline_index/Head_injury/)

## **AN EVALUATION OF THE PORTIUNCULA PAEDIATRIC ADVICE LINE (P-PAL) SERVICE**

**K O'Neill<sup>1</sup>, F Neenan<sup>1</sup>, P Cahill<sup>1</sup>**

<sup>1</sup>Paediatric Department, Portiuncula University Hospital, Ballinasloe, Ireland

**Introduction:** Bridging the gap between primary care and hospital services remains a challenge. Lengthy waiting lists create a barrier to timely patient care. The paediatric consultants at Portiuncula commenced a Portiuncula Paediatric Advice Line (P-PAL) initiative in September 2021 to offer general practitioners(GPs) and community medical doctors(CMDs) daily phone access to a consultant paediatrician. The aim of our study was to evaluate the P-PAL service six months after implementation.

**Methods:** A survey was distributed to GPs and CMDs serving the Portiuncula catchment area. A second survey was distributed to the paediatric consultants. Anonymous data were collected using an online survey tool. Patient outcome data were collated for the total calls received in the first six months of the project.

**Results:** Fifty-six calls were received in the first six months. 21(37.5%) were managed by phone with educational material and did not require attendance at the hospital. 15(26.8%) were directed to outpatient services. 10(17.9%) were seen on the paediatric day unit and deemed semi urgent. 10(17.9%) were referred to ED.

Twenty-six responses to our survey were received from GPs(24/26) and CMDs(2/26). 12/26(44%) were aware of the P-PAL service. Of these 7/12(58%) had used P-PAL. 7/7(100%) who had used the service found it useful and reported that parents were happy with the service. Improvements to P-PAL suggested longer operating hours and email capability.

Seven of eight consultants responded to our survey. All hold the P-PAL phone. 70%(5/7) of consultants think it is a useful service.

**Conclusion:** P-PALS line has been successful in providing advice over the phone to GPs and CMDs. Over a third of patients were managed with clinical information sharing by telephone. Our initiative seeks to align with the Sláintecare value of “Right care, right place, right time by the right team”. We seek to build and improve this service over the coming years.

**Thursday 15.50-15.55**  
**General Paediatrics**

## **VITAMIN D LEVEL SURVEILLANCE IN CHILDREN WITH DOWN'S SYNDROME, IN A DISABILITY SERVICE IN DUBLIN.**

**S Harte<sup>2</sup>, S Slattery<sup>1</sup>, R Barry<sup>1</sup>, J McBrien<sup>1</sup>**

<sup>1</sup>Neurodisability , CHI Temple St, Dublin, <sup>2</sup>Haematology, CHI Crumlin, Dublin, Ireland

**Objectives:** Down Syndrome (DS) is the most prevalent chromosomal abnormality in Ireland, at 20.70 per 10000 live births<sup>1</sup>. Children with DS are at higher risk of bone health co-morbidities<sup>2</sup>. Severe vitamin D deficiency results in reduced calcium absorption which can manifest as nutritional rickets, hypocalcemia and growth disturbance in the paediatric population<sup>3</sup>. Currently, there are no international guidelines of the surveillance of Vitamin D deficiency in DS. The aim of this study was to examine vitamin D status in children with DS attending a Dublin disability service. Standard of Cares: 1. 100% children to have a vitamin D level checked yearly. 2. 100% of levels above 50nmol/L.

**Methods:** research population were all children with T21 who have lived in a defined geographical location between 2002 -2020. Retrospective cohort study. Ethical approval obtained. Data was extracted from the medical notes. This data included: All vitamin D levels recorded in (nmol/L), ethnicity, gender, date of first check, month of the year, calcium, phosphate and parathyroid hormone levels (if available). We classified a Vitamin D level of <50nmol/L as insufficient and a level <30nmol/L as deficient.

**Results:** Total of 102 patients were identified. Male predominance 62/102 (60%), 40/102 female (40%). 12/102 (12%) had no recorded level. 26% (24/90) of the recorded samples were found to be insufficient or deficient. The differences between genders and age categories (0-10) and (10-20) were not found to be statistically significant. A seasonal effect was noted with 13% of the insufficient/deficient results being recorded in the Winter months.

**Conclusion:** Vitamin D deficiency among children with DS is significant and surveillance is lacking. These results fortified our existing practise to screen routinely and provide preventative advice and treatment. In order to implement change in practise there is a need for a standardised protocol to be created for the surveillance and management of Vitamin D in this cohort.

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Bastola, P. and Albert, D., 2018. How Does Down Syndrome Affect Musculoskeletal Health? - The Rheumatologist. [online] The Rheumatologist. Available at: <https://www.therheumatologist.org/article/how-does-down-syndrome-affect-musculoskeletal-health/?singlepage=1&theme=print-friendly>.

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Syndrome. International Journal of Endocrinology, 2015, pp.1-11. 6.Tomaszewska, A., Rustecka, A., Lipińska-Opałka, A., Piprek, R., Kloc, M., Kalicki, B. and Kubiak, J., 2022. The Role of Vitamin D in COVID-19 and the Impact of Pandemic Restrictions on Vitamin D Blood Content. Frontiers in Pharmacology, 13.

Thursday 16.25-16.35  
Medical/Education Management

### **NURSE-LED VIRTUAL TRIAGE CLINICS IMPROVE OUTPATIENT ACCESS AND REDUCE WAITING TIMES**

**B Devery**<sup>1</sup>, S Hussey<sup>1,2,3</sup>

<sup>1</sup>NCPG, CHI-C, Dublin, <sup>2</sup>Paediatrics, RCSI, Dublin, <sup>3</sup>Paediatrics, UCD, Dublin

**Background:** The National Centre for Paediatric Gastroenterology experiences delayed access to care, especially for less urgent and routine referrals which generate long-term wait-lists.

**Aims:** This project aimed to reduce the waiting times of children on long-term waiting lists, and to identify patients in need of more priority access to care through a nurse-led validation and triage clinic.

**Methods:** A nurse-led validation and triage (NLV) clinic was introduced in January 2021, funded by the National Treatment Purchase Fund. The CHI GI wait-list was assessed in chronological order from the 'longest waiter' to the most recent referral. Calls with the Clinical Nurse Specialist Practitioner (CNSp) were pre-scheduled and standardised questionnaires were used to assess patients. Five consultation outcome categories were defined: discharge, investigations, ; direct endoscopy referral; GI consultant review (all  $\leq 6$  months) and referral to Primary Care for urgent assessment. 'Endoscopy/ colonoscopy' outcomes were discussed with consultants. Patients requiring investigations got a follow up appointment within 8 weeks.

**Results:** A total of 385 patients were waiting for up to 5 years; 112 (29%) declined GI review and were discharged directly and 273 (71%) required NLV clinic review. The NLV clinic outcomes included discharge to primary care (35%), endoscopy referral (24%), consultant review required (35%) and 6% required further investigations. As of November 2021, all but 27 patients on the GI waiting list were within the HSE target waiting time category of <12 months.

**Conclusion:** Nurse-led validation clinics are a novel, cost efficient and effective approach to managing long waiting lists for outpatient access. Most patients did not need a consultant review. Nurse-led validation should be considered by other clinical services to improve access to care and prioritise patients in greatest clinical need.

**ESTABLISHING AN ED-PAEDIATRIC MULTI-DISCIPLINARY IN-SITU SIMULATION EDUCATION PROGRAMME FOR RESUSCITATION SCENARIOS.**

**MARCUS JEE**<sup>1,2</sup>, ELLA MURPHY<sup>1</sup>, LAURA HEFFERNAN<sup>1</sup>, SINEAD O'ORIORDAN<sup>2</sup>, JOHN GANNON<sup>2</sup>, SARAH

KYNE<sup>2</sup>, AF AOIFE FLYNN<sup>2</sup>, KG KEVIN GAUGHAN<sup>2</sup>, EH ELYASSA HAMZA<sup>2</sup>, LC LISA CORLEY<sup>2</sup>, MR MAIREAD

REIDY<sup>1,2</sup>, MAURY GUERRERO<sup>1,2</sup>, ABDUL BASIT<sup>1</sup>, MADIHA GHAZANFAR<sup>1</sup>, MOHAMMED ELMURTAGA GAFAR<sup>1</sup>,

ETHEL RYAN<sup>2</sup>, ELAINE READE<sup>2</sup>, JAMES BINCHY<sup>1</sup>, JOHN O'DONNELL<sup>1</sup>, BRIAN MCNICHOLL<sup>1</sup>

<sup>1</sup>EMERGENCY DEPT, GALWAY UNIVERSITY HOSPITAL, GALWAY, <sup>2</sup>PAEDIATRICS DEPT, GALWAY UNIVERSITY HOSPITAL, GALWAY, IRELAND

In Ireland, the Paediatric team predominantly manages medical emergencies while Emergency-Medicine manages surgical/trauma resuscitations. Under these circumstances, we established a multidisciplinary insitu simulation programme to enhance the training in paediatric emergencies. This programme aimed to share insights through practice and enhance the educational experience at all staff levels.

Scenarios were written by SpRs with a standardised format/template. Weekly rotas where paediatrics facilitated the surgical/trauma-based cases and the ED facilitated paediatric medical-based emergencies, which were supervised by Consultants. Participants, who were pre-selected, were privy to resources and relevant guidelines/algorithms. We emphasised the shared cognitive model and a “discussion style” prebrief/debrief. Each session had an average of 5 participants. Participants alternated each week; giving all members of each team an opportunity to be involved. Scenarios were conducted weekly for about 40 minutes.

A pre and post-simulation questionnaire, relating to the participant's experience and their perspective on learning outcomes was used. Due to COVID19, audio-visual recording was implemented with consent and uploaded onto YouTube(private).

Participants (N=68). 92% of participants reported the simulations improved their skillset and they felt more confident in participating in the future. 100% felt better equipped to manage Paediatric-Emergencies. 98% felt the pre-sim reading material was beneficial. 84% would like to participate again. 90% reported the experience of the simulation as enjoyable. Engagement in the video streaming was most encouraging, there was a maximum viewer of 104 (82% of staff) and an average view/video of 96.

With the complexities and intricate structure of every hospital, we recognised that developing an education programme requires the evaluation of the current system. Implementing an innovative and multidisciplinary training programme, utilizing the modality of in-situ simulation, can enhance education, broaden the participant’s knowledge, and test for latent safety hazards.

We hope that our experience may be used as foresight by others in developing their own multidisciplinary in-situ programme.

**Thursday 16.45-16.55**  
**Medical/Education Management**

**QUALITY IMPROVEMENT IN ACTION PROJECT TO IMPROVE THE EMOTIONAL WELL-BEING AND BURNOUT**

**LEVEL OF PAEDIATRIC NCHDS AT PORTIUNCULA UNIVERSITY HOSPITAL**

**L Rehman**, A Raza, A Ali, MU Ayoub, M Shahid, M Islam, R Cooke, P Curran, P Cahill, F Neenan

<sup>1</sup>Paediatrics Department , Portiuncula University Hospital , Ballinasloe, Ireland

**Aim:** To decrease the level of burnout/emotional exhaustion through QIA from a baseline of 90% to 0% among Paediatric Registrars working at PUH in one year.

**Method:** Our 12 hours shift work roster consisted of 9 Registrars. We introduced three PDSA cycles during one year. An online survey was designed to find out causes & impact of roster layout on level of burnout, emotional exhaustion and staff shortage.

Five factors of emotional well-being (happiness, motivation, tiredness, irritability and sleep quality) were highlighted. Answer options were framed using a Likert-scale ranging from 1-7. The Maslach Burnout Inventory (MBI) was used to determine the level of burnout of physicians. The survey was circulated to all Registrars at the end of three months and then analysed in between July 2021 – 2022.

**Results:** Results from September 2021 confirmed 12 hours shift work layout contributed to high levels of burnout (88%), emotional exhaustion and staff shortages due to the number and frequency of night shifts.

First PDSA cycle: Number of night shifts reduced from four to two with very little improvement at the end of December 2021.

Second PDSA cycle: From Jan 2022, new roster layout was introduced i.e. ‘ED shift-physician’ who would work alongside the 24 hour on-call physician from 12pm–10pm (busy hours), Monday to Friday. Phenomenal decline in burnout level (38%) & emotional exhaustion were seen in March 2022 results, however length and number of ED shifts (12-10pm, 5 days a week) seemed very tiring.

Third PDSA cycle: Shift length reduced from ten to eight hours in April 2022. June 2022 results revealed further decline in burnout levels to 18% and moderate improvement in motivation, happiness, irritability and sleep quality.

**Conclusion:** In one year QIA project we successfully reduced burnout levels, improved emotional exhaustion significantly and tackled staff shortages after continuous reevaluation of our roster layout and by alleviating the workload of the 24-hour on-call physician.

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Likert Scale examples for surveys

<https://www.extension.iastate.edu/Documents/ANR/LikertScaleExamplesforSurveys.pdf>

**Thursday 16.55-17.05**

**Medical/Education Management**

## **REAL LIFE EVALUATION: WHAT DOES DIGITAL TRANSFORMATION MEAN FOR FRONTLINE PAEDIATRIC JUNIOR DOCTORS?**

**B Dias<sup>1</sup>, J Finnegan<sup>2</sup>, R Finnegan<sup>3</sup>, N O'Hare<sup>4</sup>, M Hesselings<sup>5</sup>**

<sup>1</sup>Medicine Year 3, Royal College of Surgeons in Ireland (RCSI) Research Summer School (RSS), Dublin, <sup>2</sup>Lead

NCHD, Children's Health Ireland (CHI) at Crumlin, Dublin, <sup>3</sup>Lead NCHD, CHI at Temple Street, Dublin, <sup>4</sup>Chief Information Officer, CHI, Dublin, <sup>5</sup>Department of Digital Health, CHI at Temple Street, Dublin,



**AIMS:** Digital transformation in the healthcare industry has the capability to improve the working lives of frontline doctors, and their ability to treat patients in a more efficient and safe environment. Much of the current literature available is focused on the benefits of individual systems, but there is little published about the general experience of frontline staff with technology. A study was conducted at two children’s hospitals to better understand the working realities of paediatric NCHDs in relation to digital technology.

**METHODS:** A survey consisting of seven questions was circulated amongst NCHDs. Doctors responded through digital and paper based surveys, as well as face to face interviews. The questions addressed their opinions on adopting paper-free systems and any barriers to technology. Thematic analysis was used to identify commonalities amongst interview responses.

**RESULTS:** A total of 66 survey responses were collected from two hospitals in Dublin across multiple medical departments. Most doctors stated that digital technology in the clinical setting was slow (n=51, 77.3%) in addition to infrastructure being limited and outdated (n=52, 77.8%). Further assessment revealed that 89.2% (n=58) of NCHDs support the idea of a completely paperless hospital. Barriers to digital transformation include multiple user logins, limited IT support and limited digital training.

**CONCLUSION:** The study revealed that NCHDs currently face many obstacles when it comes to smooth and effective digital work. These are obstacles encountered by doctors across the country. Despite these barriers, NCHDs believe that a complete digital transformation of the workplace is the way of the future of healthcare, thereby improving patient safety and efficacy.

This study provides evidence of the importance of improving basic digital infrastructure before attempting to add the latest trends in technology. Successful digital transformation requires organisations, in Ireland and worldwide, to ensure that basic digital support and infrastructure are in place first.

### **GROWTH TRAJECTORY IN INFANTS WITH TRISOMY 21 REQUIRING CARDIAC SURGERY**

MP Michaela Pentony<sup>1</sup>, ML Marie Lavelle<sup>1</sup>, SH Sarah Hayes<sup>2</sup>, NA Noeleen Allen<sup>2</sup>, AJ Allan Jenkinson<sup>1</sup>, **OF Orla Franklin<sup>1</sup>**

<sup>1</sup>Department of Paediatric Cardiology, CHI at Crumlin, Dublin, <sup>2</sup>Department of Clinical Nutrition and Dietetics, CHI at Crumlin, Dublin

**AIMS:** Ireland has the highest incidence of Trisomy 21 or Down's Syndrome (DS) in Europe, 1 in 546 live births (1). Congenital heart disease occurs in 50% of patients with DS. The optimal time for surgical repair of complete AVSD (cAVSD) or VSD is between 3 and 6 months of life when a weight of 5 kg has been achieved (2). We aim to describe the growth trajectory and dietetic intervention strategies for infants with DS who undergo cardiac surgery.

**METHODS:** The Children's Health Ireland at Crumlin NICOR data was searched for patients with DS who had surgical repair of cAVSD or VSD between 1<sup>st</sup> January 2019-31<sup>st</sup> December 2020. Retrospective chart review was undertaken to obtain demographic, medical and nutritional data.

**RESULTS:** Sixty patients with DS met inclusion criteria. The majority of infants required high calorie supplementation pre-operatively, 52/60 (87%). Nasogastric tube (NGT) placement was required for 37/60 patients (62%) for clinical indications including inadequate volume intake (54%) and poor weight gain (38%). Patients who required NGT feeding retained some oral feeding in a combined regime in 23/37 cases (62%). Seventeen patients 17/60, (28%) were still receiving some breastmilk at surgery, however three patients in total (3/60, 5%) exclusively received breastmilk at time of surgery. On average, ten dietetic contacts were required to wean NGT post cardiac surgery. Patients with DS had significant co-morbidities including pulmonary hypertension, gastrointestinal disorders and thyroid disease.

**CONCLUSION:** Infants with DS who require cardiac surgery have significant additional nutritional needs. There is a paucity in the literature describing detailed feeding experiences in infants with DS who require cardiac surgery, despite the importance in achieving target weights for surgical intervention. The information we have acquired will enable us to accurately counsel parents regarding the expected challenges surrounding feeding their child prior to cardiac surgery.

### Oral Presentation –

- (1) Murphy J, Hoey HM, Philip M, Roche EF, Macken S, Mayne P, et al. Guidelines for the medical management of Irish children and adolescents with Down syndrome. *Ir Med J.* 2005;98(2):48-52.
- (2) Xie O, Brizard CP, d'Udekem Y, Galati JC, Kelly A, Yong MS, et al. Outcomes of repair of complete atrioventricular septal defect in the current era. *Eur J Cardiothorac Surg.* 2014;45(4):610-7.

Friday 10.40-10.50

Sub-Speciality

#### THE NEW CLASSIFICATION SYSTEM IN A PAEDIATRIC EPILEPSY COHORT: NEEDS AND

**OPPORTUNITIES** Eoin P. Donnellan<sup>1</sup>, Caroline Kehoe<sup>1</sup>, Maire Ni Chollatain<sup>1</sup>, Yvonne Hynes<sup>1</sup>, Elaine Reade<sup>1</sup>, Nicholas M.

Allen<sup>1,2</sup>

<sup>1</sup>Department of Paediatrics, Galway University Hospital, Ireland, <sup>2</sup>Department of Paediatrics, University of Galway, Ireland.

**Aims:** The ILAE recently published major position papers for new classification of seizures, epilepsies (2017), and the epilepsy-syndromes (2022).<sup>1,2</sup> There are few studies reporting the spectrum of epilepsy across different settings (none in second-level units), using this new classification. The study aimed to determine the spectrum of paediatric epilepsies using 1) three levels of diagnosis (seizure-type/epilepsy-type/epilepsysyndrome), 2) aetiologies and co-morbidities, and 3) utility of molecular diagnosis to identify “precisiontreatments”.

**Methods:** Cross sectional study of all children with epilepsy diagnosis ( $\leq 16$  years) attending a second-level unit (July 2017-July 2022). Data collection was standardised to ensure a systematic approach, consistent with ILAE criteria.<sup>1,2</sup> Ethics Committee approval was obtained.

**Results:** 355 children were included (57.5% male). Mean age of epilepsy-onset 5.6 years ( $\pm 4.22$ ). The predominant seizure-type was focal (mostly motor) in 44.8% of patients, generalised (mostly motor) in 43.9%, and unknown-onset in 7.3%. Epilepsy-classification was *focal* in 46.5%, *generalised* in 38.9%, and *combined-generalised-and-focal* in 5.9%. Specific epilepsy-syndrome-classification was determined in 39.4% of patients, spanning 23 different epilepsy-syndromes, the most common of which were *self-limited-epilepsywith-centrotemporal-spikes/SeLECTS* (8.2%-of-overall cohort), *childhood-absence-epilepsy* (7.3%), *juvenileabsence-epilepsy* (5.6%) and *infantile-spasms-syndrome* (formerly West-syndrome)(5.9%). *Aetiologyspecific-syndromes* were identified (e.g. *CDKL5-developmoental-and-epileptic-encephalopathy*).

Aetiological classification identified *genetic* (54.4%), *structural* (18.6%), *infectious* (2%), *metabolic* (1.7%) and *unknown* (34.9%). Regarding genetic-aetiology, 19.2% of-overall-cohort were

### Oral Presentation –

molecularly confirmed (12.7%/n=45 as monogenic, and 6.5% chromosomal), and 34.9% were presumed-genetic. Molecular (genetic)-based precision treatments were rarely identified. Review of neurodevelopmental comorbidities revealed global-delay (29%), severe-neurological-impairment (16.3%), isolated delay (13%), and neuropsychiatric/behavioural (23.4%). Other correlations were determined.

**Conclusion:** Applying the new classification systems identifies a large burden of genetic aetiology and neurodevelopmental co-morbidity. We noted an expansion of the phenotype in some molecular diagnosis in our cohort, but few purported molecular-based “precision-therapies”. There is a “cosmic-gap” between molecular genetic (and other) aetiologies and meaningful (molecular-based) precision-therapies, indicating major needs and challenges in epilepsy-related disorders.

1. Scheffer IE, et al. ILAE classification of the epilepsies: position paper of the ILAE Commission for Classification and Terminology. *Epilepsia*. 2017;58(4):512-21.
2. Wirrell E, et al. Introduction to the epilepsy syndrome papers. *Epilepsia*. 2022 Jun;63(6):1330-2.

**Friday 10.50-11.00**

**Sub-Specialty**

### **LILAC: AN INTEGRATED APPROACH TO DIAGNOSIS AND INTERVENTION IN AUTISTIC SPECTRUM DISORDER**

**C Duff<sup>1</sup>**, A Mulvihill<sup>2</sup>, S Ni Chadhain<sup>2</sup>, EL Bohan<sup>2</sup>, R Hogan<sup>2</sup>, R Howard<sup>2</sup>, O Flanagan<sup>1</sup>, A Lyons<sup>1</sup>  
<sup>1</sup>Paediatric Department, Galway University Hospital, Galway, <sup>2</sup>Children's Disability Network 6, Galway, Ireland

**Introduction:** The national Prevalence rate of ASD in Ireland is 1.5%<sup>1</sup>, new ASD referrals can wait up to 18 months for review by the Early Intervention Service and a further 18 months for a formal ASD assessment. In Galway City East, the EIS team combatted waiting times by establishing LILAC: an innovative new approach to the assessment, intervention and diagnosis.

**Methods:** LILAC represents Laughing, Interaction, Listening, Attention and Communication. Sessions comprise activity and strategy based interventions for children and coaching for their parents using evidence based interventions through a family centred approach. Parent education sessions are provided and children undergo a diagnostic process if deemed appropriate. LILAC runs over three months with sessions taking place weekly. The team incorporates a community nurse, occupational therapist, physiotherapist, psychologist, pre-school liaison teacher, social

### Oral Presentation –

worker, and speech and language therapist who liaise with two consultant paediatricians. At the end a diagnostic report is provided to parents, with intervention recommendations. It provides a very effective, integrated and combined intervention and diagnostic process. The pilot recruited a cohort of eight children.

**Results:** Of the eight children and families who participated in the first LILAC 7/8 received a diagnosis of ASD. Staff perceptions were overwhelmingly positive. Team members extolled LILAC as a “very effective and efficient method of interdisciplinary service delivery”. They lauded the individualized, family-centred approach to healthcare.

**Conclusion:** Further investigation of the benefits of LILAC is planned, including measurement of parental satisfaction. The programme aims to expand to regional Progressing Disability Services providing a family centred multidisciplinary approach to intervention and diagnosis of ASD.

1. Estimating Prevalence Rate of Autism Spectrum Disorders (ASD) in the Irish Population: A review of Data Sources and Epidemiological Studies Department Of Health 2018.

**Friday 11.00-11.10**  
**Sub-Specialty**

### **USE OF THE INFANTILE HAEMANGIOMA REFERRAL SCORE BY GENERAL PRACTITIONERS AND PAEDIATRICIANS AND ITS UTILITY IN OPTIMISING REFERRALS TO DERMATOLOGY**

**R Harrington, M Leahy, B Byrne, A Murphy**

<sup>1</sup>Department of Dermatology, University Hospital Galway, Galway, Ireland

**Background:** The Infantile Haemangioma Referral Score (IHReS) is a validated, decision-making tool designed to aid physicians in appropriate and timely referral of patients with infantile haemangiomas (IH) to specialist centres. Previous studies(1,2) have assessed its usefulness in identifying patients who require referral, however none have analysed its utility in identifying those who do not.

**Objective:** To assess the use of the IHReS score by GPs and paediatricians and to correlate the patients’ score with outcomes.

**Method:** Retrospective, cross-sectional analysis of all new IH referrals to a dedicated IH clinic over 6 months. The IHReS was retrospectively applied using information from referral letters and Dermatology clinic letters. Data on demographics, wait-time and outcomes were analysed.

**Results:** Forty-four patients were identified (female n=23 male n=21). Mean age at referral was 30(sd±48) weeks. Only 3(7%) referrals had recorded the patient's IHReS. Overall, 17(39%) patients did not satisfy criteria for referral when the IHReS was retrospectively applied. Of these, 10(59%) were discharged on first visit, 6(35%) were offered follow-up and one did not attend. None of the patients in this group received propranolol.

Of the 27(61%) patients who did meet the IHReS criteria, 12(44%) required propranolol, 11(41%) were monitored for potential treatment, and 4(15%) were discharged.

**Conclusion:** The IHReS is a useful tool for identifying IH patients who do not require referral to a specialist centre. It is underutilised by GPs and paediatricians and its use could reduce the number of unnecessary referrals to Dermatology.

1. Léauté-Labrèze C, Baselga Torres E, Weibel L, Boon LM, El Hachem M, van der Vleuten C, Roessler J, Troilius Rubin A. The Infantile Hemangioma Referral Score: A Validated Tool for Physicians. *Pediatrics*. 2020 Apr;145(4):e20191628. doi: 10.1542/peds.2019-1628. Epub 2020 Mar 11. PMID: 32161112
2. Chitpiromsak K, Techasatian L, Jetsrisuparb C. Utility of the Infantile Hemangioma Referral Score (IHReS) as a decision-making tool for referral to treatment. *BMJ Paediatr Open*. 2021 Sep 15;5(1):e001230. doi: 10.1136/bmjpo-2021-001230. PMID: 34604545; PMCID: PMC8444243.

Friday 11.10-11.20  
Sub-Specialty

### **CANCER CARE FOR CHILDREN, ADOLESCENTS AND YOUNG ADULTS AND THEIR PATHS TO DIAGNOSIS. AN IRISH EXPERIENCE.**

**EB Brennan**<sup>1</sup>, NOS O'Sullivan <sup>1,2</sup>, PMC McCarthy<sup>1,3</sup>, OS Smith<sup>1,3</sup>, SA Alken <sup>1,2</sup>

<sup>1</sup>Oncology/Haematology, CHI at Crumlin, Dublin, <sup>2</sup>Oncology/Haematology, St James Hospital, Dublin, <sup>3</sup>School of Medicine, UCD, Dublin, Ireland

**Introduction:** There are approximately 500,000 childhood cancer survivors currently living in Europe<sup>1</sup>. With improving survival rates of childhood and adolescent cancer, survivorship, and long-term follow-up and care for these patients has been at the forefront of focus in recent years. We aimed to explore the Irish experience of cancer care for this group.

### Oral Presentation –

**Methods:** Data was collected via anonymous online survey. Inclusion criteria were age over of 16 years and having been diagnosed with cancer between the ages of 0 and 24 years. Data including demographics, path to diagnosis, diagnosis and treatment was collected.

**Results:** Sixty-one surveys were completed, and were stratified into the 0-14-year-old group (n=28) and over 15 group (n=33) at time of diagnosis. In the 0–14-year cohort, the commonest diagnosis was leukaemia (35.7%, n=10), solid tumour (28.6%, n=8) and CNS tumour (21.4%, n=6). Lymphoma was the commonest diagnosis in the 15–26-year age group (51.5%, n=17 vs 10.7%, n=3 in 0-14yo) ( $p=0.009$ ).

Most patients (85.2%) attended a general practitioner (GP) prior to diagnosis, with 21.2% reporting >3 GP visits.

A statistically significant difference was noted between the groups as to who informed YP they had cancer; 76% of over 15s were told by healthcare professionals compared with 36% of 0-14yo ( $p=0.16$ ). Participants were invited to comment on questions answered with one respondent quoting “Nobody properly told me I just figured it out from hearing doctors talk around me”.

**Conclusion:** The management of cancer for children and adolescents spans beyond care and cure. These data reflect the first patient experience survey of this population. We must endeavour to include children and young people in discussions about their health. Children and YP with cancer require their unique psychosocial needs to be met with age-appropriate communication styles and provision of information which needs to be prioritised by healthcare professionals<sup>2</sup>. No one should overhear their cancer diagnosis.

1. Winther, Jeanette F., Line Kenborg, Julianne Byrne, Lars Hjorth, Peter Kaatsch, Leontien C. M. Kremer, Claudia E. Kuehni, et al.

“Childhood Cancer Survivor Cohorts in Europe.” *Acta Oncologica* 54, no. 5 (2015): 1–14.

doi:10.3109/0284186X.2015.1008648. 2. Mascarin M, Milano GM, Bertolotti M, Quarello P, Spinelli M, Tornesello A, Bomben F, Ferrari A. Discussing communication issues and needs with adolescents with cancer. *Tumori*. 2021 Aug;107(4):360-363. doi: 10.1177/0300891620977036.

Epub 2020 Nov 26.

PMID: 33243095.

**Friday 11.20-11.30**  
**Sub-Specialty**

**GENOTYPE AND TONGUE REDUCTION SURGERY IN BECKWITH WIEDEMANN SYNDROME**

**C POWER<sup>1</sup>, S BRACKEN<sup>2</sup>, C GALLAGHER<sup>2</sup>, D MURRAY<sup>2</sup>, S LYNCH<sup>3</sup>, SM O' CONNELL<sup>4</sup>**

<sup>1</sup>DEPT. OF GENERAL PAEDIATRICS, CHILDREN'S HEALTH IRELAND AT CRUMLIN, DUBLIN, <sup>2</sup>DEPT. OF CRANIOFACIAL SURGERY, CHILDREN'S HEALTH IRELAND AT TEMPLE STREET, DUBLIN, <sup>3</sup>DEPT. OF CLINICAL

GENETICS, CHILDREN'S HEALTH IRELAND AT CRUMLIN, DUBLIN, <sup>4</sup>DEPT. OF DIABETES AND ENDOCRINOLOGY, CHILDREN'S HEALTH IRELAND AT CRUMLIN, DUBLIN IRELAND

**Introduction:** Beckwith Wiedemann syndrome (BWS) is a rare genetic disorder caused by epigenetic alterations affecting gene expression on chromosome 11p15. It is the commonest cause of macroglossia in childhood<sup>1</sup>. 90% of children with BWS have macroglossia and ~40% undergo tongue reduction surgery (TRS)<sup>2</sup>. Indication for TRS may be multifactorial including parental preference, airway compromise and impact on speech. The genotype predicts certain phenotypic features in BWS. Tongue enlargement is reported to be more marked in individuals with Loss of Methylation at Imprinting centre 2 (IC2-LoM)<sup>3</sup>, which is seen in 50% of BWS cases, but there is a paucity of studies examining the relationship between genotype and severity of macroglossia. We examined the genotypes of children with BWS and macroglossia attending the craniofacial surgery service at Children's Health Ireland (CHI), which is the only centre nationally performing TRS, to determine any associations with requirement for TRS.

**Methods:** A search of the craniofacial surgery database for children with BWS and macroglossia over past 10 years was conducted. Medical charts were reviewed to determine outcomes. Genetic laboratory records were reviewed to determine genotype.

**Results:** 27 children with BWS and macroglossia were identified, 18 of whom required TRS. Genotype was available on 15 of these children; 7 had IC2-LoM, 4 had a mutation in CDKN1c which accounts for only 5% of BWS cases, 2 were due to mosaic paternal uniparental disomy of chromosome 11 (UPD(11)pat), and 1 due to IC1 gain of methylation. One child had negative testing despite a clinical BWS diagnosis.

**Conclusions:** All genotypes are seen in this cohort, suggesting that genotype is not a strong predictor for severe macroglossia and requirement for TRS. CDKN1c, a rare cause of BWS, is more strongly represented in this cohort. Further study is warranted to examine this possible association between CDKN1c and degree of macroglossia.



### Oral Presentation –

1. Prada CE, Zarate YA, Hopkin RJ. Genetic causes of macroglossia: diagnostic approach. *Pediatrics*. 2012 Feb;129(2):e431-7. doi: 10.1542/peds.2011-1732. Epub 2012 Jan 16. PMID: 22250026.
2. Brioude F, Kalish JM, Mussa A, Foster AC, Bliet J, Ferrero GB, Boonen SE, Cole T, Baker R, Bertolotti M, Cocchi G, Coze C, De Pellegrin M, Hussain K, Ibrahim A, Kilby MD, Krajewska-Walasek M, Kratz CP, Ladusans EJ, Lapunzina P, Le Bouc Y, Maas SM, Macdonald F, Öunap K, Peruzzi L, Rossignol S, Russo S, Shipster C, Skórka A, Tatton-Brown K, Tenorio J, Tortora C, Grønsvov K, Netchine I, Hennekam RC, Prawitt D, Tümer Z, Eggermann T, Mackay DJG, Riccio A, Maher ER. Expert consensus document: Clinical and molecular diagnosis, screening and management of Beckwith-Wiedemann syndrome: an international consensus statement. *Nat Rev Endocrinol*. 2018 Apr;14(4):229-249. doi: 10.1038/nrendo.2017.166. Epub 2018 Jan 29. PMID: 29377879; PMCID: PMC6022848.
3. Mussa A, Russo S, De Crescenzo A, Freschi A, Calzari L, Maitz S, Macchiaiolo M, Molinatto C, Baldassarre G, Mariani M, Tarani L, Bedeschi MF, Milani D, Melis D, Bartuli A, Cubellis MV, Selicorni A, Cirillo Silengo M, Larizza L, Riccio A, Ferrero GB. (Epi)genotype-phenotype correlations in Beckwith-Wiedemann syndrome. *Eur J Hum Genet*. 2016 Feb;24(2):183-90. doi: 10.1038/ejhg.2015.88. Epub 2015 Apr 22. PMID: 25898929; PMCID: PMC4717210.
4. Defabianis P, Mussa A, Ninivaggi R, Carli D, Romano F. Maxillo-Facial Morphology in Beckwith-Wiedemann Syndrome: A Preliminary Study on (epi)Genotype-Phenotype Association in Caucasians. *Int J Environ Res Public Health*. 2022 Feb 20;19(4):2448. doi: 10.3390/ijerph19042448. PMID: 35206635; PMCID: PMC8872180.

**Friday 11.30-11.40**

**Sub-Specialty**

### **MULTISYSTEM INFLAMMATORY SYNDROME IN CHILDREN IN THE REPUBLIC OF IRELAND: THE STORY SO FAR...**

**B Ramly<sup>1</sup>, B Treston<sup>1</sup>, M Lawler<sup>1</sup>, M Hassan<sup>1</sup>, K Forde<sup>1</sup>, N Linnane<sup>2</sup>, C McMahon<sup>2</sup>, O Franklin<sup>2</sup>, TR Leahy<sup>1</sup>, C O'Maoldomnaigh<sup>1</sup>, B Freyne<sup>1</sup>, P Gavin<sup>1</sup>**

<sup>1</sup>National Centre for Paediatric Infectious Diseases and Immunology , CHI Crumlin and CHI Temple Street , Dublin

<sup>2</sup>National Centre for Paediatric Cardiology, CHI Crumlin and CHI Temple Street, Dublin

**Background:** In contrast to adults, COVID-19 is a rare cause of severe disease in children. However, the emergence of Multisystem Inflammatory Syndrome in Children (MIS-C), a potentially life-threatening hyperinflammatory syndrome associated with preceding SARS-CoV-2 infection has caused considerable concern. Unlike COVID-19 infection, MIS-C is not yet a notifiable condition.

### Oral Presentation –

**Aim:** To describe the epidemiology, clinical characteristics, treatment, and outcomes of children with MIS-C in the Republic of Ireland during the first 2.5-years of the SARS-CoV-2 pandemic (April 2020-Sept 2022).

**Methods:** Prospective surveillance of MIS-C cases identified in Irish hospitals from April 2020 to September 2022. Medical records of all children diagnosed and treated for MIS-C in Children's Health Ireland were entered prospectively into a standardized electronic database. Data from children treated in other paediatric centers were obtained retrospectively. The Royal College of Paediatrics and Child Health case definition of MIS-C was used.

**Results:** From April 2020 to September 2022, 118 MIS-C cases were identified. Median age, 7 years, (range, 1 month – 15.5 years); Males 58.5%. In the first year of the pandemic from April 2020 to April 2021, 63% of cases had features of hyperinflammatory MIS-C and 37% of cases fulfilled the criteria for Kawasaki disease (KD). KD type-MIS-C cases were significantly younger (median age 2 years).

**Conclusion:** MIS-C is a newly emerged and evolving phenomenon. Mandatory reporting and ongoing surveillance of MIS-C cases will help identify those at greatest risk and determine the long-term impact of COVID-19 vaccination on incidence.

Friday 12.10-12.20

Neonatal

### **MATERNAL SARS-COV-2 DELTA VARIANT INFECTION: INCREASE IN PRETERM BIRTH AND NICU ADMISSIONS**

S Ogo<sup>1</sup>, **C Murphy**<sup>1,2</sup>, D O'Reilly<sup>1</sup>, J Donnelly<sup>3</sup>, C McCarthy<sup>1</sup>, N McCallion<sup>1,2</sup>, R Drew<sup>4,5</sup>, W Ferguson<sup>1,2</sup>

<sup>1</sup>Neonatology, Rotunda, Dublin, <sup>2</sup>Paediatrics, RCSI, Dublin, <sup>3</sup>Obstetrics and Gynaecology, RCSI, Dublin,

<sup>4</sup>Clinical Innovation Unit, Rotunda, Dublin, <sup>5</sup>Irish Meningitis and Sepsis Reference Laboratory, Children's

Health Ireland at Temple Street, Dublin, <sup>6</sup>Department of Clinical Microbiology, RCSI, Dublin

**Aims:** The SARS-CoV-2 Delta variant (B.1.617.2) caused more severe illness in pregnant women, with increased rates of maternal ICU admission. Our aim was to describe the outcomes of infants born to women with SARS-CoV-2 during pregnancy, during the period of Delta variant dominance.

**Methods:** This was a retrospective observational study. Women with SARS-CoV-2 detected during pregnancy between 5<sup>th</sup> July 2021 and 5<sup>th</sup> December 2021 (period of Delta dominance) were included. Patients were identified from in-house testing or infants transferred-in following delivery in an acute adult hospital due to maternal COVID-19. Only pregnancies completed by 31<sup>st</sup> January 2022 were included.

**Results:** 81 women were identified, 4 pregnancies ended in miscarriage/stillbirth and there were 79 liveborn infants. The mean maternal age was 30.3 years ( $\pm 6.5$  years), 30 (37%) were primigravida, 65 (80%) were Caucasian and 28 (35%) had an underlying medical condition. 17 (21%) were vaccinated, 19 (23%) were unvaccinated and in 45 (56%), vaccination status was not available. 14 (18%) women delivered preterm (<37 weeks). The median gestation at diagnosis was 36 weeks (32-38 weeks) with a median duration of 8 days (1-31) from diagnosis to delivery. 6 (7%) women required respiratory support. 30 (38%) infants were delivered by caesarean section. The median birth weight was 3.3kg (2.9 – 3.7) and gestation was 39 weeks (37.6 – 39.9). 18 (23%) infants were admitted to NICU with a median stay of 5.5 days (3-21). 36 (46%) infants were receiving breast milk at discharge.

**Conclusions:** The incidence of preterm birth (18% v 8%,  $p < 0.01$ ) and NICU admission (23% v 14%,  $p = 0.03$ ) was significantly higher among pregnancies affected by the Delta variant than the general hospital population in 2020. This finding was unique to the Delta variant and was not observed during previous studies of Wild type or Alpha variant epochs in the Rotunda Hospital.

12.30

**Neonatal DEVELOPMENT OF A NATIONAL NEONATAL HEART VALVE DONATION PROTOCOL**

**S Dempsey**<sup>1</sup>, E Shields<sup>2</sup>, D Sweetman<sup>1,3</sup>, J Murphy<sup>1,3</sup>

<sup>1</sup>Neonatal Department, National Maternity Hospital, Dublin, Ireland

<sup>2</sup>Organ Donation Transplant Ireland, HSE, Dublin, Ireland

<sup>3</sup>National Women and Infants Health Programme, HSE, Dublin, Ireland

**Aims:** To develop a national neonatal heart valve donation protocol to assist healthcare professionals to facilitate donation within their local unit where there is a confirmed neonatal death by neurological criteria death and/or cardiac criteria death. To highlight the growing need for heart valve donations and identify who is eligible to donate them.

**Method:** The initial protocol was created in a tertiary Neonatal Intensive Care Unit (NICU) in conjunction with The Mater Hospital Transplant team. This was adapted making it suitable for use in all units in Ireland. A national organ donation coordinator provided advise on how to approach families regarding heart valve donation.

**Results:** A national neonatal heart valve donation protocol was published with supporting organisational flow charts and parent information leaflets. These documents can be found both online on the National Women and Infants Health Programme (NWIHP) website and within local unit's policy folder and/or QPULSE system. A short video explaining the protocol was produced and is available on the NWIHP website. An increasing number of health care professionals are now aware of who is eligible for valve donation and how to organise it within the short 4 hours from death timeframe. The protocol has been approved by the Neonatal Clinical Advisory Group, RCPI Faculty of Paediatrics, National Women's and Infants Programme and Organ Donation Transplant Ireland.

**Conclusion:** Since the first heart valve transplant in 1967, demand has significantly outgrown supply. In 2021, thirty-eight heart valves were transplanted in Ireland, thirty-one of which were for paediatric patients in Children's Health Ireland (CHI), Crumlin. The national protocol highlights the need for heart valve donations, educates health care professionals on who is eligible for donation and how to organise this within local units. The request for heart valve donation should be considered a standard and appropriate aspect of end-of-life care.

**CURRENT MANAGEMENT OF NEONATAL ABSTINENCE SYNDROME: SURVEY OF THE**

**BRITISH ISLES S Dempsey<sup>1</sup>, J Wong<sup>1</sup>, MJ O'Grady<sup>1,2</sup>**

<sup>1</sup>Department of Paediatrics, Midland Regional Hospital, Mullingar

<sup>2</sup>Women's and Children's Health, School of Medicine, University College Dublin

**Aim:** To establish the current management of neonatal abstinence syndrome (NAS) in neonatal units throughout the United Kingdom (UK) and Ireland.

**Methods:** Postal questionnaire to 215 neonatal units in Ireland and the UK, with follow -up telephone calls for those who did not respond or underfilled questionnaires.

**Results:** 94% of all respondents treat NAS within their unit, of which 93% had a formal NAS guideline. The medium number of neonates treated annually for NAS was 5. A variety of scoring tools were described, the Finnegan Method is the most commonly used scoring tool(63%). Other scoring systems included Modified Finnegan score, Lipsitz score and Rivers score. 98% of the units who treat NAS use oral Morphine as their first line agent for opiate and polysubstance abuse with varying doses. Phenobarbitone is the predominant first line treatment for seizures associated with opiate and polysubstance withdrawal. Breastfeeding is encouraged in a majority of units for mothers taking methadone and those who are Hep B and/or C positive. Cranial Ultrasound is not routinely performed in 100% of the responding units. Follow up clinic appointments are not standardised with varying time lengths described.

**Conclusion:** The majority of units in Ireland and the UK use morphine as the first line of treatment for drug withdrawal. There is significant inconsistency in drug dosage and frequency throughout the units. This study demonstrates the paucity of research into the optimal management of NAS.

**DOES ANAKINRA (IL1 RECEPTOR ANTAGONIST) AFFECT IMMUNE DYSFUNCTION IN INFANTS WITH NEONATAL ENCEPHALOPATHY**

**A Branagan**<sup>1-3</sup>, C Stanciu<sup>4</sup>, J Isaza-Correa<sup>1,2</sup>, L Kelly<sup>1,2</sup>, M White<sup>1</sup>, J Miletin<sup>1</sup>, EJ Molloy<sup>1-3,5</sup>

<sup>1</sup>Discipline of Paediatrics, Trinity College, The University of Dublin, Dublin, <sup>2</sup>Trinity Translational Medicine

Institute, Trinity College, The University of Dublin, Dublin, <sup>3</sup>Paediatrics, Coombe Women and Infants

University Hospital, Dublin, <sup>4</sup>Paediatrics, Rotunda Hospital, Dublin, <sup>5</sup>Neonatology, Paediatrics, Children's Health Ireland, Dublin,

**INTRODUCTION:** Neonatal encephalopathy (NE) is a clinical condition describing acute brain injury in newborns, incorporating multiple diverse aetiologies. Despite routine use of therapeutic hypothermia (TH), there is a significant burden of mortality and morbidity. Therefore, there is an urgent need for the development of adjunctive treatments. The pathophysiology is based on immune dysregulation, and therefore immunomodulatory drugs may have a positive effect. Anakinra (interleukin-1 receptor antagonist) (ILRA) is one such drug. ILRA blocks both IL-1 $\alpha$  and IL-1 $\beta$  biologic activity and has been safely used in neonates.

**AIMS:** To assess the impact of ILRA on ex-vivo immune function in infants with NE.

**METHODS:** Infants with moderate-severe NE (Sarnat scoring) undergoing TH were recruited. Blood samples were taken on days 1, 3 and 7 of life and compared with healthy neonatal controls. Innate immune function was analysed by flow cytometry for CD11b (cell activation, migration) and Toll-like receptor (TLR)-4 (recognition of lipopolysaccharide, LPS) in neutrophils (CD66b+) and subpopulations of monocytes (CD14/CD16) stimulated with and without LPS, treated with and without ILRA. Samples were assessed by reverse transcription–polymerase chain reaction (PCR) for markers of inflammasome activation (NLRP3), downstream signalling of TLR4 (MYD88) and proinflammatory cytokines (IL1B)

**RESULTS:** 20 infants with neonatal encephalopathy were recruited with 36 samples assessed (day 1 (n=10), day 3 (n=18) day 7 (n=8)), along with 7 healthy controls. There were no differences in innate immune function when assessed by CD11b or TLR4 after treatment with anakinra. Neutrophil TLR4, although initially suppressed, increased by day 7, indicating dysregulated

immune responses in these patients. PCR analysis showed significant increase in IL1B in infants with NE which was not decreased by ILRA.

**DISCUSSION:** Ex-vivo treatment with anakinra does not alter immune function in infants with neonatal encephalopathy, and may not be a useful adjunctive therapy to TH in infants with NE.

Friday 12.50-13.00

Neonatal

### **TWO HANDS ARE BETTER THAN ONE: DELIVERING POSITIVE PRESSURE VENTILATION IN A NEONATAL MANIKIN MODEL**

**AL Murray**<sup>1</sup>, T Beechinor<sup>1</sup>, V Livingstone<sup>1,2</sup>, EM Dempsey<sup>1,2</sup>

<sup>1</sup>Department of Neonatology, Cork University Maternity Hospital, <sup>2</sup>University College Cork, Ireland

**Background/Aims:** Approximately 10% of newly born infants require some form of assistance to establish regular breathing at birth(1, 2). International guidelines advise the use of positive pressure ventilation (PPV) in newly born infants with bradycardia or inadequate respiratory effort(1).

Previous studies investigating mask leak in modified newborn manikin models have shown high levels of mask leak (up to 65%). Conflicting evidence exists for various hand hold techniques. (3, 4). The aim of the study was to determine if there is a difference in mask leak between one-handed (OH) and two-handed (TH) hold of the facemask when delivering PPV during neonatal resuscitation.

**Methods:** A crossover randomized trial was performed. All participants had completed their Neonatal Resuscitation Programme training. A Neopuff Infant Resuscitator used with a size 35mm facemask and premature neonatal manikin. A Respiration Respiratory Function monitor (RFM) was used to measure inflation pressures, tidal volumes, and expiratory leak via a flow sensor between the mask and the Neopuff. Two minutes of breaths were recorded for each participant for both OH and TH techniques. For TH technique, the participants performed the mask hold, while one of two researchers delivered breaths. Participants were randomly assigned to perform OH or TH technique first.

**Results:** There were 41 healthcare providers included; one was excluded from the analysis due to missing data. A total of 1350 breaths were analysed. Median mask leak was greater with one-

## Oral Presentation –

handed (median(IQR): 12.3%(5.5% to 49.1%)) compared with two-handed (median(IQR): 5.2%(2.6% to 14.6%)) technique ( $p=0.041$ ). There were no significant relationships between years of experience or professional group with one-handed or two-handed leak values.

**Conclusion:** When using positive pressure ventilation is provided in a neonatal manikin model, two-handed technique is superior to one-handed technique in reducing mask leak. Further clinical trials of OH versus TH are warranted in the delivery suite.

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**Friday 14.30-14.40**

**Neonatal**

### **IMPACT OF A NEONATAL EARLY-ONSET SEPSIS CALCULATOR ON ANTIBIOTIC USAGE IN A LEVEL II SPECIAL**

#### **CARE NURSERY IN WESTERN AUSTRALIA**

**J Gannon<sup>1</sup>, T Strunk<sup>2</sup>, N Friesen<sup>1</sup>, C Saw<sup>1</sup>**

<sup>1</sup>Paediatrics, St. John of God Midland Hospital, Perth, Australia

<sup>2</sup>Neonatal Directorate, King Edward Memorial Hospital, Perth, Australia

**Aims:** Overuse of empirical intravenous antibiotics in neonates in high-income countries (HICs) is well documented.<sup>1-3</sup> The Kaiser Permanente neonatal early-onset sepsis calculator (EOSC) is a free online evidence-based multivariate tool that has demonstrated potential to reduce antibiotic usage in this population.<sup>4</sup> The incidence of early-onset sepsis (EOS) in most HICs is 0.4 – 0.8 per 1,000 live births.<sup>5 6,7</sup> This study's objective was to evaluate the calculator's impact on antibiotic rates and length of stay in a regional level II Special Care Nursery in Western Australia.



**Methods:** A single-centre retrospective cohort study compared antibiotic prescription rates in the first 72 hours in neonates > 35 weeks gestation born over two 6-month periods: 2019 (pre-EOSC) and 2021 (postEOSC). Electronic and paper case records for mother and baby were accessed to capture required data points. Continuous data were summarised using mean and standard deviation, and categorical data were summarised using frequency distributions. Sample sizes were 951 (2019) and 1,129 (2021).

**Results:** Following EOSC implementation, antibiotic use decreased from 13.7% to 4.7% of neonates (Fisher Exact Test  $p < 0.00001$ ) without any reported negative outcomes. The proportion treated for obstetric risk factors as indication decreased from 31.5% to 18.9%, and proportion treated for clinical condition increased from 63.1% to 79.2%. Mean length of stay for all neonates born across the two cohorts decreased from 2.38 to 2.13 days. There were no culture-proven cases of sepsis in either cohort.

**Conclusions:** The EOSC significantly reduced the administration of unnecessary antibiotics, preventing potential side effects and complications for neonates. Indications for treatment were more closely aligned to clinical condition rather than obstetric risk factors and mean length of hospital stay was lower across all births. These findings highlight the value of an evidence-based algorithm to improve antimicrobial stewardship in neonates with health benefits to the individual and cost savings to the healthcare system.

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Friday 14.40-14.50

Neonatal

**ASSESSMENT OF MYOCARDIAL FUNCTION IN NEONATES CONCEIVED BY ASSISTED REPRODUCTIVE TECHNOLOGIES USING DEFORMATION IMAGING OVER THE FIRST YEAR OF AGE**

**A Smith**<sup>1</sup>, O Franklin<sup>2</sup>, E Mocanu<sup>3</sup>, N McCallion<sup>1,4</sup>, A EL-Khuffash<sup>1,4</sup>

<sup>1</sup>Neonatology, The Rotunda Hospital, Dublin, <sup>2</sup>Cardiology, Children's Health Ireland at Crumlin, Dublin, <sup>3</sup>Department of Obstetrics, School of Medicine, Royal College of Surgeons in Ireland, Dublin, <sup>4</sup>Department of Paediatrics, School of Medicine, Royal College of Surgeons in Ireland, Dublin

**Aims:** Recent data suggest that fetuses conceived by assisted reproductive technologies (ART) undergo cardiovascular remodelling resulting in altered myocardial performance. Our aim was to assess biventricular function and markers of pulmonary vascular resistance (PVR) in infants conceived by ART during over the first year of age and compare the findings with a control group conceived spontaneously.

**Methods:** This was a prospective study of 50 term infants conceived by ART and 50 infants term infants conceived spontaneously. Echocardiography was performed on Day 2, 6 months and 1 year of age. LV and RV function was measured using deformation analysis. LV eccentricity index (LV EI) and pulmonary artery acceleration time indexed to right ventricular ejection time (PAATi) were used to assess PVR.

**Results:** There was no difference in gestation ( $38.9 \pm 1.0$  vs.  $39.3 \pm 0.9$  weeks,  $p=0.08$ ), birthweight ( $3.6 \pm 0.5$  vs.  $3.6 \pm 0.4$  Kg,  $p=0.95$ ), male sex (60% vs. 46%,  $p=0.23$ ) or the rate of cesarean section (66% vs. 74%,  $p=0.51$ ) between ART infants and controls. ART infants had lower PAATi at 6 months and one year and higher LV EI throughout the study period. ART infants had lower LV strain, systolic strain rate and early diastolic strain rate throughout the study period. ART infants had lower RV free wall strain throughout the study period in addition to lower RV systolic strain

rate and early diastolic strain rate at one year of age. On subgroup linear regression analysis of the ART group egg characteristics (own vs. donor and fresh vs. frozen) and mode of ART (IVF vs. ICSI) had no influence on LV or RV function.

**Conclusion:** Infants conceived by ART present with evidence right and left ventricular systolic and diastolic dysfunction and persistent elevation of PVR over the first year of age.

Friday 14.50-15.00

Neonatal

**A RANDOMISED, CROSSOVER STUDY OF STANDARD LARYNGOSCOPE AND TWO  
VIDEOLARYNGOSCOPES  
(ACUTRONIC INFANT VIEW & C-MAC) FOR ENDOTRACHEAL INTUBATION OF NEWBORN  
MANNEQUINS**

**LE Geraghty**<sup>1,2</sup>, G Gambacorta<sup>1,3</sup>, CPF O'Donnell<sup>1,2</sup>

<sup>1</sup>Neonatal Unit, National Maternity Hospital, Dublin, <sup>2</sup>School of Medicine, University College Dublin, Dublin,

<sup>3</sup>Children's Hospital V. Buzzi, University of Milan, Milan, Italy

**Aims:** When intubating babies, standard laryngoscopes (SL) are used to look directly into the mouth (direct laryngoscopy), whereas videolaryngoscopes (VL) display the view of the larynx obtained with a camera on a screen (indirect laryngoscopy). Trainees who perform direct laryngoscopy with an Acutronic VL have greater first attempt success rate when a mentor views the screen; and trainees acquire competence more quickly with C-MAC VL than SL. We compared the performance of intubators using SL, Acutronic and C-MAC in two mannequins.

**Methods:** Crossover study – each participant used SL (Heine, Germany), Acutronic Infant View (Acutronic, Switzerland) and C-MAC (Karl Storz, Germany) to intubate a preterm (Premie Hal, Gaumard) and term (Newborn Anne, Laerdal) mannequin in random order. We recorded number of attempts taken to intubate and the duration of attempts.

**Results:** Twenty-four (6 senior, 18 trainees) doctors and 1 advanced nurse practitioner participated. Success at first attempt and duration of successful attempt were similar for the SL

and C-MAC for both mannequins. The success rate was lower and the duration of attempts longer with the Acutronic VL in the term mannequin.

**Conclusions:** Participants performed similarly using direct laryngoscopy or indirect laryngoscopy with the C-MAC when intubating mannequins. Direct laryngoscopy and C-MAC for intubation of newborns merit comparison in a randomised trial.

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**Friday 15.00-15.10**  
**Neonatal**

**HIGHER PLATELET TRANSFUSION THRESHOLDS IN PRETERM INFANTS ARE ASSOCIATED WITH WORSE OUTCOMES AT TWO YEARS CORRECTED**

**CM Moore**<sup>1</sup>, A D'Amore<sup>2</sup>, SF Fustolo-Gunnink<sup>3</sup>, C Hudson<sup>4</sup>, A Newton<sup>4</sup>, B Lopez-Santamaria<sup>5</sup>, A Deary<sup>4</sup>, R

Hodge<sup>4</sup>, A Mora<sup>4</sup>, V Venkatesh<sup>6</sup>, R Khan<sup>7</sup>, K Willoughby<sup>2</sup>, W Onland<sup>8</sup>, K Fijnvandraat<sup>3</sup>, H New<sup>4</sup>, P Clarke<sup>9</sup>, E Lopriore<sup>10</sup>, T Watts<sup>5</sup>, S Stanworth<sup>11</sup>, A Curley<sup>1</sup>

<sup>1</sup>Neonatology, National Maternity Hospital, Dublin, <sup>2</sup>Neonatology, Cambridge UH NHS Trust, Cambridge,

<sup>3</sup>Research, Sanquin, Amsterdam, Netherlands, <sup>4</sup>CTU, NHS Blood and Transplant, Bristol/London,

<sup>5</sup>Neonatology, Guys and St Thomas' NHS Trust, London, <sup>6</sup>Neonatology, Cloudnine Hospital, Bangalore, India,

<sup>7</sup>Neonatology, University Maternity Hospital Limerick, Limerick, <sup>8</sup>Neonatology, AMC, Amsterdam, Netherlands, <sup>9</sup>Neonatology, NNUH, Nottingham, UK, <sup>10</sup>Neonatology, LUMC, Leiden, Netherlands, <sup>11</sup>Haematology, Oxford, UK

**Background:** In 2019 the PlaNeT-2/MATISSE multicentre randomised controlled trial reported that a higher platelet transfusion threshold ( $50 \times 10^9/L$ ) in preterm babies born at  $<34$  weeks of gestation compared to a lower one ( $25 \times 10^9/L$ ) was associated with significantly increased mortality or major bleeding. We now report on neurodevelopmental outcomes at two years corrected age.

**Methods:** Neurodevelopmental outcomes were assessed using formal assessment tools, parent reporting assessment or healthcare provider reports. A favourable outcome was given if a child was alive at two years of age and did not have any of the following: cerebral palsy that impaired independent walking; cognitive impairment  $>2$  standard deviations below the mean; severe seizure disorder; hearing impairment not corrected by hearing aids; or bilateral visual impairment with no useful vision.

**Results:** Follow-up data were available for 92% (601 out of 653) eligible children. Of the 296 infants assigned to the higher threshold group, 147 (50%) infants died or survived with significant neurodevelopmental impairment, as compared with 120 (39%) of the 305 infants assigned to the lower threshold group (odds ratio 1.54, adjusted for gestational age and presence of intrauterine growth restriction as covariates, and centre adjusted using a random effect; 95% confidence interval, 1.092.17;  $P=0.017$ ).

**Conclusions:** Neonates randomised to a higher platelet transfusion threshold of  $50 \times 10^9/L$  compared to  $25 \times 10^9/L$  had a higher rate of death or significant neurodevelopmental impairment at two years corrected age. There is no evidence that prophylactic platelet transfusion reduces bleeding and increasing evidence that it causes harm which continues into childhood.

15.20

Neonatal

**SERUM ALANINE LEVELS IN PERINATAL ASPHYXIA AND NEONATAL HYPOXIC ISCHAEMIC ENCEPHALOPATHY**

**D Clifford**<sup>1,2</sup>, DS O'Boyle<sup>1,3</sup>, AA Garvey<sup>1,3</sup>, E Dempsey<sup>1-3</sup>, SJ Costelloe<sup>4</sup>, J Bierau<sup>5,6</sup>, GB Boylan<sup>1,3</sup>, BH Walsh<sup>2,3</sup>, DM Murray<sup>1,3</sup>

<sup>1</sup>Department of Paediatrics and Child Health, Cork University Hospital, Cork, <sup>2</sup>Neonatal Department, Cork University Maternity Hospital, Cork, <sup>3</sup>INFANT Research Centre, University College Cork, Cork, <sup>4</sup>Department of Clinical Biochemistry, Cork University Hospital, Cork, <sup>5</sup>Department of Clinical Genetics, Maastricht University Medical Centre, Maastricht, The Netherlands, <sup>6</sup>Department of Clinical Genetics, Erasmus MC University Medical Centre, Rotterdam, The Netherlands

**Aim:** Hypoxic-Ischaemic Encephalopathy (HIE) is a leading cause of neuro-disability, affecting 1-8/1000 live full-term births (1, 2). Early, accurate diagnosis can be difficult. Alanine has been identified as a potentially useful marker in predicting HIE. There are no published studies on normal cord blood alanine levels. We report alanine levels in healthy controls and those with HIE at birth and in the immediate postnatal period.

**Methods:** Prospective study of alanine in cord blood of term infants with perinatal asphyxia (PA), including those who progressed to clinical encephalopathy (HIE) were recruited at birth, with matched healthy controls. A separate cohort of term infants was recruited to quantify postnatal alanine, during the first 12 hours of life. Alanine concentration was quantified using UPLC-MS/MS. The reference interval was derived following CLSI guidelines where the robust method is advised when the sample size is smaller than 120, which would be a requirement for using the recommended non-parametric method.

**Results:** There was a significant difference in cord blood alanine concentrations; median 438mmol/L (360-516) in controls, 509 (452-543) in PA, 515 (460-611) in HIE,  $P < 0.001$ . Across grades of HIE the difference was also significant: mild 497mmol/L (439-590), moderate 545 (465-598), severe 733 (556-808),  $P = 0.015$ . The reference interval derived was 213-675mmol/L. Postnatally, levels were highest in the first 3 hours and decreased by 6-12 hours of life. Mean (SD) mmol/L alanine was 686.7 (261.6,  $n = 17$ ) in the first 3 hours of life, 648 (265.3,  $n = 6$ ) at 4-6 hours and 540 (108.6,  $n = 4$ ) at 6-12 hours.

**Conclusion:** Alanine levels are elevated in cord blood of infants with HIE, increase with grade of HIE, and remain elevated for 6 hours after delivery. We report reference intervals for cord alanine concentrations in healthy term neonates. Alanine levels may aid in the clinical decision to initiate neuroprotective therapies.

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1

General Paediatrics

### **AN INFANT WITH DANCING EYES**

**AYA ABDALLA, ALOK KUMAR**

<sup>1</sup>General Paediatrics, Kerry university hospital, Kerry, Ireland

**Background:** Nystagmus is a non painful involuntary rhythmic oscillation of the eyes that can be caused by a slow drift of fixation which is followed by a fast refixation saccade (jerk nystagmus) or a slow movement back to fixation (pendular nystagmus) invariably associated with loss of vision, grouped as infantile nystagmus which usually appears in the first 6 months of life and acquired nystagmus which appears later. With a slight predilection to European ancestry the prevalence of infantile nystagmus is 14 per 10,000.

**Case report:** A 16-week-old girl who presented with abnormal eye movement noticed by her parents. There was no illicit drugs, medications or alcohol exposure perinatally. She was a term baby delivered normally, with no complications and was vaccinated up to date

She is following her developmental milestones with no parenteral concerns

There was no family history of albinism, reduced visual acuity, optic nerve or retinal disease, metabolic or genetic disorder but her mom has nystagmus since young age and was undiagnosed. This child had anthropometric measurements were within appropriate centiles for age and sex. Eye examination showed bilaterally present red reflexes, normal funduscopy and bilateral horizontal



oscillating movements ,visual acuity and field examination was unapplicable .Her neurological examination including cranial nerves, tone, power and reflexes, cerebellar examination was unremarkable

The differential diagnoses were infantile nystagmus, spasms mutans, infantile squint syndrome and acquired nystagmus neurological syndrome.

Neuroimaging in form of cranial ultrasound and MRI was normal. Subsequently an ophthalmological referral was made for further assessment and management. Eventually the diagnosis of infantile nystagmus was made

**Conclusion:** Nystagmus is a common physical sign, seen in a large variety of ophthalmological and neurological conditions

It is important to differentiate between infantile and acquired nystagmus Neurological disease should be suspected when the nystagmus is asymmetrical or unilateral with onset after 4 months and with associated papilledema or optic atrophy.

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2

Sub-Specialty

**A QI FOLLOW UP PROJECT ASSESSING SUSTAINED REDUCTION IN USE OF INFLAMMATORY MARKERS IN IBD PATIENTS PRESENTING FOR INFLIXIMAB INFUSIONS.**

**AYA ABDALLA<sup>1</sup>**, Ths Arunan, S Keogh, AM Broderick  
<sup>1</sup>GASTROENEROLOGY ,CHI CRUMLIN DUBLIN IRELAND

**Background:** ESR and CRP are completed as part of routine bloods as a surrogate marker of disease activity for youngsters with IBD attending for regular infusions with a cost of €1656. These inflammatory markers have not been shown to be an accurate marker of disease severity in absence of clinical activity. Previously a quality improvement project was performed including an educational intervention to decrease unnecessary use of inflammatory markers in these patients. A significant decrease was demonstrated, however it was unclear whether this change was sustained past the study period or not.

**Methods:** The number of inflammatory markers completed in the month of May 2022 prior to infliximab infusions for youngsters with IBD attending Day Wards at CHI was recorded. This was then compared with data collected and audited in November 2021 as part of the initial QI project.

**Results:** 57% 77/135 of patients attending for routine infusions had either an ESR and/or CRP performed in May 2022, costing €2835. Only fifty eight percent (45/77) had elevated ESR and or CRP. €1,179 could have been saved this month.

135 patients presented to the Day Unit for infusions in May 2022 compared to 177 and 94 in November and December 2021 respectively,

57% patients were tested for inflammatory markers, compared to 97% in November prior to the intervention. However, the May 2022 figure has increased from 33% for December 2021, the month immediately after the intervention.

58% had an elevated ESR and or CRP result in May 2022 which is slightly increased from 40.5% of patients with elevated CRP and or ESR in December 2021.

**Conclusion:** There continues to be a sustained reduction over two years in the overall number of inflammatory markers being performed. However, a repeat educational intervention may be necessary to ensure the sustainability in the medium and long term.

3

General Paediatrics

### TRACHEOESOPHAGEAL FISTULA: MULTISYSTEM PAEDIATRICIAN'S GUIDE TO FOLLOW-UP

**S Agarwal<sup>1</sup>**, C Brophy<sup>1</sup>, C Eddershaw<sup>1</sup>, C Gaughan<sup>1</sup>, O Horner<sup>1</sup>, C Keenan<sup>1</sup>, J O'Grady<sup>1</sup>, D Noone<sup>1</sup>, A Branagan<sup>1,2</sup>, G Semova<sup>1,3</sup>, E Isweisi<sup>1,3</sup>, E Roche<sup>1,3,4</sup>, J Meehan<sup>1,3</sup>, E Molloy<sup>1,2,4</sup>

<sup>1</sup>Discipline of Paediatrics, School of Medicine, Trinity College Dublin, Dublin, Ireland

<sup>2</sup>Neonatology, Coombe Women and Infants University Hospital, Dublin, Ireland

<sup>3</sup>Paediatrics, Children's Health Ireland at Tallaght, Dublin, Ireland

<sup>4</sup>Trinity Research in Childhood Centre, Trinity College Dublin, Dublin, Ireland

**Aims:** Tracheoesophageal fistula (TOF) is the most common congenital abnormality of the oesophagus, occurring in ~ 1:2500–4500 live births. This article provides a concise overview by system of associated complications, illnesses, and abnormalities commonly found in children who have been treated for TOF. TOF is commonly managed by paediatric surgeons, however, there is a vast array of complications relevant to the general paediatrician when managing follow-up of children with TOF. This article provides advice for the general paediatrician on the recognition and management of such complications.

**Methods:** Gastrointestinal complications following repair include anastomotic stricture, gastroesophageal reflux disease, recurrent TOF and dysphagia. Respiratory problems include aspiration, bronchiectasis, wheeze, asthma and tracheomalacia. Congenital heart defects are common comorbidities, reported in up to 30% of patients with TOF. TOF commonly occurs as part of, or alongside syndromes and genetic associations, including the VACTERL (vertebral defects, anal atresia, cardiac defects, TOF, renal anomalies and limb abnormalities) association, CHARGE (coloboma, heart defects, atresia choanae, growth retardation, genital abnormalities and ear abnormalities) syndrome and trisomies 18 and 21. Figure 1 provides a system-based summary of the complications and abnormalities to be aware of during follow-up of TOF patients.

**Results:** Figure 1: Summary of complications and abnormalities to be aware of during follow-up of TOF patients, organised by system. (VSD- ventricular septal defect, ASD-atrial septal defect, PDA- patent ductus arteriosus).

**Conclusions:** Ultimately, a multidisciplinary approach is essential in the long-term management of these patients, with the general paediatrician playing a central role in the coordination of many aspects of their care.

4

General Paediatrics

#### 20q11-q12 DELETION SYNDROME- A CASE REPORT

AA AHAD, MP PARDY, IA AHMAD

PAEDIATRIC UNIT, UNIVERSITY HOSPITAL LIMERICK, LIMERICK, IRELAND

**BACKGROUND AND AIMS:** Chromosome 20q11-q12 deletion syndrome is a multisystemic disorder with variable features. It is characterized by global developmental delay, poor overall growth, sometimes with severe feeding difficulties, facial dysmorphism, and distal skeletal anomalies. Some patients may have hearing impairment, retinopathy, or cardiac defects.<sup>1</sup> Our aim is to report a case of 20q11-q12 deletion syndrome who in addition to the above developed infantile spasms.

**METHODS:** We describe the case with unique clinical presentation, including history and physical exam findings, laboratory workup, genetics, electro physiologic studies, radiology studies, treatment and outcome. A review of the current available literature on 20q11-q12 deletion syndrome was also undertaken.

