17th East of England School of Paediatrics and Child Health Meeting 5th July 2024

CHEERI Celebration of Trainee Works 2023-2024

(excluding abstracts without approval for sharing on website)

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Urus H, Castaldi S, Katana A, Chandershekar P.

Assessing Early Onset Neonatal Sepsis: Kaiser vs. NICE Guidelines at a District General Hospital

Background:

Early Onset Neonatal Sepsis (EONS) affects 1 in 1000 live births in the East of England. Kaiser Permanente Sepsis Calculator (KP- SRC) identifies neonates at highest risk of EONS and risk stratifies them to either direct septic screen <1 hour of life or observations for a minimum of 24 hours.

Aims:

Assessing whether use of the KP- SRC compared with NICE EONS guidelines NG195 reduced the number of neonates undergoing septic screening in the early postnatal period (<7 days).

Method:

Retrospective data was collected from admission notes on all neonates ≥34 weeks, born between November 2023 and March 2024 at Lister hospital, East and North Hertfordshire NHS Trust. Atotalof 120 neonates at risk of EONS were identified contemporaneously at birth and stratified to observation or direct screening via KP-SRC.

Results:

In total, 33/120 babies (28%) were screened for sepsis and given antibiotics. Of these, 7/33 (21%) were directly screened. 3/7 (43%) had CRP >20 vs 7/26 (27%) screened during the observation period. Compared with NICE guidelines, KP- SRC reduced numbers screened by 20% (24/120 fewer babies screened). 32/33 babies received antibiotics for 5 days or fewer; 1 baby required 7 days.No culture- positive EONS cases were identified, with no re-attendees with Late Onset Sepsis.

Conclusion:

Applying KP- SRC resulted in fewer babies being screened and receiving antibiotics, without missing EONS cases. It also resulted in fewer admissions for antibiotic treatment, reducing associated emotional distress and improving experiences for families of neonates born at Lister Hospital.

Duncan A, Burton K.

Hearing impairments and audiology follow-up in children with cerebral palsy

Aims:

This evaluation aimed to determine the prevalence of hearing loss in a cohort of children with cerebral palsy under the care of Cambridge Community Services. Existing audiology services for these children with complex needs were described and patient pathway adapted to improve patient outcomes. Timely correction of hearing impairments is crucial for this population to mitigate any barriers to their development, given the potential impact on speech acquisition and the association with intellectual disabilities (Khaydarova et al., 2021).

Methods:

We identified a cohort of children with cerebral palsy aged between 2-11 years. We documented the presence and laterality of any sensorineural or conductive hearing impairment, newborn hearing test results, and the outcomes from subsequent audiology appointments, alongside information about their CP aetiology and other associated conditions.

Results:

In total of 66 children, 11 (16.7%) had experienced hearing loss. Of these, 4 children (36.4%) were identified as having a sensorineural hearing impairment (SNHI), two of which passed their newborn hearing screen. Hearing had not been formally assessed after the neonatal period in 29 children (44%), and 7 missed planned follow-up. Children at Gross Motor Function Classification System (GMFCS) levels 4/5 had a higher proportion of hearing impairment compared to those at levels 1-3, though this was not statistically significant [p=0.27].

Conclusion:

A significant proportion of children with cerebral palsy did not have hearing reviewed after the neonatal period despite being at higher risk of impairment. Conductive and sensorineural hearing impairment may have been undiagnosed potentially impacting development. The pathway will be altered such that audiology referrals will be initiated for all children during their first encounter with the complex needs clinic, with additional referrals at six years old for ear-specific testing. Identified missed appointments will be investigated by the community paediatric team and follow up arranged.

Elangaratnam D, Barber S

Tic-Tac Tablet: Converting children aged 7 and over from liquid to tablet Medication in the Paediatric Emergency Department (PED)

The use of liquid preparation medications in the PED is high. Liquid preparations have short expiry dates, require refrigeration, can cause dental decay, can be unpalatable which leads to poor adherence. All of this leads to increased costs for the Trust and parent (liquid meds x10 more expensive than tablet). Liquid preparations are also more taxing to the environment due to high transport load and packaging and therefore a project such as this is in line with the CUH Sustainability Charter.

Previous national projects such as KidzMed and Liq2Tab in the outpatient setting has shown significant cost savings of over £40000 for switches to 36 medicines. The aim of this project is to switch 50% of eligible children aged 7 and over from liquid ibuprofen to tablet form within PED.

A baseline data analysis in September 2023 highlighted that 66 out of the 196 ibuprofen scripts were for children aged 7 and over. Out of this 60 of these children had a 200-400mg dose which is available in tablet form and was not prescribed and not dispensed. If these children had been switched, we could have saved £25 for 1 drug switch for 1 month, more than 60% of that saving comes from switching teenagers.

After approval from management team, teaching of medical and nursing teams cycle 1 was completed in November 2023. 110 out of 446 ibuprofen scripts were for children aged 7 and over. 41 of these children were given tablets therefore achieving 37% of switches. 23 of the 110 children had a prescription of 200mg but were given liquid. This needs further investigation as to what step of tic-tac tablet they achieved. Lessons learnt to date is that buy-in from permanent members of staff such as nursing and play specialist teams are key to making a cultural and operational change in a department. A lot of assumption has also been made that children should be prescribed liquid but in fact we found a lot of children were willing to try a tablet or were in fact taking tablets at home already.

Following the success of cycle 1, cycle 2 is currently in progress. Switches where possible is now being made to prednisolone and paracetamol. Over the next 6 months we will start expanding the project to Urgent Treatment Centre and the wards.

Elsefya A ,Bar M, Ahmed T, Khan R, Hamed M

Bronchiolitis management at a district general hospital's paediatric assessment unit: Bridging the gap with NICE guidelines.

Objectives:

We audited the management of acute bronchiolitis at Southend Hospital paediatric assessment unit in comparison with the NICE guideline, ultimately aiming at reducing the length of stay through minimising exposure to chest x-rays, blood gases, and avoidance of unnecessary use of bronchodilators and antibiotics.

Methods:

Data of babies and children who were clinically diagnosed with bronchiolitis were obtained from the hospital computerised patient records over six months during the winter season (October 2022 to March 2023). Management was compared to the NICE guideline NG9 in the domains of investigations and treatment.

Results:

We found 285 patients diagnosed with bronchiolitis, 5 of which were excluded due to incomplete discharge summaries. We studied 280 patients, their age distribution at presentation was 10% (n=28) below 1 month of age, 32.86% (n=92) between 1-3 months of age, and 57.14% (n=14) aged three months and above. Investigations included blood gases 6.87% (n=19) and chest x-rays 7.86% (n=22), 27.3% out of which (n=6 images) showed consolidations. Patients needing oxygen treatment were 5.71% (n=16) and high flow nasal cannula 2.5% (n=7). Salbutamol treatment was given to 8.57% (n=24) patients, while 6.07% (n=17) patients received Ipratropium Bromide. Antibiotics were given to 11.07% (n=31) patients, either intravenously or orally to treat presumed lower respiratory tract infection. Nasogastric tube feeding was given in 9.64% (n=27). Intravenous fluids were given for 5% (n=14) patients.

Conclusion:

The overall management showed good alignment with the NICE guidance as only 7.86% of the patients had chest x-rays and 6.87% of the patients had blood gases done. There is further room for improvement, especially with the use of antibiotics and bronchodilators by increasing NICE guideline awareness and promoting better adherence to them, especially during the Bronchiolitis season. This will involve educational initiatives, teaching sessions and printed flowcharts of the guideline.

Fatine Y, Dabbour S, Chetcuti Ganado C, Ali S.

Should every newborn infant be screened for jaundice before discharge from hospital?

Background:

Neonatal jaundice affects 60% of term and 80% of preterm babies, and can be harmful if left untreated. As visual identification of jaundiced infants is notoriously unreliable, our project's aim was to improve the identification of infants requiring phototherapy using a transcutaneous bilirubin (TCBR) machine prior to discharge from hospital and to tailor the monitoring of infants depending on the result. We also wanted to see if this would reduce the need for readmission and thus help relieve the administrative burden and stress associated with re-admitting jaundiced babies.

Methods:

TCBR was measured in all infants older than 24 hours of age and >35 weeks gestation on the postnatal ward. If TCBR plotted close to the treatment line on the jaundice chart, a plan to repeat TCBR was to be made in 24 hours with feeds optimised and closely monitored. Infants plotting on/above the phototherapy line were treated as per normal guidelines. Readmission data was also analysed.