**RESULTS:** The patient is a 9 months old now and initially presented with failure to thrive on background of significant antenatal history such as IUGR, antenatal renal pelvic dilation and enlarged ventricles with a choroid plexus cyst. He also had a significant early neonatal history as he was admitted at 4 hours of age with hypoglycaemia, temperature instability breathing difficulty and poor feeding. micrognathia, low birth weight, coronal hypospadias with hooded foreskin and axial hypotonia were the significant physical findings. CGH array yielded a diagnosis of 20q11-q12 deletion syndrome.

Cardiology and ophthalmology assessments were normal. Follow up renal and brain imaging were also done.

He presented with multiple seizure like episodes aged 10 weeks and was started on levetiracetam as his EEG was abnormal. At the age of 8 months, primary seizure type semiology changed such as he started having forward head drops with flexion of arms. These abnormal movements were occurring in clusters, a repeat EEG showed modified hypsarrythmia which was suggestive of infantile epileptic spasm syndrome (IESS).

**CONCLUSION:** Failure to thrive, feeding problems dysmorphic feature of micrognathia are established features of the syndrome, our case is unique as he presented with IESS, which is not reported in the literature.

## OUTCOME OF PAVLIK HARNESS USE IN DEVELOPMENTAL DYSPLASIA SCREENING CLINIC

L.O. AKINLABI<sup>1</sup>, L KIRK<sup>2</sup>, A RAHMAN<sup>1</sup>

<sup>1</sup>Department of Paediatrics, Cavan Monaghan General Hospital, Cavan, Ireland

<sup>2</sup>Department of Physiotherapy, Cavan Monaghan General Hospital, Monaghan, Ireland

**Aims:** this study aims at reporting the outcomes of infants placed in Pavik Harness (PH) in the DDH Screening Clinic of our Hospital. We also look at the risk factors most associated with Developmental Dysplasia of the Hips (DDH) referrals to Orthopaedic Service.

**Method:** Attendance and outcomes records of our Clinic for period November 2019 to October 2021 were reviewed and data extracted. Data extracted include gestation, risk factors, ultrasound scan results, Pavlik Harness (PH) use and outcome of PH use.

**Results:** 488 infants were screened in the Clinic during this period of which 27 (5.5%) had abnormal Hip Ultrasound Scan (HUSS) results of varying Graf Classifications while 461 (94.5%) had normal results.

After 8 weeks in PH, 17 (63%) infants had normal HUSS and were followed with Hip X-ray check at age six months while 10 (37%) were referred to the Orthopaedic services as Developmental Dysplasia of the Hips (DDH) needing further intervention.

The worse the Graf Classification of the Hips on the first HUSS the higher the chances of needing orthopaedic referral. Breech presentation at birth was responsible for majority of the Orthopaedic referral followed by first degree family history.

**Conclusions:** Our study of 2 years activities in our Clinic revealed 27 of the 488 patients had varying degrees of abnormalities on their Hip Ultrasound scans done between weeks 5 and 8 of life. 37% of these infants required further Orthopaedic referral after 8 weeks in Pavlik Harness while 63% improved. 60% of the referred infants were breech presentation at birth and only 10% had first degree family history.

## ASSESSMENT OF THE ADRENAL FUNCTION IN CHILDREN WITH ACUTE LYMPHOBLASTIC LEUKEMIA BEFORE AND AFTER INDUCTION THERAPY WITH CORTICOSTEROIDS

M Al Obaidani<sup>5</sup>, D Khater<sup>1,2</sup>, S Al Yaarubi<sup>1</sup>, N Al Dhawi<sup>3</sup>, I Ullah<sup>1</sup>, Y Wali<sup>1</sup>, A Saad<sup>4</sup>, M Elshinawy<sup>1,2</sup>

<sup>1</sup>Child health department , Sultan Qaboos university hospital , Muscat, Oman

<sup>2</sup>Heamatology department, Sultan Qaboos university hospital , Muscat, Oman

<sup>3</sup>Oman Medical speciality Board , Oman Medical Speciality Board , Muscat, Oman

**Aim of the Study:** To assess the adrenal gland function of children with acute lymphoblastic leukemia before and after induction therapy of corticosteroids.

**Patients and Methods:** The study involved all newly diagnosed children with ALL during the period from December 2016 till end of January 2018. Study patients with either B or T cell acute lymphoblastic leukemia were recruited from pediatric hematology/oncology ward at Sultan Qaboos University Hospital. All patients had diagnostic full blood count, included hemoglobin levels (Hb), platelets count, white blood cell count (WBC) and absolute neutrophil count (ANC), bone marrow aspirate and bone marrow trephine biopsy. They underwent initial Adrenocorticotrophic hormone (ACTH) and serum cortisol levels assessment before and after induction therapy with either prednisolone or dexamethasone.

**Results:** The mean baseline ACTH values of the studied patients were within normal range in all the studied patients. Baseline serum cortisol levels were significantly higher than their follow-up values at the end of the induction treatment.  $T=2$  and  $P \leq 0.000$ . Of note that only 23 patients had their post induction cortisol levels measured due to various reasons (patient shifted to PICU, the treating physician didn't request the test, patient left the hospital for treatment abroad and sample was insufficient).

**Conclusion:** Glucocorticoids are essential in the treatment of ALL. Adrenal insufficiency can be a major side effect of this treatment. Cortisol level assessment must be obtained after steroids discontinuation for all patients. In addition, steroids replacement therapy should be started immediately if abnormal levels were detected and follow up is required till recovery.

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## GALL STONES ON THE RISE IN PEDIATRICS POPULATION

A.A Ali, A.K Kahlil, A.K Kumar, A.C Chaudry

<sup>1</sup>Paediatrics, UHK, Tralee, Ireland

**Background:** Historically gallstones are rare in the pediatrics age group. However, over the past decades the incidence is on the rise(1). This is thought to be mainly due to the rising incidence of pediatric obesity and the survival of the sick extremely premature neonates (1).

**Aim:** To raise awareness of the increasing incidence of gallstones in pediatrics age group.

Methods:

To report a case of gallstones in a previously healthy child.

**Result:** A previously healthy 12 years old girl presented with recurrent epigastric pain for 6 months, colicky, radiating to the lateral side of the chest, lasting for 30 mins, associated with vomiting, no relieving or aggravating factors. Positive family history of IBS, Crohn's disease and gallstones. Had menarche at 10 years and having regular periods. Vitally stable and normal clinical examination. Her BMI was 24.FBC,U&E,LFT,CRP,HBA1c,coeliac screen, TFTs and hormonal profile were all within normal range. Amylase was slightly elevated. US abdomen revealed cholelithiasis. She underwent laparoscopic cholecystectomy.

**Discussion:** Population based study estimated prevalence of gallstones in children at 1.9 % (2). The most common type pigmented stones 20-40% (3) from hemolytic diseases. The prevalence of Cholesterol stones is 21% (1). They are mainly formed of bile supersaturation with cholesterol (1).The prevalence of cholesterol stones increases with age, obesity, family history, parity, female sex and early menarche(1).Other types of the stones are protein dominant stones.

The majority of children with cholelithiasis are asymptomatic (1).The signs and symptoms include upper quadrant abdominal pain with nausea and vomiting. Fever and jaundice may also be apparent. On physical examination, there may be right upper quadrant tenderness with positive Murphy's sign. All laboratory results are normal in simple cholelithiasis. Abdominal US is the investigation of choice.

Asymptomatic gallstones are managed conservatively. Cholecystectomy is recommended for symptomatic and complicated gallstones.

**Conclusion:** The incidence of gallstones is rising and pediatricians should be alert.

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## COMPLIANCE WITH MODIFIED CENTOR SCORE FOR STREPTOCOCCAL PHARYNGITIS-A QUALITY IMPROVEMENT STUDY.

IA ALI<sup>1</sup>, IF FAROMBI<sup>2</sup>

<sup>1</sup>Paediatrics Department, Our Lady of Lourdes , Drogheda, Ireland

**Abstract :** Group A Streptococcus (GAS) pharyngitis is currently the only commonly occurring form of acute pharyngitis for which antibiotic therapy is definitely indicated . Empirical antimicrobial treatment in children with sore throat and a high clinical suspicion of GAS pharyngitis will still result in significant overtreatment of non-streptococcal pharyngitis. This is costly and leading to emerging antibiotic resistance.

**Aim:** The aim of our study was to evaluate compliance with Modified Centor score, in determining the use of antibiotics in children who presented to emergency department of Our Lady Of Lourdes Hospital Drogheda , with symptoms of upper respiratory tract infection

### Methodology:

*Study duration:* 1<sup>ST</sup> May to 30<sup>TH</sup> July 2022

*Inclusion criterion:* children between age 3 to 15 years presenting to ED with symptoms suggestive of URTIs. *Standards:* Modified Centor Score Criterion for determining throat swab cultures and antibiotics prescription

**Results:** Out of 30 children,7 cases of children scored 0-1(subgroup1), 14 cases of children were scored 2-3(subgroup2)and 9 cases scored 4-5(subgroup3)

*Subgroup 1:*2 children were given oral antibiotics and no swabs were taken in any child. Adherence to guideline in terms of throat swabbing and oral antibiotics was 100% and 71% respectively.

*Subgroup 2:*4 children had their throat swab for GAS taken and 8 children were treated with antibiotics .

Adherence to guidelines was 28% and 42% respectively.

*Subgroup 3:*Throat swab for GAS culture was taken in 2 children and 8 children were given treatment with antibiotics. Adherence to guidelines was found to be 22 % and 88% respectively.

**Conclusion/Recommendations:** These findings suggest poor adherence to Modified Centor Score guidelines. Also significant number of children were treated with antibiotics despite of low score on MCS criterion, which is a leading cause of antibiotic resistance. Following recommendations were given to improve outcome.

- To avoid unnecessary antibiotics, treatment should be delayed until culture results are available. Also children with negative cultures should discontinue any antibiotics already started if clinically stable.
- To create awareness about importance of Modified Centor Score for determination of antibiotic usage.

Re-audit (prog)

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## **USE OF METERED DOSE INHALER VS NEBULISED SALBUTAMOL IN ACUTE WHEEZE IN A REGIONAL PAEDIATRIC CENTRE**

A Ali<sup>1</sup>, D Huggard <sup>1</sup>

<sup>1</sup>Paediatric , University Hospital Waterford, Waterford, Ireland

**Aims:** A validated paediatric asthma clinical scoring system (the Paediatric Respiratory Assessment Measure (PRAM)) helps classify the initial severity and clinical response of respiratory distress in children. This scoring system can facilitate the appropriate use of bronchodilators and potentially reduce hospitalisations. We assessed our current practice in UHW in treating acute wheezing against a recommended acute asthma management guideline for CHI.

**Method:** A retrospective chart review was performed targeting patients from 1 to 16 years, who received bronchodilator treatment between September 2021 and January 2022. Based on the severity (PRAM score) we categorised patients as mild, moderate or severe and correlated

whether they received bronchodilators via Metered Dose Inhaler (MDI), nebulised or both and compared with CHI guideline.

**Result:** There were 41 patients included: 14.6% (n=6) of patients were <2 years old, 58.6% (n=24) were 2-5 years, and 26.9% (n=11) were > 5 years. The majority of patients (68.3%) did not have a pre-existing asthma diagnosis. Only 7.3% of patients had the severity documented in the notes. Using the PRAM score, 70.1% (n=29) of patients were classified as mild, with the majority receiving MDI treatment initially; 68.9% (n=20). 90% (n=9) of those classed as moderate received nebulised bronchodilators. Almost all patients received oral steroids (90.2% ). Chest x-ray was requested in 19.5% (n=8) and antibiotics were prescribed for 17% (n=7). Almost 80% (n=23) classified as mild were discharged home.

**Conclusion:** This audit demonstrated poor documentation of asthma severity on initial assessment, although with a high (68.9 %) initial utilisation of bronchodilators via MDI in mild cases. Education and implementation of PRAM scoring on triage and for treatment of these patients have the potential to streamline patients based on severity and optimise management which could reduce the length of stay and admissions.

Acute Asthma – Care in the Emergency Department , CHI clinical guidelines, 2021. PRAM on line: <https://www.mdcalc.com/pediatric-respiratory-assessment-measure-pram-asthmaexacerbation-severity>

## **FAMILIAL OUTBREAK OF HAEMOLYTIC UREAMIC SYNDROME SECONDARY TO VEROTOXIGENIC E.COLI INFECTION.**

**K Amin**<sup>1</sup>, W Asif<sup>2</sup>, AP Macken<sup>1</sup>

<sup>1</sup>Department of Paediatrics, Tipperary University Hospital, Clonmel, Ireland.

<sup>2</sup>Department of Paediatrics, St Luke's Hospital, Kilkenny, Ireland.

**INTRODUCTION/AIMS:** Haemolytic ureamic syndrome (HUS) is defined by the simultaneous occurrence of microangiopathic haemolytic anaemia, thrombocytopenia, and acute kidney injury. HUS is generally preceded by a prodromal illness with abdominal pain, vomiting, and bloody diarrhoea. Diagnosis is by full blood count, peripheral blood smear, renal function studies (serum creatinine), and urinalysis. Prompt identification and management can help prevent renal failure, hypertension and/or neurological manifestations.

**METHODS:** A retrospective case study was undertaken of two cousins with HUS, examining mode of transmission, presentation, clinical course and subsequent management.

### **RESULTS:**

#### **Case 1**

Following a family barbeque a 2.5 year old boy presented with vomiting, bloody diarrhoea and dark coloured urine. He was hypertensive 120/70mmHg. Urinalysis revealed haematuria and proteinuria. His haemoglobin was 9.2g/dl, platelets 29, urea 18.2mmol/L, creatinine 51 umol/L and his urinary protein: creatinine ratio was 2150mg/mmol. His stool cultured verotoxigenic E.Coli,VT2. Subsequently his haemoglobin dropped to 7.5g/dl, warranting 2 transfusions. He was discharged on day 5 with day ward follow up.

#### **Case 2**

A 4-year-old girl, first cousin of case 1, who attended the same barbeque was admitted to hospital in a neighbouring county with similar symptoms. She was hypertensive, her urinalysis revealed haematuria and proteinuria. Her haemoglobin was 9.6g/dl, platelets 9, urea 10.0mmol/L, creatinine 100 umol/L and her urinary protein: creatinine ratio was 64mg/mmol. Her haemoglobin further dropped to 7.3g/dl, needing 3 red cell transfusions and required transfer to a Nephrology Centre for management of acute renal failure. She recovered without dialysis and discharged 2 weeks later to outpatient follow up.

**CONCLUSIONS:** These two cases illustrate the variation in disease severity and clinical course of HUS precipitated by Verotoxigenic E.coli.

## **A SURPRISING PRESENTATION OF COARCTATION OF AORTA IN A 9 YEAR OLD BOY**

**K Amin<sup>1</sup>, M Saleem<sup>1</sup>, AP Macken<sup>2</sup>, AR Herrera<sup>1</sup>**

<sup>1</sup>Department of Paediatrics, St Luke's hospital, Kilkenny, Ireland

<sup>2</sup>Department of Paediatrics, Tipperary university hospital, Clonmel, Ireland.

### **INTRODUCTION/AIMS:**

Coarctation of Aorta (CoA) usually presents in the newborn period but the age of presentation can be delayed to later in life depending on the degree of obstruction and collateral vasculature. Screening for this abnormality is key in newborn assessment facilitating prompt referral to specialist services for intervention. It also highlights the importance of careful evaluation of peripheral pulses and blood pressure in a child.

### **METHOD:**

We reviewed the case of a 9-year-old boy with a late and atypical presentation of CoA.

### **RESULTS:**

A previously well, very active and sporty 9-year-old boy presented with sudden, atypical chest pain at rest and tingling sensations in his legs and arms. His lower limb BP was >20mmHg less than that of his upper limbs. He was extremely hypertensive with his brachial arterial pressure of 160/90mmHg. Radio femoral delay was present with diminished femoral pulses. Chest examination, ECG, Chest Xray and Abdominal ultrasound were normal. Echocardiography revealed discrete coarctation of the descending aorta with severe narrowing distal to the subclavian artery, with a gradient of 65mmHg. He was also noted to have dilated head and neck vessels. A subsequent cardiac catheterization was performed with aortic stent placement, approximately 32mm into descending aorta. He was started on atenolol 25mg for his systemic hypertension.

### **CONCLUSION:**

CoA should be a differential diagnosis for any child presenting with atypical chest pain and/or hypertension. Along with careful history taking and clinical examination, assessment of the brachial and femoral pulses and pressures are key in establishing the presence or absence of CoA. Pre and post ductal pulse oximetry is a useful screening tool at birth.

**Paediatrics CONGENITAL MYASTHENIC SYNDROME TYPE 4A: A CASE REPORT AND LITERATURE REVIEW**

**A Biagini<sup>1</sup>**, K Lynam<sup>1</sup>, L Mannion<sup>1</sup>, MJ O'Grady<sup>1, 2</sup>, S O'Doherty<sup>1</sup>

<sup>1</sup>Department of Paediatrics, Regional Hospital Mullingar, Mullingar, Co. Westmeath

<sup>2</sup>Women's and Childrens Health, School of Medicine, University College Dublin, Belfield, Dublin 4 , Co. Dublin

**Aim:** To describe the clinical presentation, differential diagnosis, investigation and management of congenital myasthenic syndrome (CMS) Type 4A and review the relevant literature.

**Methods:** The patients' medical records were reviewed for clinical details and investigations. A literature review regarding CMS was conducted.

**Results:** A 19-month-old girl presented with restricted eye movements with a paralysis of upward gaze in both eyes as well as reduced movement on right lateral gaze in the right more than in the left eye and twitching of the eyelids. There was no additional clinical evidence of muscle weakness or easy fatiguability. Family history was non-contributory and parents were non-consanguineous. An ophthalmological assessment diagnosed chronic progressive external ophthalmoplegia. Preliminary investigations for mitochondrial disorders and mutations in POLG and PEO1 were negative. A gene panel identified a missense variant c.865C>T in CHRNE gene, that is linked with autosomal dominant slow channel congenital myasthenic syndrome type 4A. At age 5 the proband was noted to have developed ptosis. She is now aged 13 and has not developed any further manifestations and she is not currently on medication.

**Conclusion:** Congenital myasthenic syndromes (CMS) include a variety of genetic disorders characterized by genotypic and phenotypic heterogeneity. The impairment of the neuromuscular transmission clinically manifests as fatigable muscle weakness in all subtypes of CMS but age at onset, presenting symptoms, distribution of weakness, and response to treatment can largely vary depending on the CMS subtype.

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### SEVERE ACUTE HEPATITIS ASSOCIATED WITH RSV INFECTION. A CASE REPORT.

M Bonadonna<sup>1</sup>, S Koe<sup>1</sup>

<sup>1</sup>Paediatrics, CHI at Tallaght, Dublin, Ireland

**Introduction:** Respiratory syncytial virus (RSV) infection is the most common cause of lower respiratory tract infection in children <1 year of age. RSV is highly restricted to the respiratory epithelium and is shed apically into the lumen of the airways. In rare cases, RSV may be recovered from extrapulmonary tissues, such as liver, cerebrospinal fluid or pericardial fluid. The aim was to present a 6 years old girl diagnosed with airway infection and hepatitis during the RSV infection.

**Material and methods:** AST (aspartate aminotransferase), ALT (alanine transaminase), GGT (gammaglutamyl transferase) and bilirubin concentrations were monitored during her hospitalization and at control examination. An acute cholestatic hepatitis workup was performed.

**Results:** The child had minor symptoms of respiratory infection but had clinical signs of liver disease. On admission her liver function tests were abnormal with AST levels (2008 IU/L) greater than 50 times the upper limit of normal, ALT levels increased 77-fold (2702 IU/L), GGT 9-fold (229 IU/L) and total bilirubin 4.3fold (74 umol/L) in comparison with the upper reference limits. Two weeks after discharge there was a drastic reduction of the catalytic enzymes but AST, ALT and GGT remain mildly increased. Screening for viral hepatitis was negative. RSV infection was confirmed in the respiratory molecular PCR test. In addition her connective tissue disease screen, as mitochondrial antibodies, parietal cell antibodies, smooth muscle antibodies and titre, liver-kidney microsomal antibodies and ANCA, were all negative. Her abdominal ultrasound did not show any abnormalities in the visualized abdominal organs (liver, bile ducts, gallbladder, pancreas, spleen, kidneys).

**Conclusion:** Acute hepatitis may occur with RSV infections. There may be increase in ALP (alkaline phosphatase), AST, ALT, GGT and bilirubin. In May 2022, the European Centre for Disease prevention and control published guidelines for diagnostic testing of cases of hepatitis of unknown aetiology in children.

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## THE OUTCOME OF MEDICAL INVESTIGATIONS IN CHILDREN WITH AUTISM SPECTRUM DISORDER (ASD).

E.B Burns Beattie<sup>1</sup>, P.C Curran<sup>1,2</sup>

<sup>1</sup>Graduate Entry Medical School, University of Limerick, Limerick, Ireland

<sup>2</sup>Portiuncula University Hospital, Ballinasloe, Galway, Ireland

**Aims:** International guidelines recommend that children with autism spectrum disorder (ASD) are offered genetic tests as part of their medical assessment. The aim was to describe the outcome of medical investigations in children with ASD in our locality. Genetic investigations included chromosomal microarray, Fragile X testing (FXT) and blood tests for iron deficiency anaemia and vitamin D deficiency.

**Methods:** Ethics approval was granted by the Galway Clinical Research Ethics Committee in July 2022. We identified 150 children diagnosed with ASD through our database. 12 children were excluded for three reasons: discharged from hospital, over 16 years of age or having a recognised syndrome.

**Results:** 69 of 138 children (27 female, 111 male) offered blood tests agreed to testing, 53 (11 female, 58 male) gave consent.

6 of 49 (12.25%) children with microarray testing had chromosomal imbalances, 2 associated with ASD (chromosome15q deletion, chromosome1q21 duplication), 4 had benign copy variants. None of 42 FXT analysed were positive.

Vitamin D levels were available in 20 of 53 patients, 7 of 20 (35%) were vitamin D deficient (< 50nmol/l), range 20-43nmol/l and 3 of 40 (7.5%) had iron deficiency anaemia.

**Conclusion:** As FXT has a pick up rate internationally of 0.5-2% in ASD and our tests were negative, FXT may be better as a second-line genetic test. A chromosomal microarray positive rate associated with ASD of 4% is lower than Irish and international studies (10-20%). These genetic results may represent the investigated population or bias due to numbers tested. Vitamin D deficiency was found in 35% of children tested, a higher incidence than Irish studies of vitamin D in children without ASD (4-6%). Two had iron deficiency anaemia possibly reflecting restricted/limited diets. Results will give important information on the outcome of medical investigations and detailed dietary assessments are vital to medical assessments in children with ASD.

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### **SERVICE EVALUATION OF BLENDED DIET BY ENTERAL FEEDING TUBES**

G McElligott<sup>1</sup>, M Butler<sup>1</sup>, D Griffin<sup>1</sup>, Z Yusuf<sup>1</sup>

<sup>1</sup>Paediatrics, Cork University Hospital, Cork, Ireland

**AIMS:** The prevalence of home enteral tube feeding (HETF) is increasing in the paediatric population, with commercial enteral formula usually considered first choice. Blended diet, however, is growing in popularity, with families requesting to use home-made foods entirely or in combination with a commercial feed. This area is under researched and there are concerns that blended diet by enteral feeding tube is unsafe and nutritionally incomplete in comparison to a commercial feed. This service evaluation investigated the family experience and reviewed biochemical markers of children on blended diet by enteral feeding tube.

**METHODS:** This was a mixed methods service evaluation. It included a semi-structured questionnaire administered by a clinician over the phone to all (n=9) parents of children on blended diet by enteral feeding tube attending the Paediatric Department at Cork University Hospital. Biochemical monitoring on all nine children was reviewed pre and post commencement of the blended diet.

**RESULTS:** Nine children were included, with a mean age of 6 years and 10 months, and a mean time on blended diet of 2 years and 8 months. All children had a gastrostomy tube. Families reported subjective improvement in vomiting, diarrhoea, heart burn and reflux, sleep quality, communication and engagement, behaviour, cognitive function, and skin health. Families found hospital Dietitians to be the best source of knowledge and support. Families unanimously had a positive experience introducing blended diet. After starting blended diet children had biochemical markers within the normal range, apart from one child with a low ferritin.

**CONCLUSION:** This service evaluation demonstrates that families feel blended diet by gastrostomy clinically and holistically benefits their child. Recent guidelines by the British Dietetic Association supports Dietitians in safely using blended diet where it may be beneficial. Further research, however, is required to understand the safety profile and beneficial effects of a blended diet by HETF.

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2. BDA. Nov 2021. 'Practice Toolkit: The Use of Blended Diet with Enteral Feeding Tubes', The Association of UK Dietitians, pp 1-80.
3. Ingleby C, Yusuf Z, and Hindley C. Feb 2021. 'Service evaluation of blended diet by gastrostomy', *Dietetics Today*, pp 36-40.

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**AN UNUSUAL CASE OF EARLY GROWTH FAILURE SCHAAF-YANG SYNDROME - A CASE REPORT**  
**S CAVALLARI<sup>1</sup>, C O'SHEA<sup>1</sup>, S GALLAGHER<sup>1</sup>**

<sup>1</sup>PAEDIATRICS , UNIVERSITY HOSPITAL LIMERICK, LIMERICK , IRELAND

**BACKGROUND AND AIMS:** Schaaf-Yang Syndrome (SYS) is a rare neurodevelopmental disorder which is associated with growth failure, hypotonia, and distal joint contractures in infancy. It is caused by a genetic variant of MAGEL2 gene on chromosome 15, which may be inherited in an autosomal dominant, maternally imprinted manner, or by a de novo mutation. (1) In later childhood, behavioural abnormalities may present and there is an increased rate of autism. SYS is also associated with early feeding difficulties, which may develop into polyphagia in older children.

Our aim is to report a case of early infantile growth failure caused by SYS in a now eight year old girl.

**METHODS:** We describe the clinical presentation, including history and examination findings, laboratory work, genetics, and radiological findings. A review of the current available literature on SYS was also conducted.

**RESULTS:** The patient is a now eight year old girl who presented in infancy with hypotonia, short stature, abnormal fat distribution, contractures of hands and talipes equinovarus. At age 15 months, growth hormone deficiency was diagnosed and treatment with regular subcutaneous growth hormone was commenced. She is non-verbal, and though she can walk, the child is a wheelchair user for distance outside of the home.

SYS is associated with feeding problems in infancy, however in childhood and adolescence, obesity and food seeking behaviour is more common. Our patient is currently aged eight, and has a severe oral aversion, receiving nutrients solely via fortified supplement drinks.

She has furthermore been diagnosed with other conditions associated with SYS including hypothyroidism and autism spectrum disorder.

**CONCLUSION:** We add our case report to the literature on this rare syndrome. While an unusual cause of growth failure, SYS is worth considering in infants who present with feeding difficulties, global developmental delay, and contractures.

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## MAKING A RASH DIAGNOSIS

L Murphy<sup>1</sup>, L Corley<sup>1</sup>, I Ciocoiu<sup>1</sup>

<sup>1</sup>Department of Paediatrics, University Hospital Galway, Galway, Ireland

**Background:** Stevens-Johnson Syndrome (SJS) and Toxic Epidermal Necrolysis (TEN) are severe type IV immune mediated hypersensitivity reactions. SJS/TEN are considered a disease continuum, distinguished based upon the percentage body surface area (BSA) involved. The disease is characterised by extensive epidermal necrosis and detachment with mucous membrane involvement in 90% of cases. Incidence is rare estimated at 2-7/1,000,000 per year <sup>[1]</sup>.

**Case Report:** We present the case of a previously well 3 year-old boy with a two day history of rash. The rash started as flat erythematous macular lesions which progressed to blistering erosions. It rapidly became more extensive affecting mucosal membranes with involvement of the mouth, eyes and penis. The patient had a background of preceding upper respiratory tract infection two weeks prior. There was no temporal exposure to medication aside from paracetamol and ibuprofen, both introduced following the onset of the rash. Rapid clinical deterioration with ongoing epidermal loss ensued. The patient was subsequently transferred to Crumlin with a working diagnosis of SJS. Initially he was managed at ward level and treated with IV dexamethasone. Subsequent transfer to PICU was required in the setting of progression to TEN with >30% BSA involvement. Sedation was initiated for analgesic effect and a nasogastric tube sited for high calorie feeds. He underwent an amniotic membrane transplant for corneal protection. An extended viral panel was positive for parainfluenza type 2 , likely representing an underlying trigger for his presentation. After a 19 day PICU stay he was transferred to the ward and continued to make a good recovery. Follow up is in place with dermatology and ophthalmology.

**Discussion:** SJS/TEN is associated with significant paediatric morbidity and mortality<sup>[2]</sup>. It is therefore an important consideration and when suspected warrants prompt discussion with expert centres with access to paediatric intensive care. Our case highlights the value of interprofessional collaboration in managing multisystem complications of this rare condition.

1.Frey N, Bodmer M, Bircher A, Jick SS, Meier CR, Spöndlin J. Stevens-Johnson Syndrome and Toxic Epidermal Necrolysis in Association with Commonly Prescribed Drugs in Outpatient Care Other than Anti-Epileptic Drugs and Antibiotics: A PopulationBased Case-Control Study. *Drug Saf.* 2019 Jan;42(1):55-66. 2.Forsyth, J., Russell, E., & McPherson, T. (2022). A retrospective case series of paediatric Stevens-Johnson syndrome and toxic epidermal necrolysis: evaluation of practice using the British Association of Dermatology's National Guidelines for children and young people. *The British journal of dermatology*, 186(1), 197–198. <https://doi.org/10.1111/bjd.20741>

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Sub-Specialty

### CANTU SYNDROME; A RARE CASE OF CONGENITAL HYPERTRICHOSIS

J Heckmann<sup>1</sup>, L Corley<sup>1</sup>, A Byrne<sup>1</sup>, H Stokes<sup>2</sup>, N McGrath<sup>1</sup>

<sup>1</sup>Department of Paediatrics, University Hospital Galway

<sup>2</sup>Department of Paediatrics, Mayo University Hospital

**Background:** Congenital hypertrichosis occurs in isolation or as a component feature of an underlying syndrome. Cantú syndrome is rare and is characterised by hypertrichosis, macrosomia, osteochondrodysplasia, coarse facies and cardiac defects. It is autosomal dominant in inheritance, resulting from gain of function mutations in *ABCC9* or *KCNJ8* which regulate the widely expressed KATP channel. To date, only 150 cases of Cantú syndrome have been reported worldwide<sup>[1]</sup>. We herein describe the first reported Irish case of Cantú Syndrome.

**Case Report:** We present the case of a 2 year-old girl referred with severe hypertrichosis. She is the first child of non-consanguineous parents of South Asian ethnicity. She was born at 35 weeks gestation following a pregnancy complicated by polyhydramnios. Birth weight was 3.5kg (>99.6<sup>th</sup> centile) and her neonatal course was notable for a haemodynamically significant PDA requiring surgical closure. Hypertrichosis was generalised and present since birth. Early developmental milestones were delayed. On examination she was macrocephalic (OFC 57cm >99<sup>th</sup> centile), had coarse facies and marked generalised hypertrichosis. Systemic examination was otherwise normal. Clinical photography was performed with consent and presented to clinical genetics on whose recommendation targeted panel diagnostics was undertaken. Results confirmed heterozygosity for a likely pathogenic missense variant in the *ABCC9* gene



(c.3056C>T;p.Thr1019Ile) consistent with a diagnosis of Cantú Syndrome. Currently, no targeted treatment exists and management involves surveillance, with symptomatic management of secondary complications. The patient is linked with developmental paediatrics and since diagnosis has been referred to cardiology. The family have also been referred to genetic counselling. Ongoing management and surveillance will take place according to Cantú Syndrome guidelines<sup>[2]</sup>.

**Discussion:** This case demonstrates the importance of a multidisciplinary approach to comprehensive multisystem history and examination in yielding an underlying rare diagnosis. Our case provides increased awareness to physicians and expands on known phenotypical associations of Cantú syndrome.

1.Grange, D., Nichols, C. and Haaften, G., 2019. Cantú syndrome: Findings from 74 patients in the International Cantú Syndrome Registry. *American Journal of Medical Genetics Part C: Seminars in Medical Genetics*, 181(4), pp.658-681. 2.Grange DK, Nichols CG, Singh GK. Cantú Syndrome. 2014 Oct 2 [updated 2020 Oct 1]. *GeneReviews*<sup>®</sup> [Internet]. Seattle (WA): University of Washington, Seattle; 1993–2022. PMID: 25275207.

**Poster No 19**  
**General Paediatrics**

### **A CASE REPORT: DIABETES TYPE 1, TYPE 2 OR TYPE 3?**

**R Cullen**<sup>1</sup>, M Comerford<sup>1</sup>, H Fitzgerald<sup>1</sup>, EF Roche<sup>1</sup>, C McDonnell<sup>1</sup>, J Gibney<sup>2</sup>

<sup>1</sup>Paediatric Endocrinology, Tallaght University Hospital, Tallaght, Ireland

<sup>2</sup>Adult Endocrinology, Tallaght University Hospital, Tallaght, Ireland

**Introduction:** Diabetes mellitus is common in children and adolescents. The majority are diagnosed with Type 1 diabetes (T1D), a smaller number Type 2 diabetes (T2D) and rarely Type 3c diabetes (T3cD). The classification can prove challenging, evidenced by the following unusual presentation.

**Description:** Patient A presented age 15 with polyuria, polydipsia and weight loss. Laboratory results showed serum glucose 14mmol/L, pH of 7.34 and negative ketones, consistent with new

T1D without ketoacidosis. Treatment with multiple daily injections of insulin glargine and insulin aspart was commenced with typical T1D level doses and finger prick blood sugar monitoring.

Following complaints of abdominal pain, clinical examination revealed epigastric tenderness and hepatosplenomegaly. Investigations showed: elevation of alanine transaminase (78) and gamma glutamyl transferase (53); normal amylase (51); elevated cholesterol (5.9) and triglycerides (5.75) with diffuse hepatic steatosis on liver ultrasound.

Unusual presentation of T2D or lipodystrophy was considered. Extended liver panel; insulin autoantibodies, including IAA, GAD and ZnT8 and a monogenic diabetes screen returned negative.

Despite intensive care and Dexcom continuous glucose monitoring her HbA1c remains consistently above 9.2% (77mmol/mol). Her brother has coeliac disease, both parents T2D and father hyperlipidaemia.

Past medical history includes increased BMI (latest 30.9) requiring regular dietician input, coeliac disease and oligomenorrhoea suggestive of evolving polycystic ovarian syndrome. Episodic epigastric and right iliac fossa pain necessitated surgical follow up, subsequently an ovarian cyst was diagnosed, this failed to fully explain her symptoms.

This prompted re-exploration of the initial diagnosis of T1D and broadening of differentials including pancreatitis, consistent with T3cD. Her care was transferred to the specialist adult T3cD clinic.

**Conclusion:** Adult T3cD is increasing in prevalence, many initially misclassified as T1D or T2D. Data is limited for the adolescent population. It is important to explore T3cD as a differential, keeping specific risk factors and epidemiology in mind.

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2. Ewald, N. and Bretzel, R., 2013. Diabetes mellitus secondary to pancreatic diseases (Type 3c) — Are we neglecting an important disease?. *European Journal of Internal Medicine*, 24(3), pp.203-206.
3. Gudipathy, L. and Rickels, M. The Pancreapedia: Exocrine Pancreas Knowledge Base, 2015. Pancreatogenic (Type 3c) Diabetes.

## **INCIDENTAL FINDING OF TOXIC AMINOPHYLLINE LEVEL IN CASE OF ACUTE, SEVERE ASTHMA EXACERBATION IN OBESE PATIENT WITH KNOWN BRITTLE ASTHMA**

**J Dalmeny**, J Clarke, E Fitzgerald, B Linnane

<sup>1</sup>Department of Paediatrics, University Hospital Limerick, Limerick, Ireland

This presentation/poster examines the case of a 12 year old female with brittle asthma, who has had 13 exacerbations necessitating admission this year alone. The patient has class 1 obesity with a BMI of 32.6 kg/m<sup>2</sup> (Height: 147 cm; Weight: 70.5 kg) and received an aminophylline infusion (Loading dose: 330 mLs; Maintenance dose: 45 mLs/kg/hr) as per the local asthma exacerbation guideline in University Hospital Limerick.

A subsequent incidental finding of a toxic aminophylline level of 38 mg/L was discovered (therapeutic level: 10-20 mg/L), due to the infusion doses not being adjusted for the patient's ideal body weight for their height (40.1 kg). The ideal loading and maintenance doses for this patient would have been 200 mLs and 28 mLs/kg/hr, respectively.

After this toxic level was highlighted the maintenance dose was adjusted for ideal body weight and a subsequent level returned as 16 mg/L, within the therapeutic range. No adverse outcomes arose as a result and the prescribing error was disclosed to the patient and her mother.

This case offers multiple learning opportunities to doctors, nurses and pharmacists caring for children with severe asthma. As obesity becomes an ever more common public health concern, it is important that prescribers consider whether ideal body weight is required for dosing in paediatrics and what is the best approach to calculating same. While this patient did not exhibit any adverse effects secondary to the suprathreshold level of aminophylline, toxic levels of this drug can be associated with a myriad of adverse drug reactions including hypotension, arrhythmias and seizures.

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<https://www.sps.nhs.uk/articles/how-should-medicines-be-dosed-in-children-who-are-obese/> 4. UHL Paediatric Aminophylline IV Infusion Guideline 5. UHL Paediatric Acute Asthma Exacerbation Management Guideline

**Poster No 21**  
**General Paediatrics**

### **HYPERCALCAEMIA OF DOWN'S SYNDROME – IT'S AS EASY AS ABCD**

**(SYNDROME) P Donovan, N Goggin, A Carroll**

<sup>1</sup>Department of Paediatrics, University Hospital Waterford, Waterford, Ireland

**Background:** Hypercalcaemia of Down's syndrome is a rare under recognized condition consisting of Down's syndrome with a triad of Hypercalcaemia, Hypercalciuria and Nephrocalcinosis. This can present with varying symptoms such as feed intolerance, vomiting or faltering growth and can lead to kidney injury and even death. The hypercalcaemia and kidney failure are both responsive to dietary calcium restriction and adequate hydration.

**Case presentation:** 12-month-old girl with Trisomy 21 being followed in the outpatient setting for faltering growth. Patient having intermittent vomiting illnesses however persisting poor weight gain prompted further investigation which demonstrated raised calcium levels and evidence of kidney injury. Subsequent renal USS showed nephrocalcinosis and diagnosis was made with involvement of Endocrinology. Good response in calcium levels and weight gain was achieved through dietary restriction of calcium by changing to Locasal formula and NG supplemental hydration.

**Conclusions:** Hypercalcaemia of Down's syndrome is a likely under-recognized condition, with this being the 9<sup>th</sup> case reported cases in the literature thus far and the first reported case in Ireland. There has been one reported fatality in the UK wherein diagnosis was made on post-mortem(1). At present mechanism is unknown, however theories regarding the impact of extraneous chromosome 21 on calcium homeostasis in particular increased calcium absorption have been postulated(2). This case highlights need for early consideration for hypercalcaemia screening in patients with Trisomy 21 presenting with faltering growth or vomiting, further workup for those with Trisomy 21 found to have hypercalcaemia and prompt initiation of treatment once diagnosis made.

1. Hosdurga S, Hussain, S., & Fraser, J. Abnormal Calcium, Calcinosis, and Creatinine in Down's syndrome: Case Report. *The Physician*. 2022;7. 2. Ramage IJ, Durkan A, Walker K, Beattie TJ. Hypercalcaemia in association with trisomy 21 (Down's syndrome). *J Clin Pathol*. 2002;55(7):543-4.

## **AN AUDIT OF OXYGEN SATURATION ALARM SETTINGS IN A TERTIARY NEONATAL UNIT**

**P Donovan**<sup>1</sup>, N Redmond, A O'Sullivan, P O'Connor

<sup>1</sup>Department of Paediatrics, The Coombe Woman and Infant's Hospital, Dublin, Ireland **AIMS**

To lessen the harmful effects of hypoxia or hyperoxia, oxygen saturation alarm settings are in place in most neonatal units. We aimed to audit compliance with our local oxygen saturations alarm settings policy.

### **METHODS**

Ethical approval was obtained for this audit. It was a single center audit in a tertiary neonatal unit. Data was collected on 6 randomly selected time points over a 6-week period in March and April 2022, during a mixture of day and night shifts. All preterm and term infants in the NICU or HDU were eligible for the audit, with patients not having oxygen saturations monitoring excluded. The corrected gestation, weight, whether the infant was on oxygen or not and the oxygen alarm settings were collected. This data was compared to the unit policy and deemed compliant or not.

### **RESULTS**

A total of 63 alarm settings were included in the study. 31 were of infants <32 weeks gestation/1500g. 32 were  $\geq$ 32 weeks/1500g. 34 were on infants on oxygen with 40 occurring during the day shift. Overall compliance levels were 41% for the higher limit and 46% for the lower limit. For those in Oxygen compliance rates for higher target were 12%, with equally low levels seen in those  $\geq$ 32 weeks for their lower targets at 17%. There were no differences seen between day and night shift with compliance being between 40-50%.

### **CONCLUSION**

Our compliance rates with alarm limits were low with poor rates amongst those in oxygen with some having higher targets set at 100% which for premature infants can lead to harmful side effects of hyperoxia. This is not out of keeping with findings in other international audits(1, 2), however it does demonstrate appropriate intervention is needed. We plan to assess the effects of as educational session for neonatal nurses and reaudit post this intervention in October.

1. Harris SL, Wolff E, Beecroft G, Troughton R, Austin N. Neonatal oxygen saturation alarm compliance –under recognized source of iatrogenic harm? An audit and survey of nursing opinion. *Journal of Neonatal Nursing*. 2020;26(1):43-8.
2. Clucas L, Doyle LW, Dawson J, Donath S, Davis PG. Compliance With Alarm Limits for Pulse Oximetry in Very Preterm Infants. *Pediatrics*. 2007;119(6):1056-60.

## TOO MUCH OR TOO LITTLE, MEDICATION DOSE BANDING AT HOME

Maeve Mallon, Ben Mc Donald, **Peter Duddy**<sup>1</sup>, Jean Donnelly<sup>2</sup>

<sup>1</sup>Dept of Pharmacy, Coombe Women and Infants University Hospital; School of Pharmacy, University College Cork

<sup>2</sup>Dept of General Paediatrics, CHI at Crumlin; School of Medicine, Trinity College Dublin, University of Dublin

**Introduction:** Paracetamol and Ibuprofen are two of the most commonly used oral medications in children.

They are administered at home by parents using age-based banded dosing, as outlined in product licenses. In hospital, these drugs are prescribed precisely based on the weight of the child.

**Aim:** We aim to compare these two methods of dosing to determine if age-based dose-banding at home leads to excessive or lower doses than hospital weight-based dosing, subsequently leading to potential risks of toxicity or suboptimal dosing.

**Method:** Weights and gender of 100 children 12 years and under, admitted under General Paediatrics, CHI at Crumlin team in June were recorded. Using MS excel, the dose of Paracetamol and Ibuprofen these children would have received at home (based on most popular brand) and in hospital (based on weight) were calculated and maximum daily dose calculated for each.

**Results:** 77% children received more Ibuprofen in hospital v home, 96% received more paracetamol in hospital. On average children under 12 received 123mg more Ibuprofen daily in hospital v home and 405mg more paracetamol daily. 42% of admissions had both charted, 36% paracetamol only and none Ibuprofen only.

**Summary:** A large proportion of presentations to paediatric EDs have pyrexia as a main symptom. Agebased banded dosing while simple and safe to use may be causing children to be “under medicated” causing more presentations to over stretched EDs’. A weight-based system, may provide more clinically appropriate doses but with additional risk of dosing errors.

**Discussion:** We noted a significant discrepancy between home dosing and hospital dosing of paracetamol and ibuprofen. These discrepancies should be considered by regulatory authorities, and other relevant stakeholders in relation to the ongoing use of these medications. Of note the dosing in hospital is “off label”, with the licenced dose being dose banding.

**POST-TRAUMATIC SPINAL CORD INJURY IN THE CONTEXT OF NEUROFIBROMATOSIS****TYPE 1 C Duff, A Lyons**<sup>1</sup>Paediatric Department, Galway University Hospital, Galway, Ireland

Neurofibromatosis Type 1 (NF-1) is a rare developmental syndrome, with an incidence of approximately 1 in 3,500 live births.<sup>1</sup> It is an autosomal dominant disease. It is caused by a heterozygous mutation of the neurofibromin gene on chromosome 17q11. NF-1 patients are at a high risk of developing a neurofibroma. This may present with progressive neurological symptoms, such as pain or weakness.<sup>2</sup>

A 17-year-old boy presented with a six-month history of progressive right lower limb weakness. He had a background of NF-1, scoliosis and attention-deficit hyperactivity disorder (ADHD).

On examination, he exhibited an unsteady, antalgic gait and right foot drop. He had two recent minor head injuries: a fall from a bicycle, and a mechanical fall from standing height. A further clinical history yielded two minor head injuries in recent months: a fall from a bicycle, and a mechanical fall from standing height.

An MRI displayed a C6 right pedicle fracture. Acute spinal cord oedema extended from C5-C7, secondary to a contusion, causing spinal cord compression. This caused C5-C6 myeloradiculopathy. Full spinal precautions were immediately applied. The patient was managed conservatively with head elevation for one week. No surgical intervention was performed.

He was discharged home, wearing a Miami J collar for one month of cervical spine stabilisation. He has persistent bilateral weakness of his distal interphalangeal joints. He will require long-term outpatient follow-up with Orthopaedic Surgery and Physical Rehabilitation.

This C6 fracture was managed conservatively. There are no other documented cases of similar fractures in NF-1 patients. A recent case series of NF-1 cervical neurofibroma illustrated that surgical decompression is the mainstay of treatment.<sup>3</sup> However, this surgery can cause cervical instability, progressive deformity and neurological deterioration. These risk must be weighed against the likelihood of neurological deterioration from the increase in size of a neurofibroma.

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Arnold PM, Fehlings MG, Massicotte EM. Surgical management of cervical spine manifestations of neurofibromatosis Type 1: long-term clinical and radiological follow-up in 22 cases. *J Neurosurg Spine*. 2011 Mar;14(3):356-66. doi: 10.3171/2010.9.SPINE09242. Epub 2011 Jan 14. PMID: 21235302.

## **GASTRIC PERFORATION IN THE SETTING OF CEREBRAL PALSY AND CHRONIC SELF-INDUCED VOMITING**

**C Duff<sup>1</sup>**, A Lyons<sup>1</sup>, N Allen<sup>1</sup>, P Carroll<sup>2</sup>, E Moylett<sup>1</sup>

<sup>1</sup>Paediatric Department, Galway University Hospital, Galway, Ireland

<sup>2</sup>General Surgery Department, Galway University Hospital, Galway, Ireland

Gastric perforation is a rare paediatric surgical emergency. It is most common among preterm neonates, the aetiology is highly variable.<sup>1</sup> Perforation typically presents with severe abdominal pain and massive distension. Pathognomic radiological finding includes massive pneumoperitoneum on plain film imaging of the abdomen.<sup>2</sup> Mortality approaches 30%.<sup>3</sup> There is no association between cerebral palsy (CP) and gastric perforation.

In this case, a 14-year-old girl with quadriplegic cerebral palsy and profound intellectual disability, presented to the Emergency Department with a 12-hour history of lethargy and malaise. The patient had a 5-hour history of increasing abdominal distention. She had excessive burping, with no flatus or bowel motions in the preceding 24 hours.

On examination, the patient had a grossly distended, tense, and tender abdomen. Bowel sounds were absent. Plain film imaging of the abdomen revealed a massive pneumoperitoneum. Following an urgent surgical review, the patient underwent emergency laparotomy.

Laparotomy demonstrated a one-centimetre gastric perforation proximal to the gastro-oesophageal junction. Macroscopically, there was no evidence of surrounding gastric inflammation or ulceration. The perforation was successfully sutured.

Collateral history from her parents revealed a three-year history of daily self-induced vomiting. The patient elicited her own gag reflex for secondary gain: for example, to be sent home from school for vomiting. The patient also has severe scoliosis, causing atypical anatomy and increased intra-gastric pressure.

Gastric perforation typically presents with severe abdominal pain. However, children with profound intellectual disability may struggle to communicate this, due to a lack of verbal communication skills.<sup>4</sup> Based on collateral history alone, a diagnosis of gastric perforation in this population may be challenging.

Abdominal plain film is the most useful investigation revealing a massive pneumoperitoneum. Gastric perforation is a surgical emergency, requiring urgent laparotomy and repair. Potential complications include gastric ulceration, recurrent perforation and gastric outlet obstruction.<sup>5</sup>

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## Medical/Education Management

### ZOOM OUT: THE LEARNING EFFECTS OF FACE TO FACE PATIENT HANDOVER IN THE PAEDIATRIC DEPARTMENT

C Duff<sup>1</sup>, N Allen<sup>1,2</sup>

<sup>1</sup>Paediatric Department, Galway University Hospital, Galway, Ireland

<sup>2</sup>Department of Academic Paediatrics, National University of Ireland, Galway, Ireland

**Background:** The COVID-19 pandemic has underlined the value of face-to-face communication. While there has increased practice and governance of medical handover, there is a lack of systematic research examining junior doctors’ learning. This study aimed to identify “what is being learned” and to outline the key determinants of learning.

**Methods:** A qualitative study was performed in a tertiary paediatric hospital. Data was triangulated video observation of weekend handover reports, followed by semi-structured interviews of audience participants. Qualitative analysis generated a thematic framework describing the research aims.

**Results:** Clinical handover develops multiple domains of physicians' professional roles. Clinical knowledge is gained through complex case presentations and discussions. Narrative thinking empowers in-depth analysis, demonstrating collaboration and teamwork. An open professional culture encourages participant involvement. Learner psychological safety was a critical determinant of the perceived quality of the learning environment.

**Conclusion:** Handover report is a powerful ritualistic workplace learning event found to enhance many domains of physicians' professional practice. Expert case presentations, observation of clinical reasoning processes and a secure learning environment are key determinants of learning. Further studies comparing virtual to face-to-face handover reports would be informative.

**INTRODUCING LISA: A LOCAL EXPERIENCE AND NATIONAL SURVEY****C Duff**, L Kennedy, E Ryan, J James, A Binchy, D O'Donovan<sup>1</sup>Neonatology, Galway University Hospital, Galway, Ireland

Less Invasive Surfactant Administration (LISA) is now an established method of surfactant delivery for the treatment of respiratory distress syndrome (RDS) in preterm neonates (Polin, 2014). LISA has been endorsed by the European Consensus Guidelines group as the preferred mode of surfactant administration for spontaneous breathing infants on CPAP, and the procedure has been widely adopted throughout neonatal units in Europe (Sweet, 2019). The objective of this study was to examine Galway University Hospital's experience with LISA between February 2018 and December 2021. Using an online survey and follow-up phone calls, we determined the uptake of LISA in Irish neonatal units in 2022.

During the study period, 96 infants received surfactant via LISA. Three years after introducing the procedure, LISA became the dominant method of surfactant delivery at Galway University Hospital. By 2021, over 70% of surfactant was administered via LISA. Clinical criteria for LISA included: fraction of inspired oxygen (FiO<sub>2</sub>) requirement  $\geq 30\%$  on Continuous Positive Airway Pressure (CPAP); clinical signs of increased work of breathing, such as tachypnoea and sternal recession; and/or radiological evidence of RDS (NICE, 2019; Rigo, 2016). Tracheal catheterization was achieved exclusively by video-laryngoscopy.

However, other neonatal units in Ireland have been slow to adopt this minimally invasive surfactant administration technique. Our nationwide survey revealed only 26% (5/19) of Irish neonatal units used LISA as a method of administering surfactant. The low use of LISA in neonatal units in ROI seems to be related to a satisfaction with current surfactant delivery methods and a general lack of enthusiasm to introduce LISA, possibly due to concerns about deskilling. The use of a video laryngoscope would help units develop a LISA program, and alleviate concerns regarding deskilling, as evidence suggests that this device helps persevere and develop neonatal trainee's airway procedure skills.

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## COVID-19 PNEUMONIA IN CHILDREN AND CRITICAL CARE HOSPITAL READMISSION- A CASE OF MIXED INFECTION AND EMPYEMA

G Egan<sup>1,2</sup>, T Mustonen<sup>1</sup>, D Onyekwere<sup>1,3</sup>

<sup>1</sup>Department of Paediatrics, University Hospital Kerry (UHK), Tralee, Ireland

<sup>2</sup>South West Specialist Training Programme in General Practice, IT Tralee, Tralee, Ireland

<sup>3</sup>Senior Clinical Lecturer, Paediatrics and Child Health, UCC, Cork, Ireland

**Aim:** To highlight mixed infection as a cause of severe Covid-19 pneumonia complications in paediatric population.

**Background:** Coronavirus disease 2019 (COVID-19) is a highly contagious respiratory disease caused by the SARS-COV-2 virus which has resulted in a global pandemic. There have been over 600 million confirmed cases worldwide since December 2019 (1). It usually causes mild disease in children. Mixed bacterial infection is more common in children compared to adults- 14.5% vs 8.4% rate(2,3)

**Method:** Retrospective case review of 4-year-old boy who presented to UHK with wheeze, increased work of breathing and hypoxia. High CRP of 327.6 and neutrophilia. He tested positive for COVID-19 and human metapneumovirus on nasopharyngeal swab. He received bronchodilator nebulisers, saline bolus, cefotaxime, azithromycin, hydrocortisone/Dexamethasone and started high flow oxygen. CXR showed left lobar pneumonia/mild effusion. Transferred and admitted to PICU in OLCHC for respiratory support.

**Results:** He was transferred back from OLCHC when stabilised off Non-Invasive-Ventilation and CRP down to 25. Chest xray 2 days later showed complete white-out of left lung and CRP increased to 150. He was again transferred to OLCHC for chest drain insertion for empyema. Fluid from empyema was positive for pneumococcus on PCR. He was treated with Cefalexin and discharged 21 days after initial presentation to UHK and following 2 weeks of oral antibiotic therapy.

### Conclusions:

1. COVID-19 usually causes mild disease in children.
2. Mixed bacterial and viral infection is more common in children. In a meta-analysis by RCPCH 8.4% of adults had a co-infection and 14.5% in children.
3. Early chest drain insertion for COVID-19 related para-pneumonic effusions could prevent a readmission to OLCHC

### Recommendations:

1. There should be increased suspicion of bacterial mixed infection in severe disease.

2. COVID-19 pneumonia in children can safely be managed with NIV.
3. Prompt transfer to unit with paediatric critical care facility is recommended for best patient outcomes.

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## MY CHILD'S HEAD IS A FUNNY SHAPE! A CASE REPORT OF METOPIC CRANIOSYNOSTOSIS G Egan<sup>1, 2</sup>, A Kumar<sup>1</sup>

<sup>1</sup>Department of Paediatrics, University Hospital Kerry, Tralee, Co. Kerry

<sup>2</sup>South West Specialist Training Programme in General Practice, IT Tralee, Tralee, Co. Kerry

**Aim:** To highlight a case of craniosynostosis

**Background:** Craniosynostosis is a cranial malformation in which 1 or more cranial sutures fuse in utero. It affects 1 in every 2000-2500 births worldwide. Sagittal craniosynostosis causing scaphocephaly (boatshaped skull) is the most common type and has a 3:1 ratio of males:females. Other types include unicoronal, metopic and lambdoid craniosynostosis<sup>1</sup>.

**Methods:** This case is of a 23 month old girl who presented to outpatient clinic for follow up of metopic craniosynostosis. On exam, the child has prominent frontal bone with OFC on 50-75<sup>th</sup> centile. There were no dysmorphic facial features and developmental milestones were appropriate. She has been referred to craniofacial team in Temple Street Hospital for the last 12 months but has not yet been seen due to incomplete referral as photos of the child have not been obtained. This is due to a barrier in communication as the child's mother does not speak English.

**Results:** It is important to be aware of the complications of craniosynostosis which need immediate treatment such as increased intracranial pressure, airway obstruction and damage to optic nerve/globe<sup>2</sup>. The mainstay of management for craniosynostosis is an early referral to the craniofacial team in Temple Street Hospital, particularly when there is any suspicion of

complications such as those outlined above. The craniofacial Ireland website has detailed guidance on how to make a referral and the process thereafter. This team will decide if surgery or conservative management is most appropriate<sup>3</sup>. Follow up for these children regardless of outcome of treatment pathway includes regular outpatient review to plot OFC centile and ophthalmology consult to monitor for any complications<sup>1</sup>.

**Conclusions:** Most children with isolated suture craniosynostosis have a good prognosis however the early referral to a craniofacial centre when concerns arise about complications remains the most important management role with the best outcome for the child.

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## **AN UNEXPECTED CAUSE OF FIRST SEIZURE IN A FIVE YEAR OLD GIRL**

**A English<sup>1</sup>, A Jafar<sup>1</sup>, D Gallagher<sup>1</sup>**

<sup>1</sup>General Paediatrics, Sligo University Hospital, Sligo, Ireland

**Introduction:** We present the case of a five year old typically developing girl who presented to the emergency department by ambulance following an apparent febrile seizure in school.

**Case Report:** Our patient was a generally well five year old girl with no history of febrile seizures in early childhood. Her teacher described a focal episode lasting fifteen minutes during class. She had a recorded temperature during ambulance transfer to hospital. Initial neurological examination and vital signs were within normal range on admission. Initial impression was that of a febrile seizure. She had a further similar episode as an inpatient within the following twelve hours. Vital signs were normal following this episode. Her mother recalled that she had noticed an abnormal gait in the days preceding the event, although her gait was normal on review. She underwent brain imaging the following day given recurrent seizure episode and concerns regarding preceding abnormal gait. MRI brain revealed a left sided open lipped schizencephaly.

**Discussion:** Schizencephaly is a rare malformation of early brain development of unknown aetiology which can present with severe developmental delay, early onset epilepsy and varying degrees of physical and intellectual disability. It is important to make the diagnosis due to its association with severe epilepsy and neurodevelopmental delay. Our case demonstrates the



importance of selective brain imaging and detailed history taking in the evaluation of first episode of seizure in children. Additionally, this highlights how significant cortical brain abnormalities can present in the previously neurologically normal child at any age.

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## **SOCIAL DETERMINANTS OF HEALTH AND FALTERING GROWTH IN CHILDREN: CLINICAL CONSIDERATIONS**

**L Falcone**<sup>1</sup>, A Walsh<sup>1,2</sup>, E Molloy<sup>2</sup>, C Ni Cheallaigh<sup>3</sup>, C Hensey<sup>1,2</sup>

<sup>1</sup>Department of General Paediatrics, Childrens Health Ireland at Temple Street, Dublin, Ireland

<sup>2</sup>Faculty of Paediatrics and Child Health, Trinity College Dublin, Dublin, Ireland

<sup>3</sup>Clinical Medicine, School of Medicine, Trinity College Dublin, Dublin, Ireland

**Aim:** Social determinants of health (SDH) include socioeconomic status, education, accommodation, employment and social support networks, and access to health care. Low-income families often sacrifice a healthy diet to other budgetary demands.<sup>1</sup> As part of an outreach clinic for children at risk of social exclusion, children with faltering growth were referred and managed according to their individual presentations and local guidelines. Numerous barriers to optimal management directly rooted in SDH were identified.

**Methods:** We reviewed the management pathways of children attending the inclusion health outreach clinic in CHI at Temple Street (CHI-TS), whose primary reason for referral was concern around growth and/or nutrition. Key factors were identified as being detrimental to a positive outcome and steps were taken to overcome these during which we adapted our own clinical practise.

**Results:** Key factors identified as negatively impacting response to management of faltering growth are: 1)

Language and literacy, 2) Accommodation and kitchen facilities, 3) Food and energy costs, 4) Health literacy 5) Cultural norms, 6) Prescribing challenges, 7) Community supports, and 8) Poor dentition. Thirteen children were referred over 18 months with primary concern around growth and nutrition. All families required interpreters, 84.6% (n=11) were homeless, 76.9% (n=10) did not have access to formal primary care, 84.6% (n=11) were from the Roma community.

**Conclusions:** These results clearly demonstrate the disadvantages that children experiencing adverse SDH encounter when trying to address growth and nutritional concerns, highlighting the need for early recognition of these challenges. A holistic and integrated approach is required, including proactive involvement of community supports and tailored management plans with a

lower threshold for nutritional supplementation to be considered. Recommendation of cheap, shelf stable, long-life, high calorie foods (e.g. peanut butter, tahini), review of dentition, direct liaison with interagency community supports, and conscientious, affordable prescribing is now our standard practice.

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**BIGGER NOT ALWAYS BETTER: A CASE SERIES OF PATIENTS WITH OVERGROWTH DISORDERS.**

**A Fanning<sup>1</sup>, Z Sarani<sup>1</sup>**

<sup>1</sup>Paediatric Department, Mayo University Hospital, Castlebar, Ireland

**Aims:** To present a case series consisting of four patients with overgrowth disorders currently attending the paediatric department in Mayo University Hospital (MUH).

**Methods:** We describe three patients with hemihyperplasia and one patient with PIK3CA related overgrowth syndrome (PROS) and discuss their clinical presentation, investigations, management and review the current recommendations on tumour surveillance in this cohort.

**Results:** Overgrowth syndrome refers to a group of disorders with abnormal, excessive tissue proliferation(1). Hemihyperplasia, also known as hemihypertrophy or lateralised overgrowth, is a type of overgrowth disorder causing asymmetric growth of single or multiple organs or body regions due to an overproduction of bone or soft tissue. Hemihyperplasia can occur in isolation or be associated with a syndrome or genetic conditions such as Beckwith-Wiedemann syndrome. These conditions are particularly important to recognise due to an increased risk of tumours, particularly Wilms tumours (2, 3). Recommendations for surveillance include, referral to a clinical geneticist for evaluation, 3 monthly abdominal ultrasounds until age 7, 3 monthly serum alpha-fetoprotein measurements until age 4 and daily abdominal examination performed by the provider or parent(2,4). The specific overgrown tissue observed in patients with PROS is typically adipose or vascular, however muscular and skeletal overgrowth has been observed (5). The risk of cancer in PROS appears to be lower overall than other overgrowth syndromes, however, surveillance protocols are still considered (5). We describe three patients currently attending MUH with hemihyperplasia and one patient with PROS affecting the upper limb. We review their initial presentation, examination findings, investigations and results to date. We also discuss the current recommendations for management of these patients including tumour surveillance.

**Conclusion:** We describe three patients with hemihyperplasia and one patient with PROS presenting to MUH and discuss their management to date and review current recommendations of tumour surveillance in these patients.

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**MEASUREMENT OF OCCIPITAL FRONTAL CIRCUMFERENCE (OFC) BETWEEN PAEDIATRIC HEALTHCARE PROFESSIONALS IN UNIVERSITY HOSPITAL WATERFORD**

**RF Fitzgerald<sup>1</sup>, NC Canty<sup>1</sup>, DH Huggard<sup>1</sup>**

<sup>1</sup>Department of Paediatrics , University Hospital Waterford , Waterford , Ireland

**Introduction:** The routine measurement of OFC by healthcare practitioners is a fast and non-invasive method of measuring head size and determining brain volume.<sup>(1)</sup> In infants under the age of 2 years OFC is a reliable indicator for linear growth and can be a simple indicator of diagnostic significance.<sup>(2)</sup> Best practice according to the RCPCH clinical guideline is to take 3 measurements and record the mean.<sup>(3)</sup>

**Aim:** The aim of our study was to assess across all clinical levels i.e. paediatric consultants, registrars, SHOs, medical students and nursing staff, the method and accuracy in measurement of OFC in a live infant against a control i.e. a model infant.

**Method:** This audit involved review of OFC measurement on both a control and live infant. Parameters assessed included: position of measuring tape, circumference in cm, number of measurements. Participants were instructed to measure and document the OFC for the control and for a 6 month old recruited patient. The correct measurement for each group was defined as that taken by a consultant paediatrician.

**Results:** Within the control group n=20, 50% (n=10) took the correct number of measurements, 45% (n=9) obtained the correct measurement. Of those 25% (n=5) were identified as having both parameters correct. The range in measurement was 38.5-39.9 cm (1.4cm), standard deviation was 0.3221 cm.

Within the live patient group n=11, 54% (n=6) took the correct number of measurements and 27% (n=3) measured the OFC correctly. 10% (n=1) of those enrolled had both parameters correct. The range in measurement was 41.6-43.2 cm (1.6cm), standard deviation was 0.4665 cm.

**Discussion:** This audit demonstrates differences in OFC measurement and results which can be clinically significant. There was greater variance in measurement of OFC amongst the live infant group versus the control group. Education sessions for all clinical levels should be considered to encourage greater adherence to clinical guidelines.

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### **CHANGES IN PAEDIATRIC OPD REFERRAL PATTERNS (PRE AND POST COVID 19 PANDEMIC )**

**J Foran<sup>1</sup>, A O'Brien<sup>1</sup>, G Dugaci<sup>1</sup>, D Coghlan<sup>1</sup>**

<sup>1</sup>General Paediatrics, CHI Tallaght, Dublin, Ireland

**Aim:** To assess patterns in general paediatric outpatient referrals and triage outcomes prior to and following the COVID 19 pandemic

**Methods:** Data was extracted from the records of children's active triage (CAT) clinics for Children's Health Ireland Tallaght. This included information on presenting complaint and triage outcome over 24 weeks from January 2021 to August 2021. Data was compared to a previous pre-pandemic audit performed in 2019. Data included presenting complaint, age and triage outcome

**Results:** Data on 515 referrals over the study period were obtained. There was an increase in referrals per week between the periods (19 vs 21.5,  $p=0.15$ ). The commonest current reason for referral was recurrent urinary tract infections (UTIs)/enuresis (13.0% [ $n=67$ ]) compared to 4% of referrals in 2019 ( $n=24$ ). The percentage of referrals rejected/redirected was 29.3% ( $n=145$ ) compared to 48.5% in 2019 ( $p<0.001$ ). The highest proportion of referrals deemed urgent were for patients with lymphadenopathy (50% [ $8/16$ ]). Of all referrals 1.5% ( $8/515$ ) were deemed to warrant immediate redirection to emergency department care with 4 of these being due to growth/nutritional concerns.

**Conclusion:** There have been several changes in referral patterns and outcomes between 2019 and 2021. The increase in referrals for recurrent UTIs/enuresis may reflect recent changes in surgical service provision on site, a reduced capacity for primary care management or lockdown related epidemiological changes consistent with higher rates of functional disorders. Rejection and redirection rates reduced significantly, possibly due to increased clinical acuity, risk aversion or simply a normalisation of rates. The use of collected centralised data on nature of referrals will provide timely information to guide education of primary care and evaluation in epidemiological shifts.

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### RETROSPECTIVE ANALYSIS OF SIX YEARS OF HOSPITALISED CHILDREN WITH *E. COLI* UTI: CLINICAL FINDINGS AND OUTCOME

S Kyne<sup>1</sup>, M Glynn<sup>1</sup>, A Dore<sup>1</sup>, S Whelan<sup>2</sup>, E Moylett<sup>1</sup>

<sup>1</sup>Academic Department of Paediatrics, National University of Ireland Galway, Galway, Ireland

<sup>2</sup>National University of Ireland Galway<sup>1</sup> Department of Microbiology, University Hospital Galway, Galway, Ireland

**Background:** Urinary tract infection (UTI) is a common indication for paediatric admission. We undertook a review of hospitalised UTI cases examining clinical findings and radiological outcomes over a recent 6-year period.

**Methods:** From data retrospectively identified by University Hospital Galway, Department of Microbiology

(a 10-year review of urinary *E.coli* susceptibility patterns), we identified paediatric isolates from 2016 to 2021 inclusive and selected all admitted cases of confirmed UTI. A retrospective review of medical records was performed obtaining: demographics, clinical, laboratory, radiological data and outcomes. Data were grouped according to age and whether there was a history of prior UTI or prophylaxis. Ethical approval obtained.

**Results:** During the 6-year period, 249 children admitted with *E.coli* UTI; 30 infants <2 months, 137 >2 months to 2 years, and 82 children >2 years. The majority admitted >2 months were female (73.51%); <2 months male predominance (73%). 24.65% >2 months had a prior UTI. Clinically, by age-grouping, temperature >38°C on presentation for 90, 95 and 91%, vomiting, 30, 42 and 59%,

respectively. Average IV antibiotics duration, 4.9, 2.6 and 2.7 days; length of stay, 5.4, 3.2 and 3.5 days. *E.coli* bacteraemia (<2 years only), eight of 167 blood samples, *E.coli* meningitis (<2 months only) two of 16 CSF samples. Renal US (RUS) performed on admission for 214/249 patients, 89/214 (41.5%) demonstrated abnormalities, most frequently mild prominence/dilatation of the pelvicalyceal region. Follow-up RUS was performed in 122 cases, with 61.5% being normal/demonstrating resolution of prior findings. Scarring evident on 6/214 initial RUS, increasing to 11/122 (9%) follow-up RUS.

**Conclusion:** This 6-year retrospective study confirms fever as a consistent finding in children hospitalised with UTI. Among those <2 years, urosepsis needs to be considered in addition to meningitis in those < 2 months. From onset of specific therapy, average fever duration was longest in those > 2 years and shortest in infants < 2 months.

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#### **PREVALENCE OF NON-CARDIAC CO-MORBIDITIES IN CHILDREN WITH CONGENITAL HEART DEFECTS WHO UNDERWENT CARDIAC SURGERY IN THEIR FIRST YEAR OF LIFE**

**A Golden, D Finn**

<sup>1</sup>Paediatrics, University College Cork, Cork, Ireland

<sup>2</sup>Paediatrics, University College Cork, Cork, Ireland

**Aim:** The aim of this study is to investigate the prevalence of non-cardiac comorbidities in children who underwent cardiac surgery in their first year of life.

**Methods:** A cross-sectional study was conducted in Cork University Hospital. 30 participants were recruited through a paediatric cardiology patient list. Inclusion criteria necessitates children between the ages of 4– 17 who underwent cardiac surgery in their first year of life. Parent completed questionnaires — including a standardised Strengths and Difficulties questionnaire which serves as an emotional and behavioural screening tool — were analysed with SPSS software using descriptive and inferential statistical methods.

**Results:** 30% of participants had a genetic diagnosis of which 67% had Down Syndrome, 22% a 22q11 deletion, and 11% a Q14 deletion. 46.7% reported medical problems in areas including: ear, nose, and throat (50%), renal (14.2%), respiratory (14.2%), gastrointestinal (14.2%),



musculoskeletal (7.1%), neurological (7.1%), cardiovascular (7.1%), ophthalmological (7.1%) and, rheumatological (7.1%) conditions. 63.3% attended any one or multiple of early intervention, occupational therapy, speech and language therapy, psychology. 63.3% require special needs assistance. The scores of the Strengths and Difficulties questionnaires indicated that, 43.3% of the children had 'total difficulties' scores within the 'raised' categories suggesting increased emotional and behavioural difficulties. Having additional medical comorbidities ( $p>0.24$ ), taking regular medications ( $p>0.57$ ) or having a genetic diagnosis ( $p>0.94$ ) were not associated with increased 'total difficulties' scores.

**Conclusion:** The results depict the prevalence and variety of non-cardiac comorbidities and by doing so increases our understanding of children with congenital heart defects and the additional needs they have. **37**

### **A LEARNING SYSTEM TO EXAMINE AND IMPROVE THE CARE OF INFANTS ADMITTED WITH BRONCHIOLITIS**

F Feaheny<sup>1</sup>, T Green<sup>1</sup>, S Burke<sup>1</sup>, J Fitzsimons<sup>1</sup>

<sup>1</sup>General Paediatrics, Temple Street Children's Hospital, Dublin, Ireland

**Background:** Bronchiolitis is the most common reason for infants to be admitted to hospital making it a high-volume activity. In the majority of cases, treatment is supportive. Despite this, investigations and medications remain widely used in this cohort and there are variations in management resulting in potential harm, additional costs and increased length of hospital stay.

**Aim & Objectives:** The objective was to examine our practice in caring for these infants by following the patient journey from admission to discharge and to compare this with local and international guidelines. We aimed to identify variations in care and key points during admission where it could be improved.

**Methods/Intervention:** Infants less than 1 year admitted through ED with a clinical diagnosis of bronchiolitis were included in the study. An Excel data collection sheet was used to collect information on investigations, medications, feeding support, oxygen saturations and respiratory support on each day of admission.

**Results/Findings:** We identified a number of points during the patient journey where there were variations in care. There was a large variation in target oxygen saturations documented, ranging from 92% to 96%. Weaning of humidified high-flow nasal cannula oxygen was not standardized and differed from the CHI HHFNC guideline. There were no standard discharge criteria for length of time off oxygen required before discharge.

**Conclusions:** Based on these results, we plan to develop a single-page infographic Bronchiolitis Care Bundle focusing on the key areas of management where the most variation in care was seen. This will be displayed in clinical areas to provide information and guidance to healthcare workers. We intend to use the learning system to continuously gather data to guide quality improvement of key care bundle interventions with the aim of standardising care and ultimately reducing patient length of stay.

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## THE ACTIONS TAKEN IN RESPONSE TO PAEDIATRIC EARLY WARNING SYSTEM (PEWS) TRIGGERS IN

### WEXFORD GENERAL HOSPITAL (WGH)

S Heneghan, E Redmond, A Kalim

<sup>1</sup>General Paediatrics, Wexford General Hospital, Wexford, Ireland

**Aims:** The Paediatric Early Warning System (PEWS) is a framework to provide care to admitted children and help staff to recognise deteriorating paediatric patients<sup>1</sup>. Our aim was to investigate adherence to National PEWS Clinical Guidelines within the Paediatric Department of WGH, identifying cardiac arrest calls, transfers to ICU or a tertiary paediatric hospital.

**Methods:** All admitted paediatric patients with high PEWS were included in the study. Patient details were recorded and PEWS charts, medical and nursing notes were reviewed retrospectively. Data was recorded in a spreadsheet including identifying number, PEWS, PEWS amendments, Medical Escalation Suspension (MES), correct completion of amendments/suspension, cardiac arrest calls, transfers to WGH ICU, and transfers to a tertiary paediatric hospital.

**Results:** Over a 3 month period, 5 patients were recruited. 1 patient was excluded as the medical records could not be located. Of the remaining 4 patients, none had PEWS amendments. All 4 had

a MES in place and of these, 2 were fully completed. There were no paediatric arrest calls, no local ICU admissions and 1 transfer to PICU.

**Conclusion:** Few patients were recruited which was thought not to reflect acuity of presentations over the recruitment period. This may be due to unclear instructions on responsibility of recruitment. There was a clear preference for MES protocols over PEWS amendments, which may indicate that recruited patients had no underlying medical conditions, or that medical staff are more comfortable using suspense protocols. Only 50% of MES were correctly completed, indicating a need for further education. 25% of patients were transferred to PICU. Overall, this audit was limited due to the numbers and is likely non-reflective of true clinical practice. Future recommendations include ongoing audit, regular recruitment reminders, education sessions on PEWS amendments and MES, and consideration of a retrospective audit of a larger cohort.

1. The Irish Paediatric Early Warning System (PEWS), National Clinical Guideline No. 12 - National Patient Safety Office, Department of Health; November 2016;  
<https://assets.gov.ie/11584/b591d589d8fa4d8482ccfd8429baa0>

**POST-DUCTAL PULSE OXIMETRY FOR SCREENING OF CONGENITAL HEART DISEASE (CHD) IN THE COOMBE****WOMEN AND INFANTS UNIVERSITY HOSPITAL (CWIUH)****S Heneghan<sup>1</sup>, A O'Sullivan<sup>1</sup>, J Miletin<sup>1</sup>**<sup>1</sup>Department of Neonatology, Coombe Women and Infants University Hospital, Dublin, Ireland.

**Aims:** Post-ductal pulse oximetry has been shown to improve early diagnosis of CHD<sup>1</sup>. CHD screening guidelines were implemented in 2011. It was noted that discharge paperwork asked for “pre-ductal SpO<sub>2</sub>” instead of “post-ductal SpO<sub>2</sub>”. It was hypothesised that this documentation error resulted in inadequate CHD screening. We aimed to investigate adherence to guidelines, identify if the documentation error resulted in failure to diagnose CHD, correct the documentation, identify gaps in recommendations and improve practice.

**Methods:** 100 charts were reviewed retrospectively. Data was recorded in a spreadsheet including neonatal admissions, record of saturations, type of saturation (pre-/post-ductal), abnormal readings and incidence of 4 limb BPs.

**Results:** 94 neonates were managed on the postnatal ward, 2 in NICU and 4 in SCBU. 11 (11%) had no documented SpO<sub>2</sub> reading. Of the 89 who had SpO<sub>2</sub> readings, 64 (71.9%) were documented as pre-ductal and 24 (26.9%) were at an unspecified site. As part of a murmur workup, 1 neonate had pre- and postductal SpO<sub>2</sub> readings and 4 limb BPs. 82 SpO<sub>2</sub> readings were taken on day of discharge and 7 at another time during admission. No times were documented so age at time of recordings was unknown. There were no abnormal SpO<sub>2</sub> readings recorded. No neonates were diagnosed with CHD during hospital stay.

**Conclusion:** Besides 1 neonate who had a murmur workup, there were no documented post-ductal SpO<sub>2</sub> readings. Readings documented as pre-ductal or unspecified were likely a result of the error in paperwork but this could not be confirmed and error in documentation could have prompted staff to check pre-ductal readings. Post-audit, paperwork was amended to include post-ductal SpO<sub>2</sub> readings and staff were surveyed to assess routine clinical practice. Further recommendations include additional staff training on CHD screening, highlighting the benefit of post-ductal SpO<sub>2</sub> readings over pre-ductal and re-audit following these changes.

1. Cloete E, Gentles T, Webster D, Davidkova S, Dixon LA, Alsweiler J, Bloomfield F. Pulse oximetry screening in a midwifery-led setting with high antenatal detection of congenital heart disease. *Acta Paediatrica*. 2020;109(1):100-108.

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Neonatal

**STAFF KNOWLEDGE OF PULSE OXIMETRY SCREENING OF CONGENITAL HEART DISEASE (CHD) IN THE COOMBE WOMEN AND INFANTS UNIVERSITY HOSPITAL (CWIUH)**

**S Heneghan<sup>1</sup>, C O'Leary<sup>1</sup>, A O'Sullivan<sup>1</sup>, J Miletin<sup>1</sup>**

<sup>1</sup>Department of Neonatology, Coombe Women and Infants University Hospital, Dublin, Ireland.

**Aims:** Post-ductal pulse oximetry has been shown to improve early diagnosis of CHD<sup>1</sup>. An audit revealed that the majority of neonates had pre-ductal SpO<sub>2</sub> readings documented for CHD screening, likely due to erroneous documentation. Our aim was to investigate if SpO<sub>2</sub> readings were being incorrectly documented as pre-ductal due to erroneous documentation or if the documentation error was prompting staff to check pre-ductal readings.

**Methods:** 50 staff members were surveyed including midwives, neonatal nurses and neonatal NCHDs. Data on role, frequency of involvement in discharges, knowledge of neonates needing to be screened for CHD, and placement location of saturation probes during CHD screening was recorded in a spreadsheet.

**Results:** 26 (52%) midwives, 6 (12%) neonatal nurses, 16 (32%) neonatal NCHDs and 2 (4%) student midwives completed the survey. 37 (74%) respondents worked mostly on the postnatal wards and 11 (22%) worked mostly in neonatal units. The remainder worked in a combination of locations. 36 (72%) respondents were responsible for neonatal discharges daily and 14 (28%) discharged neonates weekly or less frequently. 47 (94%) respondents knew that all neonates need SpO<sub>2</sub> readings for CHD screening. When asked which limb SpO<sub>2</sub> readings should be obtained from in screening of CHD, 13 (26%) respondents answered either foot, 33 (66%) answered the right hand and 4 (8%) answered the left hand.

**Conclusion:** Although staff were aware of the need for CHD screening, most respondents (66%) were incorrectly taking pre-ductal SpO<sub>2</sub> readings. In combination with the error in discharge documentation, we can conclude that the majority of neonates were not being correctly screened for CHD, and those who were appropriately screened had their readings erroneously documented as pre-ductal. Further recommendations include staff training on CHD screening, highlighting the benefit of post-ductal SpO<sub>2</sub> readings over pre-ductal and re-audit following these implementations.

1. Cloete E, Gentles T, Webster D, Davidkova S, Dixon LA, Alsweiler J, Bloomfield F. Pulse oximetry screening in a midwifery-led setting with high antenatal detection of congenital heart disease. *Acta Paediatrica*. 2020;109(1):100-108.

## **RADIOLOGICAL IMAGING IN THE INVESTIGATION OF UNCOMPLICATED URINARY TRACT INFECTIONS**

### **DURING THE ACUTE ILLNESS AND AT FOLLOW UP**

**HH Hobbs<sup>1</sup>**, FOB O'Brien<sup>1</sup>

<sup>1</sup>Paediatric department, St. Luke's Hospital, Kilkenny, Kilkenny, Ireland

**Aims:** To assess compliance with the imaging schedule in the investigation of patients with uncomplicated urinary tract infection (UTI) as per the guidelines of the paediatric department of St. Luke's Hospital Kilkenny (SLK).

**Methods:** Patients admitted with a UTI between June 2021 and June 2022 were identified via the electronic discharge programme and obtained from HIPE. Data were obtained from patient charts and radiological investigations performed were identified via NIMIS.

**Results\*:** Patients under 6 months old, SLK guidelines recommend renal ultrasound scan (USS) within 72 hours of admission with a repeat renal USS at 4 – 6 months. Patients under 6 months old: 75% received a renal USS within 72 hours. 33% had follow up imaging inconsistent with guidelines.

Patients between 6 months and 2 years, the SLK guidelines recommend no imaging during acute infection and renal USS at 4 – 6 months post uncomplicated UTI. Patients between 6 months and 2 years: 100 % received a renal USS during the acute infection and 50 % of these patients had follow up imaging inconsistent with guidelines.

Patients 2 years and older, the SLK guidelines recommend no radiological imaging in an uncomplicated UTI. Patients aged 2 years and older: 100% of patients received a renal USS during the acute infection. 50 % of these patients had a follow up imaging inconsistent with guidelines.

**Conclusion:** In all cases under all age groups, 30% had imaging during the acute infection in compliance with guidelines. 0 % had follow up imaging in compliance with guidelines. Deviation from these guidelines could not be attributed to documented cases of atypical or recurrent UTI. The data collected demonstrates inconsistency and deviation from local guidelines. It is imperative

to highlight inconsistencies in practice to improve patient care and minimize investigation burden on patients and hospitals.

\*Data collection ongoing.

Management of 1st UTI in Children, St. Luke's Hospital, Kilkenny. Paediatric department. Available on SLK server.



**CHARACTERISATION OF INNATE IMMUNITY ACTIVATION IN A 2 TO 5 YEARS OLD FOLLOW-UP STUDY OF CHILDREN WHO EXPERIENCED NEONATAL ENCEPHALOPATHY**

JM Isaza-Correa<sup>1,2</sup>, E Isweisi<sup>1,2</sup>, A Melo<sup>1,2</sup>, A White<sup>3</sup>, J Wyse<sup>3</sup>, J Murphy<sup>4</sup>, C Vasseur<sup>4</sup>, A El-Khuffash<sup>5</sup>, A

Foran<sup>5</sup>, E Ní Bhraonáin<sup>6</sup>, M Daly<sup>7</sup>, J O'Leary<sup>8, 11</sup>, EJ Molloy<sup>1,2,9</sup>

<sup>1</sup>Discipline of Paediatrics, Trinity College Dublin, The University of Dublin, Dublin, Ireland

<sup>2</sup>Trinity Translational Medicine Institute, Trinity College Dublin, Dublin, Ireland

<sup>3</sup>Discipline of Statistics and Information Systems, Trinity College Dublin, Dublin, Ireland

<sup>4</sup>Neonatology, National Maternity Hospital, Dublin, Ireland

<sup>5</sup>Neonatology, Rotunda Hospital, Dublin, Ireland

<sup>6</sup>Family Support Liaison, Irish Neonatal Health Alliance, Wicklow, Ireland

<sup>7</sup>Advocacy and Policymaking, Irish Neonatal Health Alliance, Wicklow, Ireland

<sup>8</sup>Department of Histopathology, Trinity College Dublin, Dublin, Ireland

<sup>9</sup>Neonatology, Children's Health Ireland at Crumlin, Dublin, Ireland

<sup>10</sup>Paediatrics, Children's Health Ireland at Tallaght, Dublin, Ireland

<sup>11</sup>Paediatrics, Coombe Women and Infants University Hospital, Dublin, Ireland

**Aims:** Neonatal Encephalopathy (NE) results in multi-organ dysfunction which may persist in childhood. We examine whether immunological markers of inflammation are present at ages 2 to 5 years.

**Methods:** Whole blood was stimulated in the presence or absence of lipopolysaccharides (LPS) to evaluate changes in the immune responses to endotoxin. Innate immune cells response from children who experienced NE as new-born (n=21, 2-5 years old) and control children (n=11, 2-6 years old) was analysed by flow cytometry. CD66b<sup>+</sup> Neutrophile and monocyte ((CD14<sup>+</sup>/CD16<sup>+</sup>) activation was assessed by the expression of CD11b (cell activation and migration), Toll-like receptor (TLR)-4 (recognition of endotoxin/lipopolysaccharide/LPS) and cytokine production, such as Interleukin-6 (IL-6), IL-10, IL-17a, TNF $\alpha$ .

**Results:** Expression of CD11b was significantly reduced in total, classical and intermediate monocytes of LPS-treated NE children samples (n=10) compared to controls (n=10). No differences in CD11b were observed in neutrophils or non-classical monocytes in any of treatments when comparing both groups. In addition, non-significant differences were observed in neutrophils, monocytes or subpopulations of monocytes. When evaluating intracellular cytokines expression in neutrophils, IL-6 and IL-10 were increased in both untreated and LPS treated samples of NE children (n=21) compared to controls (n=11). No statistical difference was observed in the expression of IL-17a and TNF- $\alpha$  in the presence or absence of LPS or when comparing the two groups. IL-10 and IL-17a were increased at basal level and following LPS treatment in monocytes of NE children compared to controls. No significant differences were observed in monocytes for IL-6 and TNF- $\alpha$ , but a tendency to increase in the NE group.

**Conclusion:** NE children immune response seems to favour reduced activation of monocytes following LPS challenge and anti-inflammatory response mediated by cytokines. Exploring systemic inflammation in NE could lead to the development of immunomodulatory adjunctive therapies and biomarkers to predict outcomes.

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## LYMPHOCYTES PHENOTYPE AND ACTIVATION IN A 2 TO 5 YEARS OLD FOLLOW-UP STUDY OF CHILDREN

### WHO EXPERIENCED NEONATAL ENCEPHALOPATHY

JM Isaza-Correa<sup>1,2</sup>, E Isweisi<sup>1,2</sup>, A Melo<sup>1,2</sup>, A White<sup>3</sup>, J Wyse<sup>3</sup>, J Murphy<sup>4</sup>, C Vavasseur<sup>4</sup>, A El-Khuffash<sup>5</sup>, A

Foran<sup>5</sup>, E Ní Bhraonáin<sup>6</sup>, M Daly<sup>7</sup>, J O'Leary<sup>8, 11</sup>, EJ Molloy<sup>1,2,9</sup>

<sup>1</sup>Discipline of Paediatrics, Trinity College, the University of Dublin, Dublin, <sup>2</sup>Trinity Translational Medicine

Institute & Trinity Research in Childhood Centre (TRiCC), Trinity College Dublin, Dublin, <sup>3</sup>Discipline of

Statistics and Information Systems, Trinity College Dublin, Dublin, <sup>4</sup>Neonatology, National Maternity

Hospital, Dublin, <sup>5</sup>Neonatology, Rotunda Hospital, Dublin, <sup>6</sup>Family Support Liaison, Irish Neonatal Health Alliance, Wicklow, <sup>7</sup>Advocacy and Policymaking Irish Neonatal Health Alliance, Wicklow, <sup>8</sup>Department of Histopathology, Trinity College Dublin, Dublin, <sup>9</sup>Neonatology, Children's Health Ireland at Crumlin, Dublin, <sup>10</sup>The Children's Health Ireland at Tallaght, Dublin, Dublin, <sup>11</sup>Paediatrics, Coombe Women and Infants University Hospital, Dublin, Ireland.

**Aims:** Evaluate adaptive immunity markers of inflammation in children at ages 2 to 5 years who experienced Neonatal Encephalopathy (NE) to determine if inflammation persist in childhood.

**Methods:** Lymphocytes phenotype and intracellular cytokine production was analysed by flow cytometry in children ages 2-5 years who experienced NE as new-born (n=18) and control children (n=11, 2-6 years). Changes in subpopulations of lymphocytes (CD3+, CD19+, CD56+, CD4+, CD8+, TCR Vdelta 1+, TCR Vdelta 2+) were evaluated. Whole blood was stimulated without/with Lipopolysaccharides (LPS) for 20 hours, followed by 4 hours Phorbol Myristate Acetate (PMA), Ionomycin and Monensin, to evaluate intracellular cytokines (IL-17a, TNF- $\alpha$ , IFN- $\gamma$ ) production and expression of lysosomal-associated membrane protein-1 (LAMP-1 or CD107a) in subpopulations of lymphocytes (Total CD3+, iNKT,  $\gamma\delta$  T cells, CD56+).

**Results:** When assessing proportions of lymphocytes, Natural killer (NK) cells (CD56+) were seen significantly increased in NE children (n=10) compared to control (n=7). No changes in Total CD3+, B cells (CD19+) or subpopulations of T cells (CD4+, CD8+, TCR Vdelta 1+, TCR Vdelta 2+) were found between the groups. Evaluation of intracellular cytokines after stimulation, shows significant increase of degranulation marker CD107a in CD3+, iNKTs and  $\gamma\delta$  T cells in NE children (n=18) compared to controls (n=11).  $\gamma\delta$  T cells also expressed higher CD107a at basal level in the same group. TNF- $\alpha$  was seen increased in CD3+, iNKTs and  $\gamma\delta$  T cells in unstimulated samples of NE children compared to controls, and in stimulated iNKTs. IL-17a was significantly higher in iNKTs at basal level in NE compared to healthy children. No major differences in intracellular cytokines expression was found in NK cells. However, a trend of IFN- $\gamma$  increase in NKs and  $\gamma\delta$  T cells.

**Conclusion:** Increased in degranulation markers and pro-inflammatory cytokine TNF- $\alpha$  might indicate persist inflammation mediated by lymphocytic populations in NE children.

## **THE GAME OF POOS: THINK BEYOND STOOLS**

NI Jameel, OR Neylon, AN Murphy

<sup>1</sup>Department of Paediatrics, University hospital limerick, Limerick, Ireland

**Aim:** To report a case of multiple autoimmune disorders in a 3-year-old with unremarkable family or personal history.

**Method:** We describe the clinical presentation, examination findings, results of hematological, serological investigations, treatment, and outcome in our patient.

**Results:** Our patient presented to A&E with history of sudden onset of constipation since the last 6 months. Mum had exhausted Movicol along with some other laxatives and had been keeping a close eye on the diet and fluid intake. She was admitted for disimpection after a PFA showing extensive faecal loading up to the splenic flexure and sent routine bloods, TFTs, and coeliac screen as part of work up. She had an enema followed by a bowel opening and looked much brighter the next morning. Mum also felt her energy level had improved. We compared the pictures from the past few weeks and agreed with mum that she is looking brighter and sent her home with a plan to contact with the remaining results. TFTs results came the next day which showed severe hypothyroidism with TSH of > 1000 and Free T4 <1.3. We discussed the case with our endocrinologist and readmitted her. She was noted to be bradycardic, pale, very tired, dry skin, and an umbilical hernia, all of which would have let one think of hypothyroidism if it weren't for constipation being the primary complaint. She was kept in for cardiac monitoring, ECG which was normal and Eltroxin was commenced. Next week she came back strongly positive for coeliac screen with a tTG of 500. She met our dietician and gluten free diet commenced and is under regular follow-up.

**Conclusion:** Constipation is quite a common presentation in pediatrics and sometimes can cause a tunnel vision, so it is imperative to have an open mind in terms of differentials, safety netting and following through with results.

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## **GENOTYPE, PHENOTYPE CORRELATION OF INCONTINENTIA PIGMENTI; A CASE REPORT**

NI Jameel, BR Finn, AN Murphy

<sup>1</sup>Department of Paediatrics, University hospital limerick, Limerick, Ireland

**Aim:** To report a case of Incontinentia pigment in a 2-month-old infant with unremarkable family or personal history.

**Method:** We describe the clinical presentation, examinations findings, results of hematological & genetic investigation, photographs, radiological & skin biopsy findings, treatment, and outcome in our patient.

**Results:** Our patient is the second child to healthy non consanguineous Caucasian parents who presented to A&E at 6 days of age with vesicular crusted linear plaque on the lower limbs extending from left buttock to ankle with circular crust of scalp. She was diagnosed by our dermatologist with Incontinentia pigmenti which was later confirmed by genetic test showing mutation in IKBKG gene and skin biopsy. Her ophthalmological findings were also consistent of the diagnosis however she has normal visual development and good fixing and following. She had a normal MRI and cardiology assessment. She remained clinically well during her in-patient stay, feeding, robust etc. She continues to achieve all her developmental milestones and we are closely monitoring her with regular follow-up.

**Conclusion:** Long-term and close cooperation between dermatologists, pediatricians, neurologists, genetic counselors, and even dentists is crucial for better understanding of IP and prediction of the occurrence of the potential anomalies later in life.

## **CHI REFERRALS OVER 16 YRS OF AGE; WHERE DO THEY COME FROM AND WHERE SHOULD THEY GO?**

**E Jenkins**<sup>1</sup>, JB Hourihane<sup>1, 2</sup>, J Dolliver<sup>3</sup>

<sup>1</sup>General Paediatrics , CHI Temple St, Dublin , Ireland

<sup>2</sup>Paediatrics , RCSI, Dublin, Ireland

<sup>3</sup>Biology, University of Oxford, Oxford, England

**Aims:** To profile the referrals of children over 16 yrs to CHI Central Referrals Office in order to identify implantable and equitable proposals to reduce this waitlist.

**Methods:** A retrospective review of 100 referrals of children > 16 yrs to CHI was conducted by two independent clinicians. Simple demographic data was extracted to detail age at referral, source of referral, specialties referred to. Clinicians independently determined whether the clinical issue could only be managed in CHI or should be provided in CHI due to clinical complexity. The clinicians then met to explore consensus.

**Results:** 50% of referrals were > 16 yrs at time of referral. 50% referred by GPs and 30% referred by

Consultants within CHI. The largest proportion of referrals were to dermatology, surgery, orthopaedics and ENT. The majority of referrals were single issue, triaged as 'routine' or non-urgent. 9% of cases were considered clinically complex by both reviewers and only 1 case was considered a clinical issue that could only be managed in CHI.

**Conclusion:** Of 100 cases referred > 16 yrs old only 10 were considered complex or necessitating care in CHI. The review identified a number of proposals to address the waitlist: Any referral of a child >16 yrs with a non-urgent, single issue should be redirected immediately. Referrals from outside Dublin should be redirected to a regional paediatric service if appropriate care available. CHI should establish a structure to manage decisions surrounding children with clinical complexity. Transitioning care for children with long standing conditions should anticipate timely referral for additional clinical needs.

National Standard for Patient Referral Information. HIQA March 2011 Report and Recommendations on Patient Referrals from General Practice to Outpatient and Radiology Services, including the National Standard for Patient Referral Information. HIQA 2011.

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## **A RETROSPECTIVE REVIEW OF THE ANTIBIOGRAMS OF ORGANISMS CAUSING URINARY TRACT INFECTION IN A PERIPHERAL HOSPITAL SETTING**

**N Keenan<sup>1</sup>, L Nechifor<sup>1</sup>, A Leahy<sup>1</sup>**

<sup>1</sup>Paediatric Department, Cavan General Hospital, Cavan, Ireland

**Aims:** Urinary Tract Infection (UTI) is a common infection causing illness in infants and children<sup>1</sup>. Many bacteria are associated with UTIs, and these organisms have unique antibiograms which can differ depending on local sensitivities and resistance patterns<sup>2</sup>. The aim of our study was to investigate the local causative bacteria of UTIs and their susceptibilities/resistance patterns to antibiotics, to determine which antibiotic treatment may be best to start empirically. This was a re-audit of a 2016 study carried out locally and the standard the audit was investigating was the hospital's clinical guideline for the treatment of UTIs.

**Methods:** A retrospective review of paediatric patients coded as having a true UTI on the Cavan General Hospital HIPE system between January 2021-March 2022 was performed. A laboratory database search was carried out to collect information on microorganisms grown and antibiotic sensitivity/resistance patterns. Children between 3 months-16 years who had a positive urine microscopy (WCC>100, single organism culture) were included in the study. Those with high probability of specimen contamination (low WCC, high resistance pattern, sterile or mixed cultures) were excluded. Data relevant to our aims was collected and patients were anonymised according to hospital number.

**Results:** A total of 48 patients with true UTIs were identified (male=8, female=40) with a median age of 3.4 years. The most common organism isolated was E.coli (83%), Klebsiella (4.1%) and Pseudomonas (4.1%). Resistance was highest to Amoxicillin (54%) and Co-amoxiclav (46.8%) while

the antibiotics with the highest sensitivities were Gentamicin (89.4%), Nitrofurantoin (83.3%) and Trimethoprim (70.8%).

**Conclusion:** E.Coli remains the most common cause of UTIs in the paediatric population. Though antibiotics such as Amoxicillin and Co-amoxiclav are used commonly to treat this infection there are high rates of resistance and therefore, in a well child with clinical symptoms of UTI, other antibiotics such as Trimethoprim may be a more useful therapy.

1) Friedman A. Management of UTI in Children: Murky Waters. Pediatrics 2016; 138. 2) Marquez L, Palazzi DL. Antibiotic Treatment for Febrile Urinary Tract Infection: The Clock Is Ticking. JAMA Pediatr 2016; 170:834



**SEVERE PULMONARY HAEMORRHAGE SECONDARY TO GRANULOMATOSIS WITH POLYANGIITIS**

**N Keenan**<sup>1</sup>, S Coyne<sup>1</sup>, J MacMahon<sup>1</sup>, O Killeen<sup>1</sup>, EJ MacDermott<sup>1</sup> <sup>1</sup>Irish National Centre for Paediatric Rheumatology, CHI at Crumlin.

**Introduction:** Granulomatosis with Polyangiitis (GPA) is an ANCA-associated vasculitis of unknown aetiology, that is rare in the paediatric population.<sup>1</sup> Lung involvement, at presentation or with flares, is a common manifestation. It has a variable severity, the most significant of which is diffuse alveolar haemorrhage.<sup>2</sup>

**Presentation:** A previously well 15-year-old boy presented with a 3-week history of fatigue, 4 days of intermittent joint pain/stiffness; papular rash to elbows and a 1 day history of fever. He was treated for presumed reactive arthritis however remained febrile and subsequently developed watery 'black-coloured' stool, macroscopic haematuria and productive cough with haemoptysis. Bloodwork showed ongoing rise in inflammatory markers with new onset significant anaemia.

He became persistently tachycardic and developed reduced O<sub>2</sub> saturations. Examination revealed reduced air entry bilaterally with coarse crackles; diffuse arthritis; erythematous papules on elbows, shins and dorsal aspect of feet; oral ulceration and unilateral septal ulceration.

CT Thorax demonstrated a 2cm cavity in left mid zone, in keeping with extensive intra-alveolar haemorrhage.

Due to the constellation of clinical findings, the main differential was vasculitis and urgent immunological bloodwork was sent. This showed a highly positive PR3-ANCA, in keeping with a diagnosis of GPA. Given the concern for diffuse active pulmonary haemorrhage, a 3-day course of pulse IV methylprednisolone (30mg/kg) was commenced.

Despite initial improvement, on day 3 of therapy the patient was noted to be pale, lethargic with increased cough and drop in haemoglobin, raising concern for ongoing active pulmonary haemorrhage. Second line therapy with IV rituximab was commenced with good effect and the patient was discharged 2 weeks later.

**Conclusion:** Pulmonary haemorrhage in children is a medical emergency requiring prompt management. Although GPA is rare in childhood, it should be considered in the context of

unexplained haemoptysis and haemorrhage and we would strongly recommend early testing for ANCA in these patients.

1) Saenz Rios F, Devaraj S, Movva G, Movva H, Nguyen QD. Granulomatosis With Polyangiitis in a Pediatric Male. *Cureus*. 2020 Dec 13;12(12):e12055. doi: 10.7759/cureus.12055. PMID: 33447484; PMCID: PMC7802401. 2) Filocamo G, Torreggiani S, Agostoni C, Esposito S. Lung involvement in childhood onset granulomatosis with polyangiitis. *Pediatr Rheumatol*. 2017;15:28

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## **THE INCIDENCE, OUTCOME AND MANAGEMENT OF PAEDIATRIC EATING DISORDERS REQUIRING ADMISSION AT WEXFORD GENERAL HOSPITAL OVER A FIVE-YEAR PERIOD**

**S Kerswill**, M Kuhar, S Parker, A Kalim

<sup>1</sup>Department of Paediatrics, Wexford General Hospital, Wexford

**AIMS:** Eating disorders (ED) have the highest mortality and morbidity risk of all mental health disorders <sup>1, 2</sup>. The incidence of early onset eating disorders (EOED) has been on the rise <sup>3, 4</sup> especially during the COVID 19 pandemic <sup>5, 6</sup>. We set out to describe the burden of disease and trends of eating disorders requiring admission to the paediatric ward in Wexford General Hospital (WGH) over a five-year period, and to assess the management of these patients according to the MARSIPAN risk stratification tool and CHI guidelines for the management of patients with Anorexia Nervosa.

**METHODS:** This was a retrospective descriptive study. Patient details were gathered from an existing register of all paediatric mental health admissions between the ages of 5 and 18 who were admitted with an eating disorder between August 2016 and October 2021, resulting in a sample size of 30 patients. An individual folder review was conducted to assess adherence to the MARSIPAN and CHI guidelines.

**RESULTS:** A significant surge in admissions was seen during the COVID-19 pandemic with 17 admissions from January 2020 to October 2021 compared with only 7 admissions in the preceding 3 years. Preliminary results from 13 out of 30 folder reviews were as follows: 69% of patients were diagnosed with Anorexia Nervosa, 15% with an unspecified eating disorder, and 15% with Avoidant Restrictive Food Intake Disorder. 1 patient (7%) was assessed as moderate risk on MARSIPAN risk assessment, while the remainder were incompletely assessed. The CHI Guidelines were generally well adhered to.

**CONCLUSION:** Paediatric eating disorders requiring admission to WGH increased significantly over the COVID-19 pandemic. Preliminary analysis of our results show that the MARSIPAN guidelines are underutilised as a risk stratification tool. Areas for improved adherence to the CHI guidelines for the management of Anorexia Nervosa were identified

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## **A REVIEW OF PAEDIATRIC MENTAL HEALTH ADMISSIONS TO WEXFORD GENERAL HOSPITAL OVER A FIVEYEAR PERIOD**

**S Kerswill**, M Kuhar, A Kalim

<sup>1</sup>Paediatrics, Wexford General Hospital, Wexford, Wexford

**AIMS:** Mental health concerns are responsible for significant morbidity and mortality in the paediatric population <sup>1, 2</sup>. A growing number of paediatric mental-health presentations to emergency departments across Ireland have been reported in the past decade <sup>3, 4</sup> with a further spike in presentations during the COVID-19 pandemic <sup>5, 6</sup>. We set out to describe the trends in paediatric mental-health admissions to Wexford General Hospital (WGH) over a five-year period in order to better understand the epidemiology and healthcare needs of our patients and to assist with appropriate resource allocation.

**METHODS:** We conducted a descriptive, retrospective review of all mental-healthcare admissions to WGH paediatric ward from August 2016 to October 2021. All patients from the age of 5 to 18 years were included, using an existing patient register of all mental-health admissions.

**RESULTS:** 249 patients were included, the majority of which (87.5%) were female. The most common single diagnosis was overdose (22%), followed by eating disorders (10%) and suicidality (10%). 9% of patients were admitted with deliberate self-harm, 8% under section 12 and 5% with depression. 23% of patients had more than one diagnosis. 82% of patients were between 13 and 16 years of age. Admission rates peaked in 2017 with 72 admissions, declining to 52 in 2018 and 27 in 2019. Admission rates then rose to 35 patients in 2020, coinciding with the pandemic, and 45 admissions in the first 10 months of 2021.

**CONCLUSION:** 249 Paediatric mental-health admissions and a cumulative 1095 admission days were recorded in WGH over the five-year period, constituting a significant burden of disease. Accordingly, adequate mental-health resources should be allocated to this hospital, which currently relies on consultant psychiatric outreach from Waterford Hospital. Adolescent females aged 13 to 16 were at particularly high risk for hospital admission.

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## ASSESSMENT OF DRUG CARDEX DOCUMENTATION IN A PAEDIATRIC WARD A QI PROJECT

A Khan<sup>1</sup>, A Raza<sup>2</sup>, A Kalim<sup>3</sup>

<sup>1</sup>Paediatrics, Wexford General Hospital, Wexford, Ireland

**Aim:** Medical records (MR) consist of information about the patients which their doctors or care providers collect through discussion with them, third parties like relatives or through audio and visual recordings. According to Medical Council's Guide to Professional Conduct and Ethics, 2016<sup>1</sup> recommendation; "You must keep accurate and up-to-date patient records either on paper or in electronic form. Maintenance of good MR ensures that patient's assessed needs are met comprehensively."<sup>2</sup>

Our aim was to assess the current practice of MR keeping and to improve it further, according to HSE standards for Healthcare Records Management.<sup>3</sup>

**Methodology:** A retrospective review of 50 sets of paediatric admission notes from the month of November and December 2021. We collected information on a predefined proforma for documentation of: Patient Sticker, Date and Time, Route of administration, Doze mentioned, Legible prescription, Allergies mentioned, Legible Signature, Bleep number, Generic name and IMC Number.

**Results:** 98% (49/50) compliance was noted in the areas of patient Sticker, date and time, route of administration and dose. Allergies were mentioned in 66% (33/50) and signature were legible in 52% (26/50) cases. Generic name was used in 82% (41/50) and IMC number was mentioned in 88% (44/50) cases. The commonly prescribed brand names instead of the generic names were: Augmentin for Coamoxiclav, Neurofen for Ibuprofen, Ventolin for Salbutamol, Atrovent for Ipratropium, Losec for Omeprazole, Zofran for Ondansetron and Becotide for Beclomethasone.

**Conclusion:** Overall, Very good compliance (98-100%) was recorded for the record of patient sticker, route of administration, dose, date and time. It was 82 to 88 % for IMCN and Generic name. 52 % to 66% compliance was noted for allergies mentioned and legible signature. The results of this audit have been presented in the departmental meeting and the re-audit is in progress.

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V3.0

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General Paediatrics

## **SIGNIFICANCE OF GENETIC TESTING (MICROARRAY) AS FIRST LINE INVESTIGATION IN CHILDREN WITH DEVELOPMENTAL DELAY AND INTELLECTUAL DISABILITY**

**A Khan<sup>1</sup>, S Batool<sup>2</sup>, IM Riazat<sup>3</sup>**

<sup>1</sup>Paediatrics, Wexford, General Hospital Wexford, Ireland

**Background:** Current evidence recommend Microarray testing as first-tier investigation for Children with developmental Delay and moderate to severe intellectual disability. Our aim is to see the significance and diagnostic yield of Microarray testing in children with Developmental Delay presenting to Wexford General Hospital.

**Methods:** A retrospective review of randomly selected 53 patient charts was done who were advised

Microarray Testing from January 2021 to December 2021 due to underlying Disordered

Development. We categorised our patients into different groups as per their age, sex, reason for referral, dysmorphic features, any known cause for developmental delay like Autism and result of microarray.

**Results:** Out of the total 53 patients ,36 were male and 17 were female. We observed a variable results and surprisingly different types of deletions were found in few of our patients. Microarray results showed abnormality in **6 patients (11.3%)** which is highly significant. It was observed that 2 of the patients have the chromosome 16 abnormality detected that is **16p11.2**. However one patient has a **Duplication defect** while the other has a **Deletion Defect** of the same region, both these defects are associated with speech delay and Autism Spectrum Disorder. Three of our patients have micro duplication which has strong association with Autistic spectrum disorder. Chromosomal imbalance was detected in one child who has X- linked ichthyosis and delayed development .The last patient has positive connexin 26 genetics which was requested because of speech delay.

**Conclusions:** A significant chromosomal abnormality was found in patients with Autistic spectrum disorder. Most of these defects are in the form of micro duplication or deletion. Our result showed 6 out of 53 patients have underlying genetic defect and underlying disordered development. We recommend to continue Testing Microarray as the first line investigation in all patients with disordered development and moderate to severe intellectual disability.

Renuka Mithyantha , Rachel Kneen , Emma McCann , Melissa Gladstone Current evidence-based recommendations on investigating children with global developmental delay.<https://adc.bmj.com/content/102/11/1071>



**THE IMPACT AND MANAGEMENT CHALLENGES FOR STAFF WHEN CARING FOR PATIENTS WITH CHALLENGING BEHAVIORS ON ACUTE PAEDIATRIC WARDS.****A KONONNA<sup>1</sup>, C PURCELL<sup>1</sup>, P Byrne<sup>2</sup>, A Deasy<sup>2</sup>**<sup>1</sup>General Paediatrics, CHI Tallaght, Dublin, Ireland<sup>2</sup>Psychiatry, CHI TUH, DUBLIN, Ireland

**Background and Aims:** Children and Adolescents may present to acute paediatric settings with significant challenging behaviours, including aggression and self-harm. These can occur due to complex psychosocial stressors, care-related issues, mental health difficulties, or in the context of disability. Challenging behaviours can be a barrier to discharge from acute paediatric services.

The primary aim of the study was to assess the impact and management challenges facing front-line staff caring for patients with challenging behaviours admitted to the acute paediatric wards in CHI at Tallaght.

**Methods:** A self-designed 10 point questionnaire was distributed to Doctors, Nurses, and AHCPs in Paediatrics in CHI Tallaght.

**Results:** 78[CP1] participants completed the questionnaire; doctors (41%), Nurses (40%), and AHCPs (19%). 41/78 of respondents had > 5 years' paediatric experience. The majority of respondents [CP2] had either witnessed a colleague being subjected to aggressive behaviour (87%), or were themselves subjected to aggression (70%). 26% were subjected to verbal aggression alone, while 12% were verbally and physically threatened. 32/68 (47%) were subjected to both verbal and physical aggression (spitting/hitting/throwing items). Of those who witnessed or were subjected to aggressive behaviour, (36/68) 53% reported increased stress levels, and 3% took time off work to recover. The majority[CP3] of participants had no formal training to deal with aggressive patients and all respondents reported accredited training in managing challenging behaviours (e.g. MAPA/TMVA)

was urgently required. Many respondents reported additional concerns for personal safety; querying whether patients would be better cared for in alternative settings.

**Conclusion:**

Aggressive behaviour to staff represents a huge burden of care, affects healthcare worker productivity, job satisfaction and leads to undue stress. Formal training is strongly recommended and urgently needed to avoid staff burnout and stress-related absences.

RCSI (2013) THE MENTAL HEALTH OF YOUNG PEOPLE IN IRELAND

**SEPSIS RECOGNITION PRIOR TO ROLLOUT OF PAEDIATRIC SEPSIS FORM TOOL IN PAEDIATRICS - THE WEXFORD PERSPECTIVE.****FX Kyeyune<sup>1</sup>, S O'Reilly<sup>1</sup>, A Kalim<sup>1</sup>**<sup>1</sup>Paediatrics Department, Wexford General Hospital, Wexford, Ireland

**AIMS:** Sepsis is a serious abnormal inflammatory host response to confirmed or suspected infection. It is a leading cause of morbidity and mortality in children worldwide.<sup>1,2</sup> Treatment delay increases mortality. Therefore, early recognition and prompt management is key to improving survival<sup>2</sup>. The HSE's National Implementation Plan aims to improve effective recognition and management and hence reduce impact of sepsis <sup>3</sup> , <sup>4</sup>.

Aim of the audit was to assess current practice in the recognition and management of sepsis in our hospital prior to implementation of the paediatric sepsis form.

**METHODS:** Children managed as suspected sepsis in the Paediatric Emergency department and the Children's Ward were identified, from January 2021 to June 2022 prior to the roll-out of the Paediatric Sepsis Form.

**RESULTS:** 13 children were identified to have clinical and /or laboratory confirmation or suspicion of sepsis. Age range was 24 days to 9 years. All children had bloods taken (100%) including full blood count, renal and liver function tests and C-reactive protein. 61.5% had lactate checked and all values were <3.0. 92.3% had blood cultures taken. 69.2% had urine sent for culture. In 61.5% an organism was identified.

10 children (76.9%) required a fluid bolus. All received intravenous antibiotics. One was treated as sepsis with shock and was given antibiotics within 1 hour. Eleven (84.6%) had suspected sepsis and received antibiotics within 3 hours. One received antibiotics > 3 hours.

**CONCLUSION:** Following recognition of confirmed or suspected sepsis, 92.3% received antimicrobial therapy within 3 hours. The 'paediatric sepsis form'<sup>4</sup> was not completed because the tool was not part of standard clinical practice at the time. However, this baseline timely initiation

of treatment is encouraging. A re-audit of this quality improvement initiative is planned for later this year to evaluate use compliance and impact on improving early recognition of sepsis.

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General Paediatrics

### **A NARRATIVE ARC: A CASE OF ARTHROGRYPOSIS, RENAL DYSFUNCTION, CHOLESTASIS SYNDROME (ARC)**

**K Lynam**<sup>1</sup>, A Biagini<sup>1</sup>, L Mannion<sup>1</sup>, MJ O'Grady<sup>1, 2</sup>, K Gorman<sup>3</sup>, S Rizvi<sup>1</sup>

<sup>1</sup>Department of Paediatrics, Regional Hospital Mullingar, Westmeath

<sup>2</sup>Women's and Children's Health, School of Medicine, University College Dublin

<sup>3</sup>Department of Neurology, CHI at Temple St, Dublin

**Background:** Arthrogyposis - renal dysfunction - cholestasis (ARC) syndrome is a rare autosomal recessive multisystem disorder involving the liver, kidneys, skin, musculoskeletal and central nervous systems. Less than 100 patients have been reported in the literature to date. Most patients with this condition do not survive beyond one year of age.

**Case Report:** A male infant was born via LSCS for breech presentation at 39 weeks to non-consanguineous parents. APGAR scores were 7 and 8 at 1 and 5 minutes respectively. Positive pressure ventilation was required initially and admission to SCBU was required for ongoing

respiratory support. The infant had central hypotonia, a weak suck and absent Moro reflex. Lower limb contractures, particularly at the ankles were noted. Nasogastric feeds were required, however weight gain was poor.

Tertiary neurology opinion was sought for further investigation. Jaundice developed in association with pale stools. A normal biliary tree on ultrasound combined with a low GGT prompted consideration of ARC Syndrome. Kidneys were also morphologically abnormal.

Trio exome sequencing identified compound heterozygous mutations in *VPS33B*, confirming ARC Syndrome. In addition, a *de novo* mutation in the mitochondrial *MT-ND1* gene, encoding for a subunit of respiratory chain complex 1 was also reported.

Current clinical issues include progressive liver failure from cholestasis, renal tubular disorder, limb contractures, poor feeding and poor weight gain. The patient has been transferred home on nasogastric feeds. Treatment is supportive with an emphasis on maximizing quality of life. An advanced treatment care plan has been completed in conjunction with the family.

**Conclusion:** Whole exome sequencing provided a rapid diagnosis of a life-limiting illness. This has allowed for early palliative care involvement and supportive treatment to optimise his quality of life. The family will be referred for genetic counselling regarding risks for future pregnancies.

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Medical/Education Management

**HOSPITAL VS COMMUNITY? EXPLORING PAEDIATRIC TRAINEES' EXPERIENCES OF HOSPITAL-BASED PAEDIATRICS COMPARED WITH COMMUNITY PAEDIATRICS**

**K Lynam**<sup>1,2</sup>, P Cantillon<sup>2</sup>

<sup>1</sup>Department of Paediatrics, Midland Regional Hospital, Mullingar, Ireland

<sup>2</sup>School of Health Sciences, University of Galway, Galway, Ireland

**Background:** Most paediatric training programmes are focused on the development of hospital-based paediatricians who work primarily in tertiary centres or large acute urban hospitals. Given the acute sectoral orientation of training, community paediatric rotations are often described as having less 'social capital' than hospital-based rotations. Prejudice against community-based training is not founded on a strong evidence base because little is known about the differences in learning experiences between acute and community-based settings.

**Method:** A phenomenological qualitative approach was adopted for this study. Semi-structured interviews were recorded during the period April to May 2022 via telephone and were transcribed verbatim by the researcher in accordance with data protection. NVIVO software was used for the coding of data which were then analysed through template analysis.

**Results:** Seven paediatric trainees participated in the study, six female and one male. All trainees were paediatric specialist registrars; six were working in acute/hospital-based settings while one was working in an academic role. Hospital-based training was associated with a more independent, autonomous learning experience whereas training in the community context facilitated a better-supported consultant-led approach.

**Conclusion:** This study has highlighted the similarities and differences between paediatric trainees' learning experiences in the acute hospital setting and community setting. Pattern recognition in a fast-paced, adrenaline-fuelled world of pathology contrasts with the in-depth exploration of case complexity and deliberation in the reflective world of day-to-day adversities. The divide between learning in acute and community paediatrics is unlikely to change unless training programmes are designed to suit the needs of paediatric services.

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Neonatal

### **A THREE YEAR REVIEW OF BRACHIAL PLEXUS INJURIES IN A TERTIARY NEONATAL CENTRE**

**K Lynam<sup>1</sup>**, R Phipps Considine<sup>2</sup>, M White<sup>1</sup>

<sup>1</sup>Department of Neonatology, Coombe Women and Infants University Hospital, Dublin

<sup>2</sup>Department of Physiotherapy, Coombe Women and Infants University Hospital, Dublin

**Aims:** Brachial plexus injuries are reported to occur in 1.7 per 1,000 births<sup>1</sup>. Early recognition and management in the newborn period can significantly impact outcomes<sup>2</sup>. We aim to describe the experience of brachial plexus injuries in a tertiary level neonatal centre over the course of three years.

**Methods:** A retrospective review was conducted of babies born in the Coombe Women and Infants University Hospital from 1st August 2018 to 31st July 2021. Patients were identified from inpatient K2 discharge summaries and relevant data were collated from medical charts and physiotherapy records.

**Results:** Twenty babies were noted to have a brachial plexus injury during this time frame (0.8 per 1,000 babies). Median gestation was 39 weeks and 2 days gestation. Median weight was 3695g; one baby weighed over 4.5kg. Shoulder dystocia was noted at delivery in six (30%) of cases. Reduced movement of the affected arm was the presenting feature in 15 babies (75%). Presence or absence of Moro reflex was documented in all medical notes (100%); however, further detail such as presence or absence of shoulder flexion was present in four (20%) patients' medical notes.

Twenty patients (100%) were reviewed by physiotherapy within two weeks of birth. 12 patients (60%) underwent x-ray imaging with one clavicular fracture reported (5%). Full neurological function was regained by 3 months of age in all cases (range: two days - three months).

### **Conclusion**

Our results demonstrate an incidence of 0.8 per 1,000 babies. Paediatricians caring for newborn infants must have increased vigilance for this condition. While macrosomia is a reported risk factor for brachial plexus injury, only one baby (5%) in this study weighed greater than 4.5kg at delivery. Radiological imaging is required to outrule fractures. Detailed documentation of an abnormal neurological examination is vital.

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## THE COVID-19 PANDEMIC AND ITS PSYCHOLOGICAL IMPACT ON THE CHILDREN WITH CANCER: A SURVEY OF PRACTICE

KA Malik<sup>1,2</sup>, A Haider<sup>2</sup>, B Paturi<sup>2</sup>, C Crehan<sup>2</sup>

<sup>1</sup>Altnagelvin Hospital, Paediatrics and Neonatology, Londonderry, Northern Ireland

<sup>2</sup>Our Lady of Lourdes Hospital Drogheda, Ireland, Paediatrics and Neonatology, Drogheda, Co. Louth, Ireland

**Aims:** Identify the psychological impact of Covid-19 pandemic on the children with cancer, and their families in relation to the health care services in the hospital.

**Methods:** The paediatric patients registered at OLOL for their cancer treatment during the Covid-19 pandemic from 2019 till 2021 were included. The cancer patient experiences questionnaire (Iversen et al., 2012, Falcone et al., 2020, Tsamakis et al., 2020) helped us design the questionnaire. After the local review committee approval, the telephone numbers were extracted from the hospital record. After verbal consent from parents, the questionnaire was read during the phone call and filled accordingly. The data was collected, and results formulated by SPSS.

**Results:** 70% were satisfied with the information provided in relation to Covid 19 on cancer. 100% were satisfied with information related to treatment facilities during the lockdown. All agreed that organisation was supportive during the lockdown and relevant treatment facilities were provided, however 5% disagreed to the infection control measures, safe inpatient services and telephone consultations to be of any help. Most families were affected emotionally and quality of life. 45% denied that they felt vulnerable due to cancer diagnosis by pandemic and if it affected the disease itself or its management. 75-85% believed that healthcare staff were supportive. 95% disagreed that any psychological services were provided, 35% were not confident in the services provided. 10% felt better services were possible during the lockdown.

**Conclusions:** The decision makers should identify the gaps in performance during the unprecedented times of Covid 19 within the different healthcare settings, particularly the oncology services, to articulate policies for improved health care outcomes, patient satisfaction and psychological effects in any future pandemic (Levesque and Sutherland, 2020). A formal psychological support was necessary for children with cancer and their families, during the Covid-19 pandemic.

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### **BASILICATA- AKHTAR SYNDROME: A CASE REPORT OF AN ULTRA-RARE X-LINKED GENETIC MUTATION MSL3 GENE**

**KA MALIK**<sup>1,2</sup>, B Paturi <sup>2</sup>, S El badri<sup>2</sup>

<sup>1</sup>Paediatrics, Altnagelvin Hospital , Londonderry, Northern Ireland UK

<sup>2</sup>Paediatrics, Our Lady of Lourdes Hospital Drogheda Ireland , Drogheda Co.Louth

**Introduction:** Rare genetic mutations with nonspecific and overlapping presentations, causes a diagnostic challenge and their tailored management. A mutation in the MSL complex Male Specific Lethal is responsible for bulk Histone 4 Lysine 16 acetylation (H4K16ac) in mammals and flies<sup>1</sup>, resulting in an XLinked syndrome in humans affecting both sexes. It was identified in fifteen families across US, Australia and Europe as a de novo variant in Male Specific Lethal homolog 3 Homolog (MSL3)<sup>2</sup>. The most common presentation includes intellectual disabilities, hypotonia, global delay, and feeding difficulties.

**Case report:** A term male infant, born through emergency C-section due to fetal bradycardia with APGARS of 6<sup>1</sup> 7<sup>5</sup> 9<sup>10</sup>. The antenatal course was uneventful. At birth he had pronounced head lag, hypotonia, abdominal distension and treated for presumed sepsis and blood cultures were negative. He had a normal CRUSS, metabolic work up, CGH array and karyotyping. He was managed by an MDT approach. During infancy he had multiple admissions due to feeding difficulties and resulting LRTI. He had a right eye squint and developmental delay. He was further worked up including DNA, MRI brain(normal) and serum CK (mildly raised).At 17 months of age, subtle dysmorphic features were apparent including long philtrum, narrow upper lip with some tenting, narrow chest, pectus carinatum, prominence of right chest wall, less muscle bulk, and a

mild head lag. Finally at 3 years he was diagnosed as basilicata-akhtar syndrome by exome sequencing at cento gene that showed, a loss of function in MSL3 gene.

**Conclusion:** Nonspecific symptoms including hypotonia, developmental delay and intellectual disabilities remains a diagnostic dilemma, where many rare genetic mutations can result in overlapping presentations. However it is crucial to identify the rare entities to direct its future management and it's risk in future pregnancies.

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## **AN AUDIT OF COMPLIANCE OF HSE RECOMMENDATIONS AND STANDARD PRACTICES FOR HEALTHCARE**

### **RECORDS DOCUMENTATION IN THE PAEDIATRIC AND NEONATAL DEPARTMENT**

**KA Malik<sup>1,2</sup>**, E Ludusan<sup>2</sup>, A Haider <sup>2</sup>, K Hanif<sup>2</sup>, O Makinde<sup>2</sup>, J Wasif<sup>2</sup>

<sup>1</sup>Altnagelvin Hospital, Paediatrics and Neonatology, Londonderry, Northern Ireland

<sup>2</sup>Our Lady of Lourdes Hospital Drogheda, Ireland, Paediatrics and Neonatology, Drogheda, Co. Louth, Ireland

**Aim:** To assess the compliance of HSE recommendation and standard practices for the paper-based manual healthcare record.

**Methods:** The audit was approved by the local audit committee. Random charts from paediatric ward, NICU, and clinic were evaluated for the HSE standard recommendations for correct identification of username, legibility, documentation of date and time in 24hr clock, author identification, corrections and abbreviations. The data was collected on excel sheet and evaluated on the same.

**Results:** The compliance of correct identification number/label on each side of the page was 100%, 90% of documentation was clear and legible, 100 % were in permanent black ink, documentation of date in d/m/y against each entry was seen in 90% and 24 hr clock in 85%, author identification

signature 60%, author printed name 80%, author job title 50%, author bleep number/IMC 85%, deletions or alterations by single line in 33%, followed by a signature in 17%, followed by a name in capital 33%, with a counter signature in 67%, correction date and time of entry 83%, reason for amendment 67%, Corrections are made as close to the record in 83%, and compliance to abbreviations used on the HSE list was 75%.

**Conclusion:** It is crucial to recognise the gap in compliance to HSE standards of documentation in paperbased records. Most standards are followed; however, measures should be taken to improve on deletions and corrections in records. Also, education of HSE approved abbreviations should be prompted to facilitate its compliance and improve the legibility of documentations.

HSE Standards and Recommended Practices for Healthcare Records Management - QPSD D 006 3 V3.0 - Mastercopy

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## **INCREASING INCIDENCE OF PSYCHOLOGICAL NEEDS AMONG PAEDIATRIC PATIENTS**

C Mapanga, A Kalim, D Bowers, **M Wallis**

<sup>1</sup>Paediatrics, HSE-Wexford General Hospital, Wexford, Wexford

**Aim:** The prevalence of Mental Health (MH) presentations increased by 25% in the first year of the COVID-

19 pandemic, worldwide.<sup>1</sup> In Ireland, MH presentations increased from 143 to 218 (by 52.4%) in July - August 2020 compared to the same months in 2019; and from 379 to 552 (by 45.6%) in September -

December 2020, compared to the same months in 2019. <sup>2</sup>

Our objective was to assess the number, diagnoses and background of children who required in-patient admission to Wexford General Hospital and who would have benefited from onsite psychology services.

**Methods:** Retrospective chart review was done of all children under 16 years admitted to WGH from October 2020 to October 2021 with psychological needs. We included admissions with anxiety, depression, eating disorders, self-harm, psychosocial issues, newly diagnosed patients with type 1 diabetes and presentation of diabetic ketoacidosis.

**Results:** We identified 53 inpatients. Their ages ranged from 8 to 16 years. Children aged 14 to 15 years accounted for 50% of the admissions. 40% of the admissions were children with diabetes, either newly diagnosed or admitted in DKA. Eating disorders accounted for 21% of the admissions, followed by anxiety-related symptoms at 15%. Self-harm accounted for 13%, while 11% of admissions were under Section 12.

**Conclusion:** A significant number of inpatients were identified during the study period who would have benefited from onsite psychology input. Our access to community psychology is via a waiting list of up to 4 years. An onsite Psychologist will greatly improve the outcome of these children. We are aware of new approaches which may help paediatricians to identify and address psychological needs<sup>3</sup>. The Faculty of Paediatrics of the RCPI should set a national strategy for paediatrician support for child and adolescent mental health, similar to that of the RCPCH in the UK.<sup>4</sup>

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## **ACCURACY OF POINT OF CARE ULTRASOUND IN THE DIAGNOSIS OF ACUTE APPENDICITIS WITHIN A PAEDIATRIC POPULATION; SYSTEMATIC REVIEW & META-ANALYSIS**

**CA McCarrick<sup>1</sup>, K Kelleher<sup>1</sup>, R Murphy<sup>1</sup>**

<sup>1</sup>Temple Street Hospital

**Introduction:** Abdominal Pain is a frequent presentation within a paediatric population to the Emergency Department. When deliberating between differential diagnoses such as mesenteric adenitis or acute appendicitis the use of conventional abdominal ultrasound is well-established to determine the primary cause. However, more recently within the United States and Europe the use of POCUS (Point of Care Ultrasound) by a non-radiology trained physician is becoming

increasingly popular as a tool within the Emergency Department to assess whether a patient has acute appendicitis. This systematic review and meta-analysis sets out to establish the level of accuracy of POCUS in the diagnosis of acute appendicitis.

**Methods:** A systematic literature search was performed by two independent reviewers of English language databases including Pubmed, EMbase, google scholar, Scopus and Cochrane for research pertaining to the use of point of care ultrasound in a paediatric population for the detection of acute appendicitis from 2000 to 2022 inclusive. A meta-analysis was then performed of all relevant data which was selected.

**Conclusions:** Point of Care Ultrasound exhibits high sensitivity and specificity in the detection of acute appendicitis within a paediatric population. However, a key variable is operator ability. POCUS should not be a standalone test and those patients with a high clinical suspicion of acute appendicitis should undergo formal transabdominal ultrasound.

## **GENERAL PAEDIATRIC SPECIALIST CLINIC -A CLINICAL NURSE SPECIALIST DELIVERED PARENT EMPOWERMENT INITIATIVE THAT REDUCES HOSPITAL APPOINTMENTS**

**C Mc Donnell** , S Kelleher, B Scanlan, J Donnelly

<sup>1</sup>General Paediatrics , Children Health Ireland , Dublin , Ireland

**Aims:** Aim of this initiative is to expedite care following GP referral and Consultant triage. Three cohorts of patients were identified with common conditions to be managed by a CNS. This initiative allows parents/carers the opportunity to connect with an expert to offer advice and treatment if required, reducing the number of physical appointments with the Consultant. Some patients can be followed up with the General Paediatric CNS only.

**Methods:** A telephone CNS clinic was established.

96 referrals were triaged and wait listed, patients with symptoms of constipation (51%), continence issues (29%) and asthma (18%) are directed to the CNS clinic. A letter is sent to the parents and GP with contact made within ten weeks. Consultation involves education, reassurance and advice, lifestyle changes and prescription medication if required and written and online information.

Asthmatic patients are sent action plans and information about correct inhaler use and symptom management.

Records are kept of calls with parents and any advice or treatment commenced and outcome.

Follow up calls are made as required and email is provided for queries.

**Results:** During the first 8 months, 96 patients were referred to the CNS led clinic. 76% had either complete resolution or significant improvement in their symptoms following the CNS clinic. 7% of those contacted did not engage with the CNS. Following phone contact, some referrals changed triage category. Duplicates were excluded. 50% did not require Consultant appointment. The clinic empowers parents to manage their child's condition at home with healthy lifestyle changes and medication. The Health Service Executive Integrated Care Programme for Children has identified the need to strengthen links between primary and secondary care. GPs receive detailed summary of intervention. This CNS led clinic has the potential to strengthen these links between primary and secondary care into the future.

**Conclusion:** Early intervention and advice for parents along with regular follow up calls has led to a reduction in the number of consultant appointments needed.

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## **SOCIAL ADVERSITY AND HEALTHCARE PROVISION: A SNAPSHOT OF CONSIDERATIONS FOR INCLUSIVE CHILD HEALTHCARE DELIVERY IN IRELAND**

**J McGirr**<sup>1</sup>, A Walsh<sup>1,2</sup>, E Molloy<sup>2</sup>, C Ní Cheallaigh<sup>3,4</sup>, C Hensey<sup>1,2</sup>

<sup>1</sup>General Paediatrics Department, CHI at Temple Street, Dublin, Ireland

<sup>2</sup>Faculty of Paediatrics and Child Health, School of Medicine, Trinity College Dublin, Dublin, Ireland

<sup>3</sup>Inclusion Health, St. James's Hospital, Dublin, Ireland

<sup>4</sup>Clinical Medicine, School of Medicine, Trinity College Dublin, Dublin, Ireland

**Aims:** The need to provide inclusive healthcare to marginalised members of society is a national and global priority(1). RCPI reports on children experiencing homelessness and those living in Direct Provision acknowledge barriers encountered in accessing and engaging with healthcare in an Irish context(2,3). These obstacles based on social determinants of health (SDH) include education, language, transience and discrimination(4).

**Methods:** Recent national figures were reviewed to estimate the number and characteristics of cohorts of children with recognisable adverse SDH. Identification of key stakeholders in governmental and nongovernmental bodies supporting vulnerable children and families was also performed to forge working partnerships that bridge hospital and community health and social care.

**Results:** Key national figures from a child health perspective in July 2022 include 3,317 children accessing emergency accommodation(5), 3,220 children seeking international protection(6), 2,339 children homeless in Dublin(5) and 12,034 Ukrainian refugees children registered(7).

The following key stakeholders in supporting marginalised children and their families have been identified;

- 1) International Protection Accommodation Services
- 2) National Social Inclusion Office
- 3) HSE Healthlink for the Homeless Team
- 4) SafetyNet Primary Care
- 5) HSE Public Health
- 6) HSE Health Screening Team, Baleskin Reception Centre



**Conclusion:** There are significant numbers of children with recognisable adverse SDH living in Ireland. Clear need for tailored interagency healthcare for children at risk of social exclusion is evident. The requirement for integrated service delivery is recognised by CHI. A hospital-based workforce trained in the recognition and mitigation of the social determinants of health is currently in development

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## REVIEW OF URINARY TRACT INFECTION IMAGING: ROOM FOR IMPROVEMENT?

E McLaughlin<sup>1</sup>, S Kelleher<sup>1</sup>

<sup>1</sup>Department of General paediatrics, CHI at Crumlin, Dublin, Ireland

**Aim:** Urinary tract infections (UTI) in children often requires imaging such as renal ultrasonography (RUSS), +/- micturating cystourethrograms (MCUG) and dimercaptosuccinic acid (DMSA) scans.[1] NICE Guidelines encourage a more targeted approach to investigations.[2] A CHI guideline on management of 1<sup>st</sup> UTI is pending, at present a certain amount of physician discrepancy exists in the selection of follow-up tests. This review assessed follow up imaging practices by general paediatricians, with the aim of reducing unnecessary investigations, relieving pressure on radiological services.

**Method:** This is a retrospective analysis of children with UTI imaging. Patients were identified by accessing RUSS booked to investigate children with UTI.

**Results:** A sample of 50 patients with UTI imaging were reviewed. Age 2 weeks to 11 years. M:F ratio 1:2. 10% had initial RUSS as an outpatient.

**Group 1:** 23 (46%) < 6 months. 52% had normal RUSS, 61% had a follow up RUSS. 3 had DMSA scans one showed scarring, one was not indicated, one was normal. 3 MCUG scans were done, one had significant reflux, 2 were normal, 1 was not indicated.

**Group 2:** 13 (26%) > 6 months – < 3 years. 62% had abnormal RUSS, 1 infant with confirmed Pyelonephritis had no follow up imaging. 2 had appropriate DMSA scans, both normal

**Group 3:** 14 (28%) > 3 years. 86% had abnormal RUSS, 79% had follow up RUSS, 4 had DMSA scans (all abnormal), 2 had MCUGs.

**Conclusions:** Previous CHI Crumlin guidelines recommended infants have a follow up RUSS, however 40% didn't have this. There is currently no international guidance on indications for follow up RUSS. It is reassuring that 96% of the sample did not have unwarranted invasive imaging. Only 2 children had unnecessary imaging, one MCUG and one DMSA. No child over a year had an MCUG inappropriately requested.

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## SEVERE RHABDOMYOLYSIS POST COVID-19 INFECTION IN A PAEDIATRIC PATIENT

O McNerney<sup>1</sup>, R Finnegan<sup>1</sup>, N Laher<sup>2</sup>, R Tobin<sup>2</sup>, C McKenna<sup>2</sup>, A Smyth<sup>3</sup>, L Baker<sup>1</sup>

<sup>1</sup>Department of Paediatrics, Children's Health Ireland at Temple Street, Dublin, Ireland

<sup>2</sup>School of Medicine, Trinity College Dublin, Dublin, Ireland

<sup>3</sup>Department of Radiology, Children's Health Ireland at Temple Street, Dublin, Ireland

**Background:** Rhabdomyolysis is a life-threatening syndrome caused by destruction of muscle cells for a number of reasons, including viral infections. SARS-CoV-2-associated rhabdomyolysis has been reported in adults and is attributed to direct toxicity by the virus. However, there have been

few reported cases in children. We present a case of severe rhabdomyolysis in a child post COVID-19 infection.

**Case Report:** An 11-year-old girl of African origin presented to the Emergency Department with a short history of fatigue, myalgia and dark-coloured urine following symptomatic COVID-19 infection two weeks previously. On presentation she had difficulty mobilising, increasing muscle pain, and tenderness on palpation of thigh muscles bilaterally. Past medical history was significant for sickle cell trait and there was no relevant family history of note. Urine dipstick was positive for protein, blood, and bilirubin. Bloods revealed a significantly raised creatine kinase (CK) at 437,574 U/L (normal <25U/L). MRI of her thighs showed hyperintensity within muscles of both thighs in keeping with extensive myositis.

Our working diagnosis was rhabdomyolysis secondary to recent COVID-19 infection. She was treated with hyper-hydration, diuretics, and bicarbonate as reno-protective agents; strict fluid balance and serial blood testing were conducted. CK levels remained >10,000 U/L for 10 days, without renal impairment. She was discharged home on day twelve post-admission and CK levels normalised after five weeks.

**Discussion:** We highlight this condition as there are limited reported paediatric cases of SARS-CoV-2-related rhabdomyolysis with significantly raised CK levels. Furthermore, it is well recognised that sickle cell disease is a risk factor for developing rhabdomyolysis, however sickle cell trait is often considered a benign entity. This case offers further support to the suggestion in the literature that sickle cell trait may augment the myositis caused by viral infection and is worth consideration when reviewing children from certain ethnic backgrounds presenting with muscle pains.

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**A RETROSPECTIVE CROSS SECTIONAL REVIEW OF PAEDIATRIC PSYCHIATRIC PRESENTATIONS TO WEXFORD GENERAL HOSPITAL'S EMERGERGENY DEPARTMENT FROM JANUARY TO MARCH 2022**  
JB Miller<sup>1</sup>, A Kalim<sup>1</sup>

<sup>1</sup>Paediatrics Department, Wexford General Hospital , Wexford, Ireland

**Aims:** Multiple studies<sup>1, 2</sup> have shown the negative effect of Covid-19 pandemic on well-being and mental health (MH). The number of paediatric psychiatric visits to emergency departments (ED) are steadily increasing in the recent years<sup>3</sup>. Our aim was to assess the number of children presenting to the ED in Wexford General Hospital (WGH), their basic demographics, presenting symptoms and outcome.

**Method:** The data was collected for all the patients under sixteen years of age presenting to the ED with any MH concerns, from January 2022 to March 2022.

**Results:** There were 70 patients who presented with MH concerns under 18 years of age, among these 43 (62%) patients were under 16 years of age. We further analysed the data of only those patients who were under 16 years of age. 15 (35%) patients were male and 28 (65%) were females.

26 (61%) were discharged from the ED and 17 (39%) were admitted to the ward.

The presentations of the admitted patients were; 10 (56%) intentional overdose, 4 (25%) social admissions, 2 (13%) behavioural issues and 1 (6%) with suicidal ideation.

Among the patients who were discharged from the ED, 12 (48%) were sent home with no follow up, 4 (15%) were discharged to University Hospital Waterford (UHW) for further urgent psychiatric review, 6 (22%) were discharged to CAMHS and 4 (15%) were discharged home with a GP follow up.

**Conclusion:** Significant number of patients with MH concerns presented to the ED in a short time period which reflects huge burden of the disease. 17 (39%) patients were admitted to the ward for an acute medical care, but ultimately they required the input from the Psychiatrists in UHW. 4 (10%) patients were transferred out to UHW for an urgent Psychiatric review. These findings indicate the need of local Psychiatric services.