Results:

During the intervention period of 12 months, there was a cohort of 3438 live births, with 1763 babies screened with the TCBR machine. 34 infants (2%) were found to require immediate phototherapy, with some needing NICU admission. 392 babies (22%) were identified as high-risk, and either kept in hospital or discharged home with closer monitoring. 3.5% of the high-risk babies later required readmission for phototherapy, with the median length of stay shorter than the median length of stay for readmissions prior to our intervention starting. The remaining 1337 babies were discharged home with normal follow-up. We have also found a statistically significant reduction in the overall need for phototherapy in infants during our intervention period (15 per 1000 live births), compared to data collected prior to it (34 per 1000 live births).

Conclusion:

TCBR screening can identify jaundiced infants that would otherwise have been missed by clinical assessment alone. We have demonstrated that closer monitoring of high-risk babies has caused a reduction in the need for phototherapy. It would be interesting to see a wider-scale adoption of TCBR screening, but a cost-effectiveness study would need to be done first.

Holland J, Riboni-Verri G, Gautam, Peake E, Mukherjee T, Coles A, Cunniffe N

Assessing Remyelinating Measures with the Objective of Understanding Repair in Multiple Sclerosis – the first year

Introduction:
Determining the most suitable primary outcome measure is a significant barrier in the design of clinical trials testing putative remyelinating therapies for multiple sclerosis (MS). Further difficulties arise with analysing longer-term changes in such outcome measures.

Objective/Aims:
The Assessing Remyelinating Measures with the Objective of Understanding Repair study – ARMOUR – began in March 2023, with the aim to better characterise the natural history of visual outcome measures commonly applied in remyelination trials. Here we present first year summary findings.

Methods:
People with MS (pwMS) and healthy volunteers (HVs) were invited to participate through NHS clinics and wider advertising. Exclusion criteria included structural eye disease, photosensitive epilepsy and expected inability to cooperate with tests. Participants were invited to undertake 3 study visits, 6 months apart, for tests including full-field pattern reversal visual evoked potential (ffVEP), multifocal visual evoked potential (mfVEP), Sloan letter low contrast letter acuity, Cambridge Trivector Colour Vision Test and optical coherence tomography (Heidelberg, Germany). For pwMS brain magnetic resonance imaging (MRI) including magnetisation transfer imaging was performed at baseline and 12 months.

Results:
136 HVs and 54 pwMS expressed interest in participating. 7 HVs and 16 pwMS were not eligible. 69 people completed their baseline study visit, including 42 HVs (age range 8-80 years) and 27 pwMS (14-78 years). Cross-sectional analysis of baseline visit data identified abnormal ffVEP peak time P100 in 26 of 51 MS eyes (51%; ‘abnormal’ VEP defined as mean + 2SD amongst HV eyes = 122.2 ms), and abnormal mfVEP latency in 8 of 40 MS eyes (20%; mean + 2SD amongst HV eyes = 156.1 ms). Increasing age demonstrated a significant correlation with increasing VEP P100 amongst pwMS (Kendall tau = 0.33, $p<.0001$) but not amongst HVs ($t=0.15$, $p=.06$). Amongst an approximately age-matched participant cohort (35-80 years; HV n=16, MS n=21), chromatic sensitivity was significantly reduced amongst MS eyes with prolonged VEP P100 vs HV and MS eyes with normal VEP P100, in all colour axes (Kruskall-Wallis with post-hoc Dunn; protan $Z=2.18$, $p=.015$; deutan $Z=2.80$, $p=.003$; tritan $Z=3.96$, $p<.001$). In this cohort, a significant difference between the MS prolonged and normal P100 groups was observed at 1.25% contrast acuity testing ($Z=-2.21$, $p=.014$) but not at 2.5% contrast ($Z=-1.85$, $p=.032$).

Conclusions:
Higher cut-off values for ‘abnormal’ ffVEP peak time P100 may be necessary in clinical remyelination trials that include wider age ranges of pwMS where P100 prolongation is used as a recruitment criterion – especially given that with increasing age, pwMS appear more likely to have abnormal VEP P100 compared to HVs the same age. Reduced chromatic sensitivity in the Cambridge Colour Vision Test and reduced 1.25% low contrast letter acuity scores are associated with prolonged ffVEP peak time P100, indicating these tests may be useful adjuncts in trials where P100 is an outcome measure. Analysis of longitudinal data will aid determining expected test variation over 12 months and further help to inform the design of future clinical remyelination trials.

Holland J, Rodda B, Ruddle JB, Ayton LN, Jolly JK

Inner retinal layer thickening in children with inherited retinal dystrophies

Objective:
Inherited retinal dystrophies (IRDs) are the most common cause of visual impairment amongst children and the working-age population in the UK. IRDs consist of a heterogenous group of genetic conditions leading to retinal degeneration. With the rise in novel therapies for IRDs, the microstructural changes occurring in the retina in childhood need to be better understood. We aimed to characterise retinal layer thicknesses using optical coherence tomography (OCT) on a cohort of children in Australia with IRDs.

Methods:
Clinical and OCT data were acquired in a cross-sectional study of 51 children (ages 3-17) with IRDs and 64 healthy volunteers (ages 6-18) in Australia. OCT total thickness, photoreceptor layer and inner retinal thickness data were calculated for different retinal eccentricities. Participants were grouped into different IRD categories: cone (n=4), macula (n=9), rod-cone (n=27) and cone-rod (n=11) dystrophies. Data analysis was performed using R (R Core Team, 2023), RStudio (Posit Team, 2024) and the Tidyverse package (Wickham et al., 2019).

Results:
All IRD groups demonstrated significant thickening of inner retinal layers versus controls (Kruskal-Wallis H test, cones: $H(1)=50.071$, $p<.0001$; macula: $H(1)=79.249$, $p<.0001$; rod-cone: $H(1)=209.33$, $p<.0001$; cone-rod: $H(1)$, $p<.0001$). This difference appeared greater moving away from the macula in all groups, with relatively increased thickness compared to healthy volunteers more peripherally. There were no significant differences between photoreceptor complex thicknesses in IRD groups and controls except for cone-rod dystrophies ($H(1)=37.437$, $p<.0001$), where thinning was observed particularly in the macula. Total thickness was significantly reduced in cone and cone-rod IRDs compared to controls (cones $H(1)=7.369$, $p=.007$; cone-rod $H(1)=37.261$, $p<.0001$), with the greatest difference appearing in the near-macula area. Macula and rod-cone IRDs did not display a significant difference versus healthy volunteers.

Conclusions:
Conventionally, IRDs are associated with microstructural thinning on OCT, but increasing evidence supports a role for remodelling in the retina, with secondary changes in the inner retinal layers. We show some differences in the patterns of loss which may be useful in predicting disease subtypes. Longitudinal follow-up studies will be crucial to characterize the natural history of retinal remodelling in IRDs.

Holland J, Varma R, Nur Cinar E, Guobadia A, Jha S, Sarantis F, Seregni F, Ratnaike T, CHEERI Early Developmental Impairment Working Group

Improving yield and equity of investigations for early developmental impairment - a project by CHEERI

Objective:
Early developmental impairment (EDI) – where a child's acquisition of skills falls significantly behind the expected level for their age in at least two developmental domains – affects around 1-3 in 100 children under the age of 5 years. Across East Anglia, we have established there is variation in how underlying causes of EDI are investigated. We aimed to understand which tests have the greatest yield and supporting evidence base, as a step towards developing new regional guidelines helping promote more equitable investigation of EDI.

Methods:
A literature review following the Preferred Reporting Items for Systematic reviews and Meta-Analyses (PRISMA) checklist was undertaken using PubMed. Search terms included "investigation", "developmental disabilities", "diagnosis", "child" with results filtered between 2013-2023. Articles were reviewed for which tests were/were not recommended in EDI investigation and test yields.

Results:
The search returned 216 articles, of which 26 were directly relevant. One further article was included, identified from reference lists. Recommended tests: The modal 5 tests recommended for the investigation of EDI were chromosomal microarray (CMA, n=18), urine organic acids (UOA, n=10), Fragile X testing (FGX, n=9), MRI brain (n=9) and plasma amino acids (PAA, n=9). 5 articles recommended whole genome sequencing-based investigations (WGS), with 4 recommending whole exome sequencing (WES). Not recommended tests included: MRI brain (n=2) due to associated risks of general anaesthesia in this age group and potential low yield; karyotype (n=1), FISH (n=1), biotinidase (n=1) and lead (n=1), due to perceived low yield. Ammonia was 'not recommended' (n=1) due to high false positive rates. One publication from 2019 advised against WES due to high cost. Highest yield tests were (in terms of abnormalities): MRI (7.9-66%), WES (20-48.7%), WGS (21-43%), EEG (7-40.7%) and karyotype (1-36%). Yield from CMA was 3.7-33%, for FGX 1-6% and for metabolic tests (including UOA and PAA) <1%.