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**OUTCOMES OF ABDOMINAL PAIN AMONG CHILDREN PRESENTING TO EMERGENCY DEPARTMENT OF TERTIARY HOSPITAL IN IRELAND BEFORE AND DURING COVID-19 PANDEMIC.**

**GM Mineva**<sup>1,2</sup>, EC Culleton<sup>1,2</sup>, TF Falusi<sup>2</sup>, DR Ryan<sup>1,3</sup>

<sup>1</sup>University Hospital Limerick, Limerick, Ireland

<sup>2</sup>School of Medicine, University of Limerick. Limerick

<sup>3</sup>ALERT, Emergency Department, University Hospital Limerick.

**Aims:** Acute abdominal pain in paediatric patients is a common presentation to the Emergency Department (ED). The COVID-19 pandemic had a significant impact on the health care sector, leading to a decline in hospital attendance. The main aim was to compare the burden of paediatric abdominal pain one year before and after the start of the COVID-19 pandemic.

**Methods:** A retrospective cohort study was conducted in a tertiary hospital. Ethical approval was obtained. Data was collected from: January 1 - June 30 from 2019 and 2021. Patients' charts were accessed using Therefore<sup>®</sup> software. Key variables were extracted and summarised in Microsoft Excel tables. Univariate and multivariate data analysis were conducted in SPSS<sup>®</sup> software.

**Results:** 661 (2019) and 479 (2021) patient records were evaluated. There was a 28% reduction in case presentations during pandemic months. Age group, sex and attending discipline were similar for both years. Rates of imaging were significantly higher in 2021 (10.6% vs 22.5%). The overall admission rate was higher in 2021 (37%) compared to 2019 (25%,  $p < 0.001$ ). Viral gastroenteritis decreased (13%) and nonspecific diagnosis increased (46%) in 2021. Fewer children represented in 2021 (13 %).

**Conclusions:** The COVID-19 pandemic had an impact on the presentation, investigations, and admission rates of abdominal pain in the paediatric ED. Our study highlights the need for increased awareness of severe abdominal pain signs and symptoms in paediatric patients, and the importance for diagnostic accuracy. Improving patient flow through the ED and having clear guidelines for staff and parents can optimise clinical practice and improve quality of care and patient outcomes.

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04015-0

## **OUTCOMES AFTER FIRST PSEUDOMONAS AERUGINOSA (PA) ISOLATE IN CYSTIC FIBROSIS (CF) PATIENTS 2018-2022**

**AM Murphy**<sup>1</sup>, D O'Donovan<sup>1</sup>, M Aherne<sup>1</sup>, P Shanahan<sup>1</sup>, L Greene<sup>1</sup>, S Dunne<sup>1</sup>, D Mullane<sup>1</sup>, M Ni Chroinin<sup>1</sup> <sup>1</sup>Paediatric Department, Cork University Hospital, Cork, Ireland

**Aim:** To assess children with first isolate for PA with regards to number, age, modulator therapy, eradication and follow-up surveillance.

**Method:** A retrospective observational study of children with CF currently attending clinic in Cork University Hospital (CUH).

**Results:** In September 2022, 101 CF patients attended all or part of their care at CUH. The prevalence of PA in 2022 according to Leeds criteria was 49 never; 31 free; 20 intermittent; 1 chronic. Between January 2018 and September 2022, 21 had a first isolate of PA (2018 3; 2019 4; 2020 4; 2021 6; 2022 4). All isolates were from throat swabs. Age range was 6 months – 12 years. 49 patients never isolated PA with 28 of them on modulators. 21 patients had first isolate with 14 not on modulator therapy. From 2018-2022, 7 patients out of 35 had first isolate of PA in modulator group while comparatively 14 out of 35 had first isolate in nonmodulator group. All had eradication treatment with oral ciprofloxacin for 3 weeks and nebulised tobramycin for 1 month. Monthly surveillance swabs for 6 months post treatment was required as per local protocol. 58% of patients had surveillance according to this protocol. After 6 months, 13 were negative and 5 had further isolate who received a course of IV treatment for 2 weeks.

**Conclusion:** First PA isolate frequency was stable over 4 years. Interestingly there appears to be an increase of first isolates in children not on modulators compared to those on modulator therapy which may indicate a protective effect. Surveillance following first isolate could be improved. The pandemic may have affected ability of families to send surveillance swabs between clinic appointments and associated staffing deficits affected ability to ensure adequate surveillance.

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### **ASPIRING TO STANDARDS IN DEVELOPMENTAL ASSESSMENT: THE ASD AUDIT**

**AL Murray**<sup>1</sup>, LA Gibson<sup>1,2</sup>

<sup>1</sup>Paediatric Department, Cork University Hospital, Cork, Ireland

<sup>2</sup>Department of Paediatrics and Child Health, University College Cork, Cork, Ireland

**Introduction:** Autism spectrum disorder (ASD) is the most common neurodevelopmental disability<sup>1</sup>. Globally there is an increase in prevalence, with most recent Irish prevalence of 1.5%<sup>2</sup>. The definition of autism has evolved since its first description in 1943, and assessment of autism remains subject to interpretation of published guidance, with significant inter-professional

variability<sup>3</sup>. Comprehensive referral letters and adhering to best practice standards will improve communication with assessment teams and minimise unnecessary delays in assessment.

**Aims:** To audit our assessment of autism spectrum disorder in the paediatric outpatient setting, considering the NICE guidelines (2017) as the clinical standard<sup>4</sup>.

**Methods:** A retrospective audit of all children referred to the Paediatric Developmental clinic was undertaken between April-July 2022. All new referrals from 1<sup>st</sup> January 2019-1<sup>st</sup> January 2022 were examined for reason for referral. 151 new referrals were identified.

**Results:** Thirty-seven referrals with concern regarding a new diagnosis of autism between Jan 2019-Jan 2022 were examined. The mean patient age was 3.7 years.

All letters contained the details of the developmental and behavioural history (100%); most recorded medical history (97.3%), assessment of the child's social and communication skills (89.2%), and the details of home life, education and social care (89.2%). Fewer letters described the parents', /carers' concerns (83.8%). Fewer still noted the details of physical examination (82.8%) or referenced possible differential diagnosis (81.1%). The topics most in need of improvement include a systematic assessment for comorbidities (75.7%) and planning for the future for the family and education for the child (62.2%).

**Conclusion:** Assessment of young children with suspected ASD occurs commonly in Paediatric clinics.

Currently our referral letters are not fully compliant with international standards. We plan to implement an "Autism Assessment Tool" for Paediatric OPD ( based on the NICE guidelines) to minimise unnecessary delays in the referral process. Further audit is planned in 2023.

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## UNPROVOKED DEEP VEIN THROMBOSIS AND PULMONARY EMBOLISM IN PREVIOUSLY HEALTHY ADOLESCENT MALE.

TM Mustonen<sup>1</sup>, A Kumar<sup>1</sup>

<sup>1</sup>Department of Paediatrics, University Hospital Kerry, Tralee, Ireland

**Aims/Background:** Venous thromboembolism (VTE) is a rare diagnosis in the paediatric population with an incidence of up to 0.21 per 10000 children per year<sup>(1)</sup>. Most children who develop a VTE have underlying medical conditions. While pulmonary embolism (PE) is less common than deep vein thrombosis (DVT) in children, the incidence of PE is increasing which can be attributed to a variety of factors<sup>(2)</sup>.

We aim to discuss the case of a healthy, active adolescent male who developed an unprovoked DVT and PE.

**Methods:** We describe the clinical presentation, examination findings, radiological and laboratory investigations, and treatment of an adolescent male.

**Results:** This previously healthy 15-year-old boy presented to the emergency department with left leg swelling and pain with no history of trauma to the leg. Initial bloods showed an elevated d-dimer of 4.09 mg/L (reference range 0-0.5). The patient underwent a left leg compression Doppler study which found evidence of a DVT, and he was subsequently started on therapeutic tinzaparin. Although our patient was not experiencing any of the classical symptoms of a PE (dyspnoea, pleuritic chest pain and haemoptysis), we conducted a CT PA due to the significant family history of DVT and PE in younger family members, and a family history of Protein C deficiency. The CT PA demonstrated extensive emboli in the bilateral segmental arteries. At this time, he was transferred to a tertiary hospital for further management where he was switched to rivaroxaban and his thrombophilia workup is outstanding.

**Discussion/Conclusion:** PE in the paediatric population is a rare, but important condition that must be diagnosed and managed promptly. While the classical symptoms of a PE are often present in children, they may also be absent. We suggest that clinicians consider further imaging looking for PE in patients who present with a DVT, even in asymptomatic patients.

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**A RARE PRESENTATION OF ISOLATED GLUCOSURIA IN A TEENAGER WITH RECURRENT ABDOMINAL PAIN** S Nadarajan<sup>1</sup>, L Abdalla<sup>1</sup>, D Staunton<sup>1</sup>

<sup>1</sup>General Paediatrics, Mayo University Hospital, Castlebar, Mayo, Ireland

**AIM:** To describe a case of a 13 year old girl with a history of recurrent abdominal pain and associated vomiting with an incidental finding of glucosuria, in whom a mutated gene of SLC5A2 was found to provide a likely diagnosis.

**METHODS:** Description of clinical presentation, workup, diagnosis and management including a literature review of the known cases.

**RESULTS:** PM is a 13 year old girl who presented with recurrent episodes of periumbilical pain with associated vomiting, on a background of previous urinary tract infections. Positive findings on systemic examination showed a tender periumbilical region with no rebound tenderness noted. An ultrasound pelvis and kidneys then showed a focus of renal cortical scarring - particularly on the right side and a small volume of debris noted. A dimercaptosuccinic scan was subsequently performed which showed relatively equal kidney function bilaterally with the left kidney contributing 53% and the right kidney showing 47% - with reduced uptake in the lower pole of the right kidney, in keeping with the renal scarring. With the repeated presentation of glucosuria, further investigations including urinary amino acids, organic acid profile and a HbA1c were sent which were reported normal. Genetic testing for SLC5A2 mutation was then sent. This identified a likely pathogenic variant which was consistent with a genetic diagnosis of autosomal recessive renal glucosuria.

Familial Renal Glucosuria is characterised by persistent isolated glucosuria with normal blood glucose and the absence of any signs of renal tubular dysfunction<sup>1</sup>. It is a rare tubular disorder caused by mutations in SLC5A2 gene that encodes the sodium-glucose cotransporter 2 (SGLT2)<sup>2</sup>.

**CONCLUSION:** In this unexpected repeated finding of isolated glucosuria with a background of unremarkable investigations, it is important to consider a possible differential diagnosis of familial renal glucosuria - especially if diabetes mellitus has been ruled out.

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## **RAPID ACCESS FAILURE TO THRIVE CLINIC - NO FAILURE HERE**

**A Naughton**<sup>1</sup>, N Kandamany<sup>1</sup>

<sup>1</sup>Emergency Department, CHI at Temple Street, Dublin, Ireland

**Aims:** Failure To Thrive (FTT) is a common reason for presentation to the Emergency Department (ED). These babies need a thorough history, examination, medical workup and dedicated follow up with paediatrics and dietetics to ensure optimal growth. We aim to look at how many babies with FTT were seen in Temple Street Hospital (TSH) ED over a one year period, and identify how many had appropriate follow up, to make a case for a dedicated rapid access clinic.

**Methods:** Using the emergency department management software Symphony, we looked at presentations over a one year period, from 01/07/21-30/06/22. Search terms 'failure to thrive', 'ftt', 'poor weight gain', 'feeding issues' and 'poor feeding' were used. Discharge Diagnoses 'failure to thrive', 'feeding problems' and 'feeding problem of the newborn, unspecified' were also included.

**Results:** This search yielded 165 results. Applying Exclusion Criteria - children over the age of 16 weeks, and any presentation that was deemed to not be true FTT (eg. feeding problems secondary to bronchiolitis) - the final yield was 62 patients. 42% (26) of these patients were admitted from ED. 35% (22) were discharged with public health nurse referrals. 22% (14) were referred on to general paediatrics outpatient services, with hugely varying lengths of time to be seen - ranging from 6 days to 3 months.

**Conclusion:** A rapid access FTT clinic would be very beneficial for babies presenting with FTT to TSH ED. The varying, lengthy wait times for OPD appointments mean that paediatric outpatient referrals are risky for underweight babies and struggling families. The current high admission rate would also put needless excess financial burden on the health service (€878/bed/night). A rapid access OPD clinic with paediatric and dietetic service would provide a cheaper, more timely service to these fragile babies.

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## **RADIOPAQUE MARKER COLONIC TRANSIT STUDY AS TOOL TO HELP DIAGNOSE DYSMOTILITY AND TO SELECT CANDIDATES FOR FURTHER MANAGEMENT OVERSEAS.**

**RN Naveed<sup>1</sup>, MS Sani<sup>1</sup>, AH Herrera<sup>1</sup>**

<sup>1</sup>General Paediatrics, St.Luke's General Hospital, Co.Kilkenny, Ireland.

**Aim:** Neurogastroenterology and motility disorders are common and have a high health care burden. A proper assessment of the pattern of constipation can alter the clinical and management of children with constipation in roughly 50% of cases. The identification of a pattern of Slow Transit Constipation vs a pattern of Anorectal Dysfunction, will guide the decision making process and selection of the treatment. Currently if a candidate to surgical intervention is identified it will need to be transferred overseas as the surgical management of complex dysmotility disorders is not available in the Republic of Ireland. We review a case series of 20 children referred to the only dedicated gastroenterology clinic outside the Dublin area, who underwent a Radiopaque Markers Study (RMS). The goal is to identify the number who changed their initial management after getting the study completed and their progress in the following year.

**Method:** Data was collected by reviewing patients charts and radiology imaging on the NIMIS computer system. The gender, weight and length of previous treatment distribution is also reviewed. This series includes a case of severe respiratory distress due to heart compression secondary to massive abdominal distension and the identification of a severe case of colon neuropathy previously considered a behavioural disorder. Data was collected by reviewing patients charts and radiology imaging on the NIMIS computer system.

**Results:** At least 4 of the 20 cases required further assessment after results of the RMS. 1 patient was referred and treated overseas successfully.

**Conclusion:** It is not rare that children do not get a proper diagnosis and treatment plan. The Radiopaque Markers Study (RMS), because its simplicity and low-cost, may act as an effective screening tool in the process to select patients to be transferred to overseas motility services. They may have a measurable effect on selecting the most potentially effective treatment for children with complex constipation.

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**INCIDENCE OF ANTENATAL STEROID ADMINISTRATION IN PRETERM DELIVERIES:  
DOCUMENTATION IN  
NEONATAL CHART**

**RN Naveed<sup>1</sup>, BN Nguyen<sup>1</sup>, MAB Boyle<sup>1</sup>**

<sup>1</sup>Neonatology, The Rotunda Hospital, Dublin, Ireland

**AIM:** To ascertain the incidence of antenatal corticosteroid administration in the preterm population < 33 weeks and documentation in the Neonatal chart in accordance with our clinical guidelines.

**METHODS:** The audit was based in the Rotunda hospital and data was acquired in a retrospective timeframe from November 2021 to June 2022. Inclusion criteria comprised all infants born < 33 weeks of gestation. Patient information relevant to our audit included gestational age, birth weight, gender, mode of delivery, and documentation of antenatal steroid. The number of doses of steroids given was collected and the interval in hours after last dose and delivery was recorded. The information was compiled on Microsoft excel.

**RESULTS:** 95 eligible patients were included in the audit of which 52 infants (54.7%) were male and 43 (45.6%) were female. The median gestational age was 31+0 weeks. Majority of infants were born by caesarean section (73.7%). Birth weights ranged between 525 grams to 2720 grams. Antenatal steroid administration was appropriately documented in 88 (92.6%) cases. It was noted that in 24 (25.3%) cases a completed course did not occur. 7 of those had no ANS administration documented. In 11 infant greater than 24 days elapsed between second dose and delivery. 25 patients (26.3%) had no time of administration of second dose mentioned.

**CONCLUSION:** Compliance to the clinical guidelines on ANS use is high with 92.6 % of infants receiving at least one dose of antenatal steroids which is vital in preventing neonatal morbidity and mortality. Compliance to the clinical guidelines on ANS use is vital in preventing neonatal morbidity and mortality. Documentation is an integral part of this process. Attention needs to be brought to the importance of proper documentation of ANS administration.

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General Paediatrics

### INNOVATIONS IN PAEDIATRIC CARE - WHERE DO WE PUBLISH?

S Ní Neachtain<sup>1</sup>, M Salama<sup>2</sup>, L Jansen<sup>2</sup>, MB O'Neill<sup>2</sup>

<sup>1</sup>Paediatrics, University of Galway, Galway, Ireland

<sup>2</sup>Paediatrics, Mayo University Hospital, Castlebar, Ireland

**Aims:** Clinical prediction rules (CPRs) are widely used in paediatric practice for risk stratification of common presentations. (1) We aimed to analyse the impact factors of the journals that published the studies behind recent CPRs, which reflect readership and influence. Accessibility to new CPRs can aid both experienced paediatricians and medical students in their practice.

**Methods:** The clinical prediction rules used were based on a systematic review done previously by the group, which was assessing validation studies for CPRs between 2010-2019. Out of 86 CPRs from that review, 81 had attributable derivation studies. We listed the journal's respective current impact factors, and compared this to the impact factor at the time of publication. In general, impact factors higher than 10 classify as “excellent”, between 3-4 is considered “good” whilst impact factors of 1 or less are poor. (2)

**Results:** The studies were allocated into five categories based on their impact factor; <1, 1-5, 5-10, 10-15. Journals with impact factors ranging from 1 to 5 were the commonest at 53.1%. 19.8% had impact factors less than 1. 12.3% and 2.5% of the journals had factors ranging between 5-10 and 10-15 respectively. 12.3% of the studies were missing impact factors at the time of publication. Journals with the highest impact factors were The Lancet and NEJM.

**Conclusion:** In general, the studies which formed the basis the majority of the CPRs were published in journals with low or average impact factor journal. While many of the CPRs are widely used, the derivations were not originally delivered to a wide readership. For example, the PEWS original study was published in a journal with no reported impact factor in 2005 at publication, and even now is as low as 0.28. More consideration needs to be given to improve the accessibility of clinicians to validated CPRs and impactful journals.

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## RETROSPECTIVE AUDIT OF HEEADSSS SCREENING IN ADOLESCENTS ADMITTED TO THE PAEDIATRIC WARD

S O'Loughlin<sup>1</sup>, H Hanafi<sup>1</sup>, E Ruth<sup>1</sup>

<sup>1</sup>Department of Paediatrics, Children's Health Ireland at Tallaght



**Aims:** Adolescence is a time of biological growth and social transition. Mental health disorders commonly emerge during this period, and risk-taking behaviours are at their peak. In Ireland, suicide and accidents are the leading causes of youth mortality (1). It is essential that paediatric doctors carry out assessments of psychosocial risk in order to provide targeted interventions and employ prevention strategies (2). The

HEEADSSS (**H**ome environment, **E**ducation and employment, **E**ating, peer-related **A**ctivities, **D**rugs, **S**exuality, **S**uicide/depression and **S**afety from injury and violence) screening tool is used to comprehensively assess psychosocial risk and resilience factors in adolescence (3).

**Methods:** A retrospective audit of HEEADSSS screen use in adolescent inpatients aged 13-15 years of age was performed. Patients admitted from the Emergency Care Unit between April - September 2022 were included. Data was retrieved from individual medical charts which were selected at random. A HEEADSSS screen was deemed to be complete if documented in the admission note.

**Results:** Twenty-two patients were included in the analysis. The median age was 15 years (range 13-15 years). The most common reason for admission was suicidal ideation and/or deliberate self-harm (68%; n=15), followed by UTI (1), DKA (1), asthma (1), trauma (1), fatigue (1), abdominal pain (1) and ear infection (1). A HEEADSSS screen was completed for six patients (27%), of which five were admitted due to suicidal ideation and/or deliberate self-harm and one was admitted due to DKA.

**Conclusions:** This study demonstrates suboptimal implementation of the HEEADSSS screening tool for adolescent patients on admission. In order to improve compliance, a dedicated teaching session on the implementation of HEEADSSS screening was delivered to all paediatric NCHDs. To complete the audit cycle, a re-audit will be performed aiming for 90% compliance.

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## **NUTRITIONAL COMPOSITION OF DONOR BREAST MILK: EXPERIENCE IN A REGIONAL NEONATAL UNIT**

**S O'Loughlin<sup>1</sup>, R Kelly<sup>1</sup>, N Al Assaf<sup>1</sup>, R Khan<sup>1</sup>**

<sup>1</sup>Department of Neonatology, University Maternity Hospital Limerick, Limerick, Ireland

**Aims:** When maternal breast milk is unavailable, or the quantities are insufficient, donor breast milk (DBM) is the preferred alternative for premature and LBW infants. It provides immunological benefits, improves enteral feed tolerance and protects against necrotising enterocolitis when compared with formula milk (1). The nutritional composition of DBM varies depending on the time of day, stage of lactation and degree of prematurity. Fortification is required to adequately support growth and neurodevelopment (2).

Standardised fortification practices are based on assumed macronutrient reference values for human milk. Our aim was to characterise the macronutrient composition of DBM in a regional neonatal unit.

**Methods:** This prospective observational study examined the macronutrient composition of DBM received in the neonatal unit at University Maternity Hospital Limerick from May-July 2022. Values for volume(ml), fat(g/100ml), protein(g/100ml), carbohydrate(g/100ml), total sugar(g/100ml) and energy(kcal/100ml) were obtained from batch labels and analysed using descriptive statistics.

**Results:** Twenty-one batches of DBM were received. Volumes ranged from 270-1100ml, fat concentration from 1.8-6.1g/100ml, protein concentration from 0.6-1.1g/100ml, carbohydrate concentration from 5.99-15.4g/100ml, total sugar concentration from 8.7-15.9g/100ml and energy content from 44-95kcal/100ml. The median fat concentration was 3.6g/100ml, with a 54% difference between quartile 1(2.8g/100ml) and quartile 3(4.3g/100ml). The median protein concentration was 0.8g/100ml, with a 25% difference between quartile 1(0.8g/100ml) and quartile 3(1g/100ml). The median carbohydrate concentration was 8.3g/100ml, with a 5% difference between quartile 1(8.1g/100ml) and quartile 3(8.5g/100ml). The median energy content was 73kcal/100ml, with a 22% difference between quartile 1(64kcal/100ml) and quartile 3(78kcal/100ml).

**Conclusions:** There were marked variations in fat and protein concentrations, and subsequent differences in energy content. Carbohydrate concentrations were consistent across samples. In

order to provide consistent macronutrient concentrations that provide for optimal growth and development, dietician involvement is paramount (3). Advances in customised nutrition, through breastmilk analysis and individualised targeted fortification, are promising (4).

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## **DUMPING SYNDROME: A CASE OF IDIOPATHIC POST-PRANDIAL HYPERINSULINAEMIC HYPOGLYCAEMIA** COS O'Shea, SG Gallagher

<sup>1</sup>Paediatrics, University Hospital Limerick, Limerick, Ireland

**Aims:** To describe the clinical presentation and course, in a case of paediatric post-prandial hyperinsulinaemic hypoglycaemia (PPHH).

**Methods:** A 2 year old boy with a complex background history of developmental delay and NG feeding in place presented with episodes of recurrent hypoglycaemia without a precipitating cause. Observation and blood sugar monitoring on the ward revealed a pattern of post-prandial hypoglycaemic events. Critical hypoglycaemic samples were obtained following a prolonged fast. Interestingly the patient's blood sugar did not drop during the fast, but rather post cessation of the fast and after restarting feeds. Bloodwork showed glucose of 1, insulin 15.1, C-peptide of 0.36, absent ketones, and normal metabolic profile. This pattern was consistent with dumping syndrome.

**Results:** PPHH, also known as late dumping syndrome, is a well-recognised complication of gastrointestinal surgery and has been identified following fundoplication in particular<sup>1</sup>. There is little data available on idiopathic cases of PPHH in the paediatric population. Recognition of this condition is vital in young children in order to protect their neurodevelopment, which may be hindered by recurrent hypoglycaemic events. Alteration of feeds is the primary treatment, however pharmacological therapies do exist. Attaining normoglycaemia, without reliance on continuous feeds, was challenging in this case, but a priority, due to the patients age and level of activity. The goal was to achieve a bolus feeding regimen to allow periods between feeds where he could be active and stimulated, to encourage his developmental progress. In this case, modification of feeding regimen, with the addition of corn-starch, proved successful in the prevention of hypoglycaemia.

**Conclusion:** There is limited literature describing idiopathic paediatric PPHH. It remains difficult to detect and diagnose. Accurate history taking and continued monitoring are necessary to recognise PPHH. Early identification is crucial to prevent poor neurodevelopmental outcomes due to recurrent hypoglycaemia.

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## **EVALUATING THE MANAGEMENT OF FEBRILE INFANTS (LESS THAN 90 DAYS) PRESENTING TO TALLAGHT ED.**

**A OSMAN**, I Dafalla, S McKenna, R Kalichuran

<sup>1</sup>paediatric emergency medicine, Tallaght University Hospital, Tallaght, Ireland

**Aim:** The primary aim is to ensure that we are following NICE guidance on managing fever in infants( 0 to 90 days) presenting to Tallaght ED.

**Methods:** A retrospective case review of all patients aged between( 0 to 90) days, presented with fever to Tallaght ED in the period between 31/07/2021 and 01/08/2022. Data were collected from patients' electronic records to assess and review our current practice. Patients were included if they had a high body temperature measured at home or ED.

**Results:** During the study period, 150 infants attended ED with fever as a main cause of presentation. The median age was 57 days (IQR 35-70). 29 patients were  $\leq 30$  days of age, of which 25 had fever  $\geq 38.0$  °C measured at home. 16 out of the 29 patients had full septic work up done, including LP, and started on IV Antibiotics. Out of the rest, eleven patients had partial septic work up done, with no LP, and were admitted for observation. The last 2 patients were discharged home without blood tests.

On the other hand, 121 patients were 31-90 days old, 66 infants met the criteria for bloods workup. However, it was notice that blood sample were taken in 85 patients.

The total number of patients who were meeting the NICE criteria for LP was 58 infants. LP was performed in 34 patients during their presentation, while the procedure was missed in 24 patients who were qualified as per the same guidelines. Furthermore, there were 15 patients who had LP following clinical assessment. However, they were not meeting the NICE guidelines.

**Conclusion:** The audit showed that there is still rooms for improvement in our practice when it comes to following the NICE guideline in both partial and full septic workup.

NICE guideline

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### **ACUTE TUBULAR NECROSIS: A CASE OF ACUTE RENAL INJURY IN A 15-YEAR-OLD PATIENT**

**Z Palmer**, F Ahmed, AM Murphy, L Casserly

<sup>1</sup>Department of Paediatrics, University Hospital Limerick, Limerick, Ireland

**Aims:** Acute tubular necrosis (ATN) is the most common intrinsic cause of acute kidney injury (AKI)<sup>1</sup>. It is diagnosed clinically with a classical sudden and impressive increase in eGFR, serum creatinine and urea levels.

We describe the presentation of a previously healthy neurodevelopmentally normal 15-year-old Irish female, who presented to the paediatric emergency department (PED) with lethargy and fatigue following a 5-day history of persistent vomiting and poor oral intake.

**Methods:** A 15-year-old female presented to PED following 5 days' persistent vomiting and poor oral intake, including fluids. She was previously well, with no significant past medical history. There was no associated fever, diarrhoea, cough or known sick contacts.

On examination, the abdomen was soft non-tender. Cardiovascular and respiratory system examinations were unremarkable. She had dry mucous membranes, capillary refill time of 3 seconds, and no systemic signs of renal disease.

Family history was positive for unspecified renal disease, which resulted in mortality on the paternal side. There was no known family history of renal replacement therapy.

**Results:** Our patient's investigations revealed a diagnosis of AKI secondary to volume depletion. The patient was managed with IV fluid rehydration and oral fluid supplementation, with daily serum creatinine monitoring.

Phlebotomy in PED demonstrated AKI with serum creatinine of 396 umol/L (45-84 umol/L) and urea of 17.1 mmol/L (2.5-7.8 mmol/L). Subsequently, the patient's creatinine levels fell to 354 umol/L, then 91 umol/L over a period of 10 days. There was a similar proportionate decrease in urea levels.

Renal ultrasound showed bilateral increase in renal diameter, with associated loss of corticomedullary differentiation.

**Conclusions:** Following nephrology review, the patient was diagnosed with ATN and discharged home with outpatient follow-up once serum urea and creatinine levels had normalised. We contribute to the literature regarding AKI and ATN in previously healthy adolescent females secondary to volume depletion.

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## **INCIDENTAL FINDING OF RAISED ALKALINE PHOSPHATASE IN A WELL CHILD WITH BENIGN LYMPHADENOPATHY: A CASE PRESENTATION**

**S Parker**, K Rigney, S Ahmed, A Khan, M Jawad

<sup>1</sup>Paediatric Department, Wexford General Hospital, Wexford, Ireland

**Background:** The differential diagnosis of raised alkaline phosphatase (ALP) in childhood is broad, including; bone, hepatobiliary and renal disorders, drugs and transient hyperphosphatasemia (TH).<sup>1</sup> Diagnostic criteria for TH include: (i) *age < 5 years*; (ii) *presence of various unrelated symptoms*; (iii) *no evidence of bone / liver disease*; (iv) *ALP analysis showing elevations in bone +/- liver activity*; (v) *return to normal ALP values in four months*.<sup>2</sup>

**Case Report:** A 14-month-old female with scanty lymphadenopathy was referred to Paediatric OPD for review. Born at term by elective cesarean section, with no neonatal problems. At time of review, she was developmentally appropriate and thriving. Review of systems was negative. She appeared to be a well child. On examination positive findings were a small lymph node in the left posterior cervical chain and few shotty mobile lymph nodes in both inguinal areas with no involvement of other groups. No evidence of hepatosplenomegaly, macrocephaly, frontal bossing, or skeletal abnormalities. Bloods including FBC, LFTs, bone profile, CRP, CK, LDH and urate were ordered for further investigation. Results showed an elevated ALP at 2749 U/L, but were otherwise unremarkable. Due to this unexpected finding, further bloods including magnesium, vitamin D, PTH, U+E, repeat bone profile and LFTs were ordered. Results were normal excluding rickets, liver, and renal disease. Repeat ALP fell significantly (to 1600 U/L) after one week. CXR was normal.

**Discussion:** TH is relatively common and is estimated to occur in 2-5% of healthy children.<sup>3</sup> There may be a history of a mild viral condition in the recent past.<sup>3,4</sup> Affected children are asymptomatic with an unremarkable history, examination, and laboratory investigations aside from elevated ALP. Investigations should include LFTs, U+E, bone profile, vitamin D, PTH and repeat determination of ALP at intervals to determine its normalisation.<sup>5</sup> Recognizing the condition and targeting investigations appropriately will prevent undue investigations and follow up.<sup>3</sup>

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## **DELIVERY OF A NOVEL MEDICAL EDUCATION ELECTIVE DURING THE COVID-19 PANDEMIC**

**M Phipps**<sup>1,2</sup>, E Hicks<sup>2</sup>, S Sloan<sup>2</sup>, E Power<sup>2</sup>, C O'Reilly<sup>2</sup>, M Higgins<sup>3</sup>, EA Dunne<sup>2,3</sup>

<sup>1</sup>Paediatrics , CHI at Temple Street, Dublin, Ireland

<sup>2</sup>School of Medicine, University College Dublin, Dublin, Ireland

<sup>3</sup>Neonatology , The National Maternity Hospital, Dublin, Ireland

**Aims:** The COVID-19 pandemic has led to changes in the delivery of medical education across the globe. In order to reduce footfall in the Neonatal Intensive Care Unit (NICU), traditional clinical elective placements in Neonatology at our centre could not safely be facilitated. Additionally, the majority of medical education was being delivered online. We aimed to deliver a novel alternative elective placement for medical students which would help to address the needs of other medical students learning online.

**Methods:** A three week online elective in medical education for neonatology was offered to students scheduled for clinical elective. Students were introduced to 'design thinking' through online workshops. Students were tasked with identifying and providing a solution for medical education needs in neonatology in the era of covid-19.

**Results:** Students went through the steps of design thinking, identifying and defining the problem, stakeholders and exploring novel solutions. An educational video of a head-to-toe newborn examination was developed. The video was edited with a voiceover explaining the steps of the examination. Learning points were emphasized by pauses and questions to consolidate key concepts. The video was validated by neonatal clinical staff and uploaded to the student learning portal.

**Conclusion:** The COVID-19 pandemic has highlighted the importance of innovation within many spheres, including medical education. This initiative demonstrates "users as collaborators". The students participating in the elective, had been educated online during the initial phase of the

pandemic, putting them in a unique position to develop a curriculum that overcomes the associated challenges. They have learned important transferable skills (innovation, teamwork, communication, empathy, presentation skills, video editing) while the students using the video learning material will benefit from a much needed educational resource that will outlive the pandemic from which it originated.

**PROFILE OF NECROTIZING ENTEROCOLITIS IN NEAR-TERM AND FULL-TERM INFANTS:  
RETROSPECTIVE STUDY IN A TERTIARY CARE NICU**

**M Phipps**<sup>1,2</sup>, A Roy<sup>2</sup>, N De Silva<sup>2</sup>, A Zani<sup>3</sup>, AM Moore<sup>2</sup>

<sup>1</sup>Paediatrics, CHI at Temple Street, Dublin, Ireland

<sup>2</sup>Neonatology, Hospital for Sick Children, Toronto, Ontario, Canada

<sup>3</sup>Surgery, Hospital for Sick Children, Toronto, Ontario, Canada

**Aims:** Necrotizing enterocolitis (NEC) is one of the leading causes of morbidity and mortality in preterm infants, however, it is rare in more mature infants. The aim of this study was to clarify risk-factors for the development of NEC in near-term and term infants, and examine their clinical course and outcomes in order to improve future diagnosis and management.

**Methods:** Infants greater than 35-weeks gestation admitted to Sick Kids Hospital from 2000-2017 with NEC were identified. Patient records of 106 infants meeting Bell criteria Stage-II or greater were reviewed. Data was analysed using descriptive statistics, and Pearson correlations were performed to investigate the role of key variables as predictors for the development of NEC, disease severity, and outcome.

**Results:** The mean ( $\pm$ SD) gestational age was  $37\pm 1.8$  weeks, with mean birthweight of  $2721\pm 724$  grams. We confirmed that term-NEC develops among patients exhibiting one of the known risk-factors (85.9%). This included prenatal complications (57.5%); perinatal distress (51.6%); cardiac

malformations, gastroschisis, and other underlying disorders (75.5%); and introduction of enteral feeding, especially exposure to cow's milk-based formulas (36.8%), with a significant correlation between type of feeds and stage of NEC ( $r=0.74$ ,  $p<0.05$ ). The incidence of NEC increased over the study period, with 37% born in the first nine years, and 63% born in the second nine. The survival rate (91.5%) was similar to previous reports on term infants.

**Conclusion:** Greater awareness of the occurrence of NEC in term neonates and the associated risk-factors, should prompt preventative strategies, such as the promotion of breast feeding in at-risk infants.

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### **PERIODIC FEVER SYNDROMES; HOW THE COVID-19 PANDEMIC UNIQUELY FACILITATED THE DIAGNOSIS OF RARE AUTOINFLAMMATORY DISORDERS IN THE PAEDIATRIC POPULATION R**

**Power, B Linnane**

<sup>1</sup>Departments of Paediatrics, University Hospital Limerick, Limerick, Ireland

This presentation/poster will examine two different cases of children who presented with several month histories of recurrent unprovoked fevers during the COVID-19 pandemic. Each febrile episode documented lasted for several days, was cyclical in nature, with it often being possible to predict the timing of the next febrile event. Symptoms which accompanied these fevers differed between the cases but included; pharyngitis, stomatitis, cervical adenopathy, fatigue and headache. In between episodes their physical symptoms and raised inflammatory markers all returned to normal, a characteristic feature of periodic fever syndromes.

These syndromes, particularly PFAPA (periodic fever, aphthous stomatitis, pharyngitis and cervical adenitis), can be difficult to distinguish clinically. Furthermore, paediatricians face the added challenge of diagnosing these conditions considering more common illnesses, most recently COVID-19, can mimic the presenting symptoms.

Medications which have contributed to reducing the prevalence of febrile episodes for these patients include colchicine and prednisolone. Colchicine was found to be the treatment of choice in a study of children with PFAPA in Ireland. There may also be a role for biologic agents in refractory cases. Despite responding to treatment, these patients still experience recurrent febrile episodes, although these are occurring less frequently. Interestingly, another alternative treatment being considered is the benefit of tonsillectomies to reduce the recurrence/severity of these presentations.

These cases provide multiple learning opportunities and discussion points in the identification of a child with recurrent febrile illness. A phrase often used in medicine is “common things are common”, however these cases highlight the importance of considering rarer conditions when hypothesising a diagnosis.

The COVID-19 pandemic is rarely associated with positive news. However, these cases attest that every cloud has a silver lining. For without the consistent monitoring and recording of temperatures as a result of COVID, these children may have experienced misdiagnoses, a delay to diagnosis, or potentially have never been diagnosed.

1) GP194 Periodic fever- the Irish PFAPA story so far; BMJ 2) PFAPA syndrome; a review on treatment and outcome; Paediatric Rheumatology 3) Tonsillectomy for PFAPA; Cochrane Library

## **ATRAUMATIC LIMPING CHILDREN IN CROSS CITY EMERGENCY DEPARTMENTS AND UNITS**

**AA Raba**<sup>1,4</sup>, S Kasha<sup>2,4</sup>, C Card<sup>2</sup>, K Slater<sup>1</sup>, M Hassan<sup>4</sup>, P Fitzpatrick<sup>2,4</sup>, C Blackburn<sup>3,4</sup>, T Bolger<sup>1,4</sup>

<sup>1</sup>Emergency Care Unit, CHI Tallaght

<sup>2</sup>Emergency Department, CHI Temple Street

<sup>3</sup>Emergency Department, CHI Crumlin

<sup>4</sup>Urgent Care Centre, CHI Connolly

**Aim:** Clinical guidelines have been implemented to assist clinicians in making appropriate decisions about the management of children presenting with acute atraumatic limp. The aim of this study is to assess compliance with guidelines for children with acute atraumatic limp.

**Methods:** We conducted an electronic chart review for all children who presented with acute (<2 weeks) presentation of any of the following (limp; not weight bearing; leg pain or hip pain) to CHI Emergency and Urgent care sites between March and May 2022. Patients with a history of trauma were excluded.

**Results:** A total of 166 patients presented with atraumatic limp during the study period. Median age was 4 years. The majority of patients had transient synovitis (122 (73%)). There were 12 (7%) patients identified with confirmed/suspected bone/joint infection. Other diagnoses were 5 (3%) had mild acetabular dysplasia 4 (2.4%) with myositis; 2 (1.2%) had Henoch Schonlein Purpura; 1(0.6%) had Perthes disease; 1 (0.6% with Slipped upper femoral epiphysis (SUFE); and 19 (11.4%) had unspecified diagnoses. 9/12 patients who had septic arthritis/osteomyelitis presented with fever and 4/12 were not weight bearing. 11/12 had appropriate investigations and were diagnosed with infection at first presentation. 1/12 had investigations done at second presentation (2 days later) and treated as presumed septic arthritis with normal MRI. All patients with septic arthritis/osteomyelitis had abnormal blood results. 64/166 (39%) cases had blood tests done, and 24/166 (37%) cases had abnormal results. 30/66 (47%) patients had blood tests that were not indicated. A total of 97 (58%) had x-rays performed, with 13 (13%) had abnormalities identified. 41(42%) patients had x-rays and were not indicated.

**Conclusion:** Our study suggests that implementation of guidelines for atraumatic limp can be helpful in early diagnosis of patients with suspected septic arthritis/osteomyelitis. Improved adherence to these guidelines can reduce the need for unnecessary laboratory and radiological investigations.

## **NECROTIZING ENTEROCOLITIS AND SPONTANEOUS INTESTINAL PERFORATION: A TEN-YEAR RETROSPECTIVE REVIEW**

**AA Raba**<sup>1</sup>, J Coleman <sup>1</sup>, K Cunningham<sup>1</sup>

<sup>1</sup>Neonatology , The Rotunda Hospital , Dublin , Ireland

**Aim:** To identify and compare patient characteristics and potential risk factors, clinical, and laboratory features of necrotizing enterocolitis (NEC) and spontaneous intestinal perforation (SIP).

**Methods:** We retrospectively reviewed the medical records of all infants with a diagnosis of NEC (stage  $\geq$  IIA according to modified Bell's criteria) or SIP admitted to NICU, at the Rotunda Hospital, Dublin between March 2010 and March 2020.

**Results:** A total of 1224 VLBW infants were admitted to our NICU during the study period, with 130 infants included in the analysis (59 infants with NEC stage  $\geq$  IIA and 26 infants with SIP). In the NEC group, 40 infants had medical NEC (stage 2) and 19 had surgical NEC (stage 3). The overall incidence of NEC was 0.67 per 1000 live births and of SIP was 0.29 per 1000 live births. The mean gestational age of SIP group was significantly lower than that of NEC group ( $25 \pm 1.7$  vs  $27 \pm 2$  weeks,  $P=0.002$ ). Antenatal factors, birth weight, gender, and Apgar scores were not statistically different between the two groups. Infants with SIP were more often to have had UAC in place and were more often to have received surfactant, compared to NEC infants. NEC patients were exposed to a longer duration of antibiotics than infants with SIP (7 (2-11) vs 3 (2-4),  $P=0.001$ ). The median age at diagnosis was significantly younger in SIP than NEC patients (5 (2-6) vs 17 (9-28),  $P=0.0001$ ). Although the incidence of bilious aspirates, abdominal distention and discoloration were not statistically significant difference, bloody stool was more commonly noted in NEC group. Although the mortality was higher in SIP group compared to NEC, it was not statistically significant difference (10 (38%) vs 13 (22%),  $P=0.1$ ).

**Conclusion:** NEC and SIP differed in risk factors and prognosis. The early distinction of SIP from NEC could impact outcomes.

## **A RARE CASE OF LUNG AGENESIS AND HOLOPROSENCEPHALY**

Reilly, S.A.<sup>1</sup>, Collins, A.<sup>1</sup>, Devins, M.<sup>2</sup>, Allen, J.<sup>2</sup>, Byrne, S.<sup>3</sup> Ringholz., F.<sup>4</sup>, McBrien, J.<sup>5</sup>, Quigley, C.<sup>1</sup>, Smyth, M. <sup>1</sup>, Halim, A. M<sup>1</sup>, and Metwally, N. <sup>1</sup>

<sup>1</sup> *Department of General Paediatrics, CHI at Crumlin*

<sup>2</sup> *Department of Palliative Care, CHI at Crumlin*

<sup>3</sup> *Department of Neurology, CHI at Crumlin*

<sup>4</sup> *Department of Respiratory Medicine, CHI at Crumlin*

<sup>5</sup> *Department of General Paediatrics and Neurodisability, CHI at Temple Street*

**Aims:** Holoprosencephaly (HPE) is a brain malformation resulting from incomplete cleavage of the prosencephalon, which occurs between the 18<sup>th</sup> - 28<sup>th</sup> day of gestation (1). HPE classically affects both the forebrain and the face and has several well described conditions associated with it. We present the case of a young male infant with HPE and several well described associations of HPE, with an additional diagnosis of unilateral lung agenesis. A review of the literature was carried out to assess if this association between HPE and unilateral lung agenesis had been reported to date.

**Methods:** A review of the literature was carried out using four databases: PubMed, MEDLINE, Scopus and the Cochrane Database of Systematic Reviews, to identify studies and case reports describing patients with a diagnosis of holoprosencephaly and lung agenesis. The search terms “holoprosencephaly”, “HPE” and “lung agenesis”, “lung hypoplasia”, “pulmonary hypoplasia” and “lung aplasia” and “pulmonary aplasia” were used. Searches were restricted to human subjects published in any language up to September 30<sup>th</sup>, 2022. Reference lists of relevant studies were also searched.

**Results:** We present the case of a 10-month-old male who was admitted to an acute hospital in Ireland. This child presented with lobar holoprosencephaly and several well established conditions associated with this condition. Namely, central diabetes insipidus, hypothyroidism, cleft lip and palate, an underdeveloped vomer, absent right nasal floor, absent lower later nasal cartilage, left microtia, moderate atrial septal defect, recurrent apnoeic episodes and an undescended left testicle. Each of these sequelae of holoprosencephaly have been well described in the literature previously, except for lung agenesis, for which we were unable to locate any documented case reports. Genetic testing revealed a normal 46XY karyotype and microarray showed a small duplication of chromosome 11q, although this mutation is not considered the cause of his clinical presentation. Genetic testing is ongoing to establish if there is an underlying genetic diagnosis for this clinical presentation.

**Conclusion:**



To our knowledge, this is believed to be the first documented case report of unilateral lung agenesis in a patient with lobar holoprosencephaly. This report may add to the literature and enhance clinician knowledge regarding holoprosencephaly.

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## **DEVELOPMENT OF A NEONATAL TRISOMY 21 GUIDELINE: A QUALITY IMPROVEMENT PROJECT**

**K Rigney**<sup>1</sup>, D Sheehan<sup>1</sup>, A Kalim<sup>1</sup>

<sup>1</sup>Department of Paediatrics, Wexford General Hospital , Wexford, Ireland

**Background:** Trisomy 21 (T21) is the most common congenital cause of developmental disability in Ireland with a prevalence of 1 in 546 live births (approximately 110 a year), which is the highest in Europe<sup>1</sup>. Approximately five babies are born with T21 in County Wexford every year. At present there is no local guideline on the recommended investigations and management of newborns with suspected or confirmed T21.

**Aims:** To develop a local guideline for the investigation and management of newborns with suspected or confirmed T21. Our aim is to ensure that the relevant investigations and referrals have been organised promptly prior to discharge home, and to improve the transition home for the newborn and their parents.

**Methods:** A literature review of current international guidelines for T21 was undertaken. This included publications from the Down Syndrome Medical Interest Group (including neonatal and thyroid guidelines), UpToDate and BMJ Best Practice, as well as recent British Society of Haematology guidelines on Transient Leukaemia of Down Syndrome<sup>2-6</sup>. With reference to these resources, we have developed a local guideline for the investigation and management of newborns with suspected or confirmed T21.

**Result:** We have developed an up to date, evidence-based guideline for management of newborns with suspected or confirmed T21. We are currently undertaking a chart review of all babies born with T21 in our unit in the last year in order to evaluate their management prior to discharge. We then plan to present the guideline at a departmental meeting before implementing it into our practice. A repeat chart review will be performed in one year, to assess for any improvement in our management of these patients.

**Conclusion:** Our aim is that the implementation of this guideline will bring our management of patients with suspected or confirmed T21 in line with international standards.

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## VITAMIN K PRESCRIPTION & ADMINISTRATION IN A REGIONAL MATERNITY UNIT

K Rigney<sup>1</sup>, L Loughlin<sup>1</sup>, S Parker<sup>1</sup>, M Azam<sup>1</sup>

<sup>1</sup>Department of Paediatrics, Wexford General Hospital, Wexford, Ireland

**Background:** Vitamin K deficiency bleeding (VKDB) in the newborn is caused by immature liver function and low vitamin K in the placenta and breast milk<sup>1</sup>. A single intramuscular (IM) injection of vitamin K at birth effectively prevents VKDB<sup>2</sup>. According to international guidelines, vitamin K should be administered to all newborns within 6 hours of birth as a single IM dose (1mg in those weighing >1500g or 0.3-0.5 mg/kg in those <1500g or in preterm infants)<sup>2,3</sup>.

**Aim:** To assess the compliance of the time interval between birth and administration of vitamin K to newborns in Wexford General Hospital (WGH) and compare against international standards.

**Methods:** This is a retrospective cohort study. Data was collected by chart review over a one-month period. Details were obtained on demographics, vitamin K prescription and vitamin K administration. Data analysis was carried out in Microsoft Excel.

**Result:** A total of 25 charts were reviewed. Vitamin K was prescribed in 92% of cases (n=23). Clear documentation of vitamin K administration (date & signature) was seen in 76% of cases (n=19), however time of administration was only documented in 64% of cases (n=16). Among these 16 patients, the time from birth to administration ranged from 2-28 minutes with a mean time of 10.7 minutes.

**Conclusion:** Clear documentation of vitamin K administration including time was seen in 64% of patients (n=16) with a 100% compliance to the standard of administration within 6 hours within this group. This audit has identified shortfalls in the documentation of (a) prescription and (b) administration of vitamin K. Current practice in WGH is that vitamin K is prescribed in the mother's Kardex. Our recommendation is to review this along with the neonatal proforma currently in use to address inconsistent documentation, and to standardise practice in WGH to administer Vitamin-K in the delivery room within 6 hours of birth.

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## **AN EARLY COINCIDENCE: ANTIBIOTIC PRESCRIBING FOR ACUTE OTITS MEDIA IN A TERTIARY PAEDIATRIC EMERGENCY DEPARTMENT**

C Ring, D Speldewinde

<sup>1</sup>Emergency Department, Perth Children's Hospital, Nedlands, Australia

**Aims:** Acute otitis media (AOM) represents a significant number of presentations to the Paediatric Emergency Department (ED). The aim of this audit was to ascertain if local guidelines on antibiotic use in AOM were being followed.

Local clinical guidelines recommend utilising a 'watch and wait' approach for 48 hours in patients over six months with unilateral disease and no systemic features. Systemic features include fever, lethargy and vomiting. Patients from high-risk groups (immunocompromised, Indigenous background, cochlear implant, only hearing ear affected) are exceptions to this and can commence antibiotics promptly.

**Methods:** A retrospective review of charts was performed for the period January to September 2019. Included patients had an ED visit that resulted in a primary diagnosis of AOM. All patients were aged over six months. 40 encounters were reviewed, with 11 subsequently excluded as the patient had already commenced antibiotic therapy prior to ED presentation.

**Results:** 11 patients presented with symptoms for less than 48 hours and had no systemic features present. 8/11 (72.7%) were advised to monitor symptoms and were not commenced on antibiotics, while 3/11 (27.3%) were prescribed antibiotics.

18 patients had symptoms of AOM for > 48 hours and/or had systemic features. 13/18 (72.2%) were commenced on antibiotics while 5/18 (27.8%) were not.

One patient from an at-risk group attended with AOM during this period, but had already commenced antibiotics before presentation.

**Conclusions:** In keeping with local guidelines, a period of observation was usually recommended in patients with AOM with symptoms for less than 48 hours and no systemic features. This compares favourably to international rates noted in other studies. Patients who had symptoms for over 48 hours or who had systemic illness were more likely to be prescribed antibiotics in the ED.

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**BREASTFEEDING- EXPRESS YOURSELF! ATTITUDES TOWARDS BREASTFEEDING OF HEALTHCARE PROFESSIONALS WORKING IN PAEDIATRICS**

**M Ryan**<sup>1</sup>, J Lucey<sup>1</sup>, F Neenan<sup>2</sup>, C Bulfin<sup>1</sup>, O Somers<sup>2</sup>

<sup>1</sup>General Paediatrics, University Hospital Waterford, Waterford, Ireland

<sup>2</sup>General Paediatrics, Portiuncula University Hospital, Ballinasloe, Ireland

**Aims:** The aim of this study was to gauge attitudes and knowledge surrounding breastfeeding amongst healthcare workers who work with neonates in Irish hospitals.

**Methods:** An online questionnaire was distributed to healthcare working in paediatric and obstetrics departments of two university hospitals in Ireland. Respondents were asked about their profession, grade, and area of work. They were presented with a number of statements regarding breastfeeding as it related to their professional practice, and asked to express a degree of agreement from 1-5. They were then asked two questions relating to WHO guidance on breastfeeding. They were then offered a free text space to express any views or opinions they had in relation to breastfeeding in Ireland.

**Results:** There were 55 responses. Respondents encompassed doctors, midwives, and nurses (from general paediatrics and neonatal units,) in addition to medical students and nursing/midwifery students. 87% agreed that encouraging and supporting breastfeeding was an important part of their role. 93% agreed that increased rates of breastfeeding would improve child health in Ireland. When asked about the ages specified by WHO for exclusive breastfeeding and breastfeeding as part of weaning diet, 71% and 68% of respondents respectively answered correctly. A number of themes emerged in responses to the free text section. These included a shortfall in community support following discharge home, education of both the general public and healthcare workers, and an emphasis on supplementing with formula feeds.

**Conclusions:** Healthcare professionals recognise that breastfeeding is beneficial for babies, and recognise the importance of their role in encouraging and supporting breastfeeding. Many staff members however expressed lack of confidence in pragmatic elements of breastfeeding with regard to assessing individual cases. Improved education of hospital staff may help build their confidence to foster a positive attitude to breastfeeding and better provide support for breastfeeding mothers.

## DOES QUANTITY REFLECT QUALITY IN CLINICAL RESEARCH

M Salama<sup>1</sup>, L Jansen <sup>1</sup>, S Ni Neachtain <sup>2</sup>, MB O'Neill<sup>1</sup>

<sup>1</sup>Paediatrics, Mayo University Hospital, Castlebar, Ireland

<sup>2</sup>Paediatrics, University of Galway, Ireland

**Aims:** Citations are commonly used as indicators of research performance and are assumed to reflect the impact of the research or its quality (1). This study aimed to assess how citations relate to research quality by exploring the relationship between the quality of external validation of clinical prediction rules and the citation frequency of original derivation studies.

**Methods:** Clinical Prediction rules were identified based on a group's systematic review, which assessed external validation studies of CPRs between 2010-2019. 81 out of the 86 rules were selected. Rules with unpublished derivation studies were excluded (N=5), citation frequency in PubMed, Embase and CINAHL were recorded, and validation levels were assessed according to the hierarchy of evidence published by the Evidence-Based Medicine Working Group (EBMWG) (2). The mean differences between groups were measured using a double-sided independent t-test on SPSS.

**Results:** None of the CPRs had impact analysis. PECARN and PEWS were the most validated (n=7), and PECARN was the most cited CPR (n=1376), followed by Asthma predictive index (n=1179). Mean citations of original CPR studies validated as level 2 was 180, level 3 was 94.8, Level 4 was 56.2, Mean difference in citation between level 2 and level 4 was 123.7 (P value 0.002), Mean difference in citation between level 3 and level 4 was 38. (P value 0.014).

**Conclusion:** Researchers tend to cite CPR studies with higher validation levels, yet CPRs that were not validated or validated narrowly also were commonly cited. Clinicians are adopting CPRs in practice without undergoing impact analysis to confirm their utility in different populations. There is a need for improved external validation with impact analysis to change practice. Finally, research quality is a multidimensional concept that cannot only be captured by one indicator.

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Guyatt GH, Wyer PC, David Naylor C, Stiell IG, Richardson WS. Users' Guides to the Medical Literature XXII: How to Use Articles About Clinical Decision Rules [Internet]. Available from: <http://jama.jamanetwork.com/> 3-Beattie P, Nelson R. Clinical prediction rules: What are they and what do they tell us? *Australian Journal of Physiotherapy*. 2006;52(3):157–63. 4. Maguire JL, Kulik DM, Laupacis A, Kuppermann N, Uleryk EM, Parkin PC. Clinical prediction rules for children: A systematic review. Vol. 128, *Pediatrics*. 2011. 5. Wasson JH, Sox HC, Neff RK, Goldman L. Clinical prediction rules: applications and methodological standards. *N Engl J Med*. 1985;313(13):793–799 6. Kennedy, G., & Gallego, B. (2019). Clinical prediction rules: A systematic review of healthcare provider opinions and preferences. In *International Journal of Medical Informatics* (Vol. 123, pp. 1–10). Elsevier Ireland Ltd. <https://doi.org/10.1016/j.ijmedinf.2018.12.003>

**Poster No: 95**  
**General Paediatrics**

### **TRAINEE KNOWLEDGE AND ATTITUDE TOWARDS THE MANAGEMENT OF PAEDIATRIC ACUTE ATRAUMATIC LIMP: A COMPARATIVE SURVEY**

**M Shaikh Yousef<sup>1,2</sup>, E Moylett<sup>1</sup>, F Shannon<sup>2</sup>**

<sup>1</sup>Department of Paediatrics, School of Medicine, National University of Ireland, Galway, Ireland

<sup>2</sup>Department of Orthopaedics, University Hospital Galway, Galway, Ireland

**Aims:** There are various aetiologies for acute, non-trauma related, paediatric limp presenting to the Emergency Department, some requiring urgent intervention.<sup>1</sup> The trainee's ability to make an accurate initial assessment is dependent on knowledge and competency.<sup>2</sup> Given the varied levels of expertise among trainees across specialities within the Saolta hospital group, we looked to assess their knowledge with the hope of highlighting potential areas for improvement with the focus on patient care and outcome.

**Methods:** A semi-standardized, 2-month, online survey was used to assess trainee knowledge relating to acute non- trauma related, paediatric limp across orthopaedic, paediatric and adult Emergency Medicine trainees working in the Saolta Group. The online survey was created through [www.surveymonkey.com](http://www.surveymonkey.com); 19 questions: 4 demographic and training questions, 3 attitude questions, and 12 knowledge questions. Trainees were invited to participate via NCHD WhatsApp groups and departmental emails; all responses collected and analysed anonymously. SPSS, version 28 was used to analyse data. Ethical approval was obtained from the Galway University Hospitals Clinical Research Ethics Committee.

**Results:** Of the 357 invited NCHDs, 101 (28.3%) completed the survey, 45 of 153 (29.4%) interns, 23 of 74 (31.1%) paediatric, 19 of 54 (35.2%) orthopaedic, and 14 of 76 (18.4%) adult EM trainees. Paediatric trainees achieved the highest knowledge score, followed by orthopaedic trainees, EM trainees, and interns. Only thirty-eight (37.6%) respondents reported being 'comfortable' with their initial management of a child with acute atraumatic limp; 31 (30.7%) reported a 'neutral' attitude. Majority supported the need for additional training.

**Conclusion:** This report highlights apparent knowledge deficits across different speciality trainees and low confidence levels in managing cases of acute atraumatic limp. To improve the outcome of the diagnostic process, we recommend providing a diagnostic algorithm within the Emergency Department and promoting the application of conceptual knowledge through simulated scenarios.

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## **NEUROIMAGING IN CHILDREN PRESENTING WITH HEADACHE TO THE EMERGENCY DEPARTMENT.**

**Dr Landa Sharif**<sup>1</sup>, Dr Albert Soye<sup>2</sup>, Dr Evodia Ludusan<sup>1</sup>

<sup>1</sup>Paediatric emergency , Our Lady of Lourdes Hospital , Drogheda , Ireland

<sup>2</sup>General Paediatric department , Our Lady of Lourdes Hospital, Drogheda , Ireland

<sup>3</sup>Radiology Department , Our Lady of Lourdes Hospital, Drogheda , Ireland

**Introduction:** Headaches are among the top five health problems in childhood (27.4 % of paediatrics emergency department presentations). However, a small percentage of them have positive finding on the brain image. CT brain and MRI are costly, and the risk of radiation is high for the children.

**Aim:** To Compare our practice in investigating children presenting with headache with our local/NICE guidelines.

**Method:** Retrospective data collection from the patients' medical record and radiological electronic data from July 2020- December 2020).

**Result:**

Total Number of neuroimaging CT /MRI in period of 6 months is 177.

Total number of neuroimaging CT/MRI for children with headache 73 (41%).

Total number of neuroimaging for trauma is 85 (48%).

Others (orbital cellulitis/ VP shunt) 19 (11%).

Number of normal neuroimage 66/73 (90.5%)

Number of abnormal neuroimage 7/73. (9.5%)

Number of CT brain showed brain tumour (astrocytoma) =1

Number of CT brain showed hydrocephalus =1

Number of CT brain showed benign brain cyst =2

Number CT brain showed Sinusitis =3

**Recommendations:**

1. Neuroimaging (CT/MRI) is indicated for patients with red flags as per local guidelines/NICE guidelines, patients with progressive symptoms, history of trauma or other complex medical background.

2. Improve clinical assessment by taking detailed medical history and clinical examination, including neurological examination.

3. Information leaflet with lifestyle change and non-pharmacological management to be provided to all patients. Headache diary to be advised to all patients.

4. Patients with no red flag to be discharged on regular analgesia and appropriate follow up (ED clinic or Paediatrics OPD).

References : -NICE guidelines ,diagnosis and management of headache in young people .  
,<https://www.nice.org.uk> -Headaches in childhood,HSE,<https://www.hse.ie>

**HOW LOW CAN YOU GO? A REVIEW OF ADHERENCE TO HYPOGLYCAEMIA PROTOCOL IN A TERTIARY EMERGENCY DEPARTMENT**

**NB Shaughnessy**<sup>1</sup>, M Elsammak<sup>2</sup>, C Mattimoe<sup>3</sup>

<sup>1</sup>Nephrology, CHI at Temple St , Dublin, Ireland

<sup>2</sup>Chemical Pathology, CHI at Temple St, Dublin, Ireland

<sup>3</sup>Emergency Department, CHI at Temple St , Dublin, Ireland

**Introduction:** The most common causes of hypoglycaemia after the neonatal period are diabetes mellitus and idiopathic ketotic hypoglycaemia (IKH), but a number of other serious endocrine and inborn errors of metabolism should be considered and excluded. IKH is the most common cause of hypoglycaemia between 18 months and 5 years but this diagnosis can only be made after exclusion of other endocrine and metabolic disorders. If a point of care glucose is  $\leq 2.6$ mmol/l, a hypoglycaemia workup should be performed and treatment initiated. It is noted that often a full hypoglycaemia workup is not completed in the ED. Our aim is to ascertain the proportion of patients who had an incomplete screen, and which tests were most frequently not done, as well as reviewing the number who required referral to endocrine/ metabolic services.

**Methods:** We reviewed investigations performed on all patients who had a blood glucose level  $\leq 3$ mmol/L, aged between 1 month and 16 years who attended the ED in CHI at Temple St between 1/7/21 and 1/7/22.

**Results:** 67 patients had hypoglycaemia screens performed between 1/7/21 and 1/7/22 with a lab glucose of  $\leq 3$ mmol/L. 25/67 had a complete hypoglycaemia screen performed. Most common tests not performed were urinary organic acids and acylcarnitine profile. 3 patients were referred to endocrine and 5 to metabolic clinics for further investigation.

**Conclusion:** Deficiencies in adherence identified. We aim to improve services by implementing steps to enhance existing procedures including education of staff that a urine bag is sufficient to

analyse urine organic acids and highlight the importance that a paired urine sample is essential to interpret result of acylcarnitine profile and serum amino acids.

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Recommendations from the Pediatric Endocrine Society for Evaluation and Management of Persistent Hypoglycaemia in Neonates, Infants and Children. *The Journal of Pediatrics*. 2015;167(2):238-45.

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Sub-Specialty

### **A CURIOUS CASE OF RASBURICASE-INDUCED METHAEMOGLOBINAEMIA IN UNDIAGNOSED G6PD DEFICIENCY**

**NB Shaughnessy**<sup>1</sup>, E Jenkins<sup>1</sup>, S Cahill<sup>1</sup>, S McAnallen<sup>1</sup>, M Riordan<sup>1</sup>

<sup>1</sup>Department of Nephrology, CHI at Temple Street, Dublin

**Introduction:** Rasburicase is a recombinant urate-oxidase enzyme used in the treatment and prevention of acute hyperuricaemia, to prevent acute renal failure, particularly in those with haematological malignancy with a high tumour burden and at risk of rapid tumour lysis.

Methaemoglobinaemia is a serious rare adverse effect of rasburicase administration, more common in patients with G6PD deficiency. Prompt diagnosis and management can make the difference between successful recovery and significant morbidity.

**Methods:** We report the case of a 12-year-old girl with African ethnicity who developed methaemoglobinaemia following one dose of rasburicase treatment, who was later confirmed to be G6PD deficient. She presented to the ED with severe dehydration following a week-long viral illness on a background of longstanding polyuria and adipsia. Medical background is significant for hypomelanosis of Ito with severe intellectual disability. Initial investigations revealed an acute kidney injury requiring dialysis. Four hours after administration of rasburicase saturations dropped

to 85-91%, which did not respond to high flow oxygen therapy. CXR and Echo were normal. Arterial blood gas revealed high methaemoglobin levels, highest 19.9%.

**Results:** The enzymatic digestion of uric acid to allantoin by rasburicase produces hydrogen peroxide.

Hydrogen peroxide is an oxidising agent, capable of oxidising the iron ion in haem from the normal 2+(ferrous) to the 3+(ferric) state, forming methaemoglobin, which is unable to bind oxygen and haemoglobin which is functionally useless. Treatment includes supportive therapy with hydration and oxygen. Methylene blue is the primary treatment for reducing levels but is associated with haemolytic anaemia in patients with G6PD deficiency.

**Conclusion:** Methaemoglobinaemia should be included in the differential for unexplained hypoxia. Whenever possible, patients from at risk groups should be tested for G6PD deficiency prior to rasburicase administration. This is often not possible given turnaround times for test results. Patients with G6PD deficiency are especially vulnerable to rasburicase-induced methaemoglobinaemia, which may be the first clue to the underlying diagnosis in those who have not been tested.

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2. Wilson J. Rasburicase-induced methaemoglobinaemia and catastrophic oxidative haemolysis in undiagnosed G6PD deficiency. *Br J Haematol.* 2022.

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## **EVALUATION OF URINE SENSITIVITIES OF PAEDIATRIC PATIENTS TREATED FOR UTI WITH COMBINED AUDIT INTO COMPLICANE WITH REPORTED SENSITIVITIES.**

**PS Short, F O'Brien,**

<sup>1</sup>Paediatrics, St. Luke's Kilkenny, Kilkenny, Ireland

**Aim:** To identify the most common antimicrobial sensitivities and resistances in urine samples sent for urinary tract infection treatment, and to audit compliance with these sensitivities. These results would then be used to create local guidelines for UTI treatment based on the local sensitivities.

**Method:** Urine samples sent from St. Luke's Kilkenny in a 6 month period for paediatric patients were reviewed. In cases where a specific organism was grown, the sensitivities and resistances were assessed. These charts were then requested to audit the treatment plans.

**Results:** A total of 266 urine samples were reviewed. Of these 55 which grew organisms, this was rationalised to 48 cases.

Summary of most relevant information:

- 65% of urine samples sent were clean
- 54% of organisms are resistant to Amoxicillin
- 12.5% are resistant to Augmentin but if any concerns for more serious infection then 42.5% are resistant
- 15.5% are resistant to Cephalexin (11% if consider samples taken from same patient same day)
- 5% are resistant to 3<sup>rd</sup> Gen. Cephalosporin
- 0% resistance to Gentamicin
- In 9.5% of cases Amoxicillin was used first line
- In 28% of cases Augmentin (alone) was used first line
- In 33% of cases Augmentin and Gentamicin was used first line
- In 19% of cases Cephalexin was used first line
- In 4.7% of case Ceftriaxone was used first line
- 35% of cases required change of antibiotics based on the sensitivities but in only 14% of cases was this change documented as having been carried out.

**Conclusion:** Local guidelines were implemented to make Cephalexin the first line oral antibiotic for treatment of UTI's. Careful consideration would be taken over the use of Gentamicin with potential to use 3<sup>rd</sup> line cephalosporin as a single agent initially. Going forward urine sensitivities

will be more closely monitored, and changes made accordingly and documented once sensitivities have returned.

CHI Paediatric Formulary UTI Guidelines (<https://www.olchc.ie/healthcare-professionals/clinical-guidelines/antimicrobial-guidelines-2021.pdf>)



## DEVELOPING A MENTORSHIP PROGRAMME FOR CONSULTANTS ACROSS A HOSPITAL GROUP

DM Slattery<sup>1,2</sup>, C Neves Correia<sup>1</sup>, A Goldman<sup>3</sup>

<sup>1</sup>CHI Professionalism Programme, CHI, Dublin

<sup>2</sup>Respiratory and Cystic Fibrosis Unit, CHI Temple St Dublin

<sup>3</sup>Office of the Chief Medical Officer, CHI, Dublin

**Aim:** To establish a mentorship programme for newly appointed consultants across a hospital group.

**Methods:** Establishment of a mentorship programme where senior consultants mentor newly appointed consultants was identified as a priority by the CHI Professionalism Programme. Involvement in mentorship programmes has been linked with increased job satisfaction<sup>1</sup> and wellbeing<sup>2</sup>. A literature review on this topic was performed. The CHI Professionalism Programme partnered with the National Clinical Leadership Centre for Nursing and Midwifery (NCLC), which has a track record in training mentors, and a Chartered Organisational Psychologist and Executive Coach with experience in private industry and RCSI Institute of Leadership.

**Results:** Content was developed for the mentor training course. A half-day interactive training course was developed for mentors incorporating evidence-based methodology, pre-course reading material, case scenarios and role play. The training course will be evaluated with an anonymous feedback questionnaire at the end of the training. Mentors are peer nominated and identified across different specialities. A two-year commitment is requested from mentors. Mentees will meet with mentors four times per year (more if required by either party). Mentees will meet with the Psychologist and Lead for the Professionalism programme at 3 months and 9 months to identify what is going well, what is not going well, and any suggested changes to the programme.

A mentorship form has been developed to help guide the conversation between mentors and mentees, based on following pillars: career development, clinical development, training and education, research activities, and personal progress. At 12 and 24 months, mentors and mentees will complete a written evaluation of the programme with suggested amendments.

**Conclusion:** Development of a formal mentorship programme for new consultants with trained mentors and evaluation of the programme is a first for this hospital group and to the authors knowledge, is a first in public hospitals nationally.

References: 1.Ghosh R, Reio TG. Career benefits associated with mentoring for mentors: a meta-analysis. J Voc Behav 2013;83:106– 16 2.Steven A, Oxley J, Fleming WG. Mentoring for NHS doctors: perceived benefits across the personal-professional interface. J R Soc Med. 2008 Nov;101(11):552-7. doi: 10.1258/jrsm.2008.080153. PMID: 19029356; PMCID: PMC2586857.

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### **HAEMOPTYSIS IN A HEALTHY TEENAGER: WHAT'S ALL THE PUFF ABOUT? CASE REPORT AND LITERATURE REVIEW**

**A Wallace**, C Ryan, DM Slattery

<sup>1</sup>Department of Respiratory Medicine, CHI Temple ST., Dublin

**Aim:** Discuss the diagnostic work up of a teenager with intermittent haemoptysis over three years where vaping was the cause

**Methods:** Retrospective chart review was performed with investigation results discussed with experts in respiratory medicine, radiology and cardiology. A literature review was conducted.

**Results:** A 14 year old male presented to the ED following episodes of unprovoked fresh haemoptysis of increasing volume and frequency over three days on a background of fresh haemoptysis every ~ three months over three years. He vaped nicotine-based products daily for three years. Examination normal.

Investigations: normal FBC, coagulation profile, renal, liver and bone profile, chest radiograph, vasculitic and connective tissue disease screen.Covid-19 antigen test was positive. A CT thorax revealed pulmonary nodules with surrounding ground glass component and possible small arteries entering some of the foci. Differential diagnosis was Covid-19 infection or arteriovenous malformation(AVM).

Repeat CT Thorax eight weeks later demonstrated resolution of changes and no evidence of AVMs. Both the bubble study and review by ENT team were normal.

Flexible bronchoscopy with broncho-alveolar lavage (BAL) revealed erythematous mucosa, which was friable and bleed on contact. BAL was frothy and clear with only scanty haemosiderin laden macrophages, pathognomonic of alveolar haemorrhage. Literature review identified vaping causing friable mucosa at bronchoscopy.

Healthy Ireland Report (2019) identified that 18.1% of teenagers surveyed said they were currently using ecigarettes, increased from 10.1% in 2015. In USA 37% of high school seniors reported vaping in 2018, increased from 28% in 2018. E-cigarettes/vaping may cause sudden and severe lung injury ranging from sudden and severe lung injury to death.

**Conclusion:** Vaping may cause fresh haemoptysis in addition to other lung injury. Due to the high prevalence of vaping, Paediatricians should ask teenagers if they vape, what they vape, increase awareness of its serious side effects and encourage its discontinuation

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Medical/Education Management

#### **DEVELOPING A PROPOSED PATHWAY TO ADDRESS UNPROFESSIONAL BEHAVIOUR ACROSS A HOSPITAL GROUP**

**DS Slattery**<sup>1,2</sup>, CNC Neves Correia<sup>2</sup>, TW Wall<sup>3</sup>, AG Goldman<sup>4</sup>

<sup>1</sup>Respiratory Department, Children's Health Ireland at Temple St, Dublin, Ireland

<sup>2</sup>CHI Professionalism Programme, Children's Health Ireland, Dublin, Ireland

<sup>3</sup>Office of the Director of Nursing, Children's Health Ireland, Dublin, Ireland

<sup>4</sup>Office of the Chief Medical Officer, Children's Health Ireland, Dublin, Ireland

**Aim:** To develop a proposed pathway to address unprofessional behaviour (which is a patient safety issue), in staff across a hospital group.

**Methods:** The Chief Medical Officer prioritised development of a pathway to address unprofessional behaviour in staff. A literature review was performed by the CHI Professionalism Team to identify how international centres of excellence address this issue. Meetings were held

virtually with senior leaders from six international centres: Royal Children's Hospital Melbourne (Australia), Great Ormond Street Hospital (United Kingdom), Child and Adolescent Health Service in Western Australia & Perth Children's Hospital (Australia), KK Singapore Women's and Children's Hospital (Singapore), and Boston Children's Hospital (USA). Discussions focused on policies, pathways and implementation of practices to address unprofessional behaviour. Honest discussions were held on what worked well, what worked less well and how things would be done differently if starting again.

**Results:** Some common approaches are implemented across international centres of excellence. A framework was developed by the CHI Professionalism team to incorporate key components of pathways including: training for all staff in "speaking up for safety and values<sup>1</sup>" and "promoting professional accountability<sup>2</sup>", provision of dedicated additional training for staff leaders, a confidential electronic reporting system to report unprofessional behaviour, appointment of speaking up guardians and/or "go-to" persons, an agreed escalation process for addressing unprofessional behaviour (initially informal, but if behaviour persists or is egregious, it becomes formal<sup>3</sup>). A team incorporating key stakeholders across the organisation was established and discussed pathway options for the hospital group. Over 11 months, a draft pathway, modelled on international centres of excellence has been developed by key stakeholders. This is awaiting a final sign off.

**Conclusion:** An evidence based draft pathway to address unprofessional behaviour in staff across a hospital group has been developed by key stakeholders (awaiting signoff) based on international best practice.

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behaviors. Acad Med. 2007 Nov;82(11):1040-8. doi: 10.1097/ACM.0b013e31815761ee. PMID: 17971689.

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**AN AUDIT OF CURRENT PRACTICE IN THE MANAGEMENT OF HOSPITALIZED PATIENTS WITH EATING DISORDERS: A RETROSPECTIVE AUDIT USING THE MARSIPAN GUIDELINES**

**BN Toor<sup>1</sup>, M Sani<sup>1</sup>, L Mahmood<sup>1</sup>**

<sup>1</sup>Pediatric Department, St Luke's General Hospital - HSE, Kilkenny, Ireland

**Aim:** To assess whether the management of children with eating disorders in our pediatric ward (St. Lukes Hospital) is in line with the recommendations of MARSIPAN guidelines.

**Method:** A retrospective analysis of patients hospitalized with an eating disorder at the paediatric ward, St. Lukes Hospital. Over a 2-year period, Nine patients were eligible for enrollment. Clinician and nursing assessments in the selected patients' charts were audited using the MARSIPAN guidelines.

**Results:** 9 patients met inclusion criteria. The results of this study showed that while some parameters were consistently done, others had poor implementation. 100% of patients had blood pressure (sitting & standing) measurements, baseline lab investigations (such as FBC, electrolytes, urea, and creatinine) done, whereas inpatient assessment by a psychiatrist was as low as 22%. It is important to note that all patients that were not reviewed as inpatient had outpatient appointments booked. Other important risk stratifying tools such as supervised mealtime (88.9%), dietitian inpatient review (77.8%) and completed ECG (88.9%) were variable. However, a complete HEADSS assessment was poorly performed. Only 22% of the patients had a completed HEADSS assessment documented in the charts. Three quarters of the patients were monitored for re feeding syndrome with serum electrolytes checked at least twice during admission.

**Conclusion:** Whilst patients with eating disorders are challenging in a conventional paediatric ward, more needs to be done in taking a complete psychosocial review. This audit suggests that the MARSIPAN guideline was utilized consistently to stratify medical risk in patients with eating disorders admitted at St. Lukes. However, the psychosocial assessment was suboptimal. A designated regional Psychiatrist was consulted for all cases but unfortunately not available for most inpatient review.

## MANAGEMENT OF DYSTONIA IN PAEDIATRIC CEREBRAL PALSY

RT Treanor<sup>1</sup>, KNRD Rodrigues Dos<sup>1</sup>, CC Christle<sup>1</sup>, KB Balogun<sup>1</sup>, AAZ Al Zadjali<sup>1</sup>, CMCV McVeigh<sup>1</sup>, KMCD

McDonnell<sup>1</sup>, ER Roche<sup>1-3</sup>, JM Meehan<sup>1-3</sup>, EM Molloy<sup>1-3</sup>, AB Branagan<sup>1-3</sup>, GS Semova<sup>1-3</sup>, EI Isweisi<sup>1-3</sup>

<sup>1</sup>Discipline of Paediatrics, Trinity College Dublin, Dublin, Ireland

<sup>2</sup>Trinity Research in Childhood Centre, Trinity College Dublin, Dublin, Ireland

<sup>3</sup>Children's Health Ireland, Crumlin children's hospital, Dublin, Ireland

**Aims:** In children with dystonic cerebral palsy (population), is there an optimal intervention (Intervention and Comparator) to decrease severity of dystonia (Outcome)?

**Methods:** A search of Medline, the Cochrane library and EMBASE was carried out using the search terms 'cerebral palsy' AND 'management' AND 'dystonia' between the years January 2000 to September 2021.

Ten papers were included in the final data analysis after full text screening.

### Results:

Pharmaceutical: The final analysis suggests that within the pharmacological interventions, only clonidine and gabapentin have shown evidence to suggest some improvement in dystonia scores. However, this suggestion is based on one small, nonrandomised study each and thus the evidence is extremely limited. The evidence for oral baclofen and benzodiazepines could not be found despite oral baclofen being considered 1st line treatment for generalised dystonia by the AACPDm.

Neurosurgical: The evidence for ITB and DBS is more promising than the pharmacological interventions. However, in the only RCT, ITB did not associate with improvement in BADS scores. The one case report for each of orthopaedic surgery, pallidotomy and rhizotomy also prevents a reliable conclusion from being formed.

**Conclusions:** No pharmacological intervention has reliably proven to be effective in reducing dystonia in children.

ITB and DBS have demonstrated some promise in managing dystonia in cerebral palsy.

The lack of large, randomized controlled trials and a universal scoring system for dystonia are barriers to evidence-based practice and international guidelines based on a combination of expert opinion and the current evidence base are the best option for practice.

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### **BRONCHIOLITIS: EVIDENCE-BASED MANAGEMENT IN HIGH-RISK INFANTS WITH SEVERE DISEASE**

R Walsh<sup>1</sup>, L Costello<sup>1</sup>, A DiCosimo<sup>1</sup>, AM Doyle<sup>1</sup>, L Kehoe<sup>1</sup>, C Mulhall<sup>1</sup>, S O'Hara<sup>1</sup>, J Meehan<sup>1,2,3</sup>, E Isweisi<sup>1,2,3</sup>, G

Semova<sup>1,2,3</sup>, A Branagan<sup>1,3,4</sup>, E Roche<sup>1,2,3</sup>, E Molloy<sup>1,2,3,4,5</sup>

<sup>1</sup>Discipline of Paediatrics, School of Medicine, Trinity College Dublin, Dublin 2, Ireland

<sup>2</sup>Children's Health Ireland, Tallaght, Dublin, Ireland

<sup>3</sup>Trinity Research in Childhood Centre, Trinity College Dublin, Dublin 2, Ireland

<sup>4</sup>Neonatology, Coombe Women & Infants University Hospital, Dublin 8, Ireland

<sup>5</sup>Children's Health Ireland, Crumlin, Dublin, Ireland

**Aims:** To systematically review the management of infants with severe bronchiolitis in a paediatric intensive care unit (PICU) setting.



**Methods:** This systematic review utilised the PRIMSA guidelines to examine literature on the PICU management of bronchiolitis in infants <24 months of age. PubMed, MedLine, and EMBASE were the databases searched and included higher levels of evidence I, II and III.

**Results:** The National Institute for Health and Care Excellence (NICE) guidelines for the management of bronchiolitis included randomised control trials (RCTs), in which infants with cardiac and respiratory disorders which conferred a higher risk of severe disease were excluded in 88% and 78%, respectively. For the systematic review, there were 455 papers screened and 26 met the inclusion criteria. Furthermore, 19 of these studies examined respiratory interventions such as positive airway pressure and oxygen delivery. The remaining 7 studies examined: erythropoietin, caffeine, dexamethasone, protein supplementation, ribavirin, respiratory syncytial virus immune globulin, or diuretic therapy. Of the 26 studies, 20 excluded infants with high-risk conditions. Therapies showing favourable outcomes included prophylactic dexamethasone pre-extubation, heliox therapy, protein supplementation, and early diuretic use. Overall, the risk of bias was low-moderate.

**Conclusions:** Current clinical trials for bronchiolitis management frequently exclude high-risk children. RCTs are needed to determine efficacy and safety of treatments for the management of bronchiolitis in high-risk infants in a PICU setting.

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## IDENTIFIED HEALTH NEEDS OF ROMA CHILDREN THROUGH OUTREACH PAEDIATRIC SERVICES

A Walsh<sup>1,2</sup>, B Joyce<sup>1</sup>, E Molloy<sup>2</sup>, C Ni Cheallaigh<sup>2,3</sup>, C Hensey<sup>1,2</sup>

<sup>1</sup>General Paediatrics Department, Children's Health Ireland at Temple Street, Dublin 1, Ireland

<sup>2</sup>Faculty of Paediatrics and Child Health, School of Medicine, Trinity College Dublin, Ireland

<sup>3</sup>Inclusion Health Department, St. James Hospital, Dublin 8, Ireland

**Background/Aims:** In Ireland, little is known about the health of children from the Roma community with no population health data available. An estimated 5000 Roma live in Ireland. Factors influencing Roma health include low education and health literacy levels, social exclusion, cultural differences, historical trauma, and financial constraints. Effective healthcare provision is hindered by these factors and lack of equitable access to necessary services.

**Methods:** An outreach clinic for children at risk of social exclusion provided a unique opportunity to engage Roma children and to better understand their specific health care needs and social determinants of health (SDH). The service was developed in partnership with existing Roma health and community services. Interpreters were utilised to verbally communicate appointment details and transport was arranged by hospital services when needed.

**Results:** Over 17 months 49 Roma children were referred for general Paediatric assessment. 81.6% 89.8% (n=44) were homeless, 85.7% (n=42) had no formal access to primary care. 93.9% (n=46) required interpreters. Only 16.3% (n=8) were fully vaccinated. Anaemia, vitamin D deficiency, dental disease and poor growth and nutrition were commonly identified issues. 61.2% (n=30) required hospital follow-up, with many significant pathologies identified.

**Conclusions:** Roma children referred to the clinic experienced marked social adversity and had extremely limited access to primary healthcare. Services have developed as a result to promote healthcare access such as Roma cultural ambassadors attending outreach clinics, streamlined dental care pathways and opportunistic vaccination. Future planning includes specialised clinics with community liaison and a role in health promotion supporting primary care initiatives.

## **ALBUMIN USE AND MANAGEMENT OF HYPOALBUMINAEMIA: EVIDENCE-BASED PRACTICE IN NEONATES AND CHILDREN IN INTENSIVE CARE**

AC Crabtree<sup>1</sup>, MZ Zairi<sup>1</sup>, OF Fitzgerald<sup>1</sup>, EH Howie<sup>1</sup>, OM McKinley<sup>1</sup>, MK Kelly<sup>1</sup>, SO O'Beirne<sup>1</sup>, **IW Waters<sup>1</sup>**, El Isweisi<sup>1,2,3</sup>,

GS Semova<sup>1,2,3</sup>, JM Meehan<sup>1,2,3</sup>, ER Roche<sup>1,2,3</sup>, AB Branagan<sup>1,2,5</sup>, EM Molloy<sup>1,2,4</sup>

<sup>1</sup>Discipline of Paediatrics, Trinity College Dublin, Dublin, <sup>2</sup>Trinity Research in Childhood Centre, Trinity College Dublin,

Dublin, <sup>3</sup>Children's Health Ireland at Tallaght, Tallaght University Hospital, Dublin, <sup>4</sup>Children's Health Ireland at Crumlin, Cooley Road, Dublin, <sup>5</sup>The Coombe Women and Infants University Hospital, Trinity College Dublin, Dublin 8, Ireland

**Background and Aims:** Hypoalbuminaemia is a common finding in neonates and children in intensive care. However, whether there is clinical benefit to correcting serum albumin levels is debated. In adult ICU, there is sufficient evidence to support use of albumin in specific patient groups. This study sought evidence for similar recommendations in neonatal and paediatric intensive care.

**Methods:** PubMed and EMBASE were selected as the databases to be used according to PRISMA guidelines. Data was extracted based on study design and size, patient demographics and treatment methods and outcomes.

**Results:** The search identified a total of 206 articles, reduced to 158 independent articles after duplicates were removed. 10 studies demonstrated low albumin as a poor prognostic factor across an array of critically ill states in paediatric and neonatal populations. Furthermore, 14 core papers relating to the clinical impact of albumin use were identified. Exogenous albumin is not associated with clinically significant improvements in ventilatory supports or oxygen requirements and had no effect on severity of respiratory distress. There is no difference in changes to mean arterial pressure when comparing normal saline and albumin. Albumin administration is found to be safe but of no relative benefit when compared to control fluids in resuscitating paediatric patients with sepsis.

**Conclusions:** Hypoalbuminaemia is a negative prognostic factor in critically ill neonates and children. In treating hypoalbuminaemia in NICU and PICU settings, albumin is not superior to control fluids in terms of clinical outcome, is of extra cost and carries potential for adverse effects.

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## WHEN IS IT NOT NON-ACCIDENTAL INJURY? A CASE STUDY OF A GENETIC VARIANT DIFFERENTIAL FOR CONCERNING FEATURES OF INJURY

P Whooley<sup>1</sup>, D Staunton<sup>2</sup>, R Kernan<sup>3</sup>

<sup>1</sup>General Paediatrics, Mayo University Hospital, Castlebar, Ireland

<sup>2</sup>General Paediatrics, Mayo University Hospital, Castlebar, Ireland

<sup>3</sup>Endocrinology, Children's Health Ireland at Temple Street, Dublin, Ireland

**Aims:** To describe a case of a 19-month old male who presented with multiple features suspicious for injury in whom an underlying genetic variant mutation was found to provide a likely differential.

**Methods:** Description of clinical presentation, work-up, management and literature review.

**Results:** A 19-month old male presented to Mayo University Hospital in December 2021 following a witnessed seizure secondary to a fall. A CT brain demonstrated an acute left frontoparietal subdural haematoma with mass effect. He was transferred to CHI at Temple Street for neurosurgical review. The bleed was managed conservatively. This child had previously undergone an MRI brain in 2020 to investigate mild hearing loss and motor delay, which demonstrated benign

enlargement of the subdural space. A subdural bleed was present at this point, but not reported. Significant concern was raised regarding potential cause of two subdural bleeds at different timepoints. A subsequent skeletal survey identified bilateral tibial corner metaphyseal fractures. This prompted further concern regarding a causative mechanism as the child had only started walking six weeks previously with no reported trauma. This child had a strong maternal family history of Osteogenesis Imperfecta. He had a distinctive phenotype of blue sclera, macrocephaly and a persistently wide fontanelle. A genetic work-up revealed a heterozygous pathogenic missense variant in the COL1A2 gene. This variant has previously not been found in population studies but has been described in two patients with Osteogenesis Imperfecta/Ehlers Danlos Overlap syndrome<sup>1</sup>. Such a mutation, with a substitution of glycine near the N-terminus of the triple helix, has the potential to give rise to features such as blue sclerae, translucent skin, hyperextensibility, growth deficiency, hearing deficits and a variable degree of bone fragility with susceptibility to fractures.

**Conclusions:** Recognition of subtle phenotypical features associated with this mutation variant may help to guide investigations, diagnosis and subsequent management of the underlying condition.

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### **AN UNUSUAL CASE OF 'BRONCHIOLITIS'**

**D M Wildes**<sup>1</sup>, E McHugh<sup>1</sup>, D McLaughlin<sup>2</sup>, B Scanlan<sup>1</sup>

<sup>1</sup>Department of General Paediatrics, Children's Health Ireland at Crumlin, Dublin

<sup>2</sup>Department of Paediatric Surgery, Children's Health Ireland at Crumlin, Dublin

**Case:** A four-month-old term male infant with no prior medical history presented to the paediatric emergency department due to respiratory distress, decreased oral intake and lethargy. On clinical examination, he was found to be tachypnoeic, with sub/inter-costal recession. On auscultation, he was noted to have atypical, “gurgling” breath sounds. A chest radiograph (figure 1) was performed which showed almost complete whiteout of the left chest, with some subtle lobulated lucency in the left apex, consistent with air filled bowel loops. There was also evidence of mediastinal shift to the right. A nasogastric tube was inserted and the patient was transferred to the Paediatric intensive care unit. A chest and abdomen radiograph was subsequently performed showing bowel loops in the thoracic cavity.

On admission, the patient was commenced on humidified high flow nasal cannula oxygen. Echocardiography showed no evidence of pulmonary hypertension. Renal, pleural and cranial ultrasounds showed no evidence of any other anomalies consistent with a syndromic constellation. The patient underwent a Type A Bochdalek hernia repair via a left upper quadrant incision. He was extubated on day two post-operatively and ventilated spontaneously without respiratory support. This patient had an uneventful post-operative course and was discharged to the ward, and subsequently home by day six postoperation. A follow-up chest radiograph was performed showing complete resolution of the aforementioned defect (figure 3).

**Conclusion:** Congenital diaphragmatic herniae (CDH) account for some of the most common birth defects, with an estimated incidence of 2-4 per 10,000 births. Typically, the diaphragmatic herniation occurs on the left-side (up to 90%), on the right-side (up to 10%) and bilaterally in rare cases. With advances in obstetric ultrasonography, the number of diagnoses made antenatally continue to increase. CDH can present after the initial neonatal period. Clinicians should be cognisant of atypical presentations of CDH, when interpreting chest radiographs of infants in respiratory distress.



**MANAGEMENT OF ANTENATALLY DETECTED CONGENITAL ANOMALIES OF THE KIDNEYS AND URINARY TRACT (CAKUT) – A QI INITIATIVE****D.M Wildes**<sup>1,2</sup>, C O'Hagan<sup>1</sup>, M Stack<sup>2</sup>, M.A Boyle<sup>1,3</sup><sup>1</sup>Department of Neonatology, Rotunda Hospital, Dublin 1, Ireland.<sup>2</sup>Department of Paediatric Nephrology & Transplantation, Children's Health Ireland, Dublin 1, Ireland <sup>3</sup>Department of Paediatrics & Neonatology, Royal College of Surgeons in Ireland, University of Medicine & Health Sciences, 123 St. Stephen's Green, Dublin 2, Ireland.

**Introduction:** Congenital anomalies of the kidney and urinary tract (CAKUT) are the leading cause of endstage renal failure in children. There is a well-established causative relationship between CAKUT and the requirement for renal replacement therapy, which carries significant health and psychosocial burden for children and their caregivers alike. Antenatal diagnosis and early post-natal confirmation facilitates timely management. We sought to ascertain our performance as an institution in arranging appropriate follow-up and management of CAKUT. Our findings were used to develop a standard of practice, which we will implement at the Rotunda Hospital.

**Methods:** Retrospective chart review was performed of all patients with antenatally detected CAKUT identified by the Maternal-Fetal Medicine department at the Rotunda from March 2020-March 2021. Powerchart/NIMIS records were utilised for data extraction. Literature review and advice from our national paediatric nephrology unit were used to construct our guidelines.

**Results:** Of n=63 patients, 38 (60.3%) were male and 25 (39.7%) were female. Mean gestational age at detection of CAKUT was 24.87 weeks (IQR 20-32). Of all suspected cases, n=26 (41.27%) patients had postnatal confirmation of CAKUT. The most prevalent anomaly was hydronephrosis, with n=19 patients investigated for this. Just 63.2% of antenatal scans with hydronephrosis specified renal measurements. Compliance with guidelines was 89.65%. Adherence was poorest in the domains of bilateral hydronephrosis (0%) and ectopic kidneys (50%). Four terminations of pregnancy were noted in this population and six patients died in the perinatal period.

**Conclusion:** CAKUT are common in the practice of both Obstetricians and Neonatologists. It is important to include renal measurements in ultrasound reports, where hydronephrosis is concerned. Clinicians must endeavour to ensure post-natal imaging is conducted in an appropriate time-frame. A repeat audit will be conducted with our guideline in place.

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### **Sub-Specialty TRANSITIONAL CARE MODELS IN ADOLESCENT KIDNEY TRANSPLANT RECIPIENTS**

**D M Wildes**<sup>1</sup>, C S Costigan<sup>1</sup>, M Kinlough<sup>1</sup>, J Flynn<sup>1</sup>, N Dolan<sup>1, 2</sup>, M Riordan<sup>1,2,3</sup>, C Sweeney<sup>1,2</sup>, M Stack<sup>1,2</sup>, M

Waldron<sup>1,2</sup>, O Walsh<sup>3,4</sup>, K M Gorman<sup>5,6</sup>, A Awan<sup>1,3,6</sup>

<sup>1</sup>Paediatric Nephrology & Transplantation, Children's Health Ireland at Temple Street, Dublin, Ireland

<sup>2</sup>Paediatric Nephrology, Children's Health Ireland at Crumlin, Dublin, Ireland

<sup>3</sup>Paediatrics and Child Health, Royal College of Surgeons in Ireland, Dublin, Ireland

<sup>4</sup>General Paediatrics, Children's Health Ireland at Temple Street, Dublin, Ireland

<sup>5</sup>Neurology and Clinical Neurophysiology, Children's Health Ireland at Temple Street, Dublin, Ireland

<sup>6</sup>School of Medicine and Medical Science, University College Dublin, Dublin, Ireland

**Aims:** Adolescence is a time of significant change for patients, guardians and clinicians. The paediatrician must ensure patients develop the necessary skills and knowledge required to function as an independent entity, with autonomy over their own care. The transfer from paediatric to adult care carries an increased risk of graft-related complications attributable to a multitude of reasons, particularly non-adherence to immunosuppressants and poor attendance at appointments. This systematic review was conducted to ascertain the models available to clinicians caring for kidney transplant recipients, comparing their respective approaches.

**Methods:** A systematic review was performed, in keeping with PRISMA guidelines. OVID-MEDLINE/EMBASE databases were searched for studies that outlined replicable models pertaining to transitional care of paediatric kidney transplant recipients between 1946->Q3-2021. The reference lists of selected articles were also perused and experts in the field were consulted for

further eligible articles. Two investigators assessed all studies for eligibility and independently performed data extraction, discrepancies were settled by consensus.

**Results:** A total of 1121 abstracts were identified, leaving 1029 upon removal of duplicates. 51 articles were deemed appropriate for full-text review and critical appraisal. Twelve articles were deemed eligible for qualitative synthesis. Every paper utilized a different transition model. All but one model included a physician and nurse at minimum in the transition process. The involvement of adult nephrologists, medical social work, psychology and psychiatry was variable. The mean age for initiation of transition was 13.4 years (range:10–17.5). The mean age at transfer was 18.3 years (range:16–20.5).

**Conclusions:** Despite the well-established need for good transitional care for paediatric solid-organ transplant recipients, models tailored specifically for kidney transplant recipients are lacking. Further research and validation studies are required to ascertain the best method of providing effective transitional care to these patients. Transitional care should become a standardized process for adolescents and young adults with kidney transplants.

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Medical/Education Management

#### **CHILD ABUSE KNOWLEDGE AND REPORTING PRACTICES IN CORK UNIVERSITY HOSPITAL.**

**HHB Abdul Hamid, D Finn**

<sup>1</sup>Department of Paediatrics and Child Health, University College Cork.

#### **Aims**

1. To assess and compare the knowledge of child abuse presentation and reporting procedures in CUH Paediatric and Emergency Department healthcare staff.
2. To review child abuse reporting practices by looking at reporting experience, attitudes towards reporting, and likelihood to report.
3. To identify any factors affecting knowledge level and reporting practices.

**Methods:** This is a cross-sectional study done by distributing online and physical questionnaires to healthcare staff in CUH Paediatric and Emergency department who were conveniently sampled. The questionnaire was developed based on Children First National Guidance 2017 and was pilot tested on 10 medical students. For the assessment of knowledge level, correct answers are scored 1 while wrong and unsure answers are scored 0. For reporting practices, a 5-point Likert score was used and a higher score indicates a more positive reporting practice. Mann-Whitney and Kruskal-Wallis tests were done for statistical significance and correlation respectively.

**Results:** 67 responses are included in the study. The overall mean score for knowledge of child abuse presentation and reporting procedure is 11.9 out of 15. In terms of reporting experience, 79.1% of respondents have seen a suspected child abuse case but only 47.8% have experience reporting. A higher knowledge level is statistically correlated with a more positive reporting practice but no factors were identified to affect the knowledge level and reporting practices. There is no significant difference in the knowledge level and reporting practices between the two departments.

**Conclusion:** Healthcare staff in CUH Paediatric and Emergency department have a satisfactory level of knowledge and reporting practices. The level of experience, profession type, or Children First training did not affect knowledge or reporting practices.

## **THRIVING IN BST: INTRODUCTION OF A NOVEL MANDATORY HEALTH AND WELLBEING (H&W) COURSE**

### **FOR BASIC SPECIALIST TRAINEES IN RCPI, A TRAINEE-LED INITIATIVE**

**S Casey<sup>1, 2</sup>, P Carr<sup>1, 3</sup>, E Porter<sup>1, 4</sup>, N Moriarty<sup>1, 5</sup>, J Yates<sup>1, 3</sup>**

<sup>1</sup>RCPI Health & Wellbeing Committee, RCPI, Dublin, Ireland

<sup>2</sup>Paediatric SpR, RCPI, Dublin, Ireland

<sup>3</sup>Occupational Health SpR, RCPI, Dublin, Ireland

<sup>4</sup>Dermatology SpR, RCPI, Dublin, Ireland

<sup>5</sup>Obstetrics & Gynaecology Registrar, RCPI, Dublin, Ireland

**Aims:** We aimed to introduce a mandatory BST course to cover the below curriculum:

1. Identifying H&W supports
2. Setting/Achieving goals
3. Managing stress/burnout
4. Navigating interpersonal conflict
5. Troubleshooting common BST challenges

Course faculty will be senior NCHDs (SpR or equivalent), with oversight from the RCPI H&W department. The course is intended to be delivered to all 600 BST trainees in RCPI.

**Methods:** Members of RCPI H&W Committee designed the course content and delivered 2 virtual pilots in academic year 2020-2021. The RCPI H&W handbook (1) serves as pre-course reading. Course content is delivered through interactive workshops including breakout rooms, online whiteboards, scenario-based learning, and Q&A panel. Anonymous quantitative and qualitative feedback is sought from candidates after each course. Following positive feedback (see below), the course was listed as 'desirable' in 2021-2022. With further positive feedback, the course was registered as 'mandatory' for all BSTs from academic year 2022- onwards, with 16 course dates per academic year. A training pathway for inducting further faculty was developed to accommodate this.

**Results:** To date, 8 iterations of the course have been delivered. Of the candidates who provided feedback (n= 15), 93% of respondents were satisfied with the course, felt they could relate the

content to their role, and would recommend it to a colleague. Qualitative feedback included “(this was) the best course I have done so far” and “(it) should be compulsory for any trainee”.

48 faculty have subscribed to training, with 12 fully-trained faculty to date.

**Conclusion:** Candidate feedback has been overwhelmingly positive. All curriculum elements are met in precourse and in-course material. Sufficient courses have been provided to cover all BST trainees, and course dates are fully subscribed from candidate- and faculty-perspectives. The outputs of this course are now being shared across other medical training bodies in Ireland so they can develop their own iterations.

1. Fenn J et al. Looking After Your Health & Wellbeing- A Guide For Doctors (2nd edition) [online resource]. RCPI; 2021 (cited 28/09/22). Available from; <https://rcpi-live-cdn.s3.amazonaws.com/wp-content/uploads/2021/07/RCPI-Health-and-Wellbeing-Booklet-Update-2021-1.pdf>

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## EDUCATION - AN INTRINSIC COMPONENT OF PERSON-CENTRED CARE FOR CHILDREN AND ADOLESCENTS WITH SERIOUS ILLNESSES

M Colleran<sup>1</sup>

<sup>1</sup>Palliative medicine, St. Brigid's Hospice, Co. Kildare, Ireland

<sup>2</sup>Palliative medicine, Naas General Hospital, Co. Kildare, Ireland

**Background:** School is normal part of childhood and adolescence. However, children and adolescents with serious illnesses are disadvantaged by the illness itself, by associated absences from school and by lost educational opportunities. Children and adolescents with serious illnesses are disadvantaged by illness and then further impacted upon by lost educational opportunities. Participation in school effects their quality of life. However, they have poorer educational and ultimately poorer employment outcomes later.

**Method:** A review of national and international practice in providing educational support for children and adolescents with serious illnesses was done. This includes the lived experience of the author of accessing home tuition for her daughter here in Ireland.

**Results:** Options for educational support for students with significant medical conditions include home tuition, hospital school, individualised educational plan and re-entry to school after illness related absenteeism. Educational supports in Ireland include home tuition. The process of applying for home tuition in Ireland based on serious illness is complex and comprehensive. Serious illnesses sadly can lead to the death of the effected student. Teachers may need support addressing grief and bereavement in other pupils.

**Conclusion:** Children and adolescents with serious illnesses have poorer educational and ultimately lesser employment outcomes. Models of educational supports vary but can include home tuition and hospital schools. Individualised educational plans, and re-entry preparations and close collaboration between the students' parents or guardians, school and hospital school and home tutors.

**Poster No: 115**

### **REAL LIFE EVALUATION: WHAT DOES DIGITAL TRANSFORMATION MEAN FOR FRONTLINE PAEDIATRIC JUNIOR DOCTORS?**

**B Dias**<sup>1</sup>, J Finnegan<sup>2</sup>, R Finnegan<sup>3</sup>, N O'Hare<sup>4</sup>, M Hesselings<sup>5</sup>

<sup>1</sup>Medicine Year 3, Royal College of Surgeons in Ireland (RCSI) Research Summer School (RSS), Dublin, <sup>2</sup>Lead

NCHD, Children's Health Ireland (CHI) at Crumlin, Dublin, <sup>3</sup>Lead NCHD, CHI at Temple Street , Dublin, <sup>4</sup>Chief Information Officer, CHI, Dublin, <sup>5</sup>Department of Digital Health, CHI at Temple Street, Dublin,

**AIMS:** Digital transformation in the healthcare industry has the capability to improve the working lives of frontline doctors, and their ability to treat patients in a more efficient and safe environment. Much of the current literature available is focused on the benefits of individual systems, but there is little published about the general experience of frontline staff with technology. A study was conducted at two children's hospitals to better understand the working realities of paediatric NCHDs in relation to digital technology.

**METHODS:** A survey consisting of seven questions was circulated amongst NCHDs. Doctors responded through digital and paper based surveys, as well as face to face interviews. The

questions addressed their opinions on adopting paper-free systems and any barriers to technology. Thematic analysis was used to identify commonalities amongst interview responses.

**RESULTS:** A total of 66 survey responses were collected from two hospitals in Dublin across multiple medical departments. Most doctors stated that digital technology in the clinical setting was slow (n=51, 77.3%) in addition to infrastructure being limited and outdated (n=52, 77.8%). Further assessment revealed that 89.2% (n=58) of NCHDs support the idea of a completely paperless hospital. Barriers to digital transformation include multiple user logins, limited IT support and limited digital training.

**CONCLUSION:** The study revealed that NCHDs currently face many obstacles when it comes to smooth and effective digital work. These are obstacles encountered by doctors across the country. Despite these barriers, NCHDs believe that a complete digital transformation of the workplace is the way of the future of healthcare, thereby improving patient safety and efficacy.

This study provides evidence of the importance of improving basic digital infrastructure before attempting to add the latest trends in technology. Successful digital transformation requires organisations, in Ireland and worldwide, to ensure that basic digital support and infrastructure are in place first.



## **MEDICAL STUDENTS' PERCEPTION OF FEEDBACK**

**AM Fitzgerald**<sup>1</sup>, M Salama<sup>2</sup>, L Jansen<sup>2</sup>, MB O'Neill<sup>3</sup>

<sup>1</sup>Paediatrics, Ollscoil na Gaillimhe, Galway , Ireland

<sup>2</sup>Paediatrics, Mayo University Hospital , Castlebar, Ireland

**Aims:** This study aimed to evaluate medical students' experiences with feedback on clinical placement, explore their knowledge about 360-degree feedback and assess their attitudes towards this method of professional development.

**Methods:** This is a cross-sectional hospital-based survey of 80 medical students in their 3rd and 4th year in Ollscoil na Gaillimhe.

Data was collected using an online self-administered questionnaire. The survey explored the learning environment, student experience, and student opinion on feedback. A seven-point Likert scoring scale was utilised. Four additional questions were answered in a narrative format and allowed elaboration on elements of effective feedback and multimodal feedback structures.

To confirm the clarity of the questionnaire, an initial pilot study was organised among three medical students, resulting in minor questionnaire editions. The results were collected and analysed using Survey Monkey, and Microsoft XL. Answers were quoted, categorised, and analysed.

Ethical approval from the Research Ethics Committee of Mayo University Hospital and Written informed consent from participants were obtained.

**Results:** The survey response rate was 72.5%. 98.3% of the students believed that feedback improves student development, and 75.9% recognised its role in patient safety. Bedside history taking was observed only 30% of the time (mean Likert score of 3.07). Feedback was only given 50% of the time (mean Likert score of 4.6), positively skewed Likert reported by 9 (15%). Students ranked feedback an average of 4 out of 10 as a stressful experience, and only four were familiar with 360-degree feedback concept.

**Conclusion:** Medical students value feedback in their professional development and patient safety, but there is a significant disparity in the observation of bedside skills and commentary feedback in

medical training in Ireland; therefore, more constructive, standardised and timely feedback is needed.

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## PEER-LED ONLINE TUTORIALS- EXPERIENCE OF MRCPI PART TWO CLINICAL EXAMINATION CANDIDATES

**NM McGrath**<sup>1</sup>, RG Stone<sup>1</sup>, E Hayes<sup>1</sup>, S Harvey<sup>1</sup>, R Maher<sup>1</sup>, S Mullen<sup>1</sup>, C O'Neill<sup>1</sup>, M Pentony<sup>1</sup>, D Pereira<sup>1</sup>, TF Verbruggen<sup>1</sup>, C McDonnell<sup>1</sup>

<sup>1</sup>General Medicine, Children's Health Ireland, Dublin, Ireland

**Aims:** In response to infection control measures imposed during the COVID-19 pandemic, an NCHD-led initiative was developed within Children's Health Ireland to help those preparing for the MRCPI

(Membership of the Royal College of Paediatrics of Ireland) Part 2 Clinical Examination. Four cycles of the online tutorial program were delivered between October 2020 and May 2022. We aimed to determine the effectiveness of this project.

**Methods:** The tutorial program was designed according to the MRCPI Clinical Examination content blueprint. Each tutorial cycle recruited and coordinated participants through an instant messaging service. Senior NCHDs were recruited to deliver tutorials via online video conferencing. The tutorial series was delivered over a two-week period prior to each examination date. Questionnaires evaluating course expectations and effectiveness were provided prior to and following course completion.

**Results:** We delivered four tutorial program cycles, with a total 126 responses to "before" surveys and 105 responses to the "after" surveys. An international audience was achieved with participants from Ireland, Oman, Saudi Arabia and Pakistan. The majority of respondents (65%) were enrolled in RCPI Basic Specialist Training, with a smaller number (19.4%) in standalone registrar posts. An overwhelming majority of respondents (99.2%) rated the course as 'excellent' or 'very good'. Feedback from each cycle was used to adapt the program to the learning needs of both national and international participants.

**Conclusions:** This project was initiated in response to the challenges of education delivery during the COVID-19 pandemic. The responses from attendees indicate an ongoing appetite for structured, online, post-graduate education, particularly in the lead-up to clinical examinations. The peer-led nature of the initiative not only benefits attendees but also presenters, who gain teaching experience through their participation. Although not without limitations, we believe that online teaching could form an inclusive adjunct to clinical training, allowing trainees access to education, regardless of location.

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### **SIGNIFICANCE OF NEONATAL CARDIOVASCULAR EXAMINATION AND PRE-DISCHARGE SPO2 CHECK AT 24 HOURS OF LIFE.**

**A RAZA, L REHMAN, M ISLAM, M SHAHID**

<sup>1</sup>Paediatrics Department, Portiuncula University Hospital, Ballinasloe, Ireland

**Aim:** To report a rare case of Cor Triatrium in 25 hour old neonate.

**Method:** A male infant was born at 39 weeks via SVD with good APGARS. Antenatal scans including 20 weeks anomaly scans were unremarkable. There was no family history of Congenital-Heart-Disease. Baby was transferred to mother for routine postnatal care.

He was started on formula feeding and his initial postnatal course was uneventful. At 20 hours of life, his routine Day 2 examination was unremarkable. Parents were advised to wait for 24 hours to recheck cardiovascular examination as per hospital policy. At 25 hours of life, baby was noticed dusky by mother and his pre and post ductal SPO2 were in low 80's. On assessment-vitals: 36.8C, HR 170/min, RR 65/min, SPO2 pre-ductal 81% & post-ductal 83% on room air.

**Physical examination:** Central cyanosis, left ear tag, no dysmorphism or Rash

Systemic examination was unremarkable apart from Gallop heart rhythm with otherwise normal cardiovascular exam. Baby was admitted to SCBU on NCPAP (PEEP 6 and FIO2 100 %) to maintain SPO2 >90%. Nasogastric tube was inserted. Two peripheral IV lines were cited and IV fluids were commenced. After discussion with cardiologist, IV Prostaglandin infusion was commenced. Baby was transferred to tertiary-care-cardiology for further evaluation and management.

**Results:** His baseline blood investigations were normal. Four limbs BP was normal. Chest Xray showed cardiomegaly. Echocardiography revealed PROMINENT MEMBRANE IN RIGHT ATRIUM OVERLYING TRICUSPID VALVE and ASD with the impression of CORTRIARIUM. Baby was managed conservatively for 6 weeks and then an uncomplicated surgical resection of Cor Triatrium Via Dexter and ASD closure performed.

**Conclusion:** Cor triatrium is a very rare congenital heart disease that presents in association with other congenital heart defects i e TOF, ASD, VSD etc. Its prevalence is 0.1-0.4% of congenital heart diseases. 24 hours cardiovascular exam and pre discharge SPO2 is paramount for timely diagnosis of rare of CHDs to prevent morbidity and mortality.

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- 5-<https://www.hse.ie/eng/about/who/healthwellbeing/our-priority-programmes/child-health-and-wellbeing/newborn%20exam.pdf>

**INFANTILE URACHAL CYST ABSCESS- AN IMPORTANCE OF DIFFERENTIAL DIAGNOSIS****L Rehman**, A Raza, M Islam, M Shahid<sup>1</sup>Paediatrics Department, Portiuncula University Hospital, Ballinasloe, Ireland**Aim:** To report a case of 4-month-old infant with ring shaped infected periumbilical mass.**Method:** A 4-month-old male infant presented to paediatric emergency with two days history of high-grade fever, redness and swelling around umbilicus. He was born full term via SVD with no birth complication. His antenatal scans were unremarkable. He was admitted with E-coli UTI at age of two month and diagnosed with left sided mild pelvicalyceal dilation. He was commenced on prophylactic trimethoprim. His initial triage assessment revealed: Pews:5,Temp:37.9 c,RR-64, SPO2-100%, HR-144, CCRT:3 second, generalized pallor, irritability, peripheral mottling.

Systemic examination revealed a well demarcated, circular shaped, erythematic, and tendered periumbilical swelling with no signs of abdominal obstruction. Furthermore, there was no history of any umbilical discharge since birth. First impression was omphalitis/periumbilical cellulitis.

After initial oral analgesia, he was inserted two Intravenous lines, performed full septic work up as well as venous blood gas, renal and liver function tests. He was then stabilised with 20 ml/kg IV saline bolus, IV Ceftriaxone, flucloxacillin, Metronidazole and Gentamicin. He was referred to tertiary care team and had surgical drain of abcess and uncomplicated resection of Urachal cyst.

**Results:** Blood investigations showed low HB-9.6, high WBC- $38 \times 10^9$ , Neuts- $23 \times 10^9$ /L & CRP-150. PFA reported circular shaped density arising from pelvis with non-obstructive bowel pattern. USG abdomen revealed 4 by 4 cm complex collection tracking distally from the umbilicus to the superior bladder confirming infected urachal cyst.**Conclusion:** The urachus is an embryological tubular connection between the bladder dome and umbilicus which involutes at fourth to fifth month of gestation. Failure to obliterate at different anatomical locations results in various Urachal anomalies. A completely patent urachus is a rare anomaly, occurring in 1–2.5 per 100,000 deliveries. Male-to-female ratio of a patent urachus is 3:1.

Urachal cyst abscess is a rare complication of an infected patent urachal cyst. Early diagnosis and timely treatment would prevent serious complications and mortality in infants.

## **TO ASSESS COMPLIANCE WITH PULSE OXIMETRY IN NEWBORN EXAMINATION AND AVERAGE TIME SPEND BY NCHD FOR MEASUREMENT OF PRE-DUCTAL SATURATION.**

**S Ahmed**, L Loughlin, M Azam

<sup>1</sup>Pediatric, Wexford General Hospital, Wexford, Ireland

**Aims:** To assess compliance with National Newborn Examination guidelines for the measurement of post ductal saturation on day 2 examination.

**Background:** The National Newborn examination guideline recommend that Pulse oximetry (PO) should be performed in either foot prior to discharge ideally after first 24 hours to screen for congenital heart disease(CHD)<sup>1</sup>. CHD is one of the most common birth defect with incidence of 9 out of every 1000 live birth <sup>2,3</sup>. Routine newborn pulse oximetry screening identifies babies with critical congenital heart defects(CCHD) that would otherwise have been missed by antenatal ultrasound and postnatal examination <sup>4</sup>. The site of testing (post-ductal versus pre- and post-ductal) had no significant effect on sensitivity nor specificity for detection of CCHD <sup>5</sup>.

**Methodology:** A retrospective view of 31 babies born between 1/9/22 – 21/09/22 at WGH. Documentation of pre and post ductal saturation was noted on pro forma. Time to complete measurement of saturations were noted on 5 newborn examinations.

**Results:** Compliance results for documentation of both pre and post ductal saturation on appropriate page were 93%.In 6.4% of cases documentation were missed. Mean time spend by NCHD on 5 newborn examination for measurement of pre-ductal saturation was 44 seconds, and for post ductal saturation measurement it was 43.6 seconds.

**Conclusion:** overall compliance was good, 93% for documentation of both pre and post ductal saturation. Average time spent by NCHD for measurement of pre-ductal saturation was 44 seconds. In 2021, 1714 babies were born in WGH, thus we estimate that NCHD spent over 4 hours performing pre-ductal saturation.

**Recommendations:** Information about the audit will be presented in our departmental teaching with the aim to improve compliance. Measurement of pre-ductal saturation is not required therefore , neonatal Pro forma should be updated.

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**Withdrawn**

## IMPROVING NCHDS COMPLIANCE WITH LOCAL POLICY IN A TERTIARY NEONATAL UNIT

S.A Allawendy<sup>1</sup>, U Khan<sup>1</sup>, M Kenosi<sup>1</sup>

<sup>1</sup>Neonatology , Cork university maternity hospital , Cork, Ireland

**Aim:** To increase the compliance with the head circumference measurement on admission policy from 49% to 100% by 6 months in all babies admitted to the neonatal unit within 24 hours of admission.

**Methods:** we used HSE QI tools to highlight the steps of admission process and identify the aspects of improvements; our drivers were centered on communication, education, and documentation. Our interventions included education of NCHDs and midwives. Communication and documentation. Health care assistants were involved in the process.

**Results:** Compliance of NCHDs with Head circumference measurement increased from 49% to 100% in 3 months instead of 6 months.

**Discussion/Conclusion:** we successfully highlighted the importance of quality improvement initiatives as a continuous dynamic process which can offer solutions for day to day practice problems.



## **AN AUDIT OF POSTNATAL MANAGEMENT OF ANTENATALLY DIAGNOSED RENAL COLLECTING SYSTEM**

### **DILATATION OR HYDRONEPHROSIS**

**K Amin<sup>1</sup>, AD Rodriguez<sup>1</sup>, W Asif<sup>1</sup>**

<sup>1</sup>Department of Paediatrics, St Luke's hospital, Kilkenny, Ireland.

**AIMS/BACKGROUND:** To monitor compliance with standard guidelines for postnatal follow up of antenatally diagnosed renal pelvic dilatation/hydronephrosis in our hospital.

**Royal Children's Hospital Melbourne (RCH)** guidelines based on third trimester scan findings:

1. Bilateral dilatation >10mm requires USS pre discharge.
2. Unilateral (severe) dilatation >15mm or moderate-severe calyceal dilatation requires USS pre discharge.
3. Unilateral (moderate) dilatation 10.1-15mm with or without calyceal dilatation requires USS at 1-4 weeks of age.
4. Bilateral or unilateral (mild) dilatation 7-10mm or mild calyceal dilatation requires USS at 4 weeks of age.

**METHODS:** Neonates who had renal ultrasound to follow up antenatal renal pelvic dilatation in the year 2021 were audited for compliance with the RCH guidelines.

**RESULTS: 16 neonates** underwent Renal USS in the year 2021. One of the 16 neonatal ultrasounds performed were arranged as suggested by the RCH guidelines, the other 15 were not

1. 15 had a postnatal scan done in first 3 days of life.
2. 9 had normal postnatal scans.
3. 7 had abnormal neonatal ultrasound requiring follow up.
4. None of the neonates were symptomatic.
5. 10 had antenatal renal pelvis measurements recorded, 4 had antenatal dilatation recorded only without measurements, in 2 cases the dilatation was recorded verbally without written documentation.
6. Of the 10 babies whose antenatal renal pelvic measurements were recorded:
  - 1 had severe bilateral dilatation.
  - 4 had mild bilateral dilatation.
  - 3 had moderate unilateral dilatation.
  - 2 had mild unilateral dilatation.

**CONCLUSION:** We are not following the standard RCH guidelines. Notably our postnatal ultrasounds are being arranged earlier than recommended. We recommend the following:

1. Development of departmental guidelines in consensus with the Radiology department and in compliance with the RCH guidelines.
2. Re-audit in 1 year's time to monitor compliance and close the audit loop.

[https://www.rch.org.au/clinicalguide/guideline\\_index/Antenatal\\_urinary\\_tract\\_dilation/](https://www.rch.org.au/clinicalguide/guideline_index/Antenatal_urinary_tract_dilation/)

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## **ETIOLOGY, OUTCOME OF NEONATAL JAUNDICE IN WEXFORD GENERAL HOSPITAL FROM AUGUST 2021**

### **TO AUGUST 2022, AND COMPLIANCE WITH LOCAL GUIDELINES**

SB BATOOL<sup>1</sup>, ES SULIMAN<sup>2</sup>, AK KALIM<sup>3</sup>

<sup>1</sup>Paediatrics, Wexford General Hospital, Wexford, Ireland

**Aim:** Jaundic hyperbilirubineamia refers to the yellow coloration of the skin caused by accumulation of bilirubin. It is common in newborns affecting between 60% of term and 80% of preterm infants. There are two types of jaundice, Pathological and Physiological [1][2]. Current guidelines in Wexford General Hospital (WGH) includes serum bilirubin and DCT (Direct Coombs Test) within first 24 hours of life in infants with visible jaundice or risk factors and repeat SBR in 6hrs if recent levels are below 50mmol treatment line[1][3]. The aim is to review the etiology and outcomes of neonatal jaundice(NNJ) admissions and whether compliance with local guidelines is achieved for effective treatment[1]

**Methods:** A retrospective review of medical records of infants who have had NNJ screening done between August 2021 to August 2022. These records are categorised according to their risk factors, treatment and prognosis. Medical notes of these babies were also analysed. Data was collected on their hospitalization, treatment and outcomes.

**Results:** Total number of births for same period was 1714, and 14(0.8% of total births) amongst these had admission in SCBU due to NNJ. So a total of 14 babies were included in this study. 10 (71%) OF NNJ admissions were boys and 4(28%) were girls. 5 (35%) had physiological jaundice,

9(64%) had pathological NNJ. 7(50%) had maternal risk factor as the cause including ABO incompatibility, maternal age.03 (21%) were premature babies.03 (21%) had DCT positive status. 01(0.07%) had sub galeal hematoma.10 (71%) required single phototherapy, 04(28%) required double phototherapy. [1][2]There were 2(0.1%)readmissions.

**Conclusion:** Care was successfully managed in WGH SCBU with no infants experiencing additional adverse outcomes relating to diagnosis or treatment. No infants required transfer to tertiary care hospital. Local guidelines support expert treatment in relation to NNJ. Evaluation of records demonstrated a high standard of care, significant adherence and implementation to guidelines exists within the hospital.

1- Wexford General Hospital local Guidelines 2- NICE (2016) Neonatal Jaundice Clinical Guideline 98. <http://www.nice.org.uk>  
3[http://www.rch.org.au/clinicalguide/guideline\\_index/Jaundice\\_in\\_early\\_infancy/](http://www.rch.org.au/clinicalguide/guideline_index/Jaundice_in_early_infancy/)

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## AN AUDIT OF DELAYED CORD CLAMPING PRACTICES

M Benke<sup>1</sup>, F Khan<sup>1</sup>, R Khan<sup>1</sup>, N Al-Assaf

<sup>1</sup>Neonatal Unit, University Maternity Hospital Limerick (UMHL), Limerick, Limerick

**BACKGROUND:** Delayed cord clamping (DCC) following birth, unless cord integrity is compromised is physiologically beneficial for the neonate, including reduced risk of death, IVH, anaemia requiring transfusion, NEC.

**AIM:** To assess the DCC practice at UMHL including the timing and the documentation of timing of cord clamping (CC) at deliveries.

**METHOD:** We reviewed the documentations of one hundred consecutive live births between 19<sup>th</sup> February and 2<sup>nd</sup> March 2022 at University Maternity Hospital Limerick. Data was collected including demographics, maternal and neonatal risk factors and documentation of cord clamping.

**RESULTS:** Gestational age of audited babies was ranging between 35+2 and 41+6, average birth weight was 3399 g. The timing of CC was documented in 48 deliveries. The meantime of CC was 123 sec (range 40- 300 sec). In case of the 49 vaginal births, timing of CC was documented in 38 cases (77.5%) by midwives. Babies who were born by caesarean-section, CC was documented in 10 out of the 41 cases where neonatal doctor was called to the delivery and the documentation was not done by the midwives.

**CONCLUSION:** Documentation of CC happened in less than half of all live births. While midwives were mainly documenting timing of cord clamping at vaginal deliveries, timing of CC was not

documented in most of caesarean births. Doctors are not routinely documenting the timing of cord clamping, including the need of immediate cord clamping.

Re-education of staff with the new NRP 8<sup>th</sup> edition recommendations is required. We recommend adding a designated section on the birth documentation sheet to achieve full record of DCC in all eligible babies.

Re-auditing our practice on documentation of delayed cord clamping is required in 6 months' time.

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### **USING THE SERUM SODIUM CONCENTRATION AS A MARKER OF FLUID BALANCE IN INFANTS WITH NEONATAL ENCEPHALOPATHY (NE)**

**C Broderick Farrell<sup>1</sup>, J McGinley<sup>2</sup>, P Corcoran<sup>3</sup>, Indra<sup>3</sup>, JF Murphy<sup>1</sup>**

<sup>1</sup>Department of Neonatology, National Maternity Hospital, Dublin, Ireland

<sup>2</sup>National Women and Infants Health Programme, HSE, Dublin, Ireland

<sup>3</sup>National Perinatal Epidemiology Centre, NOCA, Dublin, Ireland

**Aims:** Infants with neonatal encephalopathy (NE) are at risk of fluid overload and cerebral oedema due to SIADH and ischaemic renal impairment. The standard practice is a reduction in total fluid input over the first few days after birth. In this study, we used the serum sodium (S. Na) as a measure of fluid balance in a cohort of infants with NE.

**Methods:** The S. Na concentrations on days 1, 2 and 3 of life were collected on a group of infants with NE.

The data was extracted from the Neonatal Therapeutic Hypothermia Reports in Ireland for 2019 and 2020. The S. Na levels were subdivided into 3 categories:  $\leq 124$ mmol/l (severe hyponatraemia), 125-131 mmol/l (moderate hyponatraemia) and  $>131$  mmol/l (normal range).

**Results:** There were 148 infants with NE in the study. The distribution of the number of infants in each of the three S. Na categories across days 1, 2 and 3 were as follows:

$\leq 124$ mmol/l - Day 1: 26 (17.5%) infants, Day 2: 25 (16.9%) infants, Day 3: 20 (13.5%) infants  
125-131mmol/l - Day 1: 77 (52%) infants, Day 2: 68 (45.8%) infants, Day 3: 50 (33.7%) infants  
 $>131$  mmol/l - Day 1: 42 (28.3%) infants, Day 2: 27 (24.9%) infants, Day 3: 51 (34.4%) infants  
S. Na not documented - Day 1 in 3 (2.0%) infants, Day 2 in 18 (12.1%) infants, Day 3 in 27 (18.2%) infants.

**Conclusion:** The distribution of the S. Na concentrations in this indicates that a high proportion of infants with NE were fluid overloaded over the first 3 days after birth. One in six infants were

significantly overloaded with a S. Na  $\leq 124$ mmol/l. It is recommended that the fluid intake should be more individually adjusted using the S. Na for guidance.

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## A RETROSPECTIVE REVIEW OF CAFFEINE CITRATE PRESCRIBING IN A TERTIARY NEONATAL INTENSIVE CARE UNIT

SJ Cahill<sup>1</sup>, NH Aminudin<sup>1</sup>, M Corderroua<sup>1</sup>

<sup>1</sup>Department of Neonatology, National Maternity Hospital, Dublin, Ireland

**Background:** Caffeine citrate used for neonatal apnoea of prematurity (defined as cessation of breathing for 20 seconds or  $\geq 10$  seconds with associated desaturations or bradycardia)<sup>1</sup> is also considered for neonates  $< 1.25$ kg and is used in assisting with airway extubation from the ventilator. Caffeine prescription practice varies in within neonatal units. A multi-centre cohort 15-year study reaffirmed this variation and the consensus surrounding its discontinuation.

**Aims:** To evaluate the practices associated with caffeine prescribing, in accordance with our unit protocol and international literature; particularly around duration of treatment and its discontinuation in relation to gestational age.

**Methods:** A retrospective review of all of caffeine prescriptions between October to December 2021 was performed. Patient identification was anonymised. Gestational age, respiratory support and presence of apnoea was extracted. Data collation assessed trends in prescribing practices and reviewed alongside local protocol for prescribing.

**Results:** 37 neonates were prescribed caffeine citrate. 11 patients were excluded from final analysis (redirection of care/transferred to another hospital). N=26; 7 patients had caffeine discontinued while remaining on respiratory supports. The gestational age range was 33- 40 weeks. Discontinuation regime of the patients not on any respiratory supports varied (n=19). 4 patients were on a weaning regime (dosage  $< 5$ mg/kg) versus those on weight appropriate dosing (n=15) (5-10mg/kg). Dosing at discontinuation was variable. 4 patients were on a weaning regime. 11 patients remained on caffeine beyond the recommended gestation (34 weeks). 1 patient had documented apnoea in this period.

**Conclusion:** There is variance of prescribing practice of caffeine among patients in our unit comparable to the practise in others. The discontinuation of caffeine in our patients with chronic lung disease on respiratory support with no further apnoeas has been identified. Dosing optimisation for weight increase is required regularly.

1. NMH Protocol, Caffeine Citrate. Updated 2022

## IMPACT OF EARLY ONSET SEPSIS GUIDELINE ON ADMISSIONS TO A LEVEL II NEONATAL UNIT

N Canty<sup>1</sup>, C Caird<sup>1</sup>, R Fitzgerald<sup>1</sup>, D Huggard<sup>1</sup>

<sup>1</sup>Department of Paediatrics, University Hospital Waterford, Waterford, Ireland

**Introduction:** Many high risk new-borns require admission to a neonatal intensive care unit (NICU) for a septic work up.<sup>1,2</sup> However, variation in the initiation of antibiotic use exists between NICU units.

Therefore, a risk stratified early onset sepsis (EOS) algorithm was developed to streamline clinical decisions.

**Method:** Firstly, we wanted to recognise infant groups at risk and to evaluate adherence to a local clinical practice guideline (CPG). A retrospective review of all patients admitted to the NICU for partial septic work up from the year before (July 2020-2021), and the year after CPG implementation. Enrolled neonates were suspected of having sepsis based on clinical signs or underlying risk factors as identified as outlined within the guideline. Infants who were born in a level III unit who were transferred for ongoing care were excluded from the study.

**Results:** July 2020 to June 2021 there were a total of 1765 live births, 383 requiring admission, 150 (8.49%) required PSWU of those 32 were <35/40 gestation. July 2021 to June a total 1635 infants were born in UHW 339 requiring admission 118 (7.2%) met criteria for PSWU with 34 being <35/40 gestation.

**Conclusion:** Figures showed that the introduction of the CPG reduced our admission rate for PSWU by 1.3%, the majority of whom were term infants. Overall, it decreases unnecessary antibiotic use and prevented prolonged hospitalisations<sup>1,2</sup>. Additional benefits include lowering antimicrobial resistance, avoiding medication errors<sup>2,3</sup>. Furthermore separation can have a profound emotional impact on parents, and avoiding admission should also be considered a patient safety focus within the term infant cohort.

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### **OUTCOMES FOR TERM INFANTS BORN WITH NO HEART BEAT.**

AC Clarke<sup>1</sup>, JMcG McGinley<sup>2</sup>, P Corcoran<sup>3</sup>, P McKenna<sup>2</sup>, JF Murphy<sup>1</sup>

<sup>1</sup>Department of Neonatology, The National Maternity Hospital, Holles Street, Dublin, Ireland

<sup>2</sup>The National Women's and Infant Programme, Dublin, Ireland

<sup>3</sup>The National Perinatal Epidemiology Centre, Dublin, Ireland

**Introduction:** It has been generally accepted that term infants with no heart beat by age 10 mins old have a poor prognosis. However this is based on relatively old studies. There is little data on whether the advent of therapeutic hypothermia (TH) has altered the outcomes of this group of infants. The 8<sup>th</sup> edition of the NRP that discontinuation of resuscitation should be considered after approximately 20 mins of no heartbeat.

**Methods:** We have analysed all cases of therapeutic hypothermia due to neonatal encephalopathy in the 5 years 2016-2020.

In the study period there were 356 cases of neonatal encephalopathy (NE) among 301,940 births. NE rate 1.17 per 1000 births.

Of the 356 NE cases there were 58 (16.5%) infants born with no heart beat at birth.

All received intensive resuscitation at birth and subsequently commenced on TH.

#### **Results:**

34/63 (53.9%) had no heart rate at 1 min

12/63 (19.0%) had no heart rate at 1, 5 mins

17/63 (26.9%) had no heart rate at 1, 5, 10 min

Mortality for infants with no heart rate at 1 min but present by 5 min was 8.8%

Mortality for infants with no heart rate at 1 and 5 mins but present by 10 min was 50%

Mortality for infants with no heart rate at 1, 5 and 10 mins but present after 10 min was 70.5%

**Conclusion:** The findings in this study found that the prognosis remains poor for babies who have not established a heart rate by 10 mins of age. Babies with no heart rate at 10 mins old have a 70% mortality rate. On the other hand, babies who establish a heart rate after 10 mins have a 30% survival. These findings are in keeping with the NRP recommendations about continuing resuscitation for longer than 10 mins.

## **INCIDENTAL PRE-CELIAC AXIS DEXTROSE INFUSION OF THE AORTA AND IATROGENIC TRANSIENT HYPERINSULINISM IN A NEWBORN INFANT: A REFLECTIVE REVIEW**

**J Clarke**, RK Philip

<sup>1</sup>Department of Neonatology, University Maternity Hospital Limerick, Limerick, Ireland

Hypoglycaemia is one of the commonest biochemical derangements encountered in neonatal intensive care units (NICU), affecting up to 15% of otherwise healthy neonates and is a leading cause of preventable brain injury in infants.

This poster reflects on a case involving a rare cause of transient neonatal hyperinsulinism likely caused by the pre-celiac axis, aortic infusion of dextrose to treat hypoglycaemia. In doing so this poster will examine the literature relating to this hypothesis while also making reference to other infusions that could have adverse consequences when infused arterially using an umbilical arterial catheter.

We hypothesise that the patient's hypoglycaemia may have been exacerbated through the usage of an umbilical arterial catheter (UAC) for glucose infusion. The UAC, initially thought to have been an umbilical venous catheter (UVC), was placed at T8/T9 level. High concentrations of glucose infused to the pancreatic islets could have triggered the insulin response. Based on a review of the literature, Puri et al. in 1987 documented the increased risk of hypoglycaemia associated with a UAC that was in a 'high' position (T6 through to T11) when compared to a UAC in a 'low' position. Experimental work with lambs by Cowett et al. has supported the notion that infusion of glucose solutions above the celiac trunk leads to increased insulin release and hypoglycaemia.

This case offers learning opportunities for staff working in Neonatology. While often required, the use of umbilical catheters are not without their multiple potential hazards. The need for awareness of the suitability of drugs and infusions to be used through the umbilical catheters and a novel hypothesis of an iatrogenic hyperinsulinism are highlighted. Similarly, this poster provides a reminder of the importance of reviewing central line positions on x-ray prior to usage and having an awareness and knowledge of the relevant vascular anatomy.

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## MODES OF VENTILATION IN INTUBATED PRETERM INFANTS: A SYSTEMATIC REVIEW

R Coomey<sup>1,2</sup>, L Alabbasi<sup>1,2</sup>, A Daas<sup>1,2</sup>, A Fairhurst<sup>1,2</sup>, CS Lim<sup>1,2</sup>, S Marsh<sup>1,2</sup>, J Rice<sup>1,2</sup>, N Tuite<sup>1,2</sup>, D Mockler<sup>6</sup>, E

Isweisi<sup>1-3</sup>, G Semova<sup>1-3</sup>, A Branagan<sup>1,2,5</sup>, J Meehan<sup>1-3</sup>, E Roche<sup>1-3</sup>, EJ Molloy<sup>1-5</sup>

<sup>1</sup>Discipline of Paediatrics, School of Medicine, Trinity College Dublin, the University of Dublin, Dublin, Ireland

<sup>2</sup>Trinity Research in Childhood Centre, Trinity College Dublin, the University of Dublin, Dublin, Ireland

<sup>3</sup>Children's Health Ireland, CHI Tallaght, Dublin, Ireland

<sup>4</sup>Children's Health Ireland, CHI Crumlin, Dublin, Ireland

<sup>5</sup>The Coombe Women and Infants University Hospital, Dublin Midlands Hospital Group, Dublin, Ireland <sup>6</sup>Subject Librarian, Trinity College Dublin, the University of Dublin, Dublin, Ireland

**Introduction:** Invasive ventilation continues to be a necessity for preterm infants with respiratory distress syndrome. However, ventilation is associated with significant complications, including

bronchopulmonary dysplasia and neurodevelopmental issues. In addition, it is unclear which ventilator settings are optimal to reduce morbidity and mortality in preterm infants.

**Objectives:** To compare and assess the short- and long-term outcomes of the different modes of invasive ventilation in preterm infants.

**Method:** Following the Preferred Reporting Items for Systematic Review and Meta-analysis (PRISMA), a systematic review of Embase, Web of Science and Medline was performed.

**Result:** A total of 1407 studies from the search results were imported into Covidence, 138 were assessed as part of the full screening stage and 6 papers met the final inclusion criteria. Included studies investigated Volume-based Ventilation, Pressure-based Ventilation, Synchronised Intermittent Mandatory Ventilation (SIMV), and High-frequency Ventilation (HFV). Of these, statistically significant positive evidence was seen for Pressure Support Ventilation (PSV) with Volume-Guarantee (VG), and Volume-Controlled Ventilation (VCV).

**Conclusion:** No mode of ventilation found to be superior across all outcomes and birthweights. Overall, PSV with VG and VCV had significant benefits in short- and long-term outcomes. However, these findings mainly support the need for additional trials to provide high quality evidence to guide clinical practice.

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## THE FULL BLOOD COUNT IN SCREENING ASYMPTOMATIC INFANTS FOR EARLY ONSET SEPSIS: A RETROSPECTIVE CROSS-SECTIONAL STUDY

J Coveney<sup>1</sup>, J O'Loughlin<sup>2</sup>, F Ní Ainle<sup>3,4</sup>, R Drew<sup>5,6,7</sup>, M Boyle<sup>1,8,9</sup>

<sup>1</sup>Department of Neonatology, Rotunda Hospital, Dublin, Ireland

<sup>2</sup>Department of Laboratory Medicine, Rotunda Hospital, Dublin, Ireland

<sup>3</sup>Department of Haematology, Rotunda Hospital, Dublin, Ireland

<sup>4</sup>School of Medicine, UCD, Dublin, Ireland

<sup>5</sup>Clinical Innovation Unit, Rotunda Hospital, Dublin, Ireland

<sup>6</sup>Irish Meningitis and Sepsis Reference Laboratory, Children's Health Ireland at Temple Street, Dublin, Ireland

<sup>7</sup>Dept of Clinical Microbiology, RCSI, Dublin, Ireland

<sup>8</sup>Department of Neonatology, Children's Health Ireland at Temple Street, Dublin, Ireland

<sup>9</sup>Department of Paediatrics, RCSI, Dublin, Ireland

**Aims:** To determine the impact of full blood count (FBC) parameters, taken as part of a work up for earlyonset neonatal sepsis (EOS) on infants' subsequent management.

**Methods:** A single-centre, retrospective cross-sectional study was performed. Asymptomatic infants at increased risk of EOS born in a single Irish tertiary neonatal hospital over a 2-year period (2019-2020) were identified from laboratory records. Infants, born at  $\geq 34$  weeks gestation, asymptomatic at birth, undergoing a partial septic work-up and receiving prophylactic antibiotics due to increased risk of EOS were included. Symptomatic infants were excluded. The primary outcome measure was the frequency of FBC result impacting on the duration of antibiotic therapy. Two authors (JC, MB) independently reviewed the data and decided independently if/how the FBC result effected clinical care. A third author (RD) was available to adjudicate should the two authors disagree. Secondary outcome measures included the frequency of white cell and neutrophil counts outside of the reference range, frequency of incidental diagnoses and repeated phlebotomy.

**Results:** 16,726 live-born infants were delivered during the study period. 802 (4.8%) were included. Thirteen infants (1.6% of included infants) received a prolonged course of antibiotics due to suspicion for EOS. Two of these infants had elevated white cell counts and all had normal neutrophil counts. In no case did the FBC result influence the decision to prolong the antibiotic course. Seventy-five (9.6%) of the included infants had a white cell count outside of the reference range and 42 infants (5.6%) had a neutrophil count outside the normal reference range. Fifty-eight infants (6.5%) had polycythaemia, twenty-four infants (3.0%) had thrombocytopenia, five infants (0.6%) had anaemia and five infants (0.6%) had thrombocytosis.

**Conclusions:** In a cohort of 802 infants, asymptomatic at birth and at increased risk of EOS, the FBC result did not impact on the decision to prolong the course of antibiotics for suspicion of EOS.

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**THE USE OF LMA TO SECURE THE AIRWAY OF A NEONATE FOR**

**TRANSFER** | Elamin, AR Herrera

<sup>1</sup>Paediatrics, HSE, Kilkenny, Ireland

Cardiopulmonary resuscitation consists of airway and ventilation techniques and cardiac compression. There are multiple airway techniques to achieve adequate ventilation during resuscitation. During CPR, it should be a goal to achieve the most adequate airway management using the least invasive technique and most applicable with even the most inexperienced “rescuer”. In this case, an unexpected emergency led to the use of the LMA in a neonate to achieve adequate ventilation in a difficult airway.