Conclusion:
Children with EDI commonly present to community paediatric settings and there is variability in investigating EDI. Our findings substantially differ from established practice. The review is biased towards genetic testing, 16 articles focussing on this area. Furthermore, 'abnormality' of some tests (EEG, MRI) did not necessarily correspond to diagnosis. CMA and FGX – generally performed regionally as first line investigations for EDI – had lower yield than WGS or WES. Our next steps are to collaborate with regional stakeholders to devise a practical, economically feasible, and population-appropriate investigation strategy.

Ip N, Yates R, Sheldrake F, Bradley-Russell K, Kelsall W

Service evaluation of the fetal cardiology outreach clinic

Objectives:
Our Trust is a fetal medicine centre, serving a large region. Previously, many of our patients had to travel long distances to our linked specialist cardiac centre (SCC) to receive tertiary fetal cardiology assessment. To address this issue, we established a fortnightly fetal cardiology outreach clinic (FCOC). The project's aim is to assess how this clinic has facilitated the management of expectant mothers and newborns closer to their homes.

Methods:
Women with estimated delivery dates between 21 January 2017 and 13 June 2020, who were referred for fetal cardiology review were included. Electronic patient records were reviewed, collecting data on the number of patients reviewed, the reasons for referrals, the proportion seen in the local FCOC and SCC, distances travelled by women to attend appointments, eventual cardiac diagnosis and pregnancy outcome.

Results:
Over the 3.5-year period, 161 pregnancies involving 163 fetuses were referred for fetal cardiac review. A total of 301 fetal cardiology clinic visits occurred, with 226 in FCOC and 75 at SCC. Most referrals were related to possible/confirmed congenital cardiac disease (CHD), followed by family history (FH) of CHD, genetics, extracardiac issues, increased nuchal translucency (NT), poor views, and other factors (Figure 1). Of the 163 fetuses, 12 pregnancies were interrupted (TOP) and 5 resulted in intrauterine death (IUD). The outcome for 24 babies born outside our Trust is unknown. There were 122 live births. Most babies had an eventual diagnosis of normal heart, followed by septal defects, aortic arch abnormalities, tetralogy of Fallot (TOF), hypoplastic left heart (HLH), coarctation (CoA)/hypoplastic arch, pulmonary valve abnormalities, pulmonary atresia with ventricular septal defect (PA-VSD) and other diagnoses (Figure 2). Ninety-seven of the 122 infants were born at term, 18 required Prostin, 17 were transferred to SCC shortly after birth, and 16 underwent cardiac interventions within their first month of life. Notably, about 28,000 miles were saved by travelling to our FCOC rather than the SCC. All babies, except one recommended for delivery at the SCC, were born within our Trust or at a hospital in our region.

Conclusion:
Our FCOC successfully saw almost 90% appointments, and almost all babies were born and cared for within our region, allowing families facing difficult news to be cared for and supported by the same paediatric team antenatally and postnatally. This initiative has also reduced time and stress associated with travelling and contributes to the NHS's sustainability goals.

Jha S, Nugent-Cruse E, Cholidis N

Clinical response to Low dose fenfluramine (FFA) in children with Dravet syndrome – a case series

Introduction:
Fenfluramine (FFA) is an amphetamine derivative. It was introduced as an appetite suppressant in 1970s but later withdrawn from market due to its potential association with cardiac valvulopathy on long-term use. Low dose FFA was approved in 2020 for use in Dravet Syndrome and formally recommended by NICE in June 2022 as an add-on therapy in children >2years of age, funded via the NHS.

Objective:
To assess change in mean Monthly Convulsive Seizures Frequency (MCSF) after introduction of FFA. To review any adverse effects.

Method:
A retrospective case series of 4 patients with Dravet syndrome known to a DGH
Period of observation: 6 months before and after introduction of Fenfluramine

Data collection:
Notes reviewed from clinic letters, admission documentations, emails, telephone conversations with parents and electronic seizure diary. Patients received 6-monthly ECHO to look for valvular complications. Convulsive seizures were defined as hemiclonic, tonic, clonic, generalised tonic-clonic, myoclonic and focal with clearly observable motor signs.

Results – good response:
3 patients showed reduction in MCSF with improvement in cognitive function. (Case 1: 19-year-old had 50% reduction, Case 2: 5-year-old had 53.2% reduction and Case 3: 18-year-old was free from convulsive seizures). All of them were on at least 4 antiepileptics, 3 of which were Stiripentol+Clobazam+CBD. Weaning of CBD in Case 3, led to increased seizure burden and re-introduction of CBD improved the burden. Appetite: Not affected

Results – poor response:
1 patient (Case 4: 3-year-old) showed significant increase in seizure burden, FFA stopped after 2 months. (Increase in GTCS by 900% and drops by 260%). She was only on 2 baseline antiepileptics (Stiripentol+Clobazam). She was on Keto diet with severe feeding difficulties.

All patients had normal ECHO

Conclusion:
FFA has varied response in patients with DS however, is a promising and safe alternative to other available options. It possibly works better with poly-drug therapy, especially Clobazam+CBD combination and better tolerated in older patients. However, further trials needed to confirm this hypothesis.

Limitations of the study:
Being a retrospective and observational study, some data is missing with no comparison group. Hence, observations cannot be generalized to all patients with DS. Non-convulsive seizures excluded as difficult to measure/record.

Kurup U, Palau HL, Lim DBN, Ishida M, Maharaj AV, Massoud A, Davies JH, Storr HL

A rare case of pre- and postnatal growth failure demonstrating the diverse clinical and molecular spectrum of Silver-Russell Syndrome

Case History:

A 5-year-old South Asian female patient was born at term with very low birth weight (-3.8 SDS). She exhibited short stature (height -3.9 SDS), feeding difficulties (BMI -3.0 SDS) and microcephaly (HC -4.9 SDS). Maternal height was significantly reduced (-3.5 SDS), paternal height was normal (-0.2 SDS). She had characteristic syndromic features including triangular face, high-pitched voice, and high-arched palate. She displayed developmental delay manifesting as inattention and poor motor, writing, and reading skills. Silver-Russell syndrome (SRS) was suspected but she did not fulfil Netchine-Harbison Clinical Scoring System (NH-CSS) criteria (score 3/6).

Investigations:

Investigations established normal female karyotype (46,XX) and short stature screen with elevated serum IGF-1 levels (+4.4 SDS). Testing for common molecular causes of SRS did not detect hypomethylation of chromosome 11p15 or Maternal Uniparental Disomy of chromosome 7 (UPD(7)mat). Whole exome sequencing identified a maternally inherited, heterozygous predicated damaging missense *HMGA2* gene variant (c.166A>G; p.K56E). We report the first missense mutation in the highly conserved region of *HMGA2* (2nd AT-hook adjacent), impacting DNA binding.

Treatment:

SRS is a multisystem disorder requiring early, tailored management through multidisciplinary care. The identification of a genetic cause for this patient's phenotype allowed an end to diagnostic testing, the initiation of active surveillance and referral for genetic counselling.

Conclusions and points for discussion:

SRS is rare (epi)genetic disorder characterised by pre- and post-natal growth restriction and distinct features. ~60% of cases are attributed to 11p15LOM or UPD(7)mat and frequently identified by NH-CSS criteria. Recently, rare (<5%) monogenic defects (*CDKN1C*, *IGF2*, *PLAG1*, *HMGA2*) have been implicated. Clinical SRS diagnosis (≥4/6 NH-CSS criteria) must include relative macrocephaly and prominent forehead at birth associated with being born small for gestational age, postnatal growth failure, body asymmetry and feeding difficulties in the first 2 years of life. However, its utility in diagnosing rare monogenic causes of SRS is uncertain. SRS has a varied phenotype and clinical features diminish with age, making diagnosis challenging. (Epi)genetic testing is recommended for patients scoring ≥3/6 NH-CSS criteria.

I analysed the 17 *HMGA2* cases reported to date; 35% failed to fulfil NH-CSS criteria, 71% lacked relative macrocephaly and 6% had no body asymmetry. Our patient also had atypical features (developmental delay and microcephaly).

Therefore, NH-CSS <4/6 and atypical features should not preclude clinicians from investigating SRS. Molecular diagnosis is crucial for stratification of cases and clinical management, enhancing outcomes and reducing the diagnostic odyssey for patients and families.