**Case Report:** A 29 year old primagravida presented to the labour ward at 38 weeks gestation with a baby that had static growth for 2 weeks prior to delivery. Baby was born through SVD with a weak cry, poor tone and a heart rate below 100. PPV was started within the first minute of life. At about 3 minutes of life baby’s tone improved and heart rate picked up to 130 with oxygen saturation 85%. IPPV was continued for 2 minutes with baby’s statistics improving. At 12 minutes of life baby oxygen saturation dropped to 70-80%, CPAP was commenced. Baby was stabilized and transferred to SCBU. Baby was noted to have palate malformation, double cleft, small tongue small jaw. On auscultation there was poor air entry bilaterally. Multiple intubation attempts were unsuccessful. According to the modified Mallampatie classification, this case was classified as grade 4. LMA insertion was successful within the first attempt, midazolam was used for sedation, pressure support was started (Synchronized intermittent mandatory ventilation) and adequate ventilation was started.. Subsequent to adequate ventilatory support being achieved the patient was stable for transfer.

**Conclusion:** Intubation is generally a difficult skill to attain but even more difficult to maintain. In this case, where a patient had double palate malformation, double cleft, small tongue, and micrognathia LMA insertion was successful within the first attempt, where attempt at intubation was unsuccessful.

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**AN AUDIT OF COMPLIANCE TO THE TRANSIENT TACHYPNOEA OF THE NEWBORN POLICY AT A TERTIARY MATERNITY HOSPITAL****K Flinn**<sup>1</sup>, S Cahill<sup>1</sup>, J Hattingh<sup>1</sup>, JR Purna<sup>1</sup><sup>1</sup>Department of Neonatology, National Maternity Hospital, Dublin, Ireland

**AIM:** Respiratory distress secondary to retained fetal fluid is commonly known as transient tachypnoea of the newborn (TTN). Incidence is estimated at 4.0 – 5.7 per 1000 term births and is a common reason for admission to neonatal units (NNU).<sup>1</sup> Our aim was to assess the management of babies with TTN admitted to the NNU and improve compliance with the local guideline to minimise unnecessary investigations and treatment.

**METHODS:** The standard used for comparison was the current TTN guideline found on QPulse within the hospital. A proforma was used to collect baseline data for 20 patients. All babies admitted to the NNU with a diagnosis of TTN were included. Data was analysed using the PDSA model for quality improvement with a focus to improve compliance. Data was presented locally and appropriate amendments made to the guideline according to best practice.<sup>2</sup> A subsequent cycle was completed and the data was analysed to assess for changes in compliance.

**RESULTS:** N=20; patients admitted with TTN. 80% had risk factors for TTN. 50% required respiratory support and continuous positive airway pressure (CPAP) was the most frequently used modality. On admission, 30% underwent a chest x-ray (CXR), 35% had a capillary blood gas (CBG) taken and 10% had a septic work up (SWU) in the absence of documented septic risk factors. Reaudit was completed 10 weeks following the education session and guideline review. 35% underwent a CXR, 30% had a CBG and 30% had a SWU.

**CONCLUSION:** We could find no discernible change in clinician practice following the first cycle. One possible reason is the timing of the second audit cycle coincided with the annual Non Consultant Hospital Doctor (NCHD) changeover, although the updated guideline was taught at induction and available for consultation. Further education and audit cycles are required to encourage compliance with the updated guideline.

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## **NEONATAL THERAPEUTIC HYPOTHERMIA: TIME TO ACHIEVING THE OPTIMAL TEMPERATURE**

**A Flynn**<sup>1</sup>, J McGinley<sup>2</sup>, I Campillo<sup>3</sup>, P Filan<sup>4</sup>, JF Murphy<sup>1</sup>

<sup>1</sup>Neonatal Department, National Maternity Hospital, Holles Street, Dublin, Ireland

<sup>2</sup>National Women and Infants Health Programme, HSE, Dublin, Ireland

<sup>3</sup>National Perinatal Epidemiology Centre, University College Cork, Cork, Ireland

<sup>4</sup>Neonatal Department, Cork University Maternity Hospital, Cork, Ireland

**Aims:** To obtain the maximum benefit from therapeutic hypothermia (TH) the cooling process must be commenced by 6 hours of age. The target temperature is 33-34C, maintained for 72 hours, followed by gradual rewarming over 12 hours

This study was mounted to determine whether there were any differences between infants born in tertiary NICUs and local/regional units, in reaching this target temperature range

**Methods:** Data on TH cooling patterns for infants born in the tertiary NICUs and local/regional units was obtained from the 2019- 2020 Neonatal Therapeutic Hypothermia annual reports. We determined what proportion achieved the target temperature at 6 hours.

**Results:** There were 147 TH cases. 98 TH infants were born in the 4 tertiary NICUs and 49 TH infants were born across the other 15 peripheral/regional units.

The proportion of the TH infants reaching the target temperature by 6 hours in the tertiary NICUs was 92/98 (94%). The corresponding proportion for the TH infants born in the peripheral/regional units was 36/49 (73%). P<0.01.

**Conclusion:** A greater proportion of infants born in the tertiary centres achieved the optimal temperature at 6 hours of age compared with the TH infants born in the local/regional units. In tertiary NICUs the TH infants receive active cooling (CritiCool machine) from the start. Infants in the local/regional units receive passive cooling at the start and are switched to active cooling when they are being transported by the NNTP.



The findings would suggest that it would be preferable to commence active cooling from the start in the local/peripheral units. In the first phase, active cooling should be considered in the regional NICUs where there a consultant neonatologist roster.

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## **HYPERKALAEMIA MANAGEMENT IN NEONATES: A SYSTEMATIC REVIEW**

**MS Furqan**<sup>1</sup>, N Laher, M Chidiac, Z Weber Hall, M Salama, G Ola, AD Vimal Kumar, A O'Reilly, E Isweisi<sup>1,2,3</sup>, G

Semova<sup>1,2,3</sup>, A Branagan<sup>1,2,4</sup>, J Meehan<sup>1,2,3</sup>, E Roche<sup>1,2,3</sup>, EJ Molloy<sup>1,2,3</sup>

<sup>1</sup>Discipline of Paediatrics, School of Medicine, Trinity College Dublin, Dublin, Ireland

<sup>2</sup>Trinity Research in Childhood Centre, Trinity College Dublin, Dublin, Ireland

**Background and Aims:** Hyperkalaemia is more common in extremely low birth weight infants and is associated with high mortality in the neonatal population. We aimed to systematically review literature on the treatment of hyperkalaemia in neonates.

**Methods:** A peer reviewed search was conducted using Embase, Pubmed and Cochrane databases according to PRISMA guidelines. The search identified literature on therapeutic interventions for the treatment of hyperkalaemia in newborns ( $\leq 4$  weeks of life). Data was extracted on study design, sample size, patient demographics, primary treatments and adjunctive treatments, and outcomes.

**Results:** A total of 519 titles were identified with 67 texts for full paper review. A final 10 articles met our inclusion criteria. Primary treatments that were identified included salbutamol, glucose/insulin infusion, potassium exchange resins and peritoneal dialysis. Adjunctive treatments included sodium bicarbonate and calcium gluconate. All 5 treatment methods identified showed efficacy in the reduction of serum potassium levels in neonates. Nebulized salbutamol demonstrated superior efficacy and safety in comparison to other treatments in multiple studies. Hyperkalaemia was also associated with congenital endocrine disorders, including congenital adrenal hyperplasia and pseudohypoaldosteronism, was also reported. Treatment involved the

use of hydrocortisone and fludrocortisone in conjunction with standard treatment methods of hyperkalaemia in neonates.

**Conclusion:** Improved insight into the aetiology of hyperkalaemia would assist in planning optimal management to improve outcomes.

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### **EXPLORING PRACTICE IN RE-ESTABLISHING FEEDS POST-NEC AT A SURVEY OF CURRENT PRACTICE IN IRISH NEONATAL UNITS**

**RG Gowan**<sup>1,2</sup>, RMC McCarthy<sup>1,2</sup>, NA Aminudin<sup>2</sup>

<sup>1</sup>Clinical Nutrition and Dietetics, National Maternity Hospital, Dublin, Ireland

<sup>2</sup>Neonatology, National Maternity Hospital, Dublin, Ireland

Necrotising Enterocolitis (NEC) is a serious, potentially lethal bowel disease (in very low birth weight (<1500g preterm neonates). The rate of severe NEC requiring surgery remains high (20-50%). As part of NEC management, enteral feeds are stopped for a period of time. Re-establishing feeds afterwards can be challenging. Nutritional recommendations to prevent NEC are well established, albeit approach to re-feeding post-NEC differs.

A prospective survey among neonatologists, surgeons and dietitians was conducted to explore the approach in re-establishment of enteral feeding after NEC. Following ethical approval, an online questionnaire was sent to dietitians, surgeons and neonatologists (n= 53). All responses were anonymised and reviewed. Data from 6 questions were coded. Data from one question was analysed using 'text analysis'.

19 clinicians (77% dietitians, 40% surgeons, 26% neonatologists) completed the survey. 52% (n=10) stated

“severity of the NEC and the clinical condition” influenced their decision on restarting feeds post-medical NEC, 63% (n=12) stated “upper GI losses and stoma function” influences their decision restarting feeds post-surgical NEC. 63% would advance the rate of feeds by 15-20mls /kg/d. 50% (n=9) chose donor breast milk as a substitute to maternal breast milk post-NEC whilst 32% (n=6) chose formula. 57% (n=11) would add breast milk fortifier (BMF) at 80-120mls /kg/d, 15% (n=3) once feeds were established and 15% (n=3) would never add BMF to feeds. “Feed tolerance” and “clinical condition” influenced the responders when advancing enteral feeds.

We demonstrated variations in practice in refeeding post-NEC. The difference in practice when restarting feeds remains multifactorial. A higher responder rate would make results more generalisable. As the first survey of this kind to be conducted in Ireland, the results provide useful information in re-establishing feeds post-NEC and potentially support future practice.

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### **NOT JUST A BLOCKED NOSE : A CASE OF PYRIFORM APERTURE STENOSIS**

**H Grace**<sup>1</sup>, K Cunningham<sup>1,2</sup>, M Colreavy<sup>3</sup>

<sup>1</sup>Department of Neonatology, CHI at Temple Street, Dublin, Ireland

<sup>2</sup>Department of Neonatology, The Rotunda, Dublin, Ireland

<sup>3</sup>ENT Department, CHI at Temple Street, Dublin, Ireland

**Aims** : Pyriform Aperture Stenosis (PAS) is a rare form of birth defect first described in 1989 <sup>(1)</sup> affecting the nasal opening which results from overgrowth of the maxilla. This is particularly important during the first few months of life while neonates are obligatory nose breathers.

Pyriform Aperture stenosis may be associated with a single central incisor and abnormalities of the pituitary gland and so these should be kept in mind during initial work-up.

**Methods :** We present the case of a neonate initially suspected to have choanal atresia transferred to CHI at Temple Street. JL presented with worsening respiratory distress following an otherwise normal delivery, decline in ability to feed and difficulty passing a nasogastric tube. Only a 4Fr nasogastric tube could be passed through the left nare with no such issues noted on the right side. Following review by the ENT team and CT of the nasal sinuses a diagnosis of Pyriform Aperture Stenosis was made. In this case, the neonate in question continued to have episodes of respiratory distress and required surgical intervention.

**Results :** Shortly after diagnosis and having had numerous episodes of respiratory distress, JL was brought to theatre where a partial midface degloving and drill out was performed. An NPA was placed and managed on the ward with frequent checks, humidification and saline nebulisation until the point at which it could be removed. The post-operative course was not without complications, however, and the NPA had to be replaced on several occasions due to respiratory distress. With sufficient healing, the NPA was removed successfully, allowing him to be discharged home safely.

**Conclusions :** Pyriform Aperture Stenosis is a rare but important cause of respiratory distress and feeding difficulty that should not be forgotten in the neonatal period.

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## TERMINOLOGY, DEFINITIONS, AND ELIGIBILITY CRITERIA IN TRIALS FOR NEONATAL ENCEPHALOPATHY: A SYSTEMATIC REVIEW

T Hurley<sup>1</sup>, A Branagan<sup>1,2</sup>, F Quirke<sup>3,4</sup>, E Finucane<sup>3,4</sup>, G King<sup>1</sup>, D Devane<sup>3,4</sup>, E Molloy<sup>1,2</sup>

<sup>1</sup>Department of Paediatrics and Child Health, Trinity College Dublin, Dublin, Ireland

<sup>2</sup>Department of Neonatology, Coombe Women and Infant's University Hospital, Dublin, Ireland

<sup>3</sup>Health Research Board – Trials Methodology Research Network (HRB-TMRN), University of Galway, Galway, Ireland

<sup>4</sup>School of Nursing and Midwifery, University of Galway, Galway, Ireland

**Background:** Appropriate terminology and definitions of neonatal encephalopathy (NE), hypoxic ischaemic encephalopathy (HIE), and perinatal asphyxia (PA) have been debated for 20 years. Many have advocated for NE to be used as it does not assume an aetiology, however this is not universally accepted. There is no generally accepted case definition criteria for NE/HIE/PA and eligibility criteria used in trials of therapeutic hypothermia (TH) are frequently employed. However, variation in eligibility criteria reflects variation in cohorts recruited which may partly explain differences in control group mortality. Consistent terminology and case definition would clarify the clinical entity, standardise participant criteria in future trials, and provide greater generalisability of trial results.

**Methods:** A comprehensive search was run for randomized controlled trials of interventions for the treatment of NE/HIE/PA. Any definition or criteria for NE or HIE were eligible. Outcomes included were a description of the terminology, definitions, and participant eligibility criteria. 2 reviewers independently screened the results and extracted the data. The qualitative results were synthesised in a narrative summary.

**Results:** 62 were included in the qualitative synthesis. HIE was the most frequently used descriptive term. Several studies used different terms interchangeably. Evidence of perinatal asphyxia most frequently relied on Apgar scores and evidence of acidosis. However, there was

significant variation between studies in their application. Evidence of neurologic dysfunction most frequently required reduced level of consciousness, reduced tone, and abnormal reflexes but seizures were less frequently required. The threshold age for exclusion varied frequently between studies as did the exclusion of participants with mild NE/HIE.

**Discussion:** We identified variation in terminology, major points of agreement between studies in the requirement eligibility criteria for trial participants, and major points of variance between studies. These results will inform a consensus process for developing a definition and diagnostic criteria.

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### **BIOMARKERS TO PREDICT OUTCOME IN NEONATAL ENCEPHALOPATHY: A SYSTEMATIC REVIEW AND META-ANALYSIS**

**T Hurley**<sup>1</sup>, M O'Dea<sup>1,2</sup>, A Branagan<sup>1,2</sup>, P Stewart<sup>1,2</sup>, S Tamgumus<sup>2</sup>, K Kyng<sup>3</sup>, D Mockler<sup>4</sup>, E Molloy<sup>1,2,5</sup>

<sup>1</sup>Department of Paediatrics and Child Health, Trinity College Dublin, Dublin, Ireland

<sup>2</sup>Department of Neonatology, Coombe Women and Infant's University Hospital, Dublin, Ireland

<sup>3</sup>Department of Paediatrics, Aarhus University, Aarhus, Denmark

<sup>4</sup>Medical Library, Trinity College Dublin, Dublin, Ireland

<sup>5</sup>Department of Neonatology, Children's Hospital Ireland at Crumlin and Tallaght, Dublin, Ireland

**Introduction:** Neonatal encephalopathy (NE) biomarkers are important for early diagnosis, guiding prognosis, improved understanding of the disease pathophysiology, and identification of potential therapeutic agents. Many biomarkers have been investigated in recent years as potential prognostic markers, however none are used in routine clinical practice. We examined current evidence of serum biomarkers to predict short- and long-term outcome in neonatal encephalopathy.

**Methods:** The review was prospectively registered with PROSPERO. EMBASE, PubMed, World of Science and the Cochrane Library databases were searched for studies that reported prognostic serum biomarkers in participants with NE. Participants were dichotomised by groups into normal or adverse outcome, defined as death or abnormal neurodevelopmental outcome at >12 months of age. 2 reviewers independently completed the screening and data extraction. Risk of bias was completed using QUIPS tool. The mean difference in serum biomarkers between groups was calculated using a random effects model.

**Results :** Literature search provided 3046 results. 98 studies of over 20 different biomarkers were eligible for qualitative synthesis, however only 41 studies could be included due to variation in biomarker measurement and reporting. Meta-analyses for serum interleukin (IL)-1 $\beta$ , IL-6, IL-8, IL-10, NSE, S100, S100B, and TNF- $\alpha$  to predict adverse outcome were completed. Serum IL-6, IL-8, NSE, and TNF- $\alpha$  and were all lower in participants with normal long-term outcome. Serum IL-6 and lactate were lower in participants with normal short-term outcome. However, there was substantial variability in results between included studies.

**Discussion:** Several biomarkers provide promising prognostic value in NE. Lower IL-6, NSE, and TNF- $\alpha$  were associated with normal long-term outcome in patients with NE. This relationship requires further evaluation. Many studies could not be included a MA and greater uniformity in biomarker outcome reporting would allow improved evidence synthesis.



**NUCLEATED RED BLOOD CELLS AND NEONATAL OUTCOMES: EVIDENCE-BASED PROJECT**

S Azam<sup>1</sup>, C Doherty<sup>1</sup>, M Freeney<sup>1</sup>, NS Ishak<sup>1</sup>, XL Ong<sup>1</sup>, A Murphy<sup>1</sup>, AAA Razak<sup>1</sup>, E Warren<sup>1</sup>, E Isweisi<sup>1-3</sup>, G

Semova<sup>1-3</sup>, A Branagan<sup>1,2,5</sup>, D Mockler<sup>6</sup>, J Meehan<sup>1-3</sup>, E Roche<sup>1-3</sup>, E Molloy<sup>1-5</sup>

<sup>1</sup>Discipline of Paediatrics, School of Medicine, Trinity College Dublin, Dublin, Ireland

<sup>2</sup>Trinity Research in Childhood Centre, Trinity College Dublin, Dublin, Ireland

<sup>3</sup>Children's Health Ireland at Tallaght, Dublin, Ireland

<sup>4</sup>Children's Health Ireland at Crumlin, Dublin, Ireland

<sup>5</sup>The Coombe Women and Infant University Hospital, Dublin, Ireland

<sup>6</sup>John Stearne Medical Library, Trinity Centre for Health Sciences, Trinity College Dublin, Dublin, Ireland

**Aims:** Perinatal asphyxia is a major cause of neonatal mortality and morbidity, with several complications in surviving infants including neurological disabilities and multiorgan dysfunction.<sup>1</sup> The aim of this systematic review is to explore the prognostic value of elevated nucleated red blood cells (nRBCs) released prematurely from the bone marrow in response to hypoxia and their role in predicting the neonatal outcomes of term infants (>37 weeks gestation).

**Methods:** A systematic review was conducted according to PRISMA guidelines and to survey articles in the literature, an experienced medical librarian developed a syntax for the following 5 databases: MEDLINE, EMBASE, Web of Science, Cochrane Library and ClinicalTrials.gov.

**Results:** A total of 805 studies were retrieved and underwent title and abstract screening in duplicate. Exclusion criteria were: (1) preterm neonates (<37 weeks gestation), (2) studies which identified nRBCs as an outcome of uteroplacental insufficiency (pre-eclampsia, diabetes, obesity) (3) studies using nRBCs as a biomarker for measuring the effectiveness of another intervention (such as, doppler).

The full-text of 67 articles were assessed for their eligibility, and 16 were ultimately deemed relevant to this review. This systematic review found that elevated nRBCs correspond with: 1) Poor

perinatal outcomes; 2) Hypoxic-ischaemic encephalopathy (HIE); 3) Neonatal mortality; and 4) Neonatal seizures and developmental complications.

**Conclusions:** Elevated nRBC counts may provide short term prognostic information regarding neonatal outcome. However, they are only one part of the data set and must be considered within the broader clinical context comprising clinical examination findings and other radiological or laboratory results. Further detailed research is needed in larger cohorts, to evaluate predictive cutoff values for neurodevelopmental outcome.

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## DEVELOPING A UNIFIED PROTOCOL FOR NEONATAL PULSE OXIMETRY SCREENING IN

IRELAND ST Kelleher<sup>1</sup>, O Franklin<sup>1</sup>, MA Boyle<sup>2</sup>

<sup>1</sup>Department of Cardiology, CHI at Crumlin, Dublin, Ireland

<sup>2</sup>Department of Neonatology, Rotunda Hospital, Dublin, Ireland

**Aims:** Pulse oximetry screening (POS) is a simple, sensitive, and specific method of detecting ductdependent and cyanotic congenital heart disease (CHD)[1,2]. Antenatal screening for CHD is not universally available to Irish mothers[3], making its role even more crucial. Despite, universal adoption of POS there is variation in practice between neonatal units. Our aim was to develop a unified national screening protocol.

**Methods:** Protocol development consisted of; (a) a national survey of 19 Irish maternity units using written and phone interviews, (b) an internal audit of 675 new-born infants born in June 2020 in the Rotunda Hospital to assess acceptability and compliance, (c) an extensive literature search and review of international practice. The protocol was presented to the Neonatology Clinical Advisory Group in September 2022.

## Neonatal

**Results:** The national survey demonstrated that all 19 units had adopted POS between 2006 and 2014. Protocols differed with regard to; oximetry cut-off that was considered normal, whether pre- and postductal saturations were measured routinely, and test timing with majority recommending day of discharge testing (58%). The internal review noted excellent compliance with performance of the test (99.1%) and no parental refusals. Our literature review identified that screening before 6 hours of life is associated with high false positive rates, while late screening is associated with infants presenting symptomatically prior to screening. Use of both pre- and post-ductal measurements improved sensitivity in individual studies but not in meta-analysis. High oximetry cut-offs may lead to false positives.

**Conclusion:** Our recommendations are; (a) a screen is positive if the O<sub>2</sub> saturations <96%; (b) post-ductal saturation must be performed on all babies (either foot), a pre-ductal saturation (right arm) is recommended but not essential; (c) the test should be performed at 24 hours, and not before 6 hours in case of early discharge (d) a repeat screen after 1 hour is recommended in borderline cases.

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3. Lynch Á, Ng L, Lawlor P, Lavelle M, Gardner F, Breatnach C, McMahon CJ, Franklin O (2019) Cyanotic Congenital Heart Disease Modes of Presentation and Prenatal Detection. *Ir Med J* 112 (10):1019
4. Brown S, Liyanage S, Mikrou P, Singh A, Ewer AK Newborn pulse oximetry screening in the UK: a 2020 survey. *The Lancet*. doi:10.1016/S0140-6736(20)31959-0
5. Oddie S, Stenson B, Wyllie J, Ewer AK (2019) UK consultation on pulse oximetry screening for critical congenital heart defects in newborns. *The Lancet* 394 (10193):103-104. doi:https://doi.org/10.1016/S0140-6736(19)31515-6

**SEX AND GESTATIONAL AGE IMPACT PRO- AND ANTI-INFLAMMATORY RESPONSES AMONG PRETERM NEONATES**

M McGovern<sup>1</sup>, R Finnegan<sup>1</sup>, J Kelleher<sup>2</sup>, A El-Khuffash<sup>3</sup>, J Murphy<sup>4</sup>, **LA Kelly**<sup>1, 6</sup>, AM Melo<sup>1, 6</sup>, C Greene<sup>5</sup>, EJ Molloy<sup>1-3,6</sup>

<sup>1</sup>Paediatrics, Trinity College Dublin, Dublin, Ireland

<sup>2</sup>Neonatology, Coombe Women and Childrens University Hospital, Dublin, Ireland

<sup>3</sup>Neonatology, The Rotunda, Dublin, Ireland

<sup>4</sup>Neonatology, The National Maternity Hospital, Holles St, Dublin, Ireland

<sup>5</sup>Clinical Microbiology, The Royal College of Surgeons in Ireland, Dublin, Ireland

<sup>6</sup>Trinity Translational Medicine Institute (TTMI) & Trinity Research in Child, Trinity College Dublin, Dublin, Ireland

Preterm neonatal immune cells have altered cytokine responses compared to term neonates and adults. Also, immune responses differ between male and female preterm neonates. We aimed to examine genes involved in inflammasome activation including a comprehensive array of cytokines in response to inflammatory stimuli and sex steroid hormones in male and female term and preterm neonates.

Preterm and term infants were recruited from The Coombe Women and Childrens Maternity Hospital, Dublin with written informed consent. Whole blood was incubated at 37°C for 1 hr with Vehicle, LPS, and or with Estradiol (E2) or Progesterone (P). Serum biomarkers were analysed on a plate from Meso Scale Diagnostics. 72 newborn infants were enrolled including 40 preterm neonates (18 female, 22 male) with a mean gestation at birth (mean + SD) of 28.69 +2 weeks and corrected gestation at the time of sampling was 31.85 +2.5 weeks. The mean birth weight of preterm neonates was 1089 +304.9 grams.

IL-1RA and IL-10 were higher in preterm males, compared to preterm females, in samples treated with P, suggesting some effect of sex hormones on serum concentrations of anti-inflammatory cytokines. IL-1 $\beta$  however was affected by female sex hormones and notably the effect of E2 was greatest in the preterm population and female preterm neonates appeared more responsive to the anti-inflammatory effect of progesterone. Lastly, male preterm neonates had higher IFN- $\gamma$ , IL-8 and TNF- $\alpha$  levels compared to preterm females when treated with LPS and P combined. The inflammasome genes NLRP3, AIM2, ASC and IL-1 $\beta$  showed no difference in fold change gene expression between term and preterm neonates of either sex.

Sex hormones have an important role in modulating neonatal immune responses and our findings add to the existing literature that suggests an important role for hormones in the female immune advantage. 45

### **GENDER AND INFLAMMATION IN NEONATAL ENCEPHALOPATHY**

**LA Kelly**<sup>1,5</sup>, A Branagan<sup>1,5</sup>, MI O'Dea<sup>1</sup>, J Isaza-Correa<sup>1,5</sup>, A El-Khuffash<sup>2</sup>, M White<sup>3</sup>, J Miletin<sup>3</sup>, C Greene<sup>4</sup>, EJ Molloy<sup>1,3,5</sup>

<sup>1</sup>Paediatrics, Trinity College Dublin, Dublin, Ireland

<sup>2</sup>Neonatology, The Rotunda, Dublin, Ireland

<sup>3</sup>Neonatology, The Coombe Women and Infants University Hospital, Dublin, Ireland

<sup>4</sup>Clinical Microbiology, The Royal College of Surgeons in Ireland, Dublin, Ireland

<sup>5</sup>Trinity Translational Medicine Institute (TTMI) & Trinity Research in Childhood Centre (TRICC), Trinity College Dublin, Dublin, Ireland,

Neonatal encephalopathy (NE) is a devastating condition of the term newborn infant. There is a well recognised difference in incidence and outcome of NE according to gender with a pronounced male disadvantage. The aim of this work was to delineate the immune responses between male and female infants with neonatal encephalopathy.

Blood samples were taken in the first week of life from infants with NE and treated, with either, LPS, Estradiol (E2) or E2+LPS combined. The sample was split between flow cytometry, RT-PCR and serum.

Activation of neutrophils and monocytes was evaluated by expression CD11b and Toll-like receptor-4 (TLR4). The NLRP3 inflammasome and X-linked genes were analysed between genders. Multiplex ELISA was used for cytokine analysis. Statistical analysis was carried out using GraphPad Prism V9.4.

Eighteen infants with NE were recruited ( 11 male and 7 female). On analysis of the inflammasome related proteins we found there was significant differences in IL-2 ( $p=0.04$ ), IL-8 ( $p=0.004$ ) and IL-10 ( $p=0.034$ ) production between male and female at baseline and in the expression of IL-10 upon endotoxin stimulation ( $p=0.06$ ). No differences were found between the treatments used or across genders in our flow cytometry. Both males and females have increased NLRP3 and IL1 $\beta$  following endotoxin stimulation and there are no significant differences at baseline between the sexes. ASC

gene expression and the X-linked genes, IRAK and IKK $\gamma$  were not significantly different between sexes.

We found no significant differences between male and female cohorts in terms of Inflammasome regulation and cell analysis. Downstream protein expression differed between males and females. Stimulation with the female hormone Estradiol made no difference to the responses and therefore we propose that other mechanisms such as their genetic phenotype or innate immune profiles may drive these gender difference in NE. Further mechanistic investigations are necessary alongside correlation with clinical data.

**ABNORMAL LIVER FUNCTION AND IMPROVED SURVIVAL IN A CHILD WITH SPLICE MUTATION TARP SYNDROME****M Lane**<sup>1</sup>, N Allen<sup>1,2</sup>, J Leitshwiti<sup>2</sup><sup>1</sup>Paediatrics, NUIG, Galway, Ireland<sup>2</sup>Paediatrics, GUH, Galway, Ireland

**Aims:** TARP (talipes equinovarus, atrial septal defect, Robin sequence, persistent left superior vena cava) syndrome is an X-linked disorder secondary to RBM10 gene mutations<sup>(1)</sup>. We present the case of a toddler diagnosed with TARP syndrome due to a splicing mutation c.2295+1G>A in the *RBM10* gene. We aimed to discuss the novel features seen in our case, the prevalence of previously described major, minor and additional disease characteristics and quantifying previously reported mutation types comparing with survival beyond 1.5 yrs.

**Methods:** We quantified the prevalence of minor characteristics (6 subcategories), major characteristics (8 subcategories) and additional features (6 subcategories) stratifying by survival time subgroups (< or >18 months) including our patient. We compared mutation types (splice site, frameshift, nonsense, missense and in-frame deletion) previously published and survival time (< or >18 months).

**Results:** Literature review revealed 26 previously described cases. 17/26 patients died before 18 months with 9/26 surviving beyond this.

Previously unpublished features observed in our patient were high alpha-fetoprotein, hyperbilirubinemia, and thrombocytopenia during the neonatal period.

All patients had severe cognitive delay, severe motor delay, brain malformations, neurological symptoms, airway/ pulmonary abnormalities, failure to thrive and heterogenous dysmorphic features. Other characteristics varied considerably between cases.

All 3 previous cases due to splicing mutation (incl. our case) survived beyond 18 months. In contrast, all cases due to nonsense mutation were fatal before 18 months.

**Conclusion:** TARP Syndrome is a rare, heterogenic disease. There is large interpatient variability leading to diagnostic and prognostic uncertainty. It had been considered fatal in the neonatal period and discussed as such in the literature<sup>(1)</sup>. In many recent cases, there have been improved survival rates and almost 1/3<sup>rd</sup> of cases have lived beyond 1.5 years<sup>(2)</sup>. Comparison of underlying genetic causes may indicate milder disease phenotype in splice site, missense and in-frame deletions.

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## DELAYED ONSET HYPOGLYCAEMIA IN PRETERM INFANTS

CL Loomes , DC Corcoran

<sup>1</sup>Neonatal Department , Rotunda Hospital , Dublin, Ireland

<sup>2</sup>Neonatal Department , Rotunda Hospital , Dublin, Ireland

**Background and Aim:** Preterm infants are at risk of hypoglycaemia, likely due to immature counterregulatory hormone systems and poor nutritional reserves <sup>1</sup>. Hyperinsulinaemic hypoglycaemia in infants >

35 weeks gestation is associated with longer hospital admissions, an increased cost of care and neurodevelopmental delay. However, adverse effects of hypoglycaemia in preterm infants remain unclear. Anecdotally we have noted an increase in delayed onset hypoglycaemia amongst our preterm population at the Rotunda Hospital. There is a paucity of information in the literature analysing this group. The aim of this retrospective study is to review the biochemical data, antenatal and postnatal factors effecting preterm infants with recurrent hypoglycaemia.

**Methods:**



- 1) 21 Infants born at less than 33 weeks gestation requiring a hypoglycaemic work up between 2018 and 2021 were identified using HIPE in a tertiary maternity centre.
- 2) Information relating to infant demographics, anthropology and antenatal/ postnatal factors were collected and analysed.

**Results:** Birth gestations ranged from 23+3 to 32+3. The median birth centile was the 47th ( IQR 1475). 4/21(19%) had birth weights <10<sup>th</sup> centile. First episode of hypoglycaemia on average occurred on DOL 20. In 4/21 cases glucose levels were > 3mmol/L at the time of hypoglycaemia work up. Insulin levels were undetectable in 2/20 cases. The average insulin level was 79pmol/L (IQR 48.78- 102pmol/L). B-Hydroxybutyrate <0.1 mmol/l in all cases. 13/21 (54.17%) required consultation with endocrinology. 1/21 required treatment for hyperinsulinism.

**Conclusion:** Preterm infants are at risk of developing delayed onset non-ketotic hypoglycaemia with inappropriate insulin secretion. Growth restricted infants did not appear to be disproportionately affected. Often these infants have challenging clinical courses, exacerbated by persistent issues with glycaemic control, and protracted inpatient stays. Rarely do these infants require treatment for hyperinsulinism. In the majority of cases this phenomenon was self-limiting managed with feeding regimens, and infants did not require long term endocrinology input.

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#### **IMPACT OF IMPLEMENTATION EARLY ONSET SEPSIS CALCULATOR ON SCBU ADMISSION M Mathews<sup>1</sup>, A Kalim<sup>2</sup>**

<sup>1</sup>SCBU, Maternity Department, Wexford General Hospital, Wexford, Ireland

<sup>2</sup>Consultant Paediatrician, Wexford General Hospital, Wexford, Ireland

**Aim:** Empiric antibiotic therapy can result into separation of a mother and her baby, increased admission to the Special Care Baby Unit (SCBU), difficulty in establishing the breast feeding and overall increased financial and staff work load implications for the institute.<sup>1, 2</sup>

The neonatal EOS calculator<sup>3</sup> was introduced to our SCBU in early 2021, to improve the management of these sick neonates. Guidelines<sup>4</sup> were implemented to use it in the relevant group of neonates, manage them accordingly to avoid unnecessary use of empirical antibiotics.

The aim of this audit was to assess if the introduction of EOS calculator guideline has made any difference in number of neonates requiring admission to the SCBU for IV antibiotics.

**Methods:** The data was collected, for total number of neonates born and the number of admissions to SCBU for IV antibiotics in the year 2020 and 2021. The medical notes were reviewed of the neonates who required IV antibiotics to assess them for their indications. Antibiotics were commenced in the year 2020 as per NICE guidelines for neonatal sepsis and in the year 2021 they were commenced according to the EOS calculator guidelines.

**Results:** In the year 2020 39% (124/319) of all the admitted babies required IV antibiotics, following the NICE guidelines. In the year 2021, with the implementation of EOS calculator guidelines, the number of babies who required IV antibiotics, fell down to 22% (69/315). Prematurity was the most common indication for IV antibiotics. 3% (10/319) babies were born premature in 2020 as compared to 9% (28/315) in 2021.

**Conclusion:** Although, there was significant increase in number of the neonates born premature from 3% to 9%, overall antibiotic prescription rate fell down significantly from 39% to 22%, resulting in positive patient, staff and organisational impacts.

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### AN UNUSUAL CASE OF NEONATAL JAUNDICE

D McGlacken-Byrne<sup>1,2</sup>, D Hardiman<sup>1,2</sup>, Y Ibrahim<sup>1</sup>, M Ibrahim<sup>1,2</sup>

<sup>1</sup>Department of Neonatology, University Maternity Hospital Limerick, Limerick, Ireland

<sup>2</sup>Faculty of Paediatrics, Royal College of Physicians in Ireland, Dublin, Ireland

**Case:** A boy was born at 32+4 weeks' gestation by emergent C-section, due to ultrasonographic evidence of a retroplacental haematoma and a nonreassuring cardiotocograph. The antenatal course was unremarkable. The baby was vigorous at birth and admitted to intensive care on noninvasive ventilation. Clinical examination was normal. At one hour of life, a blood gas showed bilirubin of 104, confirmed on a serum sample, above the exchange transfusion threshold. The hyperbilirubinaemia was unconjugated and the baby was DCT-negative. His blood type was AB-positive, while his mother's was A-positive. Five-fold phototherapy was commenced. The serum bilirubin fell swiftly in response; as such, exchange transfusion was withheld. His direct Coombs test (DCT), inflammatory markers, and blood culture were negative. The baby had a normal haemoglobin and reticulocyte count throughout admission. Liver function tests were normal. Haemoglobin electrophoresis, pyruvate kinase, and glucose-6-phosphate dehydrogenase screens were negative. An abdominal radiograph was normal. Of note, the infant passed a black, tarry, malodorous stool on day one of life, suggestive of melaena. Thereafter, his clinical course was stable, with no further jaundice. He transitioned to room air at one week of life, established feeds, and was discharged at 37 weeks' corrected age.

**Discussion:** Sepsis, immune-mediated haemolysis, and inherited red cell defects are important differential diagnoses of unconjugated hyperbilirubinaemia on the first day of life. In this case, the baby developed marked hyperbilirubinaemia within one hour of delivery, nearly necessitating exchange transfusion. The aetiology remains uncertain. The antenatally detected retroplacental haematoma may have contributed. In addition, the postnatal episode of melaena suggested upper gastrointestinal ulceration. Stress-induced gastrointestinal bleeding has been reported in the neonatal period, pursuant to prolonged labour or birth asphyxia.<sup>1</sup> These represent rare but important considerations in the investigation of jaundice in the first hours of life.

1 – Joshi, A., Shrestha, P. S., Dangol, S., Shrestha, N. C., Poudyal, P., & Shrestha A, A. (2016). Stress Induced Gastric Ulcers: Presenting as Massive Rectal Bleeding in a Newborn. Kathmandu University medical journal (KUMJ), 14(54), 186–189.

**DIGITAL CLUBBING AS A PRESENTING SIGN OF UNILATERAL BRONCHIECTASIS IN A 6-YEAR-OLD****D McGlacken-Byrne<sup>1</sup>**, D Butler<sup>1</sup>, O Ahmareen<sup>1</sup>, F Ringholz<sup>1</sup><sup>1</sup>Respiratory Department, Children's Health Ireland at Crumlin, Dublin, Ireland

**Case:** A six-year-old boy was brought to his GP after difficulty gripping pencils was reported at school. His GP noted bilateral finger clubbing and right-sided crepitations, and referred to the paediatric emergency department. There were no respiratory, gastrointestinal, skin or joint symptoms, and no weight loss or night sweats. There was no history of gagging or choking at any point. There was no past medical history and no significant family history. On examination, he was pale with bronchial breathing and crepitations over the right chest. The fingers and toes were severely clubbed bilaterally. Full blood count revealed microcytic anaemia (Hb 92g/L), thrombocytosis (833 x 10<sup>9</sup>/L) and neutrophilia (WCC 29.9 x10<sup>9</sup>/L, ANC 21.6 x 10<sup>9</sup>/L). CRP was 67. Urate, lactate dehydrogenase, liver function, renal profile, blood film and blood cultures were normal. Chest radiograph showed widespread unilateral consolidation; computed tomography showed severe, bronchiectasis throughout all right lung lobes. No foreign body or mass was seen. Bronchoalveolar lavage yielded growth of two agents associated with periodontal infection, *Prevotella intermedia* and *Parvimonas micra*. TB culture, viral serology, ciliary brushings, and sweat chloride concentration were normal. The child was discharged on a long-term course of oral coamoxiclav. Repeat imaging several months later showed significant interval improvement.

**Discussion:** For two reasons, this case of severe, unilateral bronchiectasis is striking. Firstly, there were no respiratory or systemic symptoms, despite widespread bronchiectatic change. Clubbing was the presenting feature. Secondly, distribution was entirely unilateral, with one normal lung. Despite no suggestive history, an overlooked episode of aspiration earlier in childhood therefore remains a possibility.

**Conclusion:** West described clubbing in 1897 as "one of those phenomena with which we are all so familiar that we appear to know more about it than we really do".<sup>1</sup> 125 years later, clubbing remains an enigmatic yet occasionally crucial portent of underlying serious disease.

1 – West S. Two cases of clubbing of the fingers developing within a fortnight and four weeks, respectively. *Trans Clin Soc London*.

1897;30:60.

**A REVIEW OF NEONATAL ADMISSIONS TO THE PAEDIATRIC INTENSIVE CARE UNIT IN CHI @ TEMPLE STREET 2019-2020****RB McGrath<sup>1</sup>**, K Carson<sup>1</sup>

<sup>1</sup>Paediatric Intensive Care Unit, Children's Health Ireland @ Temple Street, Dublin 1, Ireland

**Background:** According to international best practice, neonates that require intensive care and access to neonatal surgery or review by subspecialist teams should be cared for in a dedicated Neonatal Intensive Care Unit (NICU). At present in Ireland this is not the case and the burden of care for these neonates falls upon the already high in demand Paediatric Intensive Care Unit (PICU).

**Aims:** To describe the total number of neonatal admissions to the PICU in the period 01/07/2019-01/07/2020.

**Methods:** A retrospective electronic health record review of all infants admitted to the PICU who were <28 days corrected gestational age on the day of admission.

**Results:** In the study period there were 116 individual admissions of infants <28 days corrected gestational age of which 81 were interhospital transfers and 35 internal referrals. Mean age of admission was 19.8 days (range 0-119) with a mean corrected gestational age on admission of 37.97±4.59 weeks (range 25.57-44). This cohort utilised 20% (657/3285) of all potential PICU bed days in the period with a mean duration of admission of 5.66±6.48 days (range 1-45).

There were 46 joint admissions under the Neonatology & Surgical teams, 46 admissions under General Paediatrics, 5 under Neonatology alone, 8 under Neurosurgery, 4 under Metabolic, 3 under Endocrinology, 2 joint admissions under ENT & Neonatology and 2 admissions under ENT alone. There were 15 neonates discharged directly from PICU via interhospital transfer and 100 discharges to ward level in Temple Street.

**Conclusion:** The results of this standard setting audit and national service evaluation quantify for the first time the number of PICU bed days utilised by neonates in a calendar year as well as highlighting the clinical workload generated in PICU by these patients. These results will help to guide forward planning for further development and expansion of a neonatal High Dependency Unit in CHI @ Temple Street.

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## CHARACTERISING NEONATAL ABSTINENCE SYNDROME IN PRETERM INFANTS

CP McGrath<sup>1</sup>, GK Leon<sup>1</sup>, MA Boyle<sup>1</sup>

<sup>1</sup>Department of Neonatology, The Rotunda Hospital, Dublin, Ireland

**Aims:** To identify the nature and presentation of Neonatal Abstinence Syndrome (NAS) in preterm infants.

**Methods:** A primary analysis of the electronic health record in a tertiary neonatal unit was carried out between January – December 2021. Data was collated and analyzed with regard to demographics, antenatal drug exposure, Finnegan scoring and opioid treatment.

**Results:** Twelve preterm infants were born with antenatal exposure to maternal substance misuse. Finnegan scoring was unavailable for four infants due to intrauterine death (1), congenital anomaly (1) and missing data (2). Gestational age ranged from 31+1 to 36+6 weeks. Average birthweight was 2.06kg (range: 1.35–2.46kg) and head circumference 31.15cm (range: 29.2-33.5cm). Antenatal drug-use comprised Methadone(1), Methadone and Benzodiazepines (3), Methadone, Benzodiazepines and Heroin (3) and Other Polypharmacy (4). Six infants had urine toxicology, 4 of which were positive. Two infants required Opioid treatment.

Average Finnegan score was 2.72. The highest score obtained was 17. Time to first symptom-onset was

8.03 hours. Five infants obtained a score above six, in an average time of 30.8 hours. Central Nervous

System (CNS) disturbance was observed in all cases and Vasomotor/Respiratory/Metabolic and Gastrointestinal in 75% of cases. Tremor (Disturbed) was the most frequent symptom; present in 100% cases and 71.7% observations. This was followed by Tone, present in 87.5% cases and 30.2% observations and Tremor (Undisturbed) in 100% cases and 17.7% observations. Six categories received 0 in all observations, 4 in the Vasomotor/Respiratory/Metabolic group and 2 in the CNS group. Seven symptoms accounted for 75% of observations, all in the CNS group.

**Conclusion:** Preterm infants manifest a primarily CNS-driven withdrawal response to antenatal drug exposure. Current scoring tools may not accurately capture the nature of this response, leading to delay in recognition and treatment. A larger sample size and matched term cohort is required to identify relevant parameters for a preterm scoring system.

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## DIURETICS USE IN THE MANAGEMENT OF BRONCHOPULMONARY DYSPLASIA IN PRETERM INFANTS: A SYSTEMATIC REVIEW

E Ó Briain<sup>1</sup>, E Molloy<sup>1,5</sup>, L Alomairi<sup>1</sup>, L Coyle<sup>1</sup>, J Dowling<sup>1</sup>, J Kiernan<sup>1</sup>, L Mulkerrin<sup>1</sup>, A O'Byrne<sup>1</sup>, JC Rio Lynch<sup>1</sup>, D

Mockler<sup>2</sup>, G Fitzgerald<sup>3</sup>, E Isweisi<sup>1,4,5</sup>, G Semova<sup>1,4,5</sup>, A Branagan<sup>1,4,7</sup>, J Meehan<sup>1,4,5</sup>, E Roche<sup>1,4,5</sup>

<sup>1</sup>Discipline of Paediatrics, School of Medicine, Trinity College Dublin, Dublin, <sup>2</sup>John Stearne Medical Library, Trinity

Centre for Health Sciences, Dublin, <sup>3</sup>Library of Trinity College Dublin, Trinity College Dublin, Dublin,

<sup>4</sup>Trinity Research in

Childhood Centre, Trinity College Dublin, Dublin, <sup>5</sup>Children's Health Ireland at Tallaght, Tallaght Hospital, Dublin, <sup>6</sup>Children's Health Ireland at Crumlin, Crumlin Hospital, Dublin, Ireland

**Background and Aims:** Bronchopulmonary dysplasia (BPD) is a respiratory complication associated with neonatal prematurity. While diuretics are used in the treatment of BPD, their use is controversial owing to a lack of evidence supporting their efficacy and so there is variance in use between institutions. The aim of this review was to summarise the evidence of clinical trials studying diuretic use in BPD.

**Methods:** A systematic review of the Excerpta Medica database (EMBASE), Medline, Web of Science, and cumulative index to nursing and allied health literature (CINAHL) library databases was conducted in accordance with the preferred reporting items for systematic reviews and meta-analyses (PRISMA) guidelines. Covidence was used to screen the results and to aid in data extraction.

**Results:** Of the 430 records identified on database screening, 13 were included for analysis. Three studies examined the effects of a combination of spironolactone and chlorothiazide, two studies examined the effects of a combination of spironolactone and hydrochlorothiazide and eight examined the effects of furosemide. All studies analysed the effect of the drugs on dynamic pulmonary compliance and pulmonary resistance; therefore, these were used as comparative tools in our review.

**Conclusions:** The efficacy of diuretics for the treatment of bronchopulmonary dysplasia remains uncertain. Applying the Population, Intervention, Comparison, Outcome (PICO) approach suggests that the focused question cannot be answered using high-level evidence, because few randomised controlled trials (RCTs) were identified. Good quality, large prospective studies may lead to firmer conclusions, although the limited numbers of eligible patients make such studies difficult to perform. We conclude that further research is needed in this area, predominantly focusing on RCTs evaluating the safety and efficacy of diuretics in this population.

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#### VITAMIN D LEVELS OF NEONATES ADMITTED TO OUR NEONATAL UNIT

S O'Donnell<sup>1</sup>, M Benke<sup>1</sup>, J Tayrab<sup>1</sup>, N Al-Assaf<sup>1</sup>, R Khan<sup>1</sup>

<sup>1</sup>Department of Neonatology, University Maternity Hospital Limerick, Limerick, Ireland

**Background:** Vitamin D is an essential fat-soluble vitamin that is needed in the neonatal period for calcium homeostasis and skeletal growth. While it is widely practiced that preterm infants receive vitamin D supplements in the first year of life, there remains some debate in the best dose to be given.

**Aims:** The objective of this audit was to evaluate the vitamin D profile of the patients admitted to our neonatal unit. We also aim to assess how the type of feeding (including supplementation) impacts their levels.

**Methods:** This retrospective observational study carried out in the Neonatal Unit at University Maternity Hospital Limerick, over a one-year period (01/09/2021 – 31/08/2022). 76 patients' charts were analyzed.

Data collected included gender, gestational age, birth weight, type of feed and nutritional supplements. Serum vitamin D levels sent from the Neonatal unit over the study period were provided by the biochemistry lab.

**Results:** 58% of the patients were male and 42% female. 64% were singleton pregnancies and 36% twins.

97% were preterm and 3% term (23.7 weeks – 38.1 weeks). Birth weights ranged between 550g and 3.2kg. A total of 129 vitamin D levels were assessed from the study period. 2% of the patients

were found to be vitamin D deficient, 9% had insufficient levels, 74% had adequate levels and 16% were found to be at risk of excess with levels  $\geq 125$ nmol/L. One infant had hypervitaminosis D with a serum level of 352 nmol/L.

**Conclusions:** While vitamin D is essential for normal skeletal growth, particularly in the neonatal period, care must be taken to ensure that patients are not at risk of hypervitaminosis D which can lead to electrolyte abnormalities, GI disturbance, acute kidney injuries, seizures, and cardiac arrhythmias.

## CHRONIC NONBACTERIAL OSTEOMYELITIS WITH COMORBID ULCERATIVE COLITIS – A SINGLE CENTRE CASE SERIES

G C Larkin<sup>1</sup>, S O' Donnell<sup>1</sup>, D O' Leary<sup>1</sup>, O G Killeen<sup>1</sup>

<sup>1</sup>1. National Centre for Paediatric Rheumatology, CHI at Crumlin, Dublin, Ireland

**Background:** Chronic Nonbacterial Osteomyelitis (CNO) is a chronic relapsing inflammatory disorder of the skeletal system which affects mainly children. An association between CNO and Inflammatory Bowel Disease (IBD) has been reported, however the relationship between these two conditions is poorly described.

**Methods:** We describe the clinical course of four patients attending the National Centre for Paediatric Rheumatology with diagnoses of CNO and Ulcerative Colitis (UC).

**Results:** Four patients sharing concomitant diagnoses of CNO and UC were identified from the 87 patients with CNO (4.6% of CNO cohort). Female to male ratio was 1:1. The median age of diagnosis for CNO was 10.7 years (range 9.8 to 13 years). In 3 of 4 patients UC preceded the diagnosis of CNO, and all were in clinical remission on an anti-TNF agent (2 on Infliximab, 1 on vedolizumab and sulfasalazine). The median time to dual diagnosis was 2.08 years (range 1.08 to 3.16 years).

One UC patient had psoriasis also preceding the CNO diagnosis. None were HLAB27 positive. All had multifocal disease with a median of 2 active sites on whole body MRI imaging at time of diagnosis (2, 2, 2, 9). Two had clavicular, two had mandibular, two had pelvic bone and one had thoracic spine involvement. Immunosuppressive therapies were altered following the second diagnosis, with methotrexate added for three patients. One patient required a switch from infliximab to adalimumab.

**Conclusions:** Our study adds to the limited literature of previous reported cases of CNO and IBD. The triple diagnosis of UC/CNO/Psoriasis is only reported in 2 patients previously. Previous studies suggest a shared genetic predisposition to both conditions, others suggest that CNO can present as an extraintestinal manifestation of IBD. There may be underreporting of CNO in UC patients, as the majority are on biologic agents or attribution of CNO symptoms made to other diagnosis such as IBD arthropathy.

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## **RESUSCITATION REQUIRED POST ELECTIVE CAESAREAN SECTIONS BETWEEN 37+0 AND 38+6 WEEKS IN**

### **COOMBE WOMEN AND INFANT HOSPITAL**

**C O Neill<sup>1</sup>, S Tamgamus<sup>1</sup>, A O Sullivan<sup>1</sup>**

<sup>1</sup>Neonatology, Coombe Women and Infant University Hospital, Dublin, Ireland

**Aim:** To audit the number of personnel attending elective Caesarean sections from 37+0 to 38+6 weeks gestation, and to assess the level of resuscitation required at these deliveries.

**Methods:** A retrospective chart review of elective Caesarean sections (ECS) between 37 and 38+5 weeks gestation over 3 months (1st January 2022 to 31st March 2022) in Coombe Women and Infant's University Hospital (CWIUH).

**Results:** Over 3 months there were 369 ECS between 37+0 and 38+6 weeks gestation. Of these, 189 (51%) had a risk factor requiring additional medical personnel as indicated by NRP guidelines i.e. multiple pregnancy, congenital abnormality or maternal risk factor. Of the remaining 180 deliveries, 126 (70%) required no resuscitation/ medical intervention, 29 (16%) required suctioning/ blow by oxygen, 11 (6%) required CPAP, and 2 required IPPV (<1%). 12 charts were lost to follow up.

**Conclusions:** It is often difficult to predict outcomes at a delivery and the need for resuscitation of a neonate. International guidelines outlined in the Neonatal Resuscitation Program (NRP) have highlighted certain factors (both ante and intrapartum) which have been linked to having an increased risk of need for stabilisation or resuscitation at birth. The most recent guidelines in CWIUH for attendance at ECS were mostly in keeping with NRP guidelines. However, medical personnel attendance were required at sections from 37 to 38+6 weeks gestation, despite NRP highlighting only those <37 weeks as high risk. This audit showed that resuscitation of any form was required in less than one third of these deliveries, with the majority only requiring suctioning/ blow by oxygen. As theatre midwives are trained in NRP, it was felt that the initial stabilisation steps could be carried out without the need for further medical personnel at these deliveries and these guidelines were changed accordingly.

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## **AN AUDIT OF THE INVESTIGATION OF PROLONGED JAUNDICE IN A TERTIARY NEONATAL CENTRE**

**E O'Connell<sup>1</sup>, J Cousins<sup>1</sup>, N Clarke<sup>4</sup>, MJ White<sup>1,2,3</sup>**

<sup>1</sup>Department of Neonatology, Coombe Women and Infants University Hospital, Dublin, Ireland

<sup>2</sup>Neonatology and Paediatrics, Royal College of Surgeons in Ireland, Dublin, Ireland

<sup>3</sup>Paediatrics, University College Dublin, Dublin, Ireland

<sup>4</sup>Clinical Biochemistry, Coombe Women and Infants University Hospital, Dublin, Ireland

**Aims:** To audit adherence to guidelines of investigation of prolonged jaundice in infants presenting to the Coombe baby clinic. To observe the yield of the investigations in this cohort.

**Methods:** Laboratory data was extracted for infants 14 days or older with serum bilirubin  $\geq$  50 $\mu$ mol/L. All laboratory tests were reviewed on these infants. Fifty charts were reviewed in more detail.

**Results:** Between June 2021-May 2022, 131 infants 2 weeks or older were identified with hyperbilirubinemia, 30% (n=39) were aged over 6 weeks. Of the charts' reviewed feeding was documented in 96% (n=48); 74% (n=37) were exclusively breast fed, 18% (n=9) were combination fed and 8% (n=4) were formula fed. Stool color was documented in 80% (n=40). All 39 infants above 6 weeks had LFTs including total and direct bilirubin, 97% had an FBC and TFTs 97%, 82% had a DAT, 82% had urine microscopy and culture. All Infants aged between 14 days and 6 weeks had serum total and direct bilirubin. Further investigation in this group was dependent on age and feeding status. Pathological jaundice was identified in three infants with conjugated hyperbilirubinemia; two were diagnosed with biliary atresia. There was one diagnosis of Gilbert's syndrome. There were no confirmed UTIs, no diagnoses of hypothyroidism and no new cases of hemolytic anemia. Incidental findings were common leading to repeat testing. Urine cultures were positive in 17.5% (n=15) all were deemed to be contamination or colonization. A raised TSH was present in 16% (n=10) and neutropenia was present in 16% (n=10). ALT was above the reference range in 17 infants (27%) without liver pathology.

**Conclusions:** Adherence to guidelines was very good in this cohort. A small number of Infants with pathological jaundice were identified with conjugated hyperbilirubinaemia. A review of the guidelines is warranted to potentially reduce unnecessary investigations in this cohort.



## **NECROTISING ENTEROCOLITIS: AN OVERVIEW OF POTENTIAL BIOMARKERS AND INFLAMMATORY RESPONSES**

**M. A. Faizan**<sup>1</sup>, A Yeo<sup>1</sup>, A McMahon<sup>1</sup>, I Khalid<sup>1</sup>, S O'Neill<sup>1</sup>, P Gavigan<sup>1</sup>, M. M. Fadel<sup>1</sup>, E Molloy<sup>1-5</sup>, A Branagan<sup>1-5</sup>,  
E Isweisi<sup>1-3</sup>, G Semova<sup>1-3</sup>, J Meehan<sup>1-3</sup>, E Roche<sup>1-3</sup>

<sup>1</sup>School of medicine, Trinity College Dublin, Dublin, Ireland

<sup>2</sup>Trinity research in childhood centre, Trinity College Dublin, Dublin, Ireland

<sup>3</sup>Childrens Health Ireland at Tallaght, Tallaght hospital, Dublin, Ireland

<sup>4</sup>Childrens health Ireland at Crumlin, Crumlin hospital, Dublin , Ireland

<sup>5</sup>The coombe womens and childrens hospital, The coombe, Dublin, Ireland

**BACKGROUND:** The objective of this systematic review was to assess the diagnostic utility of potential biomarkers associated with necrotizing enterocolitis (NEC).

**METHODS:** We conducted an electronic search of the available literature using databases such as PubMed, Embase, and Cochrane, using the search terms (“necrotizing enterocolitis” OR “necrotising enterocolitis” OR “NEC”) AND (“biomarker\*” OR ‘biological marker’). Studies reporting data on the diagnostic accuracy of biomarkers for NEC were included. Results were restricted to full-text articles of studies in the English language, available to September 2021. Risk of bias was assessed using the Quality Assessment of Diagnostic Accuracy Studies 2 (QUADAS-2) tool.

**RESULTS:** 231 studies were screened, after which we were left with a yield of 27 studies for analysis. Most studies investigated the ability of a biomarker to differentiate Bell’s stage  $\geq$ II NEC from controls and Bell’s stage II from stage III. For identifying Bell's stage  $\geq$ II, we found high sensitivity and specificity for faecal calprotectin (97.14%, 100%), serum calprotectin (100%, 96.4%), a panel consisting of urine proteins CST3, PEDF, and RET4 (96%, 90%), and maternal human milk oligosaccharide DSNLT (90%, 90%) when sampled prior to or around the initial diagnosis of NEC, while IL-33 demonstrated high accuracy (100%, 94.4%) when sampled  $\geq$ 3 days post diagnosis. For differentiating between Bell’s stage II and stage III, only urinary intestinal fatty acid binding protein (I-FABPu) demonstrated high sensitivity and specificity (90%, 92%) when sampled around the time of diagnosis.

**CONCLUSION:** Calprotectin, urinary CST3, PEDF, and RET4, and maternal DSNLT demonstrate usefulness in the initial diagnosis of NEC, while IL-33 may be used for diagnostic confirmation in the follow-up and IFABPu in the early differentiation of Bell's stage II NEC from stage III. However, data supporting their clinical use is insufficient, and limitations associated with sample modality and collection may hinder their clinical use.

## **CORD BLOOD SCREENING FOR ADA-SCID IN AN IRISH MATERNITY**

**HOSPITAL L Reaney, P O' Connor**

<sup>1</sup>Neonatal Department, Coombe Women and Infants Hospital, Dublin , Ireland

**Aims:** This project aimed to audit the cord blood screening for Severe Combined Immunodeficiency due to adenosine deaminase deficiency carried out among infants born to members of the Irish Traveller community in a maternity hospital in Dublin.

**Methods:** All infants born to members of the Irish Traveller community between 1<sup>st</sup> April 2021 and 1st October 2021 were identified via demographic data collected during the booking visit. Data regarding blood sampling was collected using the hospital laboratory system and chart reviews.

**Results:** 95 infants were born to mothers who identified themselves or their partner as members of the Irish Traveller community during this period. 55 infants had cord bloods taken (57.9%). Of those who did not have cord bloods taken, eight had peripheral bloods taken. 63 infants had either cord or peripheral blood taken (66.3%). 62 had samples successfully processed (65.2%). All samples had a lymphocyte count of >1.5 and all but one were >2. 32 had no cord or peripheral blood taken (33.6%). There were nine infants whose fathers but not mothers were members of the Traveller community. Just one of these had cord bloods taken. Four had no cord bloods taken but had a peripheral FBC sent (two of these were as part of septic workups and one was a premature infant admitted to NICU). Of the 32 infants who were missed, four had a written record of consent given for cord bloods in the maternal notes. The remainder had no written record of discussion, consent or lack of consent.

**Conclusions:** One third of infants born to members of the Traveller community did not have screening for ADA-SCID performed. Infants whose fathers but not mothers were members of the Traveller community were less likely to have screening bloods taken. Documentation regarding parental consent for cord blood screening was variable.

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**THE TREATMENT OF PATENT DUCTUS ARTERIOSUS IN PRETERM INFANTS: A RETROSPECTIVE CHART REVIEW AT CUMH IN 2019-2022.**

**CR Reynolds<sup>1</sup>**, DO O'Sullivan<sup>1</sup>, RC Chioma<sup>1</sup>, ED Dempsey<sup>1</sup>, DF Finn<sup>1</sup>

<sup>1</sup>Paediatrics and Child Health, University College Cork, Cork, Ireland

**Aim:** To summarize the current management strategies for the treatment of patent ductus arteriosus (PDA) in preterm infants at Cork University Maternity Hospital (CUMH) in 2019-2020.

**Methods:** The study is a retrospective chart review of all preterm infants born <32 weeks' gestation and <1500g with an echocardiogram (ECHO) diagnosed PDA at CUMH in the years 2019-2020. Data collected on demographic characteristics, PDA-ECHO parameters, treatment method, and outcome factors such as chronic lung disease (CLD), necrotising enterocolitis (NEC), and patient death.

**Results:** The study population consisted of 62 infants. The median gestation age (GA) was 26+5, IQR (24+4,28+6), with a mean birth weight of 0.908kg (SD 0.256). Patients who received treatment (n=29) had a statistically significantly larger PDA diameter (M=2.18, SD=0.46) compared to conservative (M=1.53, SD=0.50),  $t(61) = 5.3$ ,  $p < .001$ . In the population born <27GA (n=34), 71% received treatment, on a median DOL of 12 (Q1:7, Q3:17). A COX inhibitor was used in 90% of cases (52% ibuprofen, 38% paracetamol). There is a significant relationship between a greater stage of PDA and the development of CLD,  $X^2(2, N=62) = 7.38$ ,  $p < .05$ .

**Conclusion:** A moderate to large PDA diameter and presence of an unrestrictive flow pattern, seen more common in the <27GA population, was significantly linked to the decision to treat. PDA severity was associated with an increased incidence of negative outcomes such as CLD and NEC. A prospective study would be required to evaluate the efficacy of different treatment models.

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## LONG-TERM NEURODEVELOPMENTAL OUTCOMES OF NEONATAL SEPSIS: A SYSTEMATIC REVIEW

M Salman<sup>1</sup>, L Al Azawi<sup>1</sup>, L Byrne<sup>1</sup>, A Cadden<sup>1</sup>, D Doran<sup>1</sup>, R O'Farrell Tyler<sup>1</sup>, S Ram<sup>1</sup>, AS Silver<sup>1</sup>, E Isweisi<sup>1,2,3</sup>, G

Semova<sup>1,2,3</sup>, J Meehan<sup>1,2,3</sup>, E Roche<sup>1,2,3</sup>, A Branagan<sup>1,2,4</sup>, E Molloy<sup>1,2,4</sup>, S Almajed<sup>1</sup>

<sup>1</sup>Discipline of Paediatrics, School of Medicine, Trinity College Dublin, Dublin, Ireland

<sup>2</sup>Trinity Research in Childhood Centre, Trinity College Dublin, Dublin, Ireland

<sup>3</sup>Children's Health Ireland at Tallaght, Tallaght University Hospital, Dublin, Ireland

<sup>4</sup>Paediatrics, The Coombe Women and Infants University Hospital, Dublin, Ireland

This systematic review aims to determine the long-term neurodevelopmental outcomes of neonates with sepsis, or sepsis-like illness. A systematic search of three databases, PubMed, Embase and Medline, was carried out to analyse 8 relevant studies according to the PRISMA guidelines. Analysis of these 8 studies reported neurodevelopmental outcomes as a result of both proven neonatal sepsis and sepsis-like illness. Of the 2264 participants involved, 941 neonates were identified as having diagnosed sepsis or sepsis-like illnesses. The collected data was obtained over 36 years, from 1986 to 2021. Adverse neurodevelopmental outcomes spanned the main developmental domains of gross motor, vision or hearing and social and behavioural development with cerebral palsy being reported in two studies. Overall outcomes reported were similar in both groups with neurodevelopmental impairment and cerebral palsy being reported across the studies. The results obtained were analysed according to four categories: their diagnostic criteria for sepsis, preterm vs term infants, study design and finally study outcome. This last category found greater motor and hand-eye coordination difficulties, greater emotional behavioural problems such as anxiety and depression, increased risk of cerebral palsy in both gram positive and coagulase negative staphylococci sepsis patients, decreased corpus callosum length at three months corrected age and diminished corpus callosum growth in infants who had a systemic infection. Neonatal sepsis is a life threatening condition with significant impact on the neurodevelopment of infants. Our systematic review showed that neonates surviving sepsis have a higher risk of developing long term neurodevelopmental deficits, especially gross motor deficits. Evidence is however limited by differences in study design, lack of follow up data in the long term and differences in classification.

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## TO ASSESS THE IRRADIANCE LEVELS AND EFFECTIVENESS OF PHOTOTHERAPY SYSTEMS AT ST LUKES HOSPITAL KILKENNY

M Sani<sup>1</sup>, R Naveed<sup>1</sup>, AR Diaz<sup>1</sup>, YR Diaz<sup>1</sup>, AR Herrera<sup>1,2</sup>

<sup>1</sup>Paediatrics, St. Lukes Hospital, Kilkenny, Ireland

<sup>2</sup>Associate Professor, University College Dublin, Dublin, Ireland

**Background:** Hyperbilirubinemia is still a common problem in neonates affecting over 60% to 80% of newborns during the first week of life. Phototherapy is still the gold standard which prevents

associated complications. The effectiveness of a Phototherapy System (PS) is determined by the wavelength and intensity of the light (irradiance), the BSA exposed and the distance between device and baby. LED based devices are becoming more popular in clinical practice.

Researchers have demonstrated that some PS provided suboptimal levels of irradiance and did not cover the whole BSA of a neonate (uniformity). Additionally, the distance between the light and baby was not measured in clinical practice which also affected the irradiance dosage received by neonate. We therefore assessed our PSs in order to determine their effectiveness and highlight the need for routine measurement of irradiance level/footprint and distance between device and baby.

**Materials and Method:** Two devices were evaluated. Device 1 is an LED based system BiliLux (Dräger, Lübeck, Germany) and Device 2 was fluorescent bulb-based system (Airshield Vickers Double Fluoro- lite Phototherapy unit). Spectral irradiance and uniformity/footprint levels were measured using a calibrated radio spectrometer.

**Results:** Although Device 1 provided the recommended irradiance dosage at a safe distance of 28cm to 30cm between device and baby, the spectral irradiance emitted was not uniform. The highest irradiance was received at the center while a significant reduction was delivered towards the periphery. In contrast, Device 2 did not deliver the minimum recommended  $30\mu\text{W}/\text{cm}^2/\text{nm}$  irradiance intensity at a safe distance of 28cm to 30cm but had high uniformity.

**Conclusion:** This LED based overhead phototherapy device was more effective in managing Hyperbilirubinaemia between 28cm and 30cm but only if neonate placed directly perpendicular to device. This study further illustrates the importance of measuring the irradiance levels and distance between the phototherapy device and baby in clinical practice.

**Recommendations:**

- To Prescribe/document dosage and distance of phototherapy.
- Each unit should measure its phototherapy device irradiance levels, to monitor device effectiveness.

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## AUDIT OF PHLEBOTOMY IN THE ROTUNDA HOSPITAL FOR PREMATURE BABIES LESS THAN 32 WEEKS AND 1500 GRAMS

C Stanciu<sup>1</sup>, A Raza<sup>1</sup>, AC Glynn<sup>1,2</sup>, K Cunningham<sup>1</sup>

<sup>1</sup>Department of Neonatology, Rotunda Hospital, Dublin, Ireland

<sup>2</sup>Department of Dietetics, Rotunda Hospital, Dublin, Ireland

**Aims:** Phlebotomy is performed on babies for many reasons, blood tests may be required at their initial evaluation, or as part of monitoring, assessing for illnesses and response to treatments. In particular, preterm babies whose birth weight is <1500 g and those <32/40, while they are establishing on enteral feeds and receiving TPN, undergo frequent blood testing. The aim of this audit is to evaluate frequency of blood sampling while receiving TPN and estimate volume of blood drawn.

**Methods:** Retrospective study, 6 months data (July-December 2021) of premature babies admitted to Rotunda NICU <1500g and <32/40, divided in 2 groups: group 1 (24-28/40) and group 2 (28+1- 32/40). Estimated blood volume 90ml/kg and calculated according to Hb on admission.

**Results:** Data analyzed for 44 babies: 25 babies in group 1, and 19 babies in group 2.

75% of total babies had no electrolytes on day 1.

70.8% in group 1 and 57.9% in group 2 had PN bloods early on day 2 of life.

29.2% in group 1 and 42.1% in group 2 had PN bloods day 3-4 as recommended.

92% in group 1 had 4 to 8 PN samples and 95% in group 2 had 3 to 6 PN samples in first week, only 2 samples recommended in the guideline.

Hb levels at birth were between 9.8g/dL to 21.1g/dL. Mean blood drawn 14.5% (this includes FBC, U&Es, SBRs, PN bloods with tryglicerides, gases, glucose monitoring) in first week of life, values ranging between 1.65% to 34.3%.

**Conclusion:** Most babies had PN bloods too early and too frequently.

Volume of blood drawn was as high as 34.3%.

We need to raise awareness of iatrogenic anaemia following frequently phlebotomy and multiple transfusions needed as a consequence.



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### **PREVALENCE OF HYPOGLYCEMIA IN NEWBORNS AND ADHERENCE TO THE NEONATAL HYPOGLYCEMIA MANAGEMENT GUIDLINES AT WEXFORD GENERAL HOSPITAL.**

E Suliman, S Batool, N Khan, A Kalim

<sup>1</sup>Paediatrics, Wexford General Hospital, Wexford, Ireland

**Aim:** Neonatal hypoglycemia is regarded as the most common metabolic problem in neonates, affecting 15% of infants<sup>1</sup>. Despite the high incidence, there are no international consensus guidelines and limited evidence is available on the diagnosis and management of neonatal hypoglycemia<sup>2</sup>. Neonatal hypoglycemia management guidelines were implemented in Wexford General Hospital (WGH) in April 2021 for the better management of neonatal hypoglycemia<sup>3</sup>. The aim of this study is to assess the prevalence of hypoglycemia in newborns at WGH and adherence to the guidelines.

**Method:** Data was collected including basic demographics, gestational age, causes of hypoglycemia, management and outcome of the newborn with documented hypoglycemia, and who were born from August 2021 to August 2022. A total of 21 newborns were included in this study.

**Results:** The prevalence of neonatal hypoglycemia at WGH in this duration was 1.2%. The commonest cause of hypoglycemia was maternal diabetes in 66.7%(13/21) of cases. 38% (8/21) of babies were asymptomatic. 90.5%(19/21) of neonates received formula feed initially. 62% (13/21) of newborns developed persistent hypoglycemia, for those babies intravenous (I.V) bolus of Dextrose 10% (2ml/kg) was given in 100% (13/13) cases. However, blood glucose recheck after the bolus was attempted in 61.5%(8/13) of cases. Furthermore, maintenance Dextrose 10% was continued in 84%(11/13) of cases who received the bolus. Glucose infusion rate (GIR) was calculated in 15% (2/13) of cases. Monitoring was stopped when blood glucose was  $\geq 3$ mmol for 24 hours in 38.4% (5/13) of cases.

**Conclusion:** The prevalence of neonatal hypoglycemia at WGH was high and the highest risk group for hypoglycemia were the infants of diabetic mothers. Our compliance levels were found to be

satisfactory for initial formula feed, I.V bolus and maintenance of Dextrose 10%. However, further improvement is required for the blood glucose recheck, GIR calculation and monitoring.

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### **MANAGING COAGULOPATHY IN PRETERM AND TERM NEONATES.**

**SA Taj<sup>1</sup>, S Mujahid<sup>2</sup>, R Khan<sup>1</sup>, AA Niazy<sup>1</sup>**

<sup>1</sup>Neonatal department, University Maternity hospital Limerick, Limerick, Ireland

<sup>2</sup>Paediatric clinical tutor, University of Limerick, Limerick, Ireland

**Introduction:** Abnormal coagulation profile is frequently seen in neonates especially preterms. Neonatal Haemostasis is different from older children and as well as the threshold to treat abnormal coagulation profile. Therefore, we are proposing the following guidelines to manage Neonatal Coagulopathy.

**Purpose:** This guideline has been developed to provide clarity and standardise the management of neonatal coagulopathy.

**Two tables <sup>(1,2,3,5,6)</sup> have been developed to highlight the Normal reference ranges of PT, APTT, Fibrinogen and suggested treatment thresholds levels of abnormal PT, APTT, INR, Fibrinogen in preterm and term infants.**

**High PT/INR:** If a baby had already received first dose of Vit-K after birth, then to repeat IV Vitamin-K 100mcg/kg (max dose 1mg). <sup>(2)</sup> Repeat coagulation in 6 to 24 hours<sup>(2)</sup> as indicated. If PT is still prolonged then repeat IV Vit-K 100mcg/kg (max dose 1mg) <sup>(2)</sup> and discuss with Haematologist regarding FFP and further investigations (e.g. 50:50 mixture correction test & factors analysis). If the baby is still clinically bleeding with persistently prolonged PT/INR, then Vitamin-K can be given regularly 8 hourly <sup>(BNF)</sup> and consider FFP infusion.

**High APTT:** If APTT is above treatment threshold, then consider FFP 10-20ml/kg slow infusion over 30-60 minutes <sup>(2,3)</sup> Repeat coagulation in 4 to 24 hours as clinically indicated. <sup>(2)</sup> If APTT is still prolonged, then consider repeating FFP infusion and discuss with Haematologist regarding Fibrinogen concentrate infusion especially if Fibrinogen level is low. <sup>(2)</sup> If abnormal coagulation

persists for >24hours in absence of any precipitating factors, then seek advice from Paediatric Haematologist for further management (e.g. 50:50 mixture correction test and Factors analysis) <sup>(2)</sup>

**Low Fibrinogen:** Fibrinogen Concentrate (RiaSTAP® 20mg/ml) can be administered at 1 – 3.5ml/kg <sup>(4)</sup> after discussing with Haematologist.

**Conclusion:** Understanding development of Hemostasis in preterm neonates is vital as it naturally prolongs PT, APTT, INR and Fibrinogen levels. As there was no national guideline on neonatal coagulopathy for neonates, therefore we have developed above guidelines along with normal reference range values and treatment threshold levels from best available evidence.

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2.Coagulopathy Neonatal Guidelines 2019-21 Bedside Clinical Guidelines partnership with Midwest Neonatal Operational NHS Trust 3.Coagulopathy in the Neonate Elizabeth Pilling July 2012, reviewed June 2021, Yorkshire and Humber Neonatal ODN South Clinical Guideline NHS.

These Guidelines were based on following references: a.Andrew M. Development of human coagulation system in the healthy premature infant. Blood 1988; 72; 1651-1657 b. Christensen R D et al. Reference intervals for common coagulation tests of preterm infants. Transfusion 2014;54(3);627-632 c. Neary E et al. Laboratory coagulation parameters in extremely premature infants born earlier than 27 weeks gestation upon admission to a neonatal ICU. Neonatology 2013;104(3);222-7 4.The investigation and management of neonatal haemostasis and thrombosis.

Michael D. Williams, Elizabeth A. Chalmers, Brenda E. S. Gibson. BJHaem 2002 5.Coagulation values in term and preterm infants, Robertson's Textbook of Neonatology Fourth edition. 6.Guidelines on transfusion for fetuses, neonates and older children. Helen V. New, Jennifer Berriman.. BJHaem 2016

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## **CORRELATION OF SERUM BILIRUBIN AND POINT OF CARE (POCT) BILIRUBIN IN THE NEONATAL UNIT**

**S Tamgumus<sup>1</sup>, C O'Neill<sup>1</sup>, A Doolan<sup>1</sup>**

<sup>1</sup>Department of Neonatology , Coombe Women and Infants University Hospital, Dublin , Ireland

**Aim:** We aim to establish accuracy of POCT bilirubin vs serum bilirubin taken simultaneously to decrease phlebotomy required in our neonatal population.

**Methods:** Serum bilirubin level (venous or arterial) and POCT samples were taken at the same time when clinically indicated and sent to the lab for analysis. The results were processed and reported as standard on iLab software. Results were collected prospectively, POCT bilirubin and Serum bilirubin were recorded in a spreadsheet. Following initial data collection, the setting on the POCT blood gas analyser in the Neonatal Unit was activated. Further samples were collected in the same way and results manually collected and recorded as previous. Clinicians and nursing were advised that the POCT bilirubin on analyser was currently only for data collection and not to be used to make clinical decisions during the period of study.

**Results:** 33 paired samples were taken and processed in lab initially however the laboratory staff felt there was a delay in processing samples which may affect results. Hence the bilirubin setting was active in our own gas analyser. 29 samples were taken and processed at the NICU. Between the serum and POCT samples, there was difference of 33.7 with 1 standard deviation. The mean difference between the samples was 12.1.

**Discussion:** Neonates are exposed to repetitive phlebotomy in the NICU. The POCT bilirubin unfortunately does not show a strong enough correlation to the gold standard which is serum bilirubin to replace its use. This cannot be stated for all POCT analysers and machines as there are differences within each unit and machine. However on the basis of this in our unit it was felt serum bilirubin should be used to guide treatment. This may be due to the small numbers used in our study. Previous studies with larger numbers have shown good correlation

Choi HY, Corder W, Tefera E, Abubakar KM. Comparison of Point-of-Care versus Central Laboratory Testing of Electrolytes, Hemoglobin, and Bilirubin in Neonates. *Am J Perinatol.* 2021.

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#### **NEONATAL ABSTINENCE SYNDROME: A NATIONAL SURVEY**

**S Tamgumus**<sup>1</sup>, M Boyle <sup>1</sup>, N McCallion <sup>1</sup>

<sup>1</sup>Department of Neonatology, Rotunda Hospital, Dublin, Ireland

**Aims:** To conduct a national survey to assess management of Neonatal Abstinence Syndrome (NAS) in neonatal centres throughout Ireland.

**Methods:** The survey was conducted online via SurveyMonkey and distributed via email in April 2022. A 30 question survey was developed based on current clinical practices, NAS management changes and recent literature review in consultation with neonatal consultants, ANPs and pharmacy input in the Rotunda Hospital.

**Results:** All 21 neonatal units responded to the survey. This showed that 33% of infants are monitored for the recommended 5 days of observation with 52% being monitored <5 days and at risk of withdrawing in the community and being missed.

100% of units are currently using the Finnegan scoring system however only 48% of staff are given training in using the tool.

66% of units had a non-pharmacological treatment plan, these consisted of swaddling, reducing light and noise, minimal handling, sucrose. All units used oromorph as their first line agent however frequency varied from 4-6 hourly and dosing varied from 40mcg/kg to 100mcg/kg. 66% based their dosing of oromorph on the withdrawal score. Maximum dose varied between 400mcg/kg/day to 200mcg/kg/dose 4 hourly.

57% of units had a policy of encouraging breastfeeding in mother who are on methadone/opiates. Only 24% of units had a specific parental education program for these families. There was 1 unit had a transitional care program available for NAS management.

**Conclusion:** The results of this national survey show the wide discrepancy in treatment and management strategies for infants with NAS. The infrequency of units dealing with these infants compounds this and demonstrates the benefit that a National Guideline would have in guiding care in regional centres. This is a small, vulnerable population and the development of a consensus on management and treatment could standardise care and improve outcomes.

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2. O'Grady MJ, Hopewell J, White MJ. Management of neonatal abstinence syndrome: a national survey and review of practice. *Arch Dis Child Fetal Neonatal Ed.* 2009;94(4):F249-52.
3. Holmes AP, Schmidlin HN, Kurzum EN. Breastfeeding Considerations for Mothers of Infants with Neonatal Abstinence Syndrome. *Pharmacotherapy.* 2017;37(7):861-9.

**AUDIT OF ADHERENCE TO RECENT BLOOD PRESCRIBING GUIDELINES WITHIN THE NEONATAL INTENSIVE CARE UNIT SETTING IN CORK UNIVERSITY MATERNITY HOSPITAL.****AM Urbaniak<sup>1</sup>**, F Zulfiqar<sup>1</sup>, AM Pavel<sup>1</sup>, M Kenosi<sup>1</sup><sup>1</sup>Department of Neonatology, Cork University Maternity Hospital, Cork, Cork

**Aims:** The aim of this audit was to assess adherence to new blood transfusion guidelines in the neonatal intensive care unit setting in Cork University Maternity Hospital. The secondary objectives were to compare transfusion practices before and after guideline implementation (April 2021), analysing numbers of transfusions given and neonatal clinical outcomes.

**Methods:** This is a retrospective study of transfusion practice within the CUMH neonatal unit. A list of babies who received red cell transfusion over 6 months prior to guideline update (in April 2021) and 6 months following guideline update was collated and relevant data was extracted using Cerner database. Perinatal data on neonatal and maternal factors (such as sex, gestational age, parity) was collated and analysed using Microsoft Excel.

**Results:** Guideline adherence following implementation was 56.8%. In the 6 months prior to guideline update there were 26 babies who received red cell transfusion with a total of 73 transfusions given. Post guideline update there were 22 babies, receiving a total of 37 red cell transfusions. Both groups were similar in terms of gestational age, delayed cord clamping time, maternal age and parity. Neonatal outcomes were observed to be non-inferior in the post guideline implementation group.

**Conclusion:** Since the introduction of internal neonatal transfusion guidelines in Cork University Maternity Hospital, there have been fewer red cell transfusions, while the number of babies receiving transfusion remained similar. There was no observable negative effect on outcomes in the post guideline group. These results are in line with recent systematic reviews<sup>1</sup> which have shown restrictive haemoglobin thresholds to decrease the number of neonatal transfusions given without having a negative impact on outcomes. This audit highlights the importance of internal transfusion threshold guidelines within Irish neonatal units.

1. Whyte R, Kirpalani H. Low versus high haemoglobin concentration threshold for blood transfusion for preventing morbidity and mortality in very low birth weight infants. Cochrane Database of Systematic Reviews. 2011(11)

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**Withdrawn**

## **STOPPING PHOTOTHERAPY TREATMENT IN TERM INFANTS, WHAT IS THE BEST PRACTICE? AN AUDIT OF PRACTICE AT CUMH**

**A Abrahams**, M Kenosi, I Sarwar, L Salih, C Schwartz, EM Dempsey

<sup>1</sup>Neonatal , Cork University Maternity Hospital , Cork, Ireland

'NICE' guidelines are used for reference in CUMH neonatal unit for safe phototherapy treatment of neonatal jaundice. Gestational age specific treatment threshold charts are used to guide initiation progress and cessation treatment. NICE guidelines recommend stopping lights when bilirubin level has dropped 50 micromoles under treatment line in term infants. An initial audit was completed to assess our practice against the NICE guidelines last year. Outcomes for babies who had phototherapy stopped before and after the recommended 50micromole threshold were assessed. The hypothesis was that stopping lights early would result in prolonged length of stay and rebound hyperbilirubinaemia. A re audit was completed in order to assess change, if any, to our practices over the past 12 months.

We hypothesised that stopping lights before reaching 50mmol threshold would result in increased blood sampling, rebound hyperbilirubinaemia and readmission.

**Methods:** A retrospective chart review of all infants admitted to our neonatal unit with a primary diagnosis of neonatal jaundice was performed. Babies were excluded due to the following : preterm delivery (<37 weeks), suspected sepsis treated with antibiotics on this admission, major congenital anomalies, metabolic or haematological diagnosis, DCT positive Jaundice infants or did not require phototherapy. Baseline infant characteristics and maternal risks were collected.

### **Results:**

- Total admissions 1162
- For Jaundice Management 171 (14.7% total)
- Simple Jaundice requiring Phototherapy 62
- Phototherapy continued to 50mmol threshold 45 (72.5%)

### Comparisons between groups

- 45 lights stopped after 50mmol
- 2 readmitted
- 3 rebound hyperbilirubinaemia
- 3 Increased blood letting
  
- 17 lights stopped before 50mmol threshold
- 6 readmitted
- 4 rebound hyperbilirubinaemia

- 2 increased blood letting
- 4.4% vrs 35% readmitted
- 6.6% vrs 22.5% rebound hyperbilirubinaemia
- 4.3% vrs 11.7% requiring >3 blood samples post stopping phototherapy

**Conclusions:** One in three infants required readmission, one in five had rebound hyperbilirubinaemia and one in ten required multiple blood samplings after stopping phototherapy before threshold. We should reduce length of stay, frequency blood-letting, rebound hyperbilirubinaemia and readmission by following guidelines.

Jaundice in newborn babies under 28 days Clinical guideline Published:19 May 2010 Last updated:26 October 2016



## DIFFUSE PAEDIATRIC LOW GRADE GLIOMAS OF THE BRAINSTEM – A CASE

SERIES E Ads<sup>1</sup>, E Hamza<sup>1</sup>, S Curry<sup>1</sup>

<sup>1</sup>The National Children's Cancer Service (NCCS), Children's Health Ireland at Crumlin, Dublin, Ireland

**Background:** In Ireland, 180 children receive a cancer diagnosis annually. Tumours of the central nervous system (CNS) account for 25% of cases. Paediatric low grade gliomas (PLGG), considered benign, are the most frequent type of CNS tumour diagnosed in children; however, are much less common than aggressive high grade gliomas (diffuse midline gliomas) when considering brainstem location.

**Aim:** To describe a case series of children with brainstem PLGG.

**Methods:** A retrospective chart review was performed describing 3 patients with brainstem PLGG diagnosed 2017-2021

### Results:

Case 1: 6-year old male with Neurofibromatosis type 1 (NF1) presented with longstanding dysphonia, and swallowing difficulties. Neuro-imaging revealed a non-enhancing diffuse intrinsic pontine tumour extending into right cerebellar hemisphere, involving the medulla. Clinico-radiological surveillance strategy was recommended. He is alive with stable disease five years since diagnosis.

Case 2: 2-year old female with NF1 presented with failure to thrive, vomiting, unilateral facial weakness, and hoarse voice. Neuro-imaging revealed extensive brainstem expansile, enhancing lesion, extending from posterior pons to C3. Biopsy confirmed PLGG. Adjuvant chemotherapy was commenced. He is alive with stable disease three years and six months since diagnosis.

Case 3: 3-year old female presented with squint, unsteadiness, motor regression and swallowing difficulties. Imaging revealed a non-enhancing brainstem tumour extending from the pons to right thalamus and into the medulla and upper cord. Biopsy confirmed PLGG. Treatment involved ventriculoperitoneal shunt insertion, and commencement of chemotherapy. Subsequent clinical deterioration prompted cessation of chemotherapy and urgent focal radiotherapy. She is alive with stable disease 18 months since diagnosis.

**Discussion:** Brainstem gliomas in children are predominantly high grade and are the main cause of death in children with brain tumours. Here we present a case series of children with brainstem PLGGs. Although in a precarious location with significant morbidity, these tumours are have a better prognosis than their high grade counterparts; reflecting entirely different biology and required management strategies.

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## **A REVIEW OF CHILDREN ATTENDING THE PEDIATRIC FORENSIC MEDICAL SERVICE AT CHILDREN'S HEALTH IRELAND (CHI)**

**A Alzaabi**<sup>1</sup>, C Power<sup>1</sup>, S Roe<sup>1</sup>, A Walsh<sup>1</sup>, U Murtagh<sup>1</sup>, E Curtis<sup>1</sup>, M Barrett<sup>2</sup>, S Harty<sup>1</sup>

<sup>1</sup>Laurels Clinic, CHI @Tallaght, Dublin, Ireland

<sup>2</sup>Emergency Department, CHI @Crumlin, Dublin, Ireland

**Background & Aim:** Child sexual abuse (CSA) occurs when a child is used by another person for their sexual gratification or arousal or that of others.<sup>(1)</sup> We describe the referral characteristics of patients attending the Pediatric Forensic Medical Services (PFMS) at CHI over a 1-year period.

**Methodology:** We reviewed & analyzed data from the child protection reports of the patients attending to the PFMS at CHI in 2021.

**Results:** Twenty-one children (n=21) were seen in the first 3 months, 18/21(86%) were females. One infant, 1 toddler (1-3Y), 9 children (2- 5 years), 6 children (5-11y) and 4 adolescents were seen. Referrers were Gardaí 7/21, Tusla 5/21, ED 4/21, GP 2/21 and 3/21 were inpatients. Six (28%) had forensic assessments, 5(24%) assessed for recent abuse & 10(48%) for historical abuse. Fourteen (66%) disclosed their abuse, 4 on day of assault, and 3 within 1-5 days; all of whom were assessed within 1- 5 days. Vaginal assault occurred in 8/21(35% of cases); mostly penile- 5/21(23%). Allegations were against a single perpetrator in 18/21(86%) of the cases. Perpetrators were adult males in 15/21 (70%) cases, adolescent in 2/21, child 1/21. Ten (47%) were 1<sup>st</sup> degree relatives of victims.

Twelve children (57%) had behavioral changes & 9/21 (43%) had genital symptoms. Examinations were normal in 13/21(62%) of the patients.

**Conclusion:** Patients presented with acute allegations of assault had assessments carried out within the forensic window. Majority of the victims were females, and 2-5 years of age, while the majority of the perpetrators were adult males.

Reference: Children First, National Guidance for the Protection & Welfare of Children, TUSLA, Department of children & youth affairs 2017.

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### **NEVER FORGET THE THYROID - A RARE COMPLICATION OF ACQUIRED HYPOTHYROIDISM**

**E Burke**<sup>1</sup>, C Reynolds<sup>1</sup>, R Kernan<sup>1</sup>, NP Murphy<sup>1</sup>, E Carolan<sup>1</sup>

<sup>1</sup>Department of Endocrinology, CHI Temple Street, Dublin, Ireland

**Introduction:** Acquired hypothyroidism is the most common thyroid disturbance in children, most commonly secondary to autoimmune thyroiditis. It typically presents with faltering growth, delayed bone age or classical symptoms of cold intolerance, weight gain, poor energy and constipation. In rare cases the disease leads to Van Wyk Grumbach syndrome (VWGS), characterized by isosexual precocious puberty. The mechanism is thought to be related to very high circulating thyroid stimulating hormone (TSH) levels stimulating follicle stimulating hormone (FSH) receptors on the ovaries and testes. Prompt treatment should result in normalization of biochemical and ultrasound findings.

**Case Report:** We report a previously healthy 5 year old girl who presented to the emergency department with heavy vaginal bleeding and breast budding. History revealed faltering growth, headaches, irritability, dry hair and abdominal pain. Biochemical investigation revealed TSH >750mU/L and unrecordably low free T4 <3.9pmol/L with raised oestradiol of 26pmol/L. LH was suppressed <0.7 IU/L and FSH raised 8.7IU/L. Alpha-fetoprotein was elevated to 99ku/L and BHCG was normal. Bone age was delayed by 2 years. Pelvic ultrasound showed a pubertal uterus with endometrial stripe and bilateral enlarged multicystic ovaries. Bilateral nephrocalcinosis was present with normal serum calcium and a raised urinary calcium/creatinine ratio. She was commenced on low dose thyroid hormone replacement, 25microgram daily, with incremental increase and resultant improvement in symptoms.

**Discussion:** VWGS is diagnosed by profound hypothyroidism, precocious puberty and evidence of ovarian hyperstimulation. The condition can be misdiagnosed as ovarian tumor with raised ovarian tumor biomarkers and cystic appearance on ultrasound. Timely diagnosis and treatment can prevent reduction in final adult height, cognitive decline and surgical complications of multiple ovarian cysts. Interestingly our patient had bilateral nephrocalcinosis on renal ultrasound, with normal serum calcium. Literature review found cases linking profound hypothyroidism to nephrocalcinosis, which we expect will resolve with treatment.

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Stokes, V., Nielsen, M., Hannan, F. and Thakker, R., 2017. Hypercalcemic Disorders in Children. *Journal of Bone and Mineral Research*, 32(11), pp.2157-2170.

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#### **BUROSUMAB - INITIAL IMPACT AND SAFETY OF TARGETED THERAPY FOR X-LINKED HYPOPHOSPHATAEMIC RICKETS IN AN IRISH PAEDIATRIC MDT SETTING**

**E Burke<sup>1</sup>**, C Reynolds<sup>1</sup>, H Leahy<sup>1</sup>, E White<sup>1</sup>, C McDonnell<sup>1,2</sup>

<sup>1</sup>Centre for Paediatric Rare Bone Disorders, Department of Endocrinology, CHI Temple Street, Dublin, Ireland

<sup>2</sup>Discipline of Paediatrics, School of Medicine, University of Dublin, Trinity College, Dublin

**Aims:** X-linked hypophosphataemic rickets [XLH] caused by a mutation in the PHEX gene results in bowed legs, short stature and reduced mobility. Burosumab, an anti-FGF23 monoclonal antibody, is the first targeted medication for XLH. This audit reviews the impact of the first three months of Burosumab treatment on an Irish cohort of children with XLH-rickets to ascertain if Burosumab maintains normal calcium, alkaline phosphatase, and PTH levels and achieves normalised phosphate levels without causing an adverse risk profile.

**Methods:** All eligible patients from October 2021 to January 2022 elected to move to Burosumab therapy. Biochemistry was checked before commencing Burosumab and three months later as per protocol. Mean differences were ascertained using the paired student t-test. Adverse effects were recorded at clinic or by phone. The audit was submitted for approval to the CHI Temple St audit programme.

**Results:** Eight patients (2 males, 6 female), mean age of 6.6 years [1.3–14.2 years] commenced Burosumab in the audit timeframe. Mean height and weight at baseline were 111.0 cm [83.6-159.0cm] and 27.5kg [11.4-58.6kg] respectively. All children were Vitamin D3 sufficient (mean 75nmol/L [51-98]) and discontinued standard of care (phosphate tabs QDS/alfacalcidol drops OD) one week prior to treatment. Biochemical parameters achieved the expected response by three months with a significant improvement in mean phosphate ( $p < 0.002$ , 95% C.I -0.47 to -0.16) without a notable change in mean calcium ( $p = 0.26$ , 95% C.I -0.04 to +0.11), alkaline phosphatase ( $p = 0.12$  95% C.I -32.28 to 218.03), PTH ( $p = 0.63$  95% C.I. -3.48 to 2.23) or urinary calcium:creatinine ( $p = 0.89$  95% C.I. -0.49 to 0.44). There have been no adverse events reported to date.

**Conclusion:** Burosumab is proving to be a safe, effective targeted therapy for XLH. Further studies are required to ascertain if continuous normalisation of phosphate enables a reduction in complications not seen with the previous standard of care

Padidela, R., Cheung, M., Saraff, V. and Dharmaraj, P., 2020. Clinical guidelines for burosumab in the treatment of XLH in children and adolescents: British paediatric and adolescent bone group recommendations. *Endocrine Connections*, 9(10), pp.1051-1056. (D et al., 2019) Clinical practice recommendations for the diagnosis and management of X-linked hypophosphataemia. *Yearbook of Paediatric Endocrinology*.

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## IRISH PAEDIATRIC PATIENTS WITH EOSINOPHILIC OESOPHAGITIS; A REGIONAL CENTRE EXPERIENCE A Busher<sup>1</sup>, A Rodríguez-Herrera<sup>1</sup>

<sup>1</sup>General Paediatrics, St Luke's General Hospital, Kilkenny, Ireland

**Aim:** Eosinophilic oesophagitis (EoE) is an eosinophil-rich, TH2 antigen-mediated disease of increasing pediatric and adult prevalence(1). At present, there is a paucity of data among Irish paediatric patients. The primary aim of this study was to study the demographics of patients presenting with EoE. Eosinophils are known to be implicated in TH2-mediated inflammation(2). A secondary aim of this study was to determine if patients with EoE demonstrate serum eosinophilia.

**Methods:** A retrospective chart review of patients attending the Paediatric Gastroenterology clinic at St

Luke's Hospital, Kilkenny was carried out. The diagnosis of EoE was made with histopathological features of EoE or symptoms suggestive of oesophageal dysmotility. Full Blood Count (FBC) values were examined from laboratory reporting systems.

**Results:** 17 children are currently on follow up. 70.9% of patients affected are male (n=12), 20.1% are female (n=5). The median age at diagnosis was 9.5 years (range 4-14 years). 59% (n=10) had a personal history of another atopic condition. 88% (n=16) had biopsies carried out. Biopsies were taken in Saint Luke's or CHI Crumlin. 16 of 17 patients had an FBC collected. 25% (n=4) of patients had elevated peripheral eosinophil counts (range 0.52-1.41), 6% (n=1) had low eosinophils in the presence of an otherwise normal FBC, 69% (n=11) had normal full blood counts.

**Conclusions:** From our data, EoE disproportionately affects males. This is supported in studies of adult and paediatric patients with EoE (1). Atopic rates are higher among patients with EoE compared to the general population. From our data, peripheral eosinophilia is not a reliable marker of disease activity, with most patients affected with EoE demonstrating normal peripheral eosinophil values. This study is limited by small case numbers, although EoE is a rare disorder. In light of the potentially negative long term complications of untreated EoE, more research is required to further study EoE among Irish paediatric patients.

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## **CRYPTOSPORIDIUM INFECTION IN A POST RENAL TRANSPLANT PATIENT**

**S Cahill**<sup>1</sup>, N Shaughnessy<sup>1</sup>, E Jenkins<sup>1</sup>, A Awan<sup>1</sup>, M Riordan<sup>1</sup>

<sup>1</sup>National Centre for Paediatric Nephrology, Haemodialysis and Transplantation, CHI Temple Street, Dublin

**Aims:** To describe the management and challenges of cryptosporidium infection in a post renal transplant patient.

**Methods:** A 15-year-old male presented 2-weeks post cadaveric renal transplant with a 24 hours history of malaise accompanied by frequent episodes of watery diarrhoea. He was afebrile and appeared clinically dehydrated. He complained of headache, but had no cough, coryza, rash or vomiting. His medications at the time of admission included tacrolimus, mycophenolate mofetil (MMF) and prednisolone. His initial management focused of fluid resuscitation and stool for viral and bacterial culture was obtained.

**Results:** Bacterial stool culture revealed cryptosporidium species on day 4 of our patient's illness. In an immunocompetent host, antibiotic treatment is not required to treat this illness (1), however in the immunocompromised host, treatment efficacy and value is debated. Nitazoxamide may aide recovery in these patients (2). A broad-spectrum anti-protozoal and anti-viral agent, nitazoxamide was commenced for 14 days, in combination with a 7-day-course of paromycin, an aminoglycoside antibiotic.

Immunosuppression was reduced, with serum tacrolimus levels carefully monitored and MMF and prednisolone held.

**Conclusion:** Immunosuppression alters the cellular response to infection. It is therefore imperative to consider the need to balance immunosuppression, the effective prevention of graft rejection, with the need for an adequate immune response to mount within the host. There are some cases which suggest that solid-organ transplant patients are best treated with nitazoxamide and paromycin when managing cryptosporidium infection. Limited evidence suggests GIT involvement alone can be managed with a reduction in immunosuppression alone (3).

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### **A RARE CAUSE OF MICROSCOPIC HAEMATURIA**

**S Cahill**<sup>1</sup>, N Shaughnessy<sup>1</sup>, E Jenkins<sup>1</sup>, C Sweeney<sup>1</sup>, N Dolan<sup>1</sup>, M Stack<sup>1</sup>, M Riordan<sup>1</sup>, A Awan<sup>1</sup>

<sup>1</sup>National Centre for Paediatric Nephrology, Haemodialysis and Transplantation, CHI Temple Street, Dublin

**Aims:** To explore the investigation and management of a female with microscopic haematuria

**Methods:** A two-year old girl was incidentally found to have haematuria on dipstick and subsequently, noted to have intermittent proteinuria. She was referred to nephrology for investigation. Of note, the patient had no family history of renal disease, had normal hearing and vision and was not unwell prior to detection of haematuria. She did not complain of abdominal pain. Ultrasound kidneys and urine investigations for calcium, creatinine, oxalate and citrate were carried out.

**Results:** Investigations yielded no clear cause and thus, after a period of observation with persistence of microscopic haematuria and intermittent proteinuria, a renal biopsy was performed. This revealed thrombotic microangiopathy and thin-membrane disease. There was 10% fibrosis noted and some glomeruli noted to be sclerosed. No immune complex formation was noted on immunofluorescence. Genetic testing was carried out to unify diagnosis. Testing identified an alteration in collagen 4A5 gene on the x-chromosome, consistent with an X-linked Alport's Syndrome.

**Conclusion:** Alport's Syndrome (AS) manifests because of a defect in Type IV collagen. AS is more commonly an x-linked condition, although can also manifest in an autosomal dominant manner, which can resemble thin-membrane renal disease. Our patient was very unusually a manifesting carrier of X-Linked Alport's Syndrome. The traditional form of this condition is associated with



deafness; however this is not seen in all patients with this genetic mutation. 90% of female carriers of Alport's Syndrome will exhibit haematuria on dipstick and up to 12% will exhibit renal disease by age 40<sub>1</sub>. Significant renal disease in childhood is rarely exhibited in female X-linked Alport's Syndrome. Consideration should be given to this as a differential diagnosis in females with microscopic haematuria.

1.X-linked Alport syndrome: natural history and genotype-phenotype correlations in girls and women belonging to 195 families: a "European Community Alport Syndrome Concerted Action" study. J Am Soc Nephrol. 2003 Oct;14(10):2603-10. doi: 10.1097/01.asn.0000090034.71205.74. PMID: 14514738.

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### **THE COST OF REJECTION - DEPARTMENT OF GENETICS, CHI CRUMLIN AUDIT**

JC Coleman<sup>1</sup>, KK Kavanagh<sup>1</sup>, SL Lynch<sup>1</sup>, LB Bradley<sup>1</sup>

<sup>1</sup>Department of Clinical Genetics, CHI Crumlin, Dublin, Ireland

#### **Aims:**

To evaluate the reasons for referral rejection in the triage pathway.

To identify the time and cost implications.

**Methods:** A retrospective analysis of rejected referrals consecutively triaged by one consultant was undertaken over an 18 month period (01/01/2021-30/06/2022). Calculation of costs used data from a previous study (1).

**Results:** The consultant rejected 128/1581 (8.3%) of referrals. The rejection reasons included: 75% had not included the family/patient genetic report (6% of all referrals), 10% were conditions not accepted by our service (eg hypermobility), 8% redirected to other specialities, 3% given written advice in lieu of appointment and 4% for other reasons.

Follow-up information was requested on 101/128 (78%) of rejected referrals. For 57% this was received; in 43% no response was received and these cases remain closed. Median response time was 33 days. Of those who sent back appropriate information, 39% remain on waiting-list, 50%

attended OPD or were given appropriate advice, 5% did not attend and 4% had alternative follow up pathways.

The estimated timeframe from referral to triage response is 41.5 minutes per referral. For our rejected referrals this equated to 88.5 hours (59hrs/year). Our departmental cost for managing repeat referrals is €34.80 (1). Using this as our cost of rejection letter, this equates to €4454.40 in 18 months for one consultant (€2969.60/year), or €11,878.4/year departmental cost.

**Conclusion:** The majority of referrals are rejected for non-enclosure of patient genetic reports. Many referrals would have accepted to the waiting-list if the appropriate report had been attached. This means patients at risk of genetic disorders are not accessing clinical services because the referrer isn't providing the necessary information to allow triage.

Active management of the waiting list via upfront letters is costly, with a 57% response rate. Should similar rejection rates exist in other specialities (we estimate 914/2900 public hospital consultants likely to triage referrals (2)), this would equate to a cost of €2,714,214.40/year to the HSE.

1.) Walsh, N., Malone, L. & Lynch, S.A. Duplication of referral, a tsunami of paper: how much does it cost the Irish health services?. *Ir J Med Sci* (2022). <https://doi.org/10.1007/s11845-021-02866-y>

2.) HSE online publication, available at <https://www.hse.ie/eng/staff/leadership-education-development/met/consultantapplications/rep1/consultant-establishment-qtr-1-revised-09-07-2020.pdf>, accessed on 22/09/2022

**SURVIVAL IN A FOUR YEAR OLD WITH OUT OF HOSPITAL CARDIAC ARREST: DIAGNOSTIC DILEMMA AND WHEN TO DEVIATE FROM STANDARD RESUSCITATION GUIDELINES**

**N Collins**, D Finn, O O'Mahony, M Walsh

<sup>1</sup>Department of Paediatrics and Child Health, Cork University Hospital, Cork, Ireland

<sup>2</sup>Department of Paediatric Cardiology, Children's Health Ireland @ Crumlin, Dublin, Ireland

Out-of-hospital cardiac and respiratory arrest is rare in children. When it does occur, there is high morbidity and mortality rates, with survival to hospital discharge reported in multiple studies at between 5-27%. Additionally, favourable neurological outcomes are seen in 1 in 20 patients. However, those who do survive to discharge generally do well with five-year survival rates reported at 86% in one follow-up study.

We report an index case of catecholaminergic polymorphic ventricular tachycardia (CPVT) in a four-year-old male first presenting in cardiac arrest on Christmas Eve. The case demonstrates the diagnostic dilemma that exists in identifying CPVT, and the difficulty in treating cardiac arrest secondary to ventricular arrhythmia. It also exemplifies exceptional teamwork and collaboration between parents, paramedics and specialists in many fields in ensuring a favourable outcome in a highly unlikely scenario.

A four-year-old boy presented in cardiac arrest on Christmas Eve. He required CPR, two defibrillation shocks and adrenaline via intraosseous access before ROSC was achieved. He subsequently became unstable, with alternating rhythms of pulseless ventricular tachycardia, ventricular fibrillation, pulseless electrical activity and sinus rhythm, requiring multiple defibrillation shocks, and treatment with intravenous adrenaline, lidocaine and amiodarone. He subsequently responded to combined treatment with intravenous esmolol, amiodarone and milrinone. He was transferred to cardiac intensive care, where care was gradually normalized and oral anti-arrhythmic treatment with flecainide and nadolol commenced.

This boy has remained stable with no further runs of sustained VT and no shock outputs from his AICD. At one week post discharge from hospital, repeat echocardiography revealed a two centimetre pericardial effusion, likely related to the cardiac myotomy performed at the time of AICD insertion, a not uncommon post-operative occurrence(10). This was treated with spironolactone and frusemide, as well as prednisolone and colchicine with complete resolution within two weeks. Otherwise, he remains clinically well, and has resumed normal activities.

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2. Michiels EA, Dumas F, Quan L, Selby L, Copass M, Rea T. Long-Term Outcomes Following Pediatric Out-of-Hospital Cardiac Arrest\*. *Pediatr Crit Care Med* [Internet]. 2013;14(8). Available from: [https://journals.lww.com/pccmjournal/Fulltext/2013/10000/Long\\_Term\\_Outcomes\\_Following\\_Pediatric.4.aspx](https://journals.lww.com/pccmjournal/Fulltext/2013/10000/Long_Term_Outcomes_Following_Pediatric.4.aspx)
3. Napolitano C, Priori SG BR. Catecholaminergic Polymorphic Ventricular Tachycardia. *GeneReviews*®.

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#### **OUTSIDE THE BOX AND INSIDE THE BAG: AN UNUSUAL CASE OF ACUTE HYPERCALCAEMIA**

**K Conlon<sup>1</sup>, S Sheridan<sup>1</sup>, S Alanazi<sup>1</sup>, J Hayden<sup>2</sup>, D Cody<sup>1</sup>**

<sup>1</sup>Department of Endocrinology, CHI Crumlin, Dublin, Ireland

<sup>2</sup>Department of Gastroenterology, CHI Crumlin, Dublin, Ireland

Hypercalcaemia is a relatively rare presentation in the paediatric population, typically presenting with symptoms including abdominal pain and vomiting, acute confusion and changes in cognitive status and muscle weakness, and whose complications can include cardiac arrhythmia and renal stones. Differential diagnoses for paediatric hypercalcaemia include primary hyperparathyroidism, secondary hyperparathyroidism associated with chronic kidney disease, malignancy, thyrotoxicosis, Paget disease, exogenous calcium ingestion, hypervitaminosis A, hypervitaminosis D, pheochromocytoma and familial hypocalciuric hypercalcaemia.

Here we describe an atypical case of severe acute iatrogenic hypercalcaemia with serum calcium at presentation of 6.91 mmol/L, in a 13-year-old patient with a complex medical background and a longstanding history of TPN usage. The acute hypercalcaemia was caused by accidental excessive calcium administration via TPN at 5 times the rate prescribed. The patient's symptoms at presentation included abdominal pain, vomiting, confusion and generalised weakness.

The acute presentation was managed through administration of IV fluids to achieve hyperhydration and concurrent treatment with diuretics, resulting in rapid restoration of serum calcium levels and resolution of symptoms. Initial fluid bolusing in ED saw partial resolution of serum calcium to 5.97 mmol/L, and following 24 hours of hyperhydration at 200% of total maintenance fluids resolution of serum calcium to 2.51 mmol/L was observed. This improved serum calcium was maintained when IV fluids and diuretics were stopped, with a value of 2.32 mmol/L 24 hours after stopping treatment.

This case shows the effectiveness of treatment of acute hypercalcaemia with IV fluid infusion and administration of loop diuretics, even in cases of severe hypercalcaemia. It also serves to demonstrate the wide differential when it comes to considering causes of paediatric hypercalcaemia in the acute setting. Finally, it shows the risk of iatrogenic injury for all patients undergoing medical treatment and the importance of remaining vigilant to the potential for iatrogenic injury in all patients.

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#### **CHANGE OF BIOLOGIC UNMASKS OROFACIAL GRANULOMATOSIS IN A PATIENT WITH JUVENILE IDIOPATHIC ARTHRITIS**

**S Coyne**<sup>1</sup>, N Keenan<sup>1</sup>, N Mac Eoin <sup>1</sup>, JM Mac Mahon<sup>1</sup>, EJ MacDermott<sup>1</sup>

<sup>1</sup>The National Centre for Paediatric Rheumatology, CHI at Crumlin hospital, Dublin, Ireland

**Case Introduction:** Rates of Inflammatory Bowel disease (IBD) are higher in children with Juvenile idiopathic arthritis (JIA) than the general population (1). Orofacial Granulomatosis (OFG) is a rare disorder and is increasingly recognised as a subtype of Crohn's disease (2).

**Presentation:** A 14-year-old boy with polyarticular JIA, presented with lip swelling and blistering 2hrs post first tocilizumab infusion. Lip swelling was again noted with subsequent tocilizumab infusions, raising concern for allergic reaction. Patient described onset of swelling several hours post infusion, worsening over days and notably, without full resolution. No other symptoms of IgE-mediated drug allergy, (e.g., respiratory compromise, urticaria, cardiovascular collapse) were observed. Symptoms did not resolve when tocilizumab was stopped. On further questioning, patient reported new onset diarrhoea and subjective weight loss, which began at the same time. Of note, the patient had been receiving Adalimumab prior to starting Tocilizumab. No new connective tissue symptoms such as rash, fever or hair loss were recorded. No other joints were active on rheumatological exam.

**Investigations, treatment, and progress:** On examination, the lip swelling was felt to be in keeping with OFG. In view of his other new symptoms and temporal association with cessation of anti-TNF therapy, an urgent GI review was arranged. OGD with biopsy supported a diagnosis of Crohn's disease.

**Conclusions:** This case highlights the importance of thorough history taking in the context of potential drug reaction. Tocilizumab hypersensitivity reactions are rare, and the description was not in keeping with IgE-mediated or delayed reaction. Gastrointestinal symptoms began in the 'wash out' period changing from adalimumab, an anti-TNF inhibitor that is used to treat IBD, to tocilizumab, an IL-6 inhibitor which has been reported to aggravate underlying or undiagnosed IBD (3). OFG is a known manifestation of Crohn's disease and 40% of paediatric patients with OFG have intestinal involvement (2).

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2. Lazzerini M et al. Association between orofacial granulomatosis and Crohn's disease in children: systematic review. World J Gastroenterol. 2014;20(23):7497-504.
3. Borghini R et al. Onset of Suspected Ulcerative Colitis After Treatment with Tocilizumab in Patient with Celiac Disease and Juvenile Idiopathic Arthritis, Inflammatory Bowel Diseases. 2021;27(6)76-78.

### **CRYING OVER SPILLED MILK: CHILDREN WITH IGE MEDIATED MILK ALLERGY WHO FAILED TO COMPLETE THE MILK LADDER: A CASE SERIES**

**C Cronin**<sup>1</sup>, T Ah Heng<sup>1</sup>, S Hanley<sup>2</sup>, L Flores Villarta<sup>1</sup>, A Gallagher<sup>3</sup>, M Sexton<sup>1</sup>, AM McGinley<sup>4</sup>, JOB Hourihane<sup>2</sup>, R Velasco Zuniga<sup>5</sup>, J Trujillo Wurttele<sup>1, 3</sup>

<sup>1</sup>Department of Paediatrics and Child Health, University College Cork, Cork, Ireland

<sup>2</sup>Royal College of Surgeons of Ireland, Dublin

<sup>3</sup>Department of Paediatrics, Cork University Hospital, Cork, Ireland

<sup>4</sup>The Weir Family Health Clinic, Bandon, Co. Cork, Ireland

<sup>5</sup>Hospital Universitario Río Hortega, Department of Paediatrics, Valladolid, Spain

**Aim:** The milk ladder is the cornerstone of management of IgE-mediated cow's milk protein allergy (CMPA) in Ireland. While most children achieve tolerance to cow's milk and reach the top of the ladder (1), the reasons for stopping the milk ladder and barriers to progression through the milk ladder have not been explored to date. The aim of this study was to identify children with IgE-mediated CMPA who failed to complete the milk ladder, establish the reasons for not completing the ladder, explore the challenges to completing the milk ladder.

**Methods:** Children with CMPA who completed the milk ladder between 2011 and 2021 were identified in a chart review. Children who stopped the milk ladder were contacted to complete a telephone survey.

**Results:** 8 patients were included. The mean age of CMPA diagnosis was 12.5 months, range 3 months to 27 months. The mean age at which treatment with the milk ladder was commenced was 35 months, range 7 to 58 months. Mean Skin prick tests (SPT) was 4.875mm. Mean Specific IgE for whole milk was 47.4 kIU/ml. Most parents reported that they discontinued the ladder as their child experienced multiple and often severe reactions while progressing through the ladder. Other Challenges to completing the milk ladder that parents explained included their child having multiple food allergies which prevented them from eating certain foods on the ladder, having

other atopic conditions such as eczema for which they prioritised treatment, and diagnosis with autism, which prevented them from eating new foods due to sensory processing difficulties.

**Conclusion:** This is the first study to explore possible challenges to progressing through the milk ladder. Improved parent education, more frequent follow-ups throughout treatment, methods to reduce parental anxiety and an adapted milk ladder with greater food choices may improve progression through the ladder.

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## LEARNING FROM HOME: PILOTING A VIRTUAL EDUCATIONAL INTERVENTION FOR CAREGIVERS REGARDING ANAPHYLAXIS MANAGEMENT AND ADRENALINE AUTO INJECTOR ADMINISTRATION

C Cronin<sup>1</sup>, J Trujillo Wurttele<sup>1, 2</sup>

<sup>1</sup>Department of Paediatrics and Child Health, University College Cork, Cork, Ireland

<sup>2</sup>Department of Paediatrics, Cork University Hospital, Cork, Ireland

**Aim:** Parents of children with food allergies require training from a healthcare professional in anaphylaxis management and the administration of an adrenaline auto injector (AAI) when their child is prescribed an AAI (1). As a result of COVID-19, many consultations in the allergy clinic have been conducted via telemedicine, resulting in parents not receiving anaphylaxis management training in the pediatric allergy clinic. There is therefore a need to assess the benefit of virtual anaphylaxis management training for caregivers of children with food allergies. The aim of this study is to pilot a virtual educational intervention regarding anaphylaxis management for parents of children diagnosed with food allergy.

**Methods:** Parents of children recently diagnosed with food allergy and prescribed an AAI at a telemedicine paediatric allergy clinic were contacted by telephone to conduct virtual anaphylaxis management plan training. Training was conducted over a video call. The paediatric allergy clinic anaphylaxis management plan was discussed, and AAI administration was demonstrated in the three available brands of AAI using trainer pens.

**Results:** 10 parents took part in the pilot study. All parents were satisfied with the training they received and had the opportunity to ask any questions. At least 2 weeks after receiving this training, the parents received an online questionnaire assessing their knowledge of anaphylaxis management and AAI administration to explore if they retained the knowledge they received during training, and to also collect feedback on the training they receive.

**Conclusion:** While a formal data analysis has not yet been completed, we estimate that parental knowledge of anaphylaxis management and AAI administration will be optimal, and show that virtual training for parents is an effective and safe method of anaphylaxis management training.

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### UNPROVOKED RHABDOMYOLYSIS: FIRST PRESENTATION OF MADD IN A TEENAGE BOY

**N Crowley**<sup>1</sup>, E Forman <sup>1</sup>, D O'Shea<sup>1</sup>, C Howard <sup>1</sup>, I Knerr<sup>1</sup>, J Hughes<sup>1</sup>, E Crushell<sup>1</sup>, AA Monavari<sup>1</sup>, K Gorman <sup>2</sup>, R Boruah<sup>1</sup>

<sup>1</sup>Metabolic Medicine, CHI @ Temple Street, Dublin, Ireland

<sup>2</sup>Neurology , CHI @ Temple Street , Dublin, Ireland

**Background:** Multiple Acyl Co-A Dehydrogenase Deficiency (MADD) is a rare but treatable inherited disorder of fatty acid and protein metabolism. The attenuated form of the disease can present with episodic hypoglycaemia, liver dysfunction, cardiomyopathy, progressive encephalopathy, epilepsy and/or myopathy. Diagnosis requires genetic confirmation, however acylcarnitine profile, urinary organic acids and muscle biopsy all show abnormalities indicative of MADD. Age at presentation and presenting symptoms are varied, with median time to diagnosis of 3.9 years<sup>1</sup>. We describe a teenage boy presenting with rhabdomyolysis secondary to MADD.

**Case:** A 13 year old boy presented with a three month history of progressive muscle weakness, fatigue and dyspnoea. His symptoms were progressive and at time of admission he was unable to sit up unassisted. He noted dark coloured urine in the week prior to presentation. Clinical examination revealed reduced power in all four limbs (2/5) with pronounced proximal weakness including neck extension and flexion weakness. Creatine Kinase (CK) on presentation was 34,558U/L(58-312), Aspartate Aminotransferase (AST) and Alanine Transaminase (ALT) were elevated at 4,898U/L (17-44) and 986U/L (9-44) respectively.

Acylcarnitine profile revealed a markedly elevated C14:1 ratio, typically the marker for very long chain acylco-A dehydrogenase deficiency (VLCADD), however there were also increases in other short, medium and long chain acylcarnitines. Urinary organic acids showed increased excretion of compounds consistent with MADD. Genetic analysis revealed compound heterozygous pathogenic variants in the *ETFDH* gene, consistent with MADD. He was commenced on a high carbohydrate, low fat, low protein diet and riboflavin supplementation. His CK and transaminases have normalised and his strength is improving. One month post discharge he was mobilising independently.

**Summary:** We present a case of rhabdomyolysis secondary to MADD, which responded rapidly to dietary modification. MADD is rare but can be detected with early metabolic investigations. Inborn errors of metabolism are an important differential in rhabdomyolysis.

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**MONITORING VITAMINS, TRACE ELEMENTS, BIOCHEMICAL AND HAEMATOLOGICAL MARKERS OF NUTRITION IN CHILDREN WITH NEUROLOGICAL IMPAIRMENT (NI) AT CHILDREN'S HEALTH IRELAND TALLAGHT IN 2019**

**AD Dunphy**<sup>1, 2</sup>, SMcC McCormack<sup>1</sup>, Il Idrees<sup>1</sup>

<sup>1</sup>Department of Child Development and Neurodisability, CHI at Tallaght, Dublin, Ireland

<sup>2</sup>Department of Paediatric Nutrition & Dietetics, CHI at Tallaght, Dublin, Ireland

**Aims:** Feeding difficulties are frequent in children with neurological impairments and can be associated with undernutrition, growth failure, micronutrients deficiencies, osteopenia, and nutritional comorbidities. ESPGHAN 2017 recommends the assessment of micronutrient status on an annual basis in children with neurological impairment (NI). The aim of this audit was to audit compliance of monitoring vitamins, trace elements, biochemical and haematological markers of nutrition in children with neurological impairment at Children's Health Ireland (CHI) at Tallaght in 2019.

**Methods:** 60 children with NI aged 3-16 years in 2019 were included. 30 of these children were on exclusive enteral nutrition (EEN) and 30 were on a combination of enteral nutrition (EN) and oral (PO) diet. A yes/no audit tool was used to calculate compliance rate as a percentage.

**Results:** 23% of the children on EEN and 30% on EEN and PO diet had no bloods completed at CHI Tallaght in 2019. Compliance with an annual check of renal, liver and bone profiles ranged from 56-70% in both groups. Compliance rate with full blood counts ranged from 63-70% in both groups. On average 58% of children had vitamin D checked, 13.3% vitamin B12, 58% ferritin and 12% folate. 13% of children had vitamin A and E checked. No children on a combination of oral diet and enteral nutrition had their trace elements checked. 10% of children on EEN had their zinc checked, 3% had selenium checked while 0% had a copper checked.

**Conclusion:** The results of this audit have highlighted the need for further guidance on blood monitoring particularly fat soluble vitamins and trace elements. A working group has been formed to develop a local CHI clinical guideline on monitoring of nutritional bloods in this group.

**INVESTIGATING THE NUTRITIONAL ADEQUACY OF BLENDED TUBE FEEDS IN CHILDREN WITH SEVERE NEUROLOGICAL IMPAIRMENT**

LC Craigie<sup>1</sup>, SMcC McCormack<sup>2</sup>, RK Kelly<sup>3</sup>, MG Graham<sup>3</sup>, **AD Dunphy**<sup>3</sup>

<sup>1</sup>Department of Agriculture and Food Science, University College Dublin

<sup>2</sup>Department of Child Development and Neurodisability, CHI at Tallaght

<sup>3</sup>Department of Paediatric Nutrition & Dietetics, CHI at Tallaght

**Aim:** Blended tube feeds (BTF) consist of whole foods/liquids blended to a liquid consistency and administered through an enteral feeding tube. In recent years there has been a growing interest in administering feed using this practice. The aim of this research was to assess the nutritional adequacy of BTF in children with severe neurological impairment (SNI) attending CHI at Tallaght.

**Methods:** This was a prospective cross-sectional study (n=5). The inclusion criteria were that children were enterally tube fed, <18 years old, had a diagnosis of SNI and were receiving a fully or partially BTF. Caregivers completed a four-day food diary, a seven-day stool diary, and a PEDSQL GI Symptoms Scale. Anthropometric measurements and nutritional bloods that had been taken within 6 months of the study were assessed. Food diaries were analysed using the dietary analysis software Nutritics™. Nutritional analysis was completed both including and excluding micronutrient supplementation.

**Results:** All children were receiving 90-100% of their estimated energy requirements and ≥100% of their estimated protein requirements from their BTF. In all, 75% of children were meeting their iron, iodine, zinc, copper and folate requirements. 50% of children, who were not on Vitamin D supplementation, were not meeting vitamin D requirements. The only nutritional blood levels observed to be abnormal were a low ferritin (n=1) and low zinc (n=1). Using % weight for height, 3 out of 5 children had no evidence of malnutrition. One child presented with grade 1 malnutrition (84% weight for height) and 1 child was classified as grade 2 malnutrition (75% weight for height).

**Conclusion:** This study conveys that the majority of energy, macro-nutrient, and micro-nutrient requirements can be met from BTF; however, certain deficits were noted in micronutrients. Support and guidance from dietitians is imperative to ensure adequate nutritional intake.

## REVIEW OF THE TRANSPORTATION OF IRISH HEPATOLOGY PATIENTS TO THE UK FOR HEPATOBILIARY SURGERY

R Egan<sup>1</sup>, T Fallon Verbruggen<sup>1</sup>, L Feeney<sup>1</sup>, A Broderick<sup>1</sup>, E Fitzpatrick<sup>1</sup>

<sup>1</sup>Department of Gastroenterology and Hepatology,, CHI Crumlin, Dublin, Ireland

**Aims:** The Centre for Paediatric Hepatology at CHI Crumlin provides a national service for all children with acute and chronic liver disease. Since 2007 there has been no paediatric hepatobiliary (HPB) surgical or interventional radiology (IR) support in Ireland. At present, all children requiring this expertise must be transferred to the United Kingdom (UK) or other European countries for intervention, with the majority shared with the King's College Hospital, Paediatric Liver Centre in London (1). This service is funded by the Treatment Abroad Scheme (TAS) with transport facilitated by the National Emergency Operations Centre (NEOC).

The merits of this service are outstanding, however, with the increasing HPB needs of Irish children, the current system warrants review to quantify these needs, assess the burdens it presents to our health system, children and families and to identify possible alternatives for their care.

**Methods:** Retrospective chart review of hepatology patients requiring transfer to the UK & Europe to access hepatobiliary surgical expertise from 01/09/2019 to 01/09/2022.

**Results:** Over a this period three-year period, there were a total of 77 transfers for 51 patients. Seventy-six of these flights were to the UK. Twenty-five transfers were for liver transplantation, with an additional 14 for transplant assessment. There were 13 transfers for non-transplant HPB surgeries performed including 8 Kasai's. Seventeen were transferred for IR procedures (ERCP=8). Eight transfers were for medical hepatology input within a transplant centre. Thirty nine percent (n=30) of outgoing transfers required medical support and NEOC input, with 2 transferred with IPATs on ventilatory support.

**Conclusion:** Our current reliance on external HPB surgical & IR expertise causes a considerable resource and financial burden within our health system. The financial and personal stress to families must also be considered. As the number of children within our population requiring this expertise expands further research is warranted into facilities that could be made available in Ireland.

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Withdrawn

## **IGE-MEDIATED EGG ALLERGY – A TEN YEAR REVIEW**

**A Gallagher**<sup>1,2</sup>, C Cronin<sup>1</sup>, Y Ramesh<sup>1</sup>, J Trujillo<sup>1,2</sup>

<sup>1</sup>Paediatrics and Child Health, University College Cork, Cork, Ireland

<sup>2</sup>Paediatrics, Cork University Hospital, Cork, Ireland

**Background:** Food allergy is part of a large-scale rise in allergic conditions(1). Egg allergy is the second most common food allergy in children(2) and the main cause of anaphylaxis in infants(3). Food allergy and avoidance represents a significant burden on children and their families(4).

**Aims:** A retrospective review of IgE-mediated egg allergy in Cork University Hospital to evaluate treatment strategy over a ten year period. We hope to evaluate effectiveness and identify impacting factors.

**Methods:** Ethical approval was obtained from the Clinical Research Ethics Committee. Charts from the paediatric allergy service in Cork University Hospital were reviewed. A random sample from 2008 to 2019 was selected. Inclusion and exclusion criteria defined and applied. Results analysed using STATA.

**Results:** 110 patient charts reviewed, 74 were excluded due to insufficient documentation and/or skin prick test <3mm (n=36). Mean age at diagnosis was 14 months with mean age to start the egg ladder at 22.8 months. Average duration of treatment was 24.13 months. 36 patients completed egg ladder and achieved tolerance. Patients concurrently on the milk ladder demonstrated an extended average duration on the egg ladder of 6 months. Increasing specific IgE (SplgE) to egg (p=0.045) and having other food allergies (p=0.028) also resulted in increasing duration of treatment. Skin prick test (SPT), age starting treatment and gender were not significant.

**Conclusion:** IgE-mediated egg allergy is common and current treatment strategy in Ireland using the egg ladder is relatively new(5). It appears to be successful both here and in other countries(6). SplgE influences treatment duration however SPT was not significant. Having other food allergies and being on the milk ladder also impacts on length of time to achieve tolerance. This project provides a snapshot of egg allergy. We will continue our research with a view to publishing large-scale results impacting international treatment strategy for IgE-mediated egg allergy.



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Withdrawn

## UNPLANNED EXTUBATION – A RETROSPECTIVE CHART REVIEW

S Giva<sup>1</sup>, S O'Keefe<sup>1</sup>, S Whelan<sup>1</sup>

<sup>1</sup>1. Department of Paediatric Intensive Care, Children's Health Ireland at Temple Street, Dublin 1, Ireland

**Background:** Unplanned Extubation (UE) is the unexpected and unintentional dislodgement of the endotracheal tube in mechanically ventilated patients. It is a major adverse event as it can result in airway trauma, aspiration, hypoxemia, haemodynamic instability.

**Aim:** Primary aim: to determine whether a significant proportion of patients who had an UE event did not require re-intubation and therefore reflecting the need to assess earlier timing for extubation in mechanically ventilated patients.

Secondary aims: to determine objective causes that contributed to the UE.

**Study Design and Methodology:** retrospective chart review of all UE events occurring in CHI Temple Street PICU setting between January 2019 and December 2021. We recorded the length of time to re-intubation, mode of ventilation post UE event, oral versus nasal intubation, comfort scoring and sedation.

**Results:** During January 2019 and December 2021 there were 32 UE events; of these, 50% (n=16) required re-intubation within 24 hours while 50% (n=16) were given a trial of non-invasive ventilation (nasal CPAP, HFNC or no support).

Regarding the reason for the UE event, 18.75% (n=6) had loose tapes, 12.5% (n=4) had the ET tube in incorrect position, 28.1% (n=9) had inadequate sedation, 18.75% (n=6) had multiple reasons and 21.8% (n=7) had no documented reason.

Regarding the patient's comfort prior to the UE 59.4% (n=19) had inadequate sedation characterized by a comfort score between 13 and 20.

**Conclusion:** Of the 32 patients who experienced an UE event, 50% did not require re-intubation.

A set of recommendations will be incorporated in the PICU daily morning wardround basic checklist including: assessment of the tube type and tapes, assessment of comfort scoring and

assessment of readiness for extubation. Re audit will take place in keeping with Picanet surveillance which will result in completion of the audit cycle and real time assessment of whether the interventions were effective.

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**THE GOLDEN HOUR AUDIT: A PROSPECTIVE AUDIT OF ADHERENCE IN ANTIBIOTIC ADMINISTRATION IN PATIENTS WITH FEBRILE NEUTROPAENIA.**

A Amato<sup>1</sup>, E Hamza<sup>1</sup>, E Ads<sup>1</sup>, K Flinn<sup>1</sup>, C Owens<sup>1</sup>

<sup>1</sup>Oncology, Children's Health Ireland at Crumlin, Dublin, Ireland

**AIMS:** Febrile neutropaenia (FN) is a prominent presentation to paediatric oncology units, commonly occurring following chemotherapy administration. The “Golden Hour” approach outlined by the International Surviving Sepsis Campaign’s 1-hour bundle, highlights that the early identification and management of sepsis improves outcomes. This study examines adherence within the current management of FN at Children’s Health Ireland (CHI) at Crumlin to this international standard, and reviews potential interventions in the golden hour to improve outcomes.

**METHODS:** A prospective audit of haematology and oncology patients admitted to CHI at Crumlin. Inclusion criteria are a documented fever exceeding 38 degrees Celsius, and corresponding subsequent absolute neutrophil count below  $0.5 \times 10^9/L$ . Cases were identified through daily morning handover meetings. The study included 15 presentations, chosen according to the inclusion criteria. Data was collected between 26/08/2022 and 30/09/2022, comprising the first audit cycle.

**RESULTS:** 14 children were included in the study, including 6 males and 9 females, comprising 15 FN cases. The primary endpoint is time from first documented contact with a clinical professional to antibiotics administration. This was found to be 2.51 hours with 4/11 (26.7%) receiving antibiotics within 1 hour. Mean time to peripheral blood cultures was 3.24 hours, with peripheral cultures occurring after antibiotic administration in 9/11 (81%). Initial central and peripheral blood cultures were positive in 3/11 (27%) and 2/11 (18%) episodes, respectively. Documentation of

nurse and doctor initial review times was found in 11/15 (73.3%) and 11/15 (73.3%) cases, respectively.

**CONCLUSION:** Overall compliance to commencing antibiotics within 1 hour was 26.7%. An obstacle to prompt antibiotic administration has been disinclination to commencement prior to peripheral blood cultures. Surveying of stakeholders, and interdisciplinary staff education will be required prior to another audit cycle, with the intent being to continue the study further. We aim to promote improved documentation and prioritise timely antibiotic commencement above pending investigations.

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## **GENETIC EPILEPSIES IN CHILDREN UNDER THREE IN A SINGLE CENTRE – A CASE**

**SERIES JK Hannon<sup>1</sup>, H Stokes<sup>1</sup>**

<sup>1</sup>Department of Paediatrics, Mayo University Hospital, Castlebar, Co Mayo

**Introduction:** Childhood epilepsy is influenced by multiple aetiological factors. Determination of a genetic cause can clarify diagnosis and provide recommendations for treatment. We report four cases of intractable seizures in children under three.

**Methods:** Case notes were summarised and a review of relevant literature was undertaken.

**Case 1:** A 22-month-old boy presented following a hypotonic episode. Antenatal and family history were unremarkable. He was non-verbal on presentation. Over the coming months, generalized tonic clonic, focal, prolonged absence and atonic seizures were frequently observed. He experienced significant developmental regression.

MRI brain was normal, EEG was initially normal, with progression to frequent bursts of generalized polyspike wave discharges. Genetic investigations confirmed a MBD5 gene mutation.

**Case 2:** A five-month-old developmentally normal boy presented with multiple tonic seizures characterized by screaming, extension of limbs, staring, and facial twitching. Clusters of events were noted during febrile illness. There was a family history of generalized epilepsy in a cousin. MRI demonstrated an arachnoid cyst of left middle cranial fossa, not of significance, with excess of slow activity and multifocal epileptiform discharges on EEG. Genetic studies confirmed a mosaic PCDH19 gene mutation.

**Case 3:** A 2-year-old girl was referred with multiple absence events, associated with eyelid flickering. There was rapid increase in frequency of episodes. MRI brain was normal, with generalized regular 3 Hz spike wave activity on EEG.

A unifying genetic diagnosis is awaited.

**Case 4:** A 2-year-old girl presented with focal-onset epilepsy, with lip smacking and jerking of limbs occurring on wakening. There was no family history of neurological disorder.

MRI showed multifocal cortical tubers, with EEG demonstrating continuous focal irregular slowing in the left posterior temporal region. Genetic workup revealed a heterozygous pathogenic variant in TSC2 gene.

**Conclusion:** Knowledge of specific genetic mutations inform diagnostic and therapeutic decisions and can aid prediction of neurodevelopmental outcomes, which are significantly varied in epilepsy syndromes. **195**

## **SPLENIC ABSCESS SECONDARY TO TYPHOID FEVER: A RARE COMPLICATION IN CHILDREN**

**M Hassan**<sup>1</sup>, D Rea<sup>2</sup>, O Ó Maoldomhnaigh<sup>1</sup>

<sup>1</sup>Paediatrics Infectious Diseases, Children's Health Ireland at Crumlin, Dublin, Ireland

<sup>2</sup>Paediatrics Radiology, Children's Health Ireland at Crumlin, Dublin, Ireland

### **Background:**

Salmonellae are gram-negative bacilli that can cause various clinical infections including gastroenteritis, enteric fever, bacteraemia and focal infections such as osteomyelitis. Splenic abscess is a rare complication of typhoid fever affecting children in the post antibiotics era.<sup>1-2</sup>

### **Case Report:**

We report the case of a 13-year-old female with an unremarkable past medical history. She moved to Ireland from India at 7 years of age. Upon returning from a recent trip home, she presented to the emergency department with a 2-day history of fever, bilious vomiting, diarrhoea and left upper quadrant abdominal pain. Her C-Reactive Protein was raised at 145 mg/L, had a white blood cell count of  $9 \times 10^9/L$ , microcytic anaemia and transaminitis. She was initially treated with IV Ceftriaxone and her blood culture grew Salmonella Typhi which was sensitive to cephalosporin. Her stool cultures grew Campylobacter. Despite adequate hydration and negative repeated blood cultures, she continued to have high spikes of fevers with episodes of rigors, tachycardia and hypotension that required an administration of IV boluses of fluids and escalation of the antibiotics to Meropenem.

Serial Ultrasound scans of her abdomen showed an initial heterogeneous appearance of an enlarged spleen with a small amount of free fluid surrounding it. Subsequently, a capsular hypoechoic fusiform splenic collection was observed measuring 4.3x2x2 cm. A CT scan of the abdomen confirmed the presence of an intercommunicating abscess within the spleen. Ultrasound-guided percutaneous drainage was performed from the splenic collection and a moderate amount of bloody fluid was drained. Significant clinical improvement was observed thereafter. Repeat ultrasound scans have shown multiple small gallbladder stones without

evidence of cholecystitis or cholecystolithiasis while the splenic abscess kept decreasing in size. She recovered well, completing a 5-week IV antibiotics course and a prolonged course of oral antibiotics.

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## **ENDO PERIPHERAL ARTERIAL TONOMETRY (ENDO-PAT 2000) USE IN PAEDIATRIC PATIENTS – SYSTEMATIC REVIEW**

**J Hayden**<sup>1</sup>, G O'Donnell<sup>1</sup>, I deLaunois<sup>2</sup>, C O'Gorman<sup>1,3</sup>

<sup>1</sup>Department of Paediatrics, University Hospital Limerick (UHL), Limerick, Ireland

<sup>2</sup>Medical Librarian, University of Limerick (UL), Limerick, Ireland

<sup>3</sup>Department of Paediatrics, University of Limerick (UL), Limerick, Ireland

**Objectives:** Endo Peripheral Artery Tonometry (EndoPAT-2000) is a non-invasive technology for measuring endothelial dysfunction (ED). The reactive hyperaemia index (RHI) is resulted and is low when ED is present. We aim to synthesise the literature on paediatric ED that utilised Endo-PAT analysis.

**Design:** A comprehensive systematic review was conducted from January 2015 to March 2021. The databases included Cochrane, MEDLINE EBSCO, EMBASE (Ovid), PUBMED and CINAHL EBSCO. Exclusion criteria were: 1. If a study used a different device; 2. If the study had no results. Inclusion criteria were: 1. Published in the English; 2. More than 50% of study subjects were in the paediatric age range; 3. Data relevant to paediatric age range children could be extrapolated from all data, where not all study subjects were children.

**Results:** Following the removal of duplicates, 156 articles were initially identified. Following exclusion and snow ball searching, 50 articles were included for review. We have subdivided these papers into different systems for ease of reference and have reported our findings in 6 tables: patients with type 1/2 diabetes, obesity, cardiovascular, respiratory, psychiatric conditions and miscellaneous diseases. For each, the study design, population, control group (if available), RHI results and conclusions were reported.

**Conclusions:** A number of papers using Endo-PAT for children with various chronic diseases have evidence of ED. However, in many cases, there has only been a single cohort study using Endo-PAT. Further studies are required to validate these findings and to help characterise the cardiovascular risk profile of children with chronic disease.

Additionally, longitudinal studies are required to evaluate how this ED may change as the child ages and their chronic conditions changes. Consensus on other vascular risk markers that could be included in future studies is ideal and if accomplished, this would facilitate meta-analyses of studies of relatively rare conditions.

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#### **FIRST GENERATION HYPER IGE SYNDROME IN AN 11 YEAR OLD FEMALE PRESENTING WITH ACNEIFORM RASH**

**T Hefferon<sup>1</sup>, B Byrne<sup>1</sup>, M Leahy<sup>1</sup>, M Laing<sup>1</sup>**

<sup>1</sup>Dermatology Department, University College Hospital Galway, Galway, Ireland

Hyper-IgE syndrome (HIES1) is a rare, autosomal dominant primary immunodeficiency characterised by eczema, recurrent staphylococcal skin infections, pneumonia, increased serum IgE and eosinophilia. It is caused by a heterozygous mutation in the STAT3 gene on chromosome 17q21 with variable expressivity. Non-immunological findings including characteristic facies, hyperextensibility, long bone fractures, aneurysms, craniosynostosis and dental abnormalities<sup>1</sup>. We present the case of an 11 year old girl referred to dermatology with an acneiform facial rash. Examination was consistent with papulo-pustular scarring acne of the face and back. History also

significant for recurrent MSSA bacterial lymphadenitis requiring incision and drainage over the preceding 3 years.

Past medical history was significant for the development of moderate severity infantile acne at two weeks old, mild childhood eczema, recurrent childhood otitis media staphylococcus infections resulting in grommet insertion, primary tooth retention requiring extraction and a fractured wrist at age 9.