Kurup U, Lim DBN, Palau HL, Maharaj AV, Ishida M, Massoud A, Davies JH, Storr HL. Approach to the patient with suspected Silver-Russell syndrome. *The Journal of Clinical Endocrinology & Metabolism*. 2024, dgae423
https://doi.org/10.1210/clinem/dgae423

Approach to the patient with suspected Silver-Russell syndrome

Silver-Russell syndrome (SRS) is a clinical diagnosis requiring the fulfillment of ≥ 4/6 Netchine-Harbison Clinical Scoring System (NH-CSS) criteria. A score of ≥ 4/6 NH-CSS (or ≥ 3/6 with strong clinical suspicion) warrants (epi)genetic confirmation, identifiable in ~60% patients. The approach to the investigation and diagnosis of SRS is detailed in the only international consensus guidance, published in 2016. In the intervening years, the clinical, biochemical, and (epi)genetic characteristics of SRS have rapidly expanded, largely attributable to advancing molecular genetic techniques and a greater awareness of related disorders. The most common etiologies of SRS remain loss of methylation of chromosome 11p15 (11p15LOM) and maternal uniparental disomy of chromosome 7 (upd(7)mat). Rarer causes of SRS include monogenic pathogenic variants in imprinted (CDKN1C and IGF2) and non-imprinted (PLAG1 and HMGA2) genes. Although the age-specific NH-CSS can identify more common molecular causes of SRS, its use in identifying monogenic causes is unclear. Preliminary data suggest that NH-CSS is poor at identifying many of these cases. Additionally, there has been increased recognition of conditions with phenotypes overlapping with SRS that may fulfill NH-CSS criteria but have distinct genetic etiologies and disease trajectories. This group of conditions is frequently overlooked and under-investigated, leading to no or delayed diagnosis. Like SRS, these conditions are multisystemic disorders requiring multidisciplinary care and tailored management strategies. Early identification is crucial to improve outcomes and reduce the major burden of the diagnostic odyssey for patients and families. This article aims to enable clinicians to identify key features of rarer causes of SRS and conditions with overlapping phenotypes, show a logical approach to the molecular investigation, and highlight the differences in clinical management strategies.

Welters A, Leiter SM, Bachmann N, Bergmann C, Hoermann H, Korsch E, Meissner T, Payne F, Williams R, Hussain K, Semple RK, Kummer S. An expanded clinical spectrum of hypoinsulinaemic hypoketotic hypoglycaemia. *Orphanet Journal of Rare Diseases.* 2023, 18(360): https://doi.org/10.1186/s13023-023-02954-5

An expanded clinical spectrum of hypoinsulinaemic hypoketotic hypoglycaemia

Background:

Hypoketotic hypoglycaemia with suppressed plasma fatty acids and detectable insulin suggests congenital hyperinsulinism (CHI). Severe hypoketotic hypoglycaemia mimicking hyperinsulinism but without detectable insulin has recently been described in syndromic individuals with mosaic genetic activation of post-receptor insulin signalling. We set out to expand understanding of this entity focusing on metabolic phenotypes.

Methods:

Metabolic profiling, candidate gene and exome sequencing were performed in six infants with hypoketotic, hypoinsulinaemic hypoglycaemia, with or without syndromic features. Additional signalling studies were carried out in dermal fibroblasts from two individuals.

Results:

Two infants had no syndromic features. One was mistakenly diagnosed with CHI. One had mild features of megalencephaly-capillary malformation-polymicrogyria (MCAP) syndrome, one had non-specific macrosomia, and two had complex syndromes. All required intensive treatment to maintain euglycaemia, with CHI-directed therapies being ineffective. Pathogenic PIK3CA variants were found in two individuals – de novo germline c.323G>A (p.Arg108His) in one non-syndromic infant and postzygotic mosaic c.2740G>A (p.Gly914Arg) in the infant with MCAP. No causal variants were proven in the other individuals despite extensive investigation, although rare variants in mTORC components were identified in one. No increased PI3K signalling in fibroblasts of two individuals was seen.

Conclusions:

We expand the spectrum of PI3K-related hypoinsulinaemic hypoketotic hypoglycaemia. We demonstrate that pathogenic germline variants activating post-insulin-receptor signalling may cause non-syndromic hypoinsulinaemic hypoketotic hypoglycaemia closely resembling CHI. This distinct biochemical footprint should be sought and differentiated from CHI in infantile hypoglycaemia. To facilitate adoption of this differential diagnosis, we propose the term “pseudohyperinsulinism”.

Moh Myint Shein M, Arulogun Y, Edeko E

Expressed Breast Milk Quality Improvement Initiative: Comparison of Data Before and After Implementation of Periprem Bundle

Background

Neotrips supports the Periprem initiative, which enhances neonatal care and promotes early breast milk reception in newborns. This audit evaluates Neotrips' impact on improving perinatal outcomes. The PeriPrem bundle, initiated on August 7, 2023, includes 11 interventions to improve outcomes for premature babies under 34 weeks. One key intervention is early breast milk intake by babies within six hours.

Aim

This audit aims to assess improvements in the proportion of babies receiving early breast milk within 6 and 24 hours of life from <34 weeks' gestation, and also early maternal expression of breast milk following the introduction of the PeriPrem care package at Colchester General Hospital.

Methodology

Retrospective data collection from BadgerNet was conducted, focusing on the proportion of mothers expressing milk in the first six hours and the timing of the first EBM received by neonates. Data were collected from March 2023 to August 2023 (pre-implementation) and from September 2023 to February 2024 (post-implementation). All premature babies and their mothers were included.

Exclusion Criteria:

1. Babies not born in our local unit or transferred in.
2. Neonatal deaths within 6 hours after birth.

Results

Timeline	Pre-Implementation 25 (100%)	Post-Implementation 16 (100%)
EBM within 6hrs	6 (24%)	6 (37.5%)
EBM within 24hrs	16 (64%)	11 (68.7%)
Mother Expression within 6hrs	13 (52%)	10 (62.5%)

Conclusion

After six months of implementing the preterm bundle, we observed an improvement, though a basic Chi-squared test showed it was not statistically significant: $\chi^2(1, N=41)=0.29$, $p=0.59$ (not significant at $p<0.05$). To increase breastfeeding adoption, we plan to enhance education and awareness among staff, parents, and families regarding the bundle and EBM pack.

Myint Zu Khine H, Gupta S, Merchant N

False beeps, True alarm: Preliminary results of a project aimed at improving compliance with alarm limits on saturation monitors in a level II NICU

Objectives:

- To check the compliance of oxygen saturation targets on monitors in the neonatal unit with the standard saturation targets according to gestational age.
- To demonstrate tangible improvement in compliance by running more than one PDSA cycle with an aim to achieve 100% conformity with the established criteria.

Methods:

The guideline used for the purpose of this project was the East of England Neonatal ODN guideline. Data was gathered through random point checks performed on monitors in the neonatal unit.

> *Inclusion Criteria:* Babies of all gestational ages admitted to NICU who are attached to saturation monitors irrespective of whether they are on respiratory support or self-ventilating in air.
>
> *Exclusion Criteria:* Babies admitted to the unit but not requiring saturation monitoring or those with congenital heart diseases.

Results – PDSA cycle 1:

15 episodes were recorded for babies receiving care in the unit during point checks done in the months of April and May 2023. There was very low compliance seen with the EOE ODN guideline with only 19% of saturation monitors found to be adherent with the given standards. Interestingly, this was seen to translate into a higher corresponding oxygen saturation in babies compared to the target saturation range for their gestational age. The results were discussed locally and a practice development nurse for the neonatal service was involved. This led to the implementation of various suggested interventions including staff training, handover reminders and stricter safety checks to improve the conformity standards.

Results – PDSA cycle 2:

A repeat PDSA cycle was run in 4 months' time to assess the effectiveness of remedial actions taken and to measure changes from the previous practice, if any. Data was again collected from 15 baby episodes. There was a marked improvement in the compliance seen with 80% of saturation monitors meeting the EOE ODN standards. However, due to the turnover of the trainee cohort, continuity of these measures could not be ensured. This was reflected in initial results from the data collected for 3rd PDSA cycle which demonstrated a fall in compliance rates to 50%

Conclusions:

- Setting the correct alarm limits on the saturation monitors might help the babies to maintain saturation in the recommended target range and prevent hyperoxaemia, which will be particularly beneficial in preterm babies.
- Maintaining correct upper and lower limits on oxygen saturation monitors can often be overlooked by both nursing and medical teams in busy neonatal units.
- Simple interventions like handover reminders, nursing education and stricter shift safety checks were seen to be effective in trying to achieve the aim of 100% compliance with standards.
- However, these measures can tend to be short-lived unless supplemented with more permanent strategies.
- Further suggested action points to achieve compliance include attaching printed and laminated sheets with standard alarm limits to all saturation monitors in use and including random alarm compliance checks as a part of medical and nursing grand rounds.

Filby L, Padilla Yap Z, Philipp K, Herath D

Recurrent Chesty symptoms, think beyond basics

Introduction and Objectives:

Agammaglobulinaemia is an inherited immune deficiency marked by reduced antibody levels in the blood due to the absence of specific lymphocytes in both blood and lymph. These antibodies play a crucial role in the immune response, defending against bacteria, viruses, and other external threats.

This case report emphasizes the need for medical professionals to consider agammaglobulinaemia when a child exhibits recurrent chest infections. The goal is to foster early diagnosis, initiate prompt treatment, and prevent complications or oversight in children with this condition, which can be life-threatening.

Methods:

This case report discusses a child who repeatedly experienced pneumonia, leading to several hospitalizations in a district general hospital. Since the age of 5 months , the child had a persistent cough coupled with numerous chest infection episodes, conjunctivitis and otitis media. Subsequent tests revealed decreased immunoglobulins and a lack of B lymphocytes, necessitating a referral to a specialized immunology centre for further evaluation.

Results:

Patient was hospitalized for pneumonia, accompanied by elevated infection markers. Despite prolonged courses of oral antibiotics after discharge, the patient showed no significant improvement. Persistent symptoms of chronic cough remained evident during clinical reviews. Immunological testing revealed low IgG levels (<0.3), absence of CD19+ lymphocytes (B cells), but normal counts of CD3+ (T cells), CD56+ (NK cells), and naive/memory cells. Additionally, the patient demonstrated no response to primary vaccinations, including those for Haemophilus Influenza, Pneumococcal, and Tetanus. Genetics test confirmed X-linked agamaglobinemia – BTK variant

Treatment was initiated with immunoglobulin replacement therapy (IgRT) that required monthly infusions and Prophylactics antibiotics. It's noteworthy that Primary Agammaglobulinemia is a rare condition primarily affecting males, though a few cases in females have been reported.

Conclusion:

This case underscores the significance of considering agammaglobulinaemia in children who frequently experience chest infections, particularly during seasons when school-aged children are more susceptible. Recognizing this condition promptly ensures timely care for children with primary immunodeficiency. Early treatment, which is lifelong, can prevent severe, potentially fatal infections.

Radford E, Yakob R, Tan H-K, Gerety S, Hurles M and MAVE/SGE Project Team

Prospectively generating functional data for every coding single nucleotide variant in SLC2A1 could transform our ability to make diagnoses of GLUT1-deficiency syndrome

Genetic sequencing is a powerful diagnostic tool in rare disease. However, correctly interpreting candidate genetic variants remains challenging. Where there is conflicting or insufficient evidence of a variant's effect, it is termed a variant of uncertain significance (VUS). VUS prevent access to targeted treatments, prenatal diagnosis, inclusion in clinical trials, and can cause emotional turmoil for families. The number of VUS is rising rapidly, over 70% of missense variants in ClinVar are VUS. VUS are becoming a major impediment to optimal care. Uncertain or incorrect assessment of variant function can also reduce the power of clinical trials.

Early, fast, accurate diagnosis is particularly important where there is a targeted treatment which alters a child's outcomes. Evidence is accumulating across diverse conditions that such treatments are often most efficacious if provided early in the disease course. As we start to consider expanded screening for such conditions, there is an imperative to improve our ability to interpret the associated genetic variants.

One such condition is GLUT1-deficiency syndrome, caused by loss of function genetic variants in the gene SLC2A1 which encodes the CNS glucose transporter. We employ deep mutational scanning to experimentally determine the functional consequence of every possible coding single nucleotide variant (SNV), codon deletion and observed frameshift variants in SLC2A1 - generating a comprehensive, prospectively generated 'variant effect map'.

To date, we have functionally characterised over 4500 SLC2A1 variants. 99% of synonymous variants appear functionally normal in our assay, while 99% of nonsense variants appear to be non-functional, demonstrating that the assay correctly classifies variant function. Using clinically ascertained pathogenic variants as true positives, and variants observed in population databases and clinically ascertained as benign as true negatives, we estimate 90-95% sensitivity and 99% specificity of these data for GLUT1 deficiency syndrome. We are currently exploring whether our quantitative measurement of variant effect in vitro correlates with patients' clinical phenotypes.

For greatest clinical impact, this approach will need to be scaled across many neurodevelopmental disorders, as these conditions are individually rare. Such direct functional characterisation of genetic variation at scale has the potential to transform clinical diagnosis. We will present our deep mutational scanning approach, progress on SLC2A1, and our broader strategy to address VUS.

Ratnaike TE, Elkhateeb N, Lochmüller A, Gilmartin C, Schon K, Horváth R, Chinnery PF. Evidence for sodium valproate toxicity in mitochondrial diseases: a systematic analysis. *BMJ Neurology Open* 2024;**6:**e000650. https://doi.org/10.1136/bmjno-2024-000650

Evidence for sodium valproate toxicity in mitochondrial diseases: a systematic analysis

Background:

We aimed to determine whether sodium valproate (VPA) should be contraindicated in all mitochondrial diseases, due to known VPA-induced severe hepatotoxicity in some mitochondrial diseases.

Methods:

We systematically reviewed the published literature for mitochondrial DNA (mtDNA) and common nuclear genotypes of mitochondrial diseases using PubMed, Ovid Embase, Ovid Medline and MitoPhen databases. We extracted patient-level data from peer-reviewed articles, published until July 2022, using the Human Phenotype Ontology to manually code clinical presentations for 156 patients with genetic diagnoses from 90 publications.

Results:

There were no fatal adverse drug reactions (ADRs) in the mtDNA disease group (35 patients), and only 1 out of 54 patients with a non-POLG mitochondrial disease developed acute liver failure. There were fatal outcomes in 53/102 (52%) POLG VPA-exposed patients who all harboured recessive mutations.

Conclusions:

Our findings confirm the high risk of severe ADRs in any patient with recessive POLG variants irrespective of the phenotype, and therefore recommend that VPA is contraindicated in this group. However, there was limited evidence of toxicity to support a similar recommendation in other genotypes of mitochondrial diseases.

Ratnaike TE, Paramonov I, Matalonga L, Solve-RD consortium, Horváth R

Mitochondrial DNA disease discovery through Solve-RD

Background:
Solve-RD, a European initiative funded by the European Commission, aims to diagnose patients with undiagnosed rare diseases through next-generation sequencing. The project, operating collaboratively between data analysis and interpretation teams, recruited patients into rare disease groups through European Reference Networks (ERNs).

Mitochondrial diseases are rare, heterogeneous, and challenging to diagnose. MitoPhen is adatabase developed to capture all published phenotypes across pathogenic mitochondrial DNA (mtDNA) variants in the form of human phenotype ontology (HPO) terms. It can be used to perform phenotype similarity scoring to identify patients with similar phenotype profiles in large, rare disease cohorts such as Solve-RD, thereby improving our ability to detect patients with mtDNA diseases.

Objectives:
1. Summarize Solve-RD data on patients with rare mtDNA variants.
2. Develop an integrated phenotype-genotype approach for diagnosing mtDNAdiseases in a large, rare disease cohort.

Methods:
MToolbox was used to detect rare mtDNA variants. We separately looked for pathogenic mtDNA variants listed in MitoPhen with a heteroplasmy level >1% and read depth >5x. Affected patients with HPO data were considered in the final analyses. Phenotype similarity scoring between probands in MitoPhen and affected Solve-RD par9cipants was performed using R software.

Results:
24160 samples were analysed from 23057 individuals, with 14571 being affected, from 13317 families. The MToolbox analysis with filters applied resulted in 227 affected individuals with 76 rare mtDNA variants and HPO data available. This included 48 previously diagnosed individuals. 21 individuals were solved with mtDNA variants, 16 individuals received nuclear gene diagnoses, 8 individuals had candidate mtDNA variants, 49 individuals had reportable findings (for example homoplasmic variants such as m.11778G>A or m.1555A>G which have variable penetrance). 134 individuals overall remain unsolved. The phenoype similarity score was high in 37 individuals (including previously solved), 28 patients (76%) had variants which were diagnostic or likely causative of their phenotype. Variants of uncertain significance were reported in five samples with high phenotype similarity scores and have been reported back to the data interpretation team to request more information from recruiting clinicians. Overall, 29 individuals were newly diagnosed or found to have likely causative variants, based on phenotype and genotype analyses.

Conclusion:
The Solve-RD dataset has confirmed or likely mtDNA disease diagnoses in 70/13317 affected families (0.53%), recruited into four ERNs: ERN-EpiCARE, ERN-ITHACA, ERN-RND, ERN-NMD. The collaborative approach to continually re-analyse the genotypic and phenotypic data has been vital in feeding back likely pathogenic variants to recruiting clinicians.

Reid ES, Leiter SM, Silverwood H, Cunnington A, Ranson K, Brown J, Noone M. Implementation of pre- and post-ductal oxygen saturation screening in babies born in a district general hospital. *Archives of Disease in Childhood - Education and Practice* 2024;**109**:147-150.

Implementation of pre- and post-ductal oxygen saturation screening in babies born in a district general hospital

What is already known on this topic:
Non-invasive pre- and post-ductal oximetry screening of newborns has previously been shown to increase detection rates and reduce mortality of critical congenital heart disease.

Objectives:
This quality improvement project aimed to implement strategies to achieve universal screening in a district general hospital over a 12-month period. Babies referred to the neonatal team due to abnormal screening results and patients diagnosed with congenital heart disease were reviewed.

Methods:
Eight different interventions including IT infrastructure changes, education, regular reinforcement, updates to handover sheets and case discussion were undertaken.

Results:
In the first four weeks of the project, a weekly average of 69% of babies had their pre- and post-ductal oxygen saturations recorded. In the last four weeks of the project, an average of 95% of babies were screened and there was a significant run chart shift with 17 consecutive weeks above the median.

The number of babies referred for review by the neonatal team was low overall (23 per 1000 babies screened). Whilst the majority of babies escalated to the neonatal team either had normal repeat saturations or a non-cardiac problem e.g. neonatal sepsis or respiratory distress syndrome, one child with congenital heart disease was identified. An echocardiogram revealed hypoplastic left heart syndrome.
Conclusion:

Multi-professional working was essential for successful implementation of this screening programme. Open listening and discussion was crucial to identify small interventions which combined, had a significant impact. We feel that regular reinforcement was crucial, with feedback on the case identified and opportunities to ask questions. This was particularly important for groups where there is significant staff turnover (e.g. junior paediatric doctors).

Following the completion of this quality improvement project, pulse oximetry screening has now become the standard of care at our hospital. We hope that by sharing our experience, other hospitals may be able to implement this screening to reduce neonatal morbidity and mortality.

Rupa Sarder B, Luca I, Kelsall W

The impact of telephone consultations in triaging babies with a family history of heart disease

Objective:
A number of babies are referred to clinics conducted by paediatricians with expertise in cardiology (PEC) because of a family history of heart disease1. We compare two cohort studies looking at these referrals following the implementation of new referral guidelines, education packages for all staff conducting NIPE and the impact of telephone consultations on triaged referrals.

Method:
We conducted a retrospective study by gathering data from electronic medical records. The 2 study periods were: September 2019 to February 2020 and September 2022 to February 2023. We compared the number of face-to-face (F2F) and telephone consultations (TC) in the two cohorts.

Results:
Comparison of 2 cohorts is shown in the table below. Referrals originated from various sources, including the post-natal ward (51%), NICU (16%), transitional care (14%), the delivery unit (7%), and the midwifery-led unit (10%). Most of these patients (78%) were referred by neonatal doctors. Among those with a family history, nearly half (38 out of 77) underwent telephone consultations, with the majority being discharged (27 out of 38). Fewer patients were referred over six months (149 compared to 167). The median waiting time was decreased (59 days compared to 66 days).

Time period	1/9/19-28/2/20	1/9/22-28/2/23
Total patient	171	149
Excluded	4	0
Included	167	149
Median (range) waiting time(days)	66	59
Number of patients had telephone consultation	14 (8%)	42(28%)
Family history Number of patients (percentage)	76 (45.2%)	77 (51%)
Heart murmur	45 (26.8%)	32 (21%)
NICU (antenatal and postnatal diagnoses)	46 (28%)	40 (26%)

Conclusion:
- Telephone consultations have proven to be a safe and efficient method for safely reducing the number of infants requiring F2F clinic review. This was achieved when the family history was not clearly identified in the original referral; the first-degree relative did not require any intervention, and concerns in more distant relatives were not valid. This intervention reduces the number of unnecessary clinics and travel for families. It also frees up clinic space for other F2F consultations.
- A small number of face-to-face appointments were required following a TC; these appointments were scheduled more appropriately.
- The new guidelines, education packages and reduced number of babies seen inappropriately in the clinic, have allowed better utilisation of clinic capacity.

Seregni F, Taylor J, Toulmin H, Wong HS, Chetcuti-Ganado C

FROM NICU TO SCHOOL: involving education and raising awareness of long-term outcomes of prematurity and neonatal morbidity

Aims/Objectives:

Neonatal complications including prematurity and hypoxic ischaemic encephalopathy are associated with long term neurodevelopmental needs which can be subtle and not apparent until school age (1). Opportunities for early recognition and support can be missed in children without moderate or severe neurodisability. Teachers and parents have primary responsibility for long-term support and raising their awareness is essential (2). The overall aim of the project is to foster better collaboration between education, health and parents to improve early identification and support of school age developmental needs in children discharged from NICU.

Methods:

Phase 1:
Survey to Special Educational Needs Coordinators (SENCo) to identify gaps in skills and knowledge. The survey was distributed to 240 SENCOs and 74 replies were received (return rate 30.4%).

Phase 2 (in progress):
- Training for SENCOs to address gap in knowledge and suggestions of strategies to implement in school.
- Development of information material for parents with multidisciplinary input from neonatologists, community paediatricians, physiotherapists, occupational therapists, teachers and parents.

Phase 3:
Dissemination via regional neonatal network, local community paediatric services and schools.

Results:

Knowing the neonatal history of a child was rated highly (median of 8/10) in terms of its impact on teaching and supporting the pupils. Despite this, only 11% of respondents collected information about neonatal history at school entrance. 100% of the staff were not aware of ‘Premature Infants Skills in Mathematics’ (PRISM) training or ‘Prem Aware Award’. 92% of respondents stated they were interested in having more training.

Conclusions:

The results of the survey are helping to tailor the training for SENCOs and increasing the uptake of the ‘prem aware award’ scheme. Multidisciplinary work involving education and parents’ empowerment are key priorities to allow for early identification and support for NICU graduates at school age.

Seregni F, Coghill J, Wong HS. School-age developmental needs in children born preterm. *Paediatrics and Child Health* 2023, **33**(9):P253-P258
https://doi.org/10.1016/j.paed.2023.06.001

School-age developmental needs in children born preterm

Children born preterm are at risk of developing neurodevelopmental, cognitive and mental health needs. There is an inverse correlation between gestational age and those needs. Difficulties with communication and language, executive function, learning, coordination, behaviour, and mental health tend to become more obvious as the social and academic demand increases in later childhood. By school age, children born preterm without severe neurodisability have often been discharged from neonatal follow-up programmes and opportunities for early recognition are missed. Long term follow-up with enhanced developmental surveillance as well as raising awareness of the consequences of prematurity in school are essential in identifying the needs of children born preterm, so that timely interventions can be implemented. In this article, we draw attention to the school-age developmental needs of children born preterm and the implications for clinical care for this population.

Sinthuraj V, Shah A, Ayers K, Khanna N

Dexamethasone vs prednisolone in treating acute episodes of viral induced wheeze and asthma

Background and Aims:

Asthma is a common cause of illness and death among children. According to The British Thoracic Society, corticosteroids are often recommended acutely, with prednisolone being the preferred choice to prevent hospitalisation. However, prednisolone has a short half-life and an unpleasant taste. On the other hand, dexamethasone has a longer half-life and is more palatable. Recent studies have indicated that dexamethasone may be an effective alternative to prednisolone for the treatment of viral-induced wheeze and asthma, as it is clinically non-inferior.

Our study investigated whether a single dose of Dexamethasone is a viable alternative to a 3-day daily Prednisolone 1mg/kg course, considering palatability, cost-effectiveness, and nursing perspectives.

Methods:

To identify children who were admitted to the hospital and treated with oral steroids, inclusion and exclusion criteria were established for those over two years of age with viral-induced wheeze or asthma. Data was collected retrospectively from electronic patient records and drug charts. Additionally, a survey was created to gather the nursing team's perspectives and the results analysed.

Results:

Of the 57 patients, 8 were excluded, leaving 46 for analysis. It was noted that 24% of patients who received prednisolone vomited it, while reattendance rates remained the same for those who were given dexamethasone or prednisolone. The survey of 21 nurses revealed that 85% felt prednisolone takes longer to prepare. 80% of the respondents reported that children vomit after taking prednisolone over 50% of the time; in contrast, only 19% of respondents reported this for dexamethasone. Notably, not all nurses would ask doctors to re-prescribe prednisolone if they vomited.

Overall, 100% of nurses preferred dexamethasone due to its tolerability compared to prednisolone. Cost analysis revealed no significant difference in using dexamethasone vs prednisolone. Dexamethasone is a more cost-effective option as it does not require discharge medication, eliminating the need for nurses to check and validate it.

On 8th December 2024, a stat dose of Dexamethasone 0.6mg/kg was implemented instead of prednisolone for viral-induced wheeze and asthma. Following the implementation of dexamethasone, 40 patients were identified, and none of them experienced vomiting. Additionally, the reattendance rate was reduced to 7.5%, down from 13% with prednisolone.

Conclusion:

From a palatability, cost-effective, and nursing perspective, dexamethasone appears to be a suitable alternative to prednisolone. After implementing dexamethasone, we will repeat the study to see if dexamethasone remains the preferred choice in treating acute viral-induced wheeze and asthma.

Walder E, Stevens J, Rasheed A, Li T, Salam H

Keep Calm and Communicate – Integrating simulated communication scenarios to improve confidence of paediatric trainees entering registrar roles

Objectives:
Good communication skills are an essential component of paediatric practice and are part of the RCPCH curriculum. There is good evidence supporting the translation of clinical skills learnt during simulation into clinical practice, but the impact of simulation on communication skills is less well known.

Our objective was to enhance the scope of paediatric simulation training days in the East of England Deanery by integrating scenarios focused purely on communication skills. We aimed to conduct virtual simulation sessions to provide practice for challenging scenarios to improve communication efficacy.

Methods:
The EoE Paediatric Simulation Committee conduct simulation days aimed at trainees stepping up into registrar roles. The “Virtual Ready for Reg” day exclusively focuses on communication skills and includes scenarios focused on conflict resolution, safeguarding, and handover skills. The day was conducted virtually via Zoom. Candidates were divided into breakout rooms with a facilitator each and were given specific scenarios to manage. Each candidate would be given an opportunity to lead the scenario with their facilitator followed by a debrief. At the end of each session, candidates were asked to share their learning points with the whole group. Pre- and
post-course feedback was collected.

Results:
66.67% of attendees found the sessions to be “very useful” and 33.33% found it to be “extremely useful” to their clinical practice with 100% of candidates agreeing the course was appropriate to their level of training. The use of breakout rooms followed by group discussions was found to be useful by the candidates and facilitators in conducting these simulated scenarios on a virtual platform. Of the various scenarios included, candidates found those focusing on safeguarding and handover to tertiary teams to be most useful. 100% of candidates fed back that they would recommend the course to their colleagues.

Conclusions:
Implementing a simulation-based study day focused on communication skills has been a well-received addition to our regional training days. By including common scenarios that paediatric registrars are faced with in their daily practice, candidates were provided an opportunity to practice and improve their skills in a safe environment. The use of a virtual platform made the course more accessible to the trainees in our deanery.

Going forward, to enhance the educational experience we aim to include multisource assessment to better assess different aspects of each scenario. There is also potential to conduct these days in person to increase the authenticity of the scenarios and to account for verbal and non-verbal skills.

Stevens J, Walder E, Rasheed A, Li T, Salam H

Mind the Youths: Integrating Mental Health into Low Fidelity Simulation for Paediatric Trainees

Objectives:
CAMHS is a key area of paediatrics that requires targeted knowledge and skills. It encompasses challenging and unpredictable scenarios where confidence and sensitivity are necessary, but trainees may feel hesitant due to lack of training. In England, the number of children with a mental health condition is increasing with 1 in 5 children affected in 2023 - up from 1 in 8 in 2017. The number of paediatric admissions with a primarily mental health cause has also increased in recent years with approximately 12% of inpatient beds occupied by children admitted with a mental health problem. Paediatric trainees will encounter these children with increasing frequency and must be equipped appropriately to do so. In addition, there is increased emphasis on CAMHS in the new RCPCH curriculum.

The East of England ST1-4 simulation committee aimed to increase trainee confidence and competency in mental health scenarios through integration into low-fidelity simulation days at the start of training and in registrar transition.

Methods:
Working with a consultant in CAMHS, we developed mental health scenarios and integrated them into two simulation days – ST1 simulation and “Ready for Registrar” days. We utilised low fidelity, face-to-face simulation with actors portraying teenaged patients and their parents. Facilitators were senior clinicians in general paediatrics and CAMHS. We evaluated the effectiveness and value of these scenarios utilising pre- and post-session questionnaires.

Results:
Prior to the sessions, 59% of trainees reported feeling unconfident in managing children with mental health conditions and 31% felt only somewhat confident.
Following the sessions, there was a clear increase in confidence reported by trainees. 85% of trainees felt either confident or very confident in managing mental health conditions, and no trainees reported feeling unconfident. 100% of trainees found the mental health scenario extremely useful – the same, if not better, usefulness as reported for traditional medical scenarios, and 100% reported they would recommend the course to a colleague.

Conclusions:
Our results demonstrate that mental health simulation is effective in increasing trainee confidence in this vital area of practice. We recommend including mental health simulation as an important tool when planning simulation teaching for paediatricians and recommend the engagement of specialists in CAMHS to guide sessions and debrief participants. We demonstrate that individual scenarios and dedicated mental health simulation days can add value. We will continue to use these scenarios and collect further data in sessions later this year.

Sundralingam K, Frank D, Egyepong J

A case report of neonatal coronavirus

Background:
The outbreak of COVID-19 was declared a public health emergency by the World Health Organization on 30/1/2020. We report a case of neonatal infection at Luton hospital secondary to maternal COVID 19 positive status and its subsequent consequences. Currently, there is only limited research in regards to neonatal COVID-19 and thus needing more studies.

Case report:
An expectant mother required intubation and ventilation in ITU secondary to severe respiratory failure and sepsis, at gestational age 26 weeks. Moreover, she was noted to be COVID 19 positive. She underwent an emergency caesarean section at 26 weeks gestation due to the onset of uterine contractions whilst under general anaesthesia. Incomplete magnesium sulphate and steroid therapies were provided prior to labour. Unremarkable antenatal scans except small for gestational age. Normal maternal serology was noted. There were no other risk factors for sepsis.

Birth:
The preterm male neonate [named “M”] was born in a breech position, with a birth weight of 0.87 kg and head circumference of 25 centimetres. APGAR [appearance, pulse, grimace, activity, respiratory effort] scores were 1^{4}, 5^{7}, 10^{8}. After placing the newborn inside the vygon bag, invasive positive pressure ventilation [IPPV] was provided. He was successfully intubated with 2.5 Fr endotracheal tube [ETT] at 6.5 cm length at 7 minutes of life.

Neonatal unit admission:
Post-transfer to the neonatal unit [NNU], M was commenced on pressure control-assist control [PC-AC] ventilation mode. Initial observations included temperature 36.7*C, heart rate [HR] 171 bpm, respiratory rate [RR] 71, saturation 99% on volume guarantee [VG] PEEP 5 PIP5 FiO_2 21%, mean BP 26, glucose 3.8 mmol/L. Thereafter, he received curosurf [surfactant] via ETT on the neonatal unit [NNU].

Respiratory system:
M was successfully extubated to continuous positive airway pressure [CPAP] 7 cm H_2O FiO_2 21% on day 3. He was reported as COVID-19 positive on days 4, 7, 10 and 17 on nasopharyngeal swabs. M was re-intubated on day 8 due to desaturation [12%]. Thereafter, a chest drain was indicated for right sided pneumothorax on day 8. High-frequency oscillatory ventilation (HFOV) was commenced in view of worsening respiratory distress syndrome [RDS]. Noted pulmonary haemorrhage from bloody ETT aspirates.

Cardiovascular system:
Day 2 echocardiogram demonstrated a small patent ductus arteriosus [PDA] for which paracetamol was given. A large PDA with left to right shunt was reported on Day 13. Multiple blood components [red blood cells, platelets, fresh frozen plasma, and cryoprecipitate] were transfused for anaemia, thrombocytopenia and haemorrhage.

Infection:
This neonate received intravenous benzylpenicillin and gentamicin for early infection risk post-delivery. Meropenem and flucloxacillin were commenced on day 8 due to suspected sepsis [pyrexia 38.2*C]. Blood and cerebrospinal cultures were negative.

Fluids/feeding:
Initial intravenous dextrose was altered to total parenteral nutrition and trophic feeds at a high-risk protocol.

Neurology:
Cranial ultrasounds demonstrated right periventricular flare and grade 2 intraventricular haemorrhage [IVH]. Other sources of haemorrhage were abdominal as evidenced by bloody nasogastric aspirates.

Outcome:
Unfortunately, M passed away aged 17 days old due to clinical deterioration especially in respiratory and vascular systems.

Discussion:
Early onset of neonatal COVID 19 infection was diagnosed due to the poor initial condition and rapid deterioration. M presented with respiratory distress, temperature instability, non-specific opacities on CXR, thrombocytopenia, abnormal coagulation screen, pneumothorax, pulmonary haemorrhage and bloody NGT aspirates which are part of the multi-system inflammatory neonatal COVID 19 syndrome.

The most common symptoms noted in infected infants are pyrexia, cough, rhinorrhoea, respiratory distress, poor feeding, lethargy, vomiting, diarrhoea, rash and oedema. Moreover, abnormal CXR findings of non-specific opacities and ground-glass changes were observed. Other studies have observed that leukocytosis, thrombocytopenia, hyperlactatemia, raised C-reactive protein, and lymphopenia occur.

Conclusion:
This was our first case of vertical transmission of COVID 19 resulting in devastating consequences. It was previously deduced that newborns may be infected with COVID 19 due to the immaturity of their immune systems. Although, the vertical transmission of SARS-CoV-2 seems uncommon due to lack of viremia and the non-overlapping expression of ACE-2 and transmembrane serine protease. Even as this pandemic continues to progress, our knowledge about neonatal COVID-19 infections and their clinical implications remains scarce.

Tao CJ, Gibby A, Burrows K, Johnson JA, Patil S, Polychronakis T. P15 Predictors of obstructive sleep apnoea in children with obesity. *BMJ Open Respiratory Research* 2023;**10**:doi: 10.1136/bmjresp-2023-BSSconf.26

Predictors of obstructive sleep apnoea in children with obesity

Introduction:

Obstructive sleep apnoea hypopnoea syndrome (OSAHS) is common in children with obesity and is associated with long-term morbidity. Understanding risk factors that predispose to OSAHS in children with obesity may help improve targeted screening with sleep studies and is likely to lead to early detection and intervention.

The aim of this multicentre retrospective case-cohort study was to assess how age, sex, BMI, and adenotonsillar hypertrophy presence correlate with OSAHS diagnosis in children with obesity.

Methods:

This was a retrospective review of medical notes of children with obesity, as defined by WHO (BMI-z-score >3 for children 0–5 yo, BMI-z-score >2 for children >5 yo) referred to three regional hospitals for sleep study between January 2020 and June 2023. Children with significant co-morbidities such as trisomy 21 or neuro-disability were excluded.

Results:

46 children (16 female: 30 male) with median age (range) of 9 years (2–16) and median BMI-z-score of 3.34 (2.12–7.67) were included in the analysis. 18 had adenotonsillar enlargement. 12 had history of adenotonsillectomy. Mean (standard deviation) Obstructive Apnoea Hypopnoea Index (OAHI) was 3.198 events/hr (3.918). 19 children had normal OAHI, 16 had mild OSAHS and 11 had moderate to severe OSAHS. Diagnosis of OSAHS was independent of age. Boys had significantly higher mean OAHI than girls (4.073 Vs 1.556 respectively, p=0.009). BMI-z-score moderately correlated with OAHI, (rho=0.362, p=0.040). There was no difference in mean BMI-z-score between normal, mild, and moderate-severe OSAHS groups (p=0.116). OSAHS was more common in children with adenotonsillar enlargement (Odds Ratio=10.5, 95% CI of 2.15–51.281, p=0.002)

Discussion:

Male sex, adenotonsillar enlargement, and higher BMI-z-score are associated with OSA diagnosis in children with obesity.

Tipton C, Polychronakis T

Preschool wheeze and response to Haemophilus influenzae type B conjugate vaccine

Haemophilus influenzae type b (Hib) conjugate vaccine is highly effective against invasive disease, reduces Hib nasopharyngeal carriage and produces a strong immune response in healthy children. Persistent bacterial airways infection is a driving factor behind neutrophilic asthma and preschool wheeze (PSW). Evidence suggests there are beneficial effects of Hib vaccination to the development and control of preschool asthma.

Our aim was to assess the response to primary Hib vaccination in children with PSW. 57 children (20 female) with a median age (range) 3 years (1-5) and diagnosis of PSW had Hib antibody concentration measured.

History of atopy	25/57 (43.9%)
Family history of atopy	34/57 (59.6%)
Long term azithromycin	36/57 (63.2%)
Moderate/ High dose inhaled corticosteroid	29/57 (50.9%)
admission to paediatric intensive care	8/57 (14%)
Bronchoalveolar lavage (BAL)	32/57 (56.1%)
Positive bacterial culture BAL	20/32 (62.5%)

Median (range) Hib antibody concentration was 1.46 mg/L (0.05-12.3) which is lower than reported titres in healthy vaccinated children in this age group. Suboptimal response (<1mg/L) was common (40.4%) and was independent of age, sex, atopy, treatment and BAL growth. 14% had very low titres (<0.1 mg/L). Children with neutrophilic BAL inflammation were more likely to have a suboptimal Hib vaccine response compared to those with mixed neutrophilic and eosinophilic picture (OR 3.50, 95% CI 0.472 - 25.902, p=0.212).

Suboptimal response to Hib vaccine is common in children with preschool wheeze. Booster vaccination is likely to be beneficial.

Ip N, Trigg A, Macdougall C, Bohatshchek M

Is it necessary to follow up all babies with weakly positive direct antiglobulin test (DAT)?

Objectives:
Unnecessary neonatal appointments cause inconvenience and anxiety for families and are a poor use of resources. Our 2019 local guideline recommended that all asymptomatic DAT+ babies should have a full blood count (FBC) and reticulocyte count checked at 2-3 and 6-8 weeks to exclude anaemia. Two Plan-Do-Study-Act (PDSA) cycles were conducted in 2019 and 2023 to determine:

1. If infants with weakly positive DAT could be safely excluded from follow-up.
2. If any infants requiring transfusion were missed after changing the guideline.
3. Any associated cost savings.

Methods:
Infants aged less than 1 month who were DAT-positive from 1 January 2017 to 15 December 2018 (cycle 1) and from 1 January 2021 to 31 December 2022 (cycle 2) were identified by the laboratory. Data regarding jaundice, anaemia, treatment received and follow-up were extracted from electronic patients records. The project was registered in the Trust's Quality and Safety Information System.

Results:
In PDSA cycle 1, 143 DAT+ neonates were identified. Sixteen infants developed anaemia (haemoglobin < 100 g/L), of which five required transfusion. Anaemia was uncommon in infants with weakly positive DAT. Based on our data, additional risk factors for anaemia were identified, including prematurity, need for intravenous immunoglobulin/exchanged transfusion and presence of other haemolytic anaemia. Consequently, we determined that it was safe to excluded DAT 0.5/1 + term infants with no features of active haemolysis or other known reasons for haemolysis from follow-up. This would equate to 95 infants in this cohort. The guideline was changed accordingly.

In PDSA cycle 2, 108 neonates with positive DAT at any grade were identified. Seven developed anaemia of which two required transfusion. All infants with DAT 2+ or above were followed up. Sixty-five infants in this cohort did not require follow-up due to the change in the guideline which previously each would have required at least 2 reviews. Over 2 years, we avoided at least 130 phlebotomy appointments and 130 telephone appointments: a cost saving of over £31,500 according to our Trust's tariff payment system. No infants who developed anaemia requiring transfusion were missed.

Conclusion:
It is safe to exclude term infants with 0.5/1+ DAT and no other risk factors for haemolysis/anaemia from follow up. This reduces the financial burden of follow-up and increases the availability of clinic time. Additionally, many babies were spared from painful procedures and unnecessary travel which also reduced carbon footprint.

Vadlamani A

Innovation in the treatment of complex epilepsy

Objective:

To establish whether a benefit exists for the use of K.Vita when treating patients with complex drug resistant epilepsy.

Methods:

A case series of twelve patient (age 7-17yrs) that have been trialled on K.Vita at the Child Development Centre, Addenbrooke's Hospital, and establishing whether their trial on K.Vita resulted in a change in seizure frequency (SF). Other parameters of note included tolerance and compliance to K.Vita treatment, as well as the time between the last SF recording before, and the first SF recording after the commencement of K.Vita. A review of the current literature was also conducted on both PubMed and Google Scholar with search terms 'K.Vita' and 'Epilepsy', with a total of 5 papers included in the review.

Results:

5/12 patients experienced a decrease in seizure frequency, with an average reduction in seizure frequency of 61.2%. Pre-K.vita seizure frequency (A), and post-starting K.vita seizure frequency (B) were compared using a t-test to compare log(A) and log(B). This decrease was not statistically significant (P=0.0854). 10/12 patients had values for A and B, the t-test comparing log(A) and log(B) was also not significant (P=0.2631).

Conclusion:

Due to the nature of drug-resistant epilepsy, these patients have significant difficulty in gaining control of their seizures, with many already having tried other forms of ketogenic diet (KD). From the small group that have currently been trialled on K.Vita, the results were not significant, however, I do believe that more trials should occur, with clear evaluation of seizure frequency at the appointment starting K.Vita, as well as the follow up.

Varma R, Holland J, Ratnaike TE, CHEERI EDI