Due to the burden of infection the patient was referred to immunology in Crumlin. Investigations revealed mild neutropenia, marginally elevated IgA and IgM, normal eosinophils, marginally elevated IgE, normal lymphocyte subsets, intact complement pathways and normal oxidative burst test. Next generation sequencing were significant for STAT3 gene mutation consistent with HIES. As there is no family history this likely represents a de novo mutation. Patient is currently on prophylactic azithromycin and low dose isotretinoin with good effect .

This case represents a patient with both immunological and non-immunological features of HIES1, marginally elevated IgE, absence of eosinophils and negative family history.

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## **IMPROVING DOCUMENTATION FOR CHILDREN WITH SEVERE NEUROLOGICAL IMPAIRMENT IN CHILDREN'S HEALTH IRELAND: A QUALITY IMPROVEMENT PROJECT**

**DH Hillick<sup>1</sup>, SMC Mc Cormack<sup>1</sup>, ML Ledden<sup>1</sup>, ZE Elbishari<sup>1</sup>, JB BALFE<sup>1</sup>**

<sup>1</sup>Paediatric Neurodisability, Children's Health Ireland, Dublin, Ireland

**Background:** Children with severe neurological impairment(SNI) have high healthcare needs, attend multiple specialities and have frequent interactions with healthcare and disability services.

When children with SNI are admitted to hospital, this often occurs out-of-hours and involves staff that are unfamiliar with them. Comprehensive summaries are often non-existent in the available healthcare records, medication records may not be up-to-date and locating all the relevant information from multiple sources is time constraining and difficult. There is a risk that the absence of accessible clinical information may impact on quality of care provided.

**Aims/Objectives:** We aim to improve the care provided to children with SNI and their families by focusing on reducing: 1.the burden on clinicians to locate key information, 2. the need for parents to repeat background details, and 3.medication and transcription errors.

**The QI project:** Our multidisciplinary quality improvement team completed the Lean Six Sigma Green Belt Certification. Using a data collection template-proforma of what was deemed critical-to-quality(CTQ) information, time taken to capture background information(through paper charts and electronic systems) and the number of absent details were measured on the records of a sample of children with SNI. Acute admission notes were also analysed, and the number of missing data points compared to the template.

**Outcome:** Seven patient records were reviewed. The average time taken to find background information was 22 minutes, a time cap of 25 minutes was placed. 62% of CTQ background information was complete. Acute admission notes had 42% completeness of CTQ information.

**Future Plan:** A two-page SNI patient summary proforma has been developed with a planned phased rollout in 2022/2023.We aim for each family to have a copy, and an electronic copy to be available to relevant healthcare professionals on the secure hospital computer system with ultimate integration to the electronic health record.This will be regularly updated electronically at each outpatient clinic and inpatient admission.

## **AN EVALUATION OF THE YIELD FROM MICROBIOLOGICAL TESTING IN CHILDREN WITH SEVERE NEUROLOGICAL IMPAIRMENT (SNI) WITH RESPIRATORY TRACT INFECTIONS**

**N Howard-James<sup>1</sup>, S McCormack<sup>1, 4</sup>, B Elnazir<sup>2, 3</sup>, D McDonald<sup>1, 3</sup>**

<sup>1</sup>Department of Child Development and Neurodisability , CHI Tallaght, Dublin, Ireland

<sup>2</sup>Respiratory Department, CHI Tallaght, Dublin, Ireland

**Background and Aims:** Recurrent respiratory tract infection is a frequent concern in the care of Children with Severe Neurological Impairment (SNI), and is one of the most common causes for hospitalisation in this group. There is limited data available on the ideal investigative process to maximise yield and diagnosis of causative respiratory pathogens to guide treatment.

**Methods:** This evaluation reviewed the investigations carried out on children aged 5-10 years with SNI who presented to CHI at Tallaght with symptoms suggestive of respiratory tract infection in the past 5 years.

**Results:** Of 22 patients fulfilling this criteria, 11 had a presentation with respiratory tract infection during the study period. Number of infections per patients ranged from 1-5, with a mean of 2.2 infections per patient. Viral NPA was taken in 7 (28%), viral throat swab in 6 (24%) and bacterial throat swab in 4 (16%). Sputum samples were not obtained in any patients, stool was obtained in 6 (24%) and blood cultures obtained in 11 (44%). Chest radiograph was performed in 84% of cases, of which 61% were abnormal. None of the patients with a bacterial swab or blood culture taken yielded a positive result. Of the 11 patients who had viral testing performed, a viral organism was identified in 8 (72%). The most common organism isolated was rhino-enterovirus, followed by RSV.

**Conclusion:** Respiratory tract infection is a common presentation with variation in the investigative process. Viral pathogens predominated in testing, although this may be due to difficulties in sputum exploration in this group. This review will be extended to children with SNI of all ages to further characterise the yield and aetiologies.

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#### **GENOTYPE, PHENOTYPE CORRELATION OF SCN2A MUTATION; A CASE REPORT**

**NI Jameel, AN Murphy**

Paediatrics, University hospital limerick, Limerick, Ireland

**Aim:** To report a case of SCN2A mutation induced seizure in a 2-month-old infant with unremarkable family or personal history beside maternal history of recurrent miscarriages and multiple attempts of IVF.

**Method:** We describe the clinical presentation, examination findings, results of hematological, metabolic & genetic investigation, video footage, EEG, radiological findings, treatment, and outcome in our patient.

**Result:** Our patient is the first child to healthy non consanguineous Caucasian parents and was brought in by parents following an episode of arm flexion/stiffening, unresponsiveness, pallor, shallow breathing followed by 45-60 minutes of lethargy. Initial blood work up including ECG was normal with no dysmorphic features. The baby was admitted for observation and later in HDU had two similar events at which stage working diagnosis was migratory epilepsy of infancy and commenced on phenobarbital and Keppra. Investigation including epilepsy panel, metabolic screen, CSF neurotransmitters, microarray and karyotyping was sent. Genetic results later confirmed the diagnosis of SCN2A mutation. Meanwhile our patient continued to have intermittent focal seizure episode and eventual seizure control was achieved via Tegretol. At the age of 8 months our patient is developmentally normal and seizure free for over 4 months and we continue to closely monitor him.

**Conclusion:** This patient exhibited a novel phenotype for heterozygous SCN2A variants, and the case provides novel insights into the genotype–phenotype correlation for SCN2A variants.

## A CASE SERIES OF TUBO-OVARIAN ABSCESES COMPLICATING PELVIC INFLAMMATORY DISEASE IN A PAEDIATRIC CENTRE.

KJ Kelleher<sup>1</sup>, N Murphy<sup>2</sup>, S Awadalla<sup>3</sup>, C O'Maoldomhnaigh<sup>1</sup>

<sup>1</sup>Infectious Diseases, CHI @ Temple Street, Dublin, Ireland

<sup>2</sup>Gynecology, CHI @ Temple Street, Dublin, Ireland

<sup>3</sup>General Surgery, CHI @ Temple Street, Dublin, Ireland

**Aims:** Pelvic inflammatory disease (PID) and tubo-ovarian abscesses (TOAs) are unusual in paediatrics. We aim to highlight the variable population, presentation and aetiology of PID/TOA in children.

**Methods:** Retrospective chart review.

### Results:

Case 1: 15 year-old girl presented with two weeks of abdominal pain unresponsive to cephalexin. Two ultrasounds on presentation showed a moderate volume of complex free-fluid in the pouch-of-Douglas. One was reported as suspicious for either appendicitis or TOA, the second favoured a ruptured haemorrhagic cyst. She was therefore discharged. She represented, now symptomatic for four weeks with severe abdominal pain, radiating to the right shoulder, vomiting, diarrhoea and fainting. The deterioration coincided with an unusually heavy and painful menses. She disclosed being sexually active without barrier contraception. Repeat ultrasound demonstrated a pyosalpinx with free fluid in the abdomen. She was treated with two weeks of broad-spectrum antibiotics. STI screening was diagnostic of *Chlamydia trachomatis* and *Neisseria gonorrhoea* but negative for HIV, Hepatitis B/C and Syphilis.

Case 2: A 14 year-old girl had a background of vaginal atresia, duplex kidney and recurrent UTIs. Her vaginal atresia presented as haematometrocolpos and haematosalpinx. She had a normal karyotype without mosaicism or SRY gene detection. She had vaginal reconstruction surgery two years previously with secondary amenorrhoea leading up to her presentation. She attended with one day of severe abdominal pain, fever, vomiting, diarrhoea and vaginal bleeding. Ultrasound and CT were concerning for a tuboovarian abscess. She made a good recovery clinically and radiologically, completing a four week long extended-spectrum antibiotic course.

**Conclusion:** TUOs are a severe complication of PID, typically occurring in young sexually active women as an ascending infection caused by the organisms of case 1. In those not sexually active, ascending infection usually occurs during menses due to hormonal and cervical mucous changes, and retrograde menstruation(1) as was likely in our second case. Increased awareness may reduce future delayed diagnoses.

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## **SAVING THE GOOD EYE: ENDOPTHALMITIS PRESENTING IN A 5 YEAR OLD BOY WITH A HISTORY OF**

### **RELAPSED RETINOBLASTOMA**

**KJ Kelleher**<sup>1</sup>, S Farrell<sup>2</sup>, B Freyne<sup>1</sup>

<sup>1</sup>Infectious diseases, CHI @ Temple street, Dublin, Ireland

<sup>2</sup>Ophthalmology, CHI @ Temple street, Dublin, Ireland

**Aims:** To highlight a rare case of post-intravitreal chemotherapy-associated endophthalmitis in a child with no vision in the unaffected eye.

**Method:** Retrospective chart review

**Results:** A five year old boy had a background of bilateral group D retinoblastoma at age 1 year old, secondary to a de novo heterozygous pathological variant in the RB1 gene (RB1 c.585G>A p.(Trp195Ter)). He completed initial chemotherapy uneventfully but with the total loss of vision in his left eye and partial vision-loss in his right eye. Aged 4 years, he experienced a late recurrence of retinoblastoma in his betterseeing right eye requiring intravitreal chemotherapy.

He attended his local centre with eye-pain and erythema, which began 36 hours after receiving intravitreal chemotherapy. He was treated initially with steroid eye-drops and non-steroidal anti-inflammatories with transient improvement of symptoms. On day three of symptoms his



oncologist recommended attendance of our tertiary centre due to progressing eye pain, erythema, chemosis and reduced visual acuity.

Dense fibrous plaques were seen intraocularly and he underwent emergent surgery for anterior chamber tap and vitrectomy. Treatment consisted of empiric intravitreal ceftazidime and vancomycin, and seven days of systemic ciprofloxacin, clindamycin and rifampicin. Topical ofloxacin, atropine 1% and prednisolone eye drops were also applied. The fluid samples from the anterior chamber was sterile, however the vitreous fluid grew a pure growth of *Streptococcus gordonii*. At discharge and follow up his vision was improving but had not returned to his baseline.

**Conclusion:** Endophthalmitis is a medical emergency with poor visual prognosis. *Streptococcus spp.* are common causes of both endogenous and post-intravitreal injection endophthalmitis(1). Post-intraocular injection or surgery, it is important to be aware of the permanent risk to vision infection carries and to refer emergently to tertiary services any such vulnerable, symptomatic patients. Facemasks, silence and iodine preparation may help reduce the risk of oral flora contamination during intraocular injection(2).

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## ACCESS TO URINE TOXICOLOGY TESTING IN IRISH EMERGENCY DEPARTMENTS

SK Kyne<sup>1</sup>, LH Heffernan<sup>2</sup>, MJ Jee<sup>2</sup>, ER Ryan<sup>1,3</sup>

<sup>1</sup>Paediatric Department, University Hospital Galway, Galway, Ireland

<sup>2</sup>Emergency Department, University Hospital Galway, Galway, Ireland

<sup>3</sup>Neonatology Department, University Hospital Galway, Galway, Ireland

**Introduction:** The use of illicit drugs in Ireland is on the increase<sup>1</sup>. Urine toxicology tests are sometimes used in Emergency Departments to test for the presence of illicit drugs.

**Aims:** We wanted to evaluate access to urine toxicology testing across Emergency Departments in Ireland. We also enquired about potential barriers to accessing urine toxicology testing.

**Methods:** We decided to evaluate access to urine toxicology in all 24-hour Emergency Departments. This accounts for 28 hospitals, including paediatric facilities, adult facilities, and facilities that treat paediatric and adult patients.

We used the Irish Trainee Emergency Research Network (ITERN) to distribute our survey via mailing list to all the Emergency Departments in the Republic of Ireland. We used Google Forms and Excel to compile our survey results.

**Results:** We surveyed all 28 Emergency Departments in Ireland. In the 2 hospitals with just paediatric patients, both had point-of-care testing in the Emergency Department. In the other hospitals, just 14 out of the remaining 26 hospitals had access to urine toxicology in the Emergency Department (either through point of care testing or the laboratory). 10 of these hospitals had no access to urine toxicology testing in the institution or through an external lab. Barriers to accessing urine toxicology were cost and the need to link with senior clinicians or other hospital teams before obtaining testing or obtaining results.

**Conclusion:** With the increasing use of illicit drugs in Ireland<sup>1</sup>, we wanted to study how many Emergency Departments in Ireland had access to urine toxicology. The results vary nationally, and the barriers to testing differ between centres.

1. Mongan D, Millar SR, Galvin B. The 2019-20 Irish National Drug and Alcohol Survey: Main Findings.

**MAINTENANCE IMMUNOSUPPRESSION OPTIMISATION IN PATIENTS POST RENAL TRANSPLANT FOLLOWED**

**UP BY THE NEPHROLOGY DEPARTMENT IN CHILDRENS HEALTH IRELAND TEMPLE STREET**

**RM Mannion<sup>1</sup>, MR Riordan<sup>1</sup>, AA Awan<sup>1</sup>**

<sup>1</sup>Nephrology Department, CHI Temple Street, Dublin , Ireland

The goal of immunosuppression is to minimise the risk of graft rejection, while minimizing drug side effects. The primary side effects of immunosuppression medications post renal-transplant are infection, nephrotoxicity, and malignancy. The most common viral infections are EBV, CMV and BK1.

Participants included all patients followed up by the nephrology department in CHI Temple Street. Data was collected from EMED online database, and chart review. Data variables included demographic information, date of transplant, primary diagnosis, height, weight, medications and serology blood tests.

There were 54 patients included, 26 male and 28 female. The most common primary diagnoses were renal dysplasia, congenital nephrotic syndrome and obstructive uropathy. 53 patients were on tacrolimus and 1 patient was on Ciclosporin. 34 patients were on Mycophenolate Mofetil (MMF), and 16 were on Azathioprine. 4 patients were on neither antimetabolic agents. Of the 34 on MMF half were on Cellcept solution and half were on Myfortic tablets. Of those on Cellcept there was 13 who dosage had fallen below optimal prescription. Of these patients 9 had negative virology for CMV, EBV, and BK. The other 4 had low grade EBV viraemia previously. Of those on Myfortic there was 8 who dosage had fallen below optimal prescription. Of the 8 there were 4

patients with EBV viraemia previously. Of the 16 patients on Azathioprine there was one patient below optimal dosing.

A proportion of this cohort were under the optimal dosing of antimetabolite maintenance immunosuppression dosing. This study also excluded viral infections such as EBV, CMV and BK as the reason for reduction in immunosuppression. We have optimised the dose of MMF in these patients and we have introduced this as a standard part of annual review to ensure optimal immunosuppression regime.

(1) McCaffrey J, Bhute VJ, Shenoy M. CMV, EBV, JCV and BKV infection and outcome following kidney transplantation in children initiated on a corticosteroid-minimisation immunosuppressive regimen. *Pediatr Nephrol*. 2021 Oct;36(10):3229-3240. doi: 10.1007/s00467-021-05047-2. Epub 2021 Apr 6. PMID: 33825043.

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#### **MULTI-DISCIPLINARY MANAGEMENT OF RETT SYNDROME: AN AUDIT OF COMPLIANCE WITH CONSENSUS GUIDELINES**

**R McCarthy**<sup>1</sup>, S McCormack<sup>1</sup>, D McDonald<sup>1</sup>

<sup>1</sup>Neurodisability, CHI @ Tallaght, Dublin, Ireland

**Aims:** Our aim was to compare our clinical practice against pre-established guidelines that outline how best to care for these children. *Fu et al*<sup>(1)</sup> published a comprehensive breakdown of the management of RETT syndrome throughout childhood and in to adult years, and this will serve as a template for us to audit our own multi-disciplinary management.

**Methods:** As our cohort of patients in this group was quite small, we were able to quickly identify the patients in this study through our own records. We then retrospectively examined their medical documents to establish their overall management to date and compared this to the current literature.

**Results:** In total we identified 6 patients in our cohort who had a confirmed diagnosis of RETT syndrome. We developed multiple categories in which to evaluate whether we were meeting international standards on care, such as proper documentation of genetic results, referral to appropriate medical and surgical subspecialties, involvement of our multi-disciplinary colleagues (physio, OT, Dietetics), and clear community care follow up. Areas that scored well included MDT involvement (100%), frequent developmental checkups (100%), and certain subspecialty involvement such as orthopedics (83%) and neurology (83%), clear medication documentation (100%), and appropriate community care (100%). Some areas which scored poorly included clear documentation of original genetic results (33%), regular tanner staging (33%), audiology assessments (16%), and referrals to certain subspecialties such as Urology (0%).

**Conclusions:** While we appear to be meeting several of our goals for the holistic management of these children, there are some areas which require closer attention. What is clear from this study is that the care of these children is extremely complex and requires a great deal of collaboration on all sides to optimize their quality of life.

1) Fu C, Armstrong D, Marsh E, Lieberman D, Motil K, Witt R, Standridge S, Nues P, Lane J, Dinkel T, Coenraads M, Hehn JV, Jones M, Hale K, Suter B, Glaze D, Neul J, Percy A, Benke T. Consensus guidelines on managing Rett syndrome across the lifespan; Volume 4, Issue 1. BMJ Paediatrics Open. BMJ Journals

## **BLENDED DIET FOR ENTERAL TUBE FEEDING IN YOUNG PEOPLE - A SYSTEMATIC REVIEW OF THE BENEFITS AND COMPLICATIONS**

**S McCormack**<sup>1,2</sup>, K Patel<sup>2,3</sup>, C Smith<sup>4</sup>

<sup>1</sup>Department of Child Development and Neurodisability, CHI at Tallaght, Dublin, Ireland

<sup>2</sup>Paediatrics and Child Health, Brighton and Sussex Medical School, Brighton, UK

<sup>3</sup>Department of Paediatrics, Royal Alexandra Children's Hospital, Brighton, UK

<sup>4</sup>Department of Paediatric Dietetics, Royal Alexandra Children's Hospital, Brighton, UK

**Background and Aims:** Interest and use of blended diets (BD) for young people who are tube fed has significantly increased in the last decade, driven primarily by the desires of motivated caregivers. This review identified, appraised, and synthesised the available evidence on the benefits and complications of the BD versus commercial feeds.

**Methods:** A systematic review following PRISMA guidance and registered with PROSPERO was conducted across PubMed, Embase, CINAHL, Scopus and Cochrane up to August 2022. Inclusion criteria: English language studies including (1) children, (2) original research (interventional and observational) and (3) examination of BD outcomes. Exclusion criteria were (1) unoriginal research or case reports, (2) focus on feeding management, preparations, or attitudes and (3) comparing commercial blends only. Data was synthesised using a formal narrative synthesis approach established by Popay et al, using the Mixed Methods Appraisal Tool.

**Results:** 806 database results were identified and 61 were sought for retrieval. Full text article review revealed 7 eligible studies, involving 267 participants (age range 9 months - 26 years). Studies reported differences in GI symptoms (n=222), medication use (n=99), growth (n=189), complications or adverse events (n=91). The studies varied to an extent that deemed them

collectively unsuitable for meta-analysis or other pooled statistical analysis. The results indicate towards positive outcomes, particularly in gastrointestinal symptom control with few reports of mild adverse events in the included studies.

**Conclusion:** There is a paucity of data in this area and much heterogeneity in included studies, but the available literature points towards positive outcomes. This is an important and highly relevant topic and more primary research, ideally using standardised reporting, is required to answer the key questions.

## COMMUNICATION METHODS USED BY CHILDREN WITH SEVERE NEUROLOGICAL IMPAIRMENT- A REVIEW OF DESCRIPTIONS FROM CLINIC LETTERS

M Ledden<sup>1</sup>, S McCormack<sup>1,2</sup>, S Kelly<sup>1</sup>, D McDonald<sup>1,3</sup>, J Balfe<sup>1,3</sup>

<sup>1</sup>Department of Child Development and Neurodisability, CHI at Tallaght, Dublin, Ireland

<sup>2</sup>Department of Paediatrics, National University of Ireland, Galway, Ireland

<sup>3</sup>Department of Paediatrics, Trinity College Dublin, Ireland

**Background and aims:** Children with Severe Neurological Impairment (SNI) communicate in various ways including the use of assistive technologies. However, clinicians often do not consider this in their clinical review in hospital and often describe children in terms of what they cannot do rather than how they communicate. This project aimed to review the descriptions of communication methods used by children with SNI from clinic letters from the last review summary.

**Methods:** The initiative was registered with the TUH Quality Improvement department. Patients with SNI aged from 5 to 16 years were identified from the departmental database. Clinic summary letters were retrospectively reviewed to evaluate the communication methods described. Communication descriptors were categorised as: no mention, verbal, non-verbal, gesture, facial expression, assistive device, or a combination of methods.

**Results:** 56 patient records were reviewed. 30 (54%) had no mention of the child's method of communication at any point in the letter. 26 (46%) mention communication methods, of those 8 (31%) noted the child was verbal, 6 (23%) used "non-verbal", 7 (27%) mentioned the use of assistive technology devices, 6 (23%) communicate through gesture and 5 (19%) mentioned facial expression. 8 (31%) records mention more than one communication method.

**Conclusions:** Children with SNI communicate using a wide variety and combination of methods and there is great variation in the documentation of these methods used. Following this evaluation, the team are more aware of the descriptors used and clinic proformas were adapted to incorporate a more holistic approach to communication of children with SNI.



## DEVELOPING A COMMON DATASET FOR CHILDREN WITH SEVERE NEUROLOGICAL IMPAIRMENT – A

### SCOPING REVIEW AND INTERNATIONAL DELPHI STUDY

S McCormack<sup>1,2</sup>, N Allen<sup>2</sup>, A Mahony<sup>1</sup>, A Collins<sup>1</sup>, J Balfe<sup>1,3</sup>, D McDonald<sup>1,3</sup>

<sup>1</sup>Department of Child Development and Neurodisability, CHI at Tallaght, Dublin, Ireland

<sup>2</sup>Department of Paediatrics, National University of Ireland, Galway, Ireland

<sup>3</sup>Department of Paediatrics, Trinity College Dublin, Ireland

**Background and Aims:** Children with Severe Neurological Impairment comprise a small but significant cohort of children who have rarely been studied together previously. The absence of data is a barrier to meaningful planning for resource allocation for health and social care into adulthood. Their diagnoses often exclude this group from other research, and without a common dataset and core outcomes little progress can be made towards research to inform and manage morbidity and identify risk to longevity. Key to advancing research on children with SNI is developing this common dataset for international collection of baseline and longitudinal data.

**Methods:** An international steering committee was formed and the project was registered with the Core

Outcome Measures for Effectiveness Trials COMET initiative ([www.comet-initiative.org/Studies/Details/1976](http://www.comet-initiative.org/Studies/Details/1976)). The scoping review utilised the PRISMA extension for scoping reviews as a guide. Searches were conducted of PubMed, Embase, CINAHL Plus and Scopus until February 2022 to identify datasets, elements, registry variables and outcomes relevant to this population. The initial draft dataset was fed back to the steering committee and the Delphi study designed and consensus set in line with the Core Outcome Sets- Standards for Development and Reporting (COS- STAD/ STAR).

**Results:** Database searches revealed 1766 results. Relevant variables were extracted from 37 records and a total of 187 variables were included in the initial dataset. 71 participants from 9 countries across 7 disciplines completed at least 1 round of the Delphi process. Participants were also invited to suggest extra variables. Following 3 rounds of Delphi surveys, 22 variables formed the core “minimum” dataset and 87 formed the secondary “additional” dataset.

**Conclusions:** This international common dataset will be central to future comparative and collaborative research. Ultimately, this will better inform expectant care and will support the development of evidence based clinical care pathways and guidelines.

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### **ANTIMICROBIALS IN THE TREATMENT OF SEPTIC ARTHRITIS AND OSTEOMYELITIS: A SINGLE CENTRE EXPERIENCE AND AUDIT**

**BM Melley**<sup>1</sup>, FA Alberghina<sup>1</sup>, ML Lawler<sup>2</sup>, CE Egan<sup>1</sup>, KT Tedford<sup>3</sup>

<sup>1</sup>Orthopaedic Surgery, CHI Crumlin, Dublin, Ireland

<sup>2</sup>Infectious Disease, CHI Crumlin, Dublin, Ireland

<sup>3</sup>Pharmacy, CHI Crumlin, Dublin, Ireland

**Aims:** To compare choice and dose of antimicrobials prescribed in osteomyelitis (OM) and septic arthritis (SA) in children with the recommendations in the CHI (Children's Health Ireland) formulary antimicrobial guidelines.

To compare admission work-up for OM and SA with that recommended by BOAST (British Association of Orthopaedics Standards for Trauma) guidelines on 'Management of Children with Acute Musculoskeletal Infection'.

To highlight the organisms isolated from cases of SA and OM in children.

**Methods:** This began as an audit project in which data regarding choice of antimicrobial and dose in treatment of OM and SA in a paediatric centre was audited against the CHI formulary. In addition, this led to an observation of how these conditions were managed in comparison to BOAST guidelines on musculoskeletal infection in children. The organisms isolated were reviewed where available.

#### **Results:**

CHI formulary: On first cycle there was 54.5% compliance with antibiotic choice as per the CHI formulary. This increased to 80% on reaudit.

Boast Guideline Audit:

- 100% of patients had full blood count and C Reactive protein blood sampling on admission - 47% of patients had blood cultures and ESR blood sample taken on admission (prior to starting antibiotics)
- While 64.7% had an MRI on admission, only 27.3 % of these were performed within 48 hours - 88.2% had an XR of the affected joint

**Conclusion:** Prescribing in Paediatric practice is significantly different to that of adult practice as choice of antimicrobial in musculoskeletal infection changes based on age group. This highlights the need for guidelines/formulary to be on hand and user-friendly in order to optimise treatment of septic arthritis and osteomyelitis from the time of admission. The isolation of *Kingella* species in the above cases also highlights the need for prescription of Cefazolin in those under five years of age.

1. Brown DW, Sheffer BW. Pediatric Septic Arthritis: An Update. *Orthop Clin North Am.* 2019;50:461-70.
2. Ireland CsH. Children's Health Ireland Antimicrobial Guidelines. 23rd May 2022 ed. Clinibee2021. 68.
3. Orthopaedics BAo. The Management of Children with Acute Musculoskeletal Infection 2022. Standards for Trauma. 1 ed: British Association of Orthopaedics; 2022.

**VTE PROPHYLAXIS AND ANTICOAGULATION IN THE PAEDIATRIC POPULATION - STICKING TO SAFETY: A**

**QUALITY IMPROVEMENT PROJECT**

**BM Melley**<sup>1</sup>, SF Forde<sup>2</sup>, HW Walshe<sup>3</sup>, SB Bradley<sup>3</sup>, CE Egan<sup>1</sup>, KL Lavelle<sup>3</sup>, BN Nolan<sup>4</sup>

<sup>1</sup>Orthopaedic Surgery, CHI Crumlin, Dublin, Ireland.

<sup>2</sup>General Paediatrics, CHI Crumlin, Dublin, Ireland.

<sup>3</sup>Pharmacy, CHI Crumlin, Dublin, Ireland.

<sup>4</sup>Haematology, CHI Crumlin, Dublin, Ireland.

**Aims:**

- To determine how many patients, on a single evening in a Paediatric Centre, were on an anticoagulant drug.
- To determine whether the indication for the prescription of the anticoagulant is clearly documented.
- To highlight whether there be a need for change in the prescribing practice with regard to anticoagulants in paediatric patients

**Methods:** On a single day, the drug kardex of each inpatient on a single evening was reviewed for the presence of anticoagulant drugs. The prescription of an anticoagulant was recorded, along with the age and gender of the patient, the speciality they were admitted under, the presence of a documented indication, the indication itself and whether or not there was anything on the front of the drug Kardex to indicate, at a glance, that the patient was on an anticoagulant.

**Results:** A total of 139 drug kardexes were reviewed, across 10 wards. These included 38 surgical patients, 94 medical patients and 7 joint admissions. 11.5% (16/139) of patients were on an anticoagulant drug. Indication for the prescription of an anticoagulant was documented in the

kardex for 1 patient. There were no alerts noted on the front of the relevant kardexes to indicate that the patient was on an anticoagulant.

**Conclusion:** 11.5% of inpatients (excluding PICU) on a given day were being treated with an anticoagulant drug. Alongside the inherent risk of these drugs, the lack of any alerts or triggers to healthcare professionals looking after these patients, particularly on call, poses a significant clinical risk.

For this reason, an alert sticker has been developed and is planned to be rolled out across CHI hospitals to help improve patient safety in the area of anticoagulation.

The Association of Paediatric Anaesthetists of Great Britain and Ireland guidelines committee on thromboprophylaxis in children -

Prevention of Peri-operative Venous Thromboembolism in Paediatric Patients

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#### **VTE PROPHYLAXIS IN PAEDIATRIC ORTHOPAEDICS - OUT OF OUR COMFORT ZONE?: A SURVEY STUDY**

**BM Melley**, KV Vejsbjerg, CE Egan

<sup>1</sup>Orthopaedic Surgery, CHI Crumlin, Dublin, Ireland.

**Aims:** To determine the comfort level of members of the Paediatric Orthopaedic Surgery team (both NCHDs and consultants) in identifying patients in need of VTE prophylaxis and in prescribing same.

**Methods:** A survey was carried out among consultants and NCHDs via survey monkey.

**Results:** While 79% of participants had previously had a patient requiring VTE prophylaxis, only 10.5% were comfortable in prescribing this treatment. 58% of participants were not aware that there is currently no guideline in CHI (Children's Health Ireland) for VTE prophylaxis in the paediatric population. 37% of participants correctly identified (when listed) all risk applicable risk factors for VTE.

**Conclusion:** While VTE is much less common in the paediatric population when compared with the adult population, there are still a significant number off paediatric population who will demonstrate risk factors for VTE when undergoing Orthopaedic surgery. For this reason, in conjunction with the results outlines here, there is a need for more guidance in VTE prophylaxis prescribing in children along with education and support identifying the relevant risk factors.

1. Carrillo LA, Kumar A, Harting MT, Pedroza C, Cox CS, Jr. Venous thromboembolism risk factors in a pediatric trauma population. *Pediatr Surg Int.* 2019;35:487-93. 2. Landisch RM, Hanson SJ, Cassidy LD, Braun K, Punzalan RC, Gourlay DM. Evaluation of guidelines for injured children at high risk for venous thromboembolism: A prospective observational study. *J Trauma Acute Care Surg.* 2017;82:836-44. 3. Mahajerin A, Petty JK, Hanson SJ, Thompson AJ, O'Brien SH, Streck CJ, et al. Prophylaxis against venous thromboembolism in pediatric trauma: A practice management guideline from the Eastern Association for the Surgery of Trauma and the Pediatric Trauma Society. *J Trauma Acute Care Surg.* 2017;82:627-36.

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## TARGETED ANTIEPILEPTIC TREATMENT IN GENETIC EARLY INFANTILE EPILEPTIC ENCEPHALOPATHY

S Mullen, A Fisher, D Webb, S Byrne, M O'Regan

<sup>1</sup>Department of Neurology, CHI at Crumlin, Dublin, Ireland

**Background:** A newborn girl was admitted for evaluation of suspected seizures following a pregnancy complicated by abrupt growth arrest at 36/40 necessitating an emergency caesarean section. Her events emerged on day 4 of life.

**Case Description:** The child had witnessed events appearing shortly after admission. Event semiology included sudden arousal, cry, tonic posturing with head deviation, hemifacial spasm and myoclonic jerks.

Diagnostic testing for infectious and metabolic aetiologies was normal or negative. EEG showed a burst suppression pattern and two ictal events were captured characterized by early diffuse attenuation followed by right hemispheric periodic discharges. MRI was suggestive of periventricular leukomalacia.

Seizure frequency increased rapidly throughout admission peaking at 80 seizures/day and were accompanied by an altered mental state. Treatment with Phenobarbitone, Levetiracetam, Vigabatrin, Zonisamide and a ketogenic diet were ineffective. Rescue doses of benzodiazepines and Paraldehyde had a limited and transient effect. However, a rescue dose of phenytoin attenuated her seizures for 24 hours.

Trio exome sequencing analysis showed a de novo pathogenic variant in the SCN2A gene associated with a gain of function at the voltage gated Na channel 1.2. Following this result, the patient was commenced on Phenytoin prophylaxis and rapidly became seizure free. She regained oral feeding within 2 weeks.

**Conclusion:** Targeted treatment may have a dramatic impact on epilepsy syndromes with onset in early infancy and early genetic diagnosis should be sought.

## **POST-OPERATIVE ANTIBIOTIC MANAGEMENT IN CHILDREN WITH ACUTE APPENDICITIS ; AN AUDIT OF COMPLIANCE**

**PM Mwesigye**, LW Wrenne, AR Rowan, ST Tormey

<sup>1</sup>Surgery, University Hospital Limerick, Limerick, Ireland

**AIMS:** Recommendations for post-operative antibiotics for acute appendicitis were published by the World Society of Emergency Surgery Jerusalem(WSES). Implementation of practice recommendations is often difficult.

The purpose of this audit was to measure post-operative antibiotic compliance in the pediatric population at the University Hospital of Limerick.

Secondly, we wanted to assess any barriers to proper antibiotic prescribing and make actions to improve appropriate and correct usage.

**METHODS:** We completed a retrospective review of children who underwent appendectomies starting from an initial sample in October 2021 to December 2021. This data collected over 3 months constituted 45 patients.

Data used in the analysis included date of birth, weight, type of procedure (open vs laparoscopic), complications, type and duration of antibiotics used.

**RESULTS:** 45 patients were included with age ranging from 4-16yrs. 53.3% had uncomplicated appendicitis, 42.2% complicated appendicitis and 4.4% with other pathologies diagnosed intra-operatively. It was found that 24.4% had no weight documented in their drug charts, 20% had incorrect doses of antibiotics charted for their weight and 24.4% had unclear doses. 4.4% cases exceeded the recommended duration of 7 days antibiotics. Many patients with uncomplicated appendicitis received antibiotics (55.5%) despite published recommendations as did those with complicated appendicitis (42.2%)

**CONCLUSION:** Acute appendicitis is the most common surgical emergency in children but early diagnosis and treatment remains challenging due to atypical clinical features and difficulty of obtaining a reliable history and physical examination (1).



There is continued prescription of post-operative antibiotics in excess of published recommendations indicating need for improvement and education. Specific pediatric dosage education can help decrease medication errors.

Over treatment leads to potential unnecessary increased length of stay, costs and medication errors (2). A re-audit of this data will take place this year at same time period (October 2022-December 2022) to complete the audit cycle.

1. Almaramhy, Hamdi Hameed. "Acute Appendicitis in Young Children Less than 5 Years: Review Article." *Italian Journal of Pediatrics*, vol. 43, no. 1, 2017. 2. Di Saverio, Salomone, et al. "WSES Jerusalem Guidelines for Diagnosis and Treatment of Acute Appendicitis." *World Journal of Emergency Surgery*, vol. 11, no. 1, 2016.

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### **WHEN IS A ZEBRA NOT A ZEBRA?**

**Ciara O'Neill**<sup>1</sup>, A O'Donoghue<sup>1</sup>, S Crowe<sup>1</sup>

<sup>1</sup>Paediatric Intensive Care, CHI at Crumlin, Dublin, Ireland

**Background:** During the pandemic we admitted children and adolescents with symptoms and signs suggestive of myocarditis. Most patients had a recent history of exposure to COVID19 infection, and had recent pyrexia, joint or muscle pain, and lethargy. Myocarditis secondary to viral infection was described during this period. There were also concerns around an association of myocarditis with vaccination against coronavirus. Referrals from around Ireland were often made through the infectious diseases team. This may have contributed to diagnostic bias for some presentations.

**Case Presentation:** Two patients age 12 and 14 years had symptoms of cardiac illness including shortness of breath on exertion, dizziness and fatigue. The first child had a maculopapular rash and oral blisters. His history was short, with normal health the preceding week. He was COVID positive at presentation to ED. The second teenager presented to another hospital with chest pain on a short background of lethargy and shortness of breath. His family had COVID infection in the 2

weeks before his illness. He had a cardiac arrest in hospital and required support with an intra-aortic balloon pump, inotropes and mechanical ventilation. Both patients had pre-existing cardiac pathology. The diagnosis was obscured initially due to an overlap of symptoms consistent with covid infection.

**Conclusion:** The unusual situation of a pandemic shifted diagnostic focus onto viral illness, and may have made the diagnosis of rare illnesses more challenging.

## THE DOCUMENTATION OF AND ADHERENCE TO THE LOW RISK ANKLE RULE (LRAR) PAEDIATRIC GUIDELINE WHEN ORDERING XRAYS FOR ANKLE TRAUMA IN A PAEDIATRIC EMERGENCY

DEPARTMENT C O'Hagan, T Bolger

<sup>1</sup>Tallaght Paediatric Emergency Department, CHI at Tallaght, Dublin 24, Ireland

**Aims:** The aim of this audit was to firstly assess whether there is appropriate documentation of clinical findings when dealing with ankle trauma in the Emergency Department. We analysed if clinical guidelines such as the LRAR are being adhered to when ordering ankle xrays. From the significant findings found on the xrays we look at whether the LRAR is a good tool at detecting clinically important fractures.

**Methods:** In this audit we looked at 100 ankle x-rays carried out in our emergency department. We looked at whether there is documentation of the LRAR having been applied and whether the guideline was followed. We also analysed any significant findings on the x rays which have the LRAR clearly documented.

**Results:** Our results showed that out of the 100 x rays analysed, 95 of them were for ankle trauma. Of these, 89% had the LRAR or examination findings consistent with the LRAR documented. 11% had no documentation. Only 8% made direct reference to the LRAR.

58 ankle x rays were completed that the LRAR would have deemed unnecessary based on examination findings. Of these 58 x-rays, 88% were formally reported as normal xrays with no findings. The other 12% had findings on the xrays- 6 out of 7 of these being cortical avulsion fractures of the distal lateral fibula- a fracture that is not clinically important and can be managed as a sprain<sup>1</sup>. The other was a slight cortical irregularity of unknown significance.

**Conclusion:** The conclusion is that the LRAR is a valuable screening tool for ankle trauma that is poorly understood and applied by the staff working in our emergency department. It has been again demonstrated in this Audit that the LRAR is safe to implement and would save greatly on radiology resources and improve efficiency in the emergency department<sup>2</sup>.

1. Boutis K, Grootendorst P, Willan A, et al. Effect of the Low Risk Ankle Rule on the frequency of radiography in children with ankle injuries. CMAJ 2013;185:E731–8. 2. 2015 Oct;31(10):685-7. doi: 10.1097/PEC.0000000000000474.

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Withdrawn

## ATYPICAL PRESENTATION OF RARE CENTRAL CONGENITAL HYPOTHYROIDISM

N O'Hanrahan<sup>1</sup>, E Hayden<sup>1</sup>, A McCrann<sup>1</sup>, N Schoenmakers<sup>2</sup>, EF Roche<sup>1,3</sup>

<sup>1</sup>Department of Paediatric Endocrinology, CHI Tallaght, Dublin, Ireland

<sup>3</sup>Department of Paediatrics, TCD, Dublin, Ireland

<sup>4</sup>, Wellcome Trust-Medical Research Council Institute of Metabolic Science, Add, Cambridge, UK

**Introduction:** Loss of function mutations in IGSF1 cause a rare form X-linked Central Congenital Hypothyroidism. In Ireland, newborn screening for Congenital Hypothyroidism relies on elevated levels of TSH and therefore cases of Central Hypothyroidism will evade detection. A high index of suspicion and post-natal monitoring of thyroid function in relatives is required.

**Case:** In this case we describe a patient with a novel IGSF1 mutation causing Central Hypothyroidism. This patient posed a particular diagnostic difficulty as thyroxine (T4) levels were normal up to 7 weeks of age, prompting extended evaluation of anterior pituitary function. In addition, elevated prolactin levels were noted in this patient whereas hypoprolactinaemia has more commonly been described. This case illustrates the importance of maintaining a high index of suspicion for IGSF1 mutation in the context of a family history.

## **SIMPSON-GOLABI-BEHMEL SYNDROME TYPE 2 - A CASE REPORT**

**M Pardy**<sup>1</sup>, A Ahad<sup>1</sup>, S Gallagher<sup>1</sup>

<sup>1</sup>Paediatric Unit, University Hospital Limerick, Limerick , Ireland

**BACKGROUND AND AIMS:** Simpson-Golabi-Behmel syndrome type 2 is a very rare x-linked overgrowth syndrome. It is associated with pre and post natal overgrowth, an increased risk of tumours, a wide range of congenital abnormalities, craniofacial features, neurological dysfunction, primary ciliary dyskinesia and impaired intellectual development. Our aim is to report a case of Simpson-Golabi-Behmel syndrome type 2 in an 8 year old boy.

**METHODS:** We describe the clinical presentation, including history and physical exam findings and genetics. A review of the current available literature on Simpson-Golabi-Behmel syndrome type 2 was also undertaken.

**RESULTS:** The patient is an 8 year old boy who initially presented at 2 years old with overgrowth, both height and weight >99.6th centile, and developmental delay, both speech and gross motor, on a background history of macrosomia (Birth weight 5.25kg), patent ductus arteriosus, exomphalos and recurrent respiratory tract infections. Initial genetic testing which included microarray, BWS, EZH2, CDKN1C and a 13 gene panel for overgrowth syndromes were all normal. Subsequently, trio exome analysis detected a pathogenic variant in the OFD1 gene consistent with a diagnosis of an OFD1 related developmental disorder. This pathogenic variant was not present in either of the parents. On further review with a Consultant Clinical Geneticist it was diagnosed as Simpson-Golabi-Behmel syndrome type 2. He is currently being investigated for primary ciliary dyskinesia as there is an association with this syndrome.

**CONCLUSION:** This is a case of a very rare x-linked overgrowth syndrome in an 8 year old boy. On review of the literature there have been very few cases ever reported (approximately 20) and the majority of these cases reported infant mortality due to hydrops fetalis and respiratory complications.

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## **MEDICATION MANAGEMENT AND ECG SCREENING IN CHILDREN WITH ADHD**

MP Michaela Pentony<sup>1</sup>, **ZA Zainab Ali**<sup>2</sup>, KM Kieran Moore<sup>2</sup>, FM Fiona McNicholas<sup>2</sup>, AJ Adam James<sup>1</sup>

<sup>1</sup>Department of Paediatric Cardiology, CHI at Crumlin, Dublin, Ireland

<sup>2</sup>Department of Psychiatry, CHI at Crumlin, Dublin, Ireland

## AIMS

1. Quantify the proportion of referrals sent to Crumlin Cardiology Department for cardiac screening prior to commencement or modifying ADHD medication and comparing to NICE international guidelines
2. Quantity the number of ECGs conducted among youth with ADHD which detected a clinically significant abnormality.

**METHODS:** A prospective audit was performed over a 6-month period, from November 2021 – April 2022 inclusive. The Crumlin OPD triage letters, ECG remail address (ecg.review@olhc.ie) and walk-in ECG service was screened for all referrals pertaining to ECG/Cardiology opinion for children prior to commencing or modifying medication for children with ADHD. Each referral was coded against NICE guideline to determine the degree of clinical details given, and whether these concerns met criteria for referral. Reported abnormalities, recommended management and correspondence were recorded.

**RESULTS:** Ninety one referrals were received in total during the 6 month audit period. There was no indication other than cardiac in 53/91 (58.2%) of referrals. Twenty six (26/91, 28.5%) of referrals met NICE criteria for referral for cardiology opinion including concern regarding clinical symptoms, personal or family history of cardiac disease. There were 76 referrals for which outcome was available. Seventy one patients, 71/76 (93.4%) had normal ECGs with no cardiology follow up required. Five referrals (6.5%) had abnormalities which required a cardiology opinion in OPD. Two were incidental findings with no contraindication to ADHD medication. Three cases (3/76, 3.9%) required a cardiology OPD appointment, all of which met NICE criteria for referral.

**CONCLUSION:** Unnecessary screening ECGs and routine cardiology outpatients referrals prior to commencement ADHD medications lead to unnecessary delays in treatment for children with ADHD. Data obtained will aid creation of a national guideline for healthcare professionals in keeping with best practice recommendations with indications for tertiary subspecialist cardiology opinion clearly outlined.

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### SCHAAF-YANG SYNDROME: A PRADER WILLI MIMIC

S Quinn<sup>1</sup>, K Kavanagh<sup>1</sup>, L Bradley<sup>1</sup>, J Kenny<sup>1</sup>

<sup>1</sup>Clinical Genetics, CHI Crumlin, Dublin, Ireland

**Introduction:** Schaaf-Yang syndrome (SYS) is a rare, genetic, neurodevelopmental disorder. SYS has an overlapping phenotype with Prader-Willi Syndrome (PWS), including central hypotonia, behavioural and feeding difficulties, developmental delay and respiratory distress. SYS may have distinctive features including joint contractures, dysmorphic features and endocrinopathies. SYS is an autosomal dominant condition caused by a pathogenic variant in *MAGEL2* within the PWS critical region on chromosome 15q11.2, inherited by maternal imprinting.

**Case Presentation:** We report the case of a 5-year-old boy referred with multiple comorbidities. He was born at term following an uncomplicated antenatal course. Postnatally, he requires intubation for respiratory distress and central hypotonia. He had persistent feeding difficulties, subsequently necessitating gastrostomy insertion. He has significant global developmental delay, behavioural issues and remains non-verbal. Examination demonstrated dysmorphic features including retrognathia, anteverted nares, long philtrum and prominent eyebrows. Trio-exome sequencing identified a de novo pathogenic variant in *MAGEL2* on 15q11.2. Due to the location of this gene on the PWS critical region, there is significant clinical overlap between PWS and SYS phenotypes.

**Discussion:** Clinical suspicion of SYS should be confirmed by molecular genetic testing. However, due to the rarity of this condition and the overlapping phenotype with PWS, it can remain unidentified. When presented with possible PWS cases, it is important to consider alternative diagnoses and additional advanced molecular genetic testing. There is a wide phenotypic spectrum depending on the pathogenic variant, with one particular variant identified as fatal or severe. Whole gene deletions of *MAGEL2* are associated with a milder phenotype than other variants.



**Conclusion:** Identification of SYS can prove challenging due to the broad phenotype, similarities to PWS and non-specific antenatal findings. However, it is important to always consider it as a potential diagnosis in unconvincing cases of PWS. A definitive diagnosis of SYS will initiate genetic counselling for families and enable introduction of required supports.

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## BY HOOK OR BY CROOK- PROVING THE DIAGNOSIS OF MOSAIC TRISOMY 16

S Quinn<sup>1</sup>, K Kavanagh<sup>1</sup>, B O'Hici<sup>1</sup>, J Kenny<sup>1</sup>

<sup>1</sup>Clinical Genetics, Children's Health Ireland, OLCHC, Dublin, Ireland

MT16 has a variable phenotype with features reported including IUGR, heart defects, genital anomalies, craniofacial anomalies and variable developmental delay.

We present the case of a boy with Mosaic Trisomy 16 (MT16) that represented a significant diagnostic challenge. Referral of a Gravida 3 Para 2 woman to the Clinical Genetics service was made at 27+4 weeks' gestation. Fetal ultrasound scan at 22 weeks showed suspected transposition of the great arteries, ventricular septal defect, micrognathia, dilated echogenic bowel and cleft palate.

NIPT Harmony was unremarkable as was the TORCH screen. Amniocentesis undertaken at 23 weeks showed discordant results; the uncultured ArrayCGH had an altered log ratio for chromosome 16 suggestive of MT16, however this was not identified on ArrayCGH or the G-band karyotype on cultured cells. qfPCR was likewise unremarkable.

After birth, the baby was noted to have cholestatic hepatitis, with a choledochal cyst, a small VSD with mild TR, micropenis and cryptorchidism. He also had Pierre-Robin sequence and cleft palate. Liver biopsy was suggestive of 3-beta-hydroxy-delta-5-C27-steroid oxidoreductase deficiency but sequencing of HSD3B7 was negative.

Extensive genetic testing was undertaken to try and investigate the antenatal diagnosis of MT16. Buccal smear, skin biopsy and blood samples with karyotype and interphase FISH found no evidence of MT16. Finally, the diagnosis was confirmed by molecular testing confirming maternal uniparental disomy of chromosome 16 (heterodisomy) [upd (16)mat] thus providing evidence of trisomy rescue and an explanation for the phenotype.

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### **SEPSIS IN CHILDREN – ARE WE ON TRACK?**

**M.E. Rademan<sup>1</sup>**, A. Walsh<sup>1</sup>, N. Clarke<sup>2</sup>

<sup>1</sup>Paediatric Intensive Care Unit, CHI at Temple Street, Dublin, Ireland

<sup>2</sup>Assistant director of nursing and group sepsis lead, CHI Cross Site, Dublin, Ireland

**Aims:** Sepsis remains the leading cause of mortality worldwide<sup>1</sup> and it is estimated to account for the majority of deaths in children, under 5 years of age. The Surviving Sepsis Campaign (SCC)<sup>2</sup>, for the management of septic shock and sepsis associated organ dysfunction in children, created

international evidence-based guidelines for the management and care of children, with sepsis and septic shock. The Irish National Paediatric Sepsis Guidelines<sup>3</sup> was launched in September 2021. The aim of this audit was to benchmark the management of children admitted to the paediatric intensive care unit (PICU) with sepsis and septic shock against these guidelines.

**Methods:** All children admitted to PICU in 2022 were prospectively screened for infection and sepsis. Data was entered into a pre-designed spreadsheet using Microsoft Excel

**Results:** From January to September 2022, we identified 100 children, who met the criteria. In 68% of cases, the patients were transferred from other hospitals. The Sepsis Six care bundle was only completed in 53% of patients. Septic shock was diagnosed in 20% of patients and 15% of these patients required inotropic support. Antimicrobial cover given was given in 90% of patients but appropriate administration was only documented in 73%, within the one hour timeframe. Viral bronchiolitis was diagnosed in 30% of patients and pathogens were appropriately identified in 76% of the patients.

**Conclusion:** This audit has identified that sepsis is being recognised early, due to well-established management guidelines in the paediatric population, but the timing and initial management is poorly documented. We have also learned that a large group of the paediatric population presents with viral sepsis that remains a significant burden on our healthcare facilities. This project will be used to better practices and to assist in the early identification and prompt treatment of sepsis, by improving the use of specific sepsis forms and early referral, where necessary.

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## INTRODUCTION OF ORAL IMMUNOTHERAPY IN PAEDIATRIC POPULATIONS WITH IgE-MEDIATED EGG ALLERGY: A REVIEW OF THE LITERATURE

Y Ramesh<sup>1</sup>, C Cronin<sup>1</sup>, J Trujillo<sup>1,2</sup>

<sup>1</sup>Department of Paediatrics and Child Health, University College Cork, Cork, Ireland

<sup>2</sup>Department of Paediatrics, Cork University Hospital, Cork, Ireland

**Aims:** The aim of this study is to combine and appraise the literature on Oral Immunotherapy (OIT) methods that have been trialed to improve tolerance in paediatric populations with IgE egg allergy and their outcomes.

IgE-mediated egg allergy is commonly found in the paediatric population. Traditionally, management of egg allergy was through egg avoidance. The Irish College of General Practitioners and other paediatric societies across the globe recommend reintroducing egg into the diet to improve tolerance and quality of life. OIT has been emerging as a potential method to induce desensitisation and even long-term tolerance in children with egg allergy, with many trials reporting significant differences after its implementation.

**Methods:** A search strategy based on the objectives of this review was used to conduct electronic searches on PubMed and Wiley Online Library. After the application of filters, 152 articles were obtained and after application of the inclusion and exclusion criteria, ten articles satisfying the criteria of this review were identified.

**Results:** Six studies reported achievement of desensitisation or tolerance in more than 50% of the intervention group after OIT. Several reported immunological changes in the intervention group including decreased IgE levels, increased IgG4, and decreased SPT sizes. Allergic reactions were seen across studies, however, two studies reported a reduction in reactions over time. The patient's baseline IgE levels and egg tolerance, as well as the duration of the protocol, use of antihistamines, and participant compliance emerged as factors affecting the outcomes of these studies.

**Conclusion:** Although different OIT methods were utilised by each of these studies, most indicated that OIT has a role in improving the dose of egg tolerated by allergic patients. Future research needs to elicit whether the allergic reactions during OIT protocols are an acceptable risk and identify predictive markers for success with OIT.

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### **LIVING WITHOUT PAIN. CASE SERIES OF PATIENTS WITH HEREDITARY SENSORY AND AUTONOMIC NEUROPATHIES IN A CANADIAN TERTIARY CARE CENTRE**

**R Reddi**<sup>1</sup>, GA Horvath<sup>2</sup>

<sup>1</sup>School of Medicine, Royal College of Surgeons Ireland, Dublin, Ireland

<sup>2</sup>Department of Pediatrics, BC Children's Hospital, Vancouver, Canada

**Aims:** Hereditary sensory and autonomic neuropathies (HSAN) are a group of heterogeneous genetic disorders with a predominant feature of slow progressive loss of multimodal sensation and autonomic dysfunction. The primary objective of this study was to determine the clinical and chemical manifestations associated with hereditary sensory and autonomic neuropathies. The secondary aim of this study was to introduce two novel genes that have been discovered to be associated with HSAN that have not been mentioned in previous literature.

**Methods:** In a retrospective cohort, all patients with HSAN (Jan 2000 to June 2021) were reviewed. Collected data consisted of patients' demographics, clinical characteristics, medications being taken, imaging such as x-rays and MRIs, as well as co-morbidities

**Results:** 8 patients were included in this study. The average age at diagnosis was  $3.19 \pm 2.83$  years. Insensitivity to pain (100%), dysautonomia (100%), global development delay (87.5%), emesis (62.5%) and self-injury (62.5%) were the most prevalent manifestations of HSAN in this cohort. The most common comorbidities seen were GERD (50%), obstructive sleep apnoea (37.5%), ADHD (37.5%) and iron deficiency (37.5%).

**Conclusion:** HSANs are a diverse group of diseases, characterized by profound distal sensory loss, acral mutilations and variable autonomic disturbances. Studies examining the long-term course of HSAN may help improve treatment and prognosis.

**Poster No:**  
**Sub-Specialty**

## PAEDIATRIC CUSHING DISEASE: A RARE PRESENTATION IN PAEDIATRICS

C Reynolds, E Burke, N Murphy, R Kernan, E Carolan

<sup>1</sup>Department of Paediatric Endocrinology, Children's Health Ireland, Temple Street, Dublin, Ireland

**Case Report:** A previously healthy 11 year old girl presented with a 3 year history of growth failure, weight gain, acne and abdominal striae. There were no headaches, visual disturbances or exogenous glucocorticoid use. On examination, weight 62kg (98<sup>th</sup> – 99<sup>th</sup> centile), height 132.1cm (<2<sup>nd</sup> centile), BMI 35.7kg/m<sup>2</sup>, BP 132/60mmHg. She was cushingoid in appearance with a moon-shaped face, acne, central and post nuchal adiposity with significant striae rubrae on her abdomen and arms. She had pubic hair

(Tanner III) and prepubertal breast development (tanner I). Given the clinical evidence of hypercortisolaemia, Cushing syndrome was suspected. Initial screening tests showed increased 24-h urinary free cortisol excretion (1,417ml/24h) and loss of cortisol circadian rhythm confirming Cushing syndrome. An initial low dose dexamethasone suppression test demonstrated appropriately suppressed cortisol level of 25nmol/L at 48h. CRH stimulation test reported an exaggerated response of cortisol (180%) and ACTH (92%). MRI brain reported a microadenoma in superior aspect of anterior pituitary gland. Inferior Petrosal Sinus Sampling(IPSS) reported central to peripheral ACTH gradient consistent with central ACTH secretion. She was diagnosed with ACTH-dependent Cushing syndrome and commenced on Metyrapone to achieve normalisation of serum cortisol, followed by selective microadenomectomy. She is currently in remission on physiological hydrocortisone dosing, with no other hormone deficiencies. She is demonstrating good catchup growth but BMI remains elevated.

**Discussion:** Cushing's disease (CD) is the commonest form of ACTH-dependent Cushing's syndrome. It is a rare clinical diagnosis in paediatric patients caused by an ACTH-secreting pituitary

corticotroph adenoma. It is associated with significant morbidity in children. Early diagnosis remains challenging as salient features of CD are often overlooked especially in overweight children. Rapid normalisation of serum cortisol is the primary aim of treatment to prevent adverse effects of prolonged hypercortisolaemia on growth and development. Post-treatment management presents challenges for optimisation of growth, pubertal development and body composition.

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**THE COLLAPSED INFANT– A RARE CAUSE: FOOD PROTEIN-INDUCED ENTEROCOLITIS SYNDROME (FPIES) S Sheridan<sup>1</sup>, N Jameel<sup>1</sup>, AM Murphy<sup>1</sup>**

<sup>1</sup>Department of Paediatrics, University Hospital Limerick, Limerick, Ireland

**Aims:** Food protein-induced enterocolitis syndrome (FPIES) is a non-IgE mediated food allergy. It's presentation is variable from vomiting and lethargy to severe metabolic derangement and shock. A chronic form exists with recurrent vomiting, diarrhoea and/or failure to thrive (1).

Our aim is to report the case of an infant who presented in a collapsed state due to FPIES.

**Methods:** We describe the clinical presentation, results of haematological, metabolic and radiological investigations, management and outcome of our patient to date.



**Results:** A 4 week old term male infant presented to the emergency department resuscitation area with hypovolaemic shock on a background of diarrhoea and vomiting from birth and two days of lethargy and poor feeding. He is the firstborn child of healthy Libyan parents, with an unremarkable perinatal history. He was severely acidotic (pH 6.8) with hyperchloraemia (141mmol/L), hypernatraemia (150mmol/L) and raised inflammatory markers with neutrophilia ( $18.66 \times 10^9/L$ ), lymphocytosis ( $16.22 \times 10^9/L$ ) and a raised CRP (79). He was initially treated for sepsis. Subsequent investigations revealed methaemoglobinaemia (24%) and hyperammonaemia (118  $\mu\text{mol/L}$ ). He was transferred to PICU and managed with intravenous methylene blue and exchange transfusion until his methaemoglobin levels normalised. His amino acid and acylcarnitine profile reflected a malnourished child with low amino acids and ketosis. Further metabolic and radiological investigations were normal. A diagnosis of FPIES was made and he was commenced on extensively hydrolysed formula. Currently, age 12 months, he is developmentally normal and thriving. He will need an in-hospital cow's milk protein trial in the future.

**Conclusions:** The commonest causes of collapse in an infant include sepsis, cardiovascular, metabolic and surgical pathology. We suggest that the diagnosis of FPIES also be considered after other diagnoses are excluded. This diagnosis has a good prognosis and most cases resolve by 5 years of age (1).

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## **A FAMILIAL CASE OF LEIGH DISEASE IN AN INFANT – A CASE REPORT**

S Sheridan<sup>1</sup>, N Jameel<sup>1</sup>, AM Murphy<sup>1</sup>

<sup>1</sup>Department of Paediatrics, University Hospital Limerick, Limerick, Ireland

**Background and Aims:** Leigh disease, also known as subacute necrotizing encephalomyelopathy, is an early-onset, progressive neurodegenerative disease. It is a genetic condition resulting in failure of the mitochondrial respiratory chain that leads to disability and death in early childhood(1,2).

Our aim is to present the case of a male infant with this condition.

**Methods:** We describe the clinical presentation, results of metabolic, genetic and radiological investigations, management and outcome of our patient to date.

**Results:** A term infant male was the third child born to healthy consanguineous parents of Pakistani ethnicity, currently living in Ireland in asylum seeker accommodation. He was well postnatally. His family history was significant for a sister who died age 11 months from complications of Leigh disease. At 9 months of age he presented with developmental regression and failure to meet milestones. On examination he was severely hypotonic with no head control. He was found to have a raised serum lactate at 4.8mmol/l with a mild compensated metabolic acidosis. He had a normal echo, chest x-ray, abdominal ultrasound and ophthalmology exam. His MRI brain findings were consistent with Leigh disease showing bilateral symmetrical areas of increased T2 signal within the putamina, subthalamic nuclei and substantia nigra of the midbrain. At 13 months of age he smiled but was unable to sit, roll or support himself when prone. He had significant oral aversion and weighed 8kg (2nd-9th centile for age). His treatment included nasogastric feeding with high energy formula and coenzyme Q10. Age 15 months he died after suffering a cardiorespiratory arrest following an aspiration event.

**Conclusions:** As an inherited condition this diagnosis has significant implications for this family. There is no cure for Leigh disease, which is ultimately fatal. The management is primarily supportive.

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## **A REVIEW OF PRECOCIOUS PUBERTY PRESENTATIONS IN A TERTIARY PAEDIATRIC ENDOCRINOLOGY**

### **CENTRE PRE- AND POST-COVID PANDEMIC**

**S Sheridan<sup>1</sup>, S Alanazi<sup>1</sup>, E Somers<sup>1</sup>, D Cody<sup>1</sup>**

<sup>1</sup>Department of Paediatric Endocrinology, CHI at Crumlin, Dublin, Ireland

**Aims:** Precocious puberty is defined as the onset of secondary sexual characteristics ie. breast development or menarche before the age of 8 years in girls and testicular development before 9 years in boys(1). Treatment with GnRH analogues is aimed at preserving final adult height potential and reducing psychological stress(2). Our aim is to evaluate the cases of precocious puberty diagnosed in a tertiary paediatric endocrinology centre before and after the onset of the covid-19 pandemic to assess whether patients presented in a more advanced stage of puberty.

**Methods:** A retrospective chart review of the 40 patients diagnosed with precocious puberty in our centre between June 2018 and January 2022. Data collected included tanner stage at presentation and onset of menses. Children diagnosed up to 22 months before and after April 2020 were compared.

**Results:** 16 patients (2 male, 14 female) were diagnosed with precocious puberty between June 2018 and March 2020, 64% (n=9) of females were postmenarchal at diagnosis. 24 patients (all

female) were diagnosed with precocious puberty between April 2020 and January 2022, 41.6% (n=10) were postmenarchal. Tanner stage at diagnosis was categorised into early (stage 2/3) and advanced (stage 4/5). A chi-square test of independence was performed to examine whether the post-covid group were more likely to present in the advanced stage group however the relation between these variables was not significant  $\chi^2(1, n=40) = 0.07, p = .79$ , similarly postmenarchal rates between the groups were not significant  $\chi^2(1, n=39) = 1.81, p = .18$ .

**Conclusions:** Our study did not find a statistically significant difference in the rates of patients presenting in advanced puberty following the pandemic, however the incidence of precocious puberty in our clinic has increased by 50% (16 to 24 patients) in consecutive 22 month periods. Future studies could explore this finding by collecting further epidemiological information about these cohorts.

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**A NON CLASSICAL CASE OF A CLASSICAL PROBLEM****PA Stewart<sup>1</sup>**, C Reynolds<sup>1</sup>, N Murphy<sup>1</sup><sup>1</sup>Endocrinology Department, Temple Street CHI, Dublin, Ireland

**Aims:** Congenital adrenal hyperplasia (CAH) occurs in 0.07 per 1,000 live births. If promptly diagnosed and treated then prognosis is good. Our aim is to highlight the importance of recognising this common and serious condition.

**Methods:** A retrospective review with complete case ascertainment of a severe case of classical CAH presenting to the emergency department of Temple Street in 2022.

**Results:** A 15 day old, ex term infant, firstborn of non-consanguineous parents was noted to have 8% weight loss since birth and to have intermittent bradycardia and spells of ventricular tachycardia on monitoring. His parents reported a history of poor feeding, vomiting and diaphoresis. Examination showed a non dysmorphic male infant with pallor, sunken fontanelle and axillary, nipple and scrotal pigmentation. Biochemistry revealed severe hyperkalaemia, hyponatraemia and metabolic acidosis.

Stress doses of hydrocortisone are given for suspected CAH. Hypovolaemia was managed with 0.9 % saline and electrolyte disturbance was managed with calcium gluconate, insulin and dextrose and salbutamol. Abdominal ultrasound demonstrated prominent right adrenal and enlarged and gyriform appearing left adrenal. Once the infant was stabilized electrolyte disturbance was corrected, oral feeds were recommenced and hydrocortisone was gradually weaned to physiological replacement doses. Fludrocortisone and 30% saline supplements were commenced.

Cortisol value was surprisingly 548nmol/L pre treatment, 17OHP was markedly elevated as was renin supporting the diagnosis of CAH. Urinary steroid profile confirmed CAH due to 21-hydroxylase deficiency.

**Conclusion:** Screening for CAH is not currently part of national newborn screening in Ireland. Salt wasting CAH classically presents late in male infants with vomiting and weight loss and may be life threatening. Severe hyperkalaemia with electrocardiographic changes indicated urgent treatment to stabilize the myocardium and shift potassium from vascular space into the intracellular space.

While this infant was extremely unwell, atypically the pre-treatment cortisol value was within normal limits and the weight loss was less dramatic than the electrolyte abnormality.

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Sub-Specialty

## **SPONGIOTIC DERMATITIS WITH SYSTEMIC SYMPTOMS ASSOCIATED WITH SIROLIMUS USE IN A CHILD WITH AUTOIMMUNE LYMPHOPROLIFERATIVE SYNDROME**

**J Trayer**<sup>1</sup>, F Browne<sup>2</sup>, M O'Sullivan<sup>3</sup>, TR Leahy<sup>1</sup>

<sup>1</sup>Immunology Department, Children's Health Ireland, Crumlin, Ireland

<sup>2</sup>Dermatology Department, Children's Health Ireland, Crumlin, Ireland

<sup>3</sup>Pathology Department, Children's Health Ireland, Crumlin, Ireland

**Case Presentation:** The patient initially presented at 11 months of age with hepatosplenomegaly, lymphadenopathy, anaemia and thrombocytopenia. He was found to be heterozygous for a pathogenic variant in the *FAS* gene (c.685\_686delTT; p.Leu229Glufs\*2) and was diagnosed with Autoimmune Lymphoproliferative Syndrome (FAS-ALPS). He was commenced on sirolimus at diagnosis.

He presented to the emergency department at 15 months old acutely unwell, febrile and septic appearing. There was a widespread rash with confluent erythema and adherent fine crusting (See images). He was treated with broad-spectrum antimicrobial therapy in addition to topical corticosteroids and emollients. Sirolimus was discontinued due to suspected severe infection. The patient clinically improved and intravenous methylprednisolone was added once he was afebrile and inflammatory markers were downtrending. His rash improved with desquamation of his hands and feet. The patient was restarted on Sirolimus and discharged home.

No infectious cause was identified on investigations. A skin biopsy demonstrated subacute spongiotic dermatitis with a neutrophilic infiltration. Three days after restarting sirolimus his rash returned though he remained clinically well. Based on the skin biopsy findings and lack of an

infective cause sirolimus was suspected as a possible cause and was discontinued. An immediate improvement was seen with complete resolution two weeks later. He was switched to mycophenolate mofetil for long-term immunosuppression.

**Case Discussion:** Rash is a commonly reported side effect of sirolimus with a wide spectrum of severity<sup>1</sup>. Acneiform dermatitis is the most commonly seen rash though cases of severe rash necessitating cessation of sirolimus have been reported including cases of leukocytoclastic vasculitis, lower limb oedema with ulceration and palmoplantar peeling with spongiosis<sup>2-5</sup>. The rash in this case differs from those described previously in the literature. Sirolimus should be considered as a potential cause of rash in children. Severe cases may necessitate discontinuation of sirolimus and substitution with alternative immunosuppressant medication.

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**Poster No 31**  
**Sub-Specialty**

**ATYPICAL RASH IN A CHILD WITH AUTOSOMAL DOMINANT ANHIDROTIC ECTODERMAL DYSPLASIA WITH IMMUNODEFICIENCY**

**S Hoolahan**<sup>1</sup>, J Trayer<sup>2</sup>, TR Leahy<sup>2</sup>, I Farombi-Oghuvbu<sup>1</sup>

<sup>1</sup>Department of Paediatrics, Our Lady of Lourdes Hospital, Drogheda

<sup>2</sup>Department of Immunology, Children's Health Ireland at Crumlin, Dublin

**Case Description:** A female infant was born at term and in good condition to unrelated Irish parents. Antenatal ultrasound suggested intrauterine growth restriction and the birth weight was 2.6kg. In the first few days of life she was noted to have leucocytosis and a shiny, ichthyotic rash predominantly on her hands, soles of her feet and scalp. The rash evolved to become pustular and she was treated with intravenous antibiotics and acyclovir.

The combination of rash, low birth weight and lymphocytosis suggested a possible inborn error of immunity. A skin biopsy was performed, showing interface vacuolar dermatitis reminiscent of graft versus host disease and consistent with Omenn syndrome or maternal engraftment.

Genetic testing demonstrated a pathologic mutation causing gain of function in the *NFKBIA* gene (c.95G>A; p.Ser32Asn heterozygote). A diagnosis of autosomal dominant EDA-ID was made based on these results and the patient commenced on prophylactic antimicrobials and immunoglobulin replacement. She has subsequently undergone haematopoietic stem cell transplant (HSCT) at ten months of age with a significant improvement in her rash.

**Case Discussion:** EDA-ID typically presents with cutaneous manifestations including skin dryness, eczema, ichthyosis, sparse hair and abnormal teeth in addition to heat intolerance caused by anhidrosis and an increased susceptibility to infection<sup>1</sup>. This case is unusual as the rash seen was more typical of that seen in patients with Omenn syndrome. The histological features of focal spongiotic and lichenoid dermatitis associated with histiocyte-rich inflammation have not been previously described in a child with AD-EDA-ID.

AD-EDA-ID is a rare condition and genotype-phenotype relationships are being identified. In particular, missense mutations at serine 32 (as seen in this case) have been associated with a more severe disease phenotype<sup>2,3</sup>. This case serves to highlight that more severe cutaneous manifestations can occur in patients with this mutation than have been previously reported.

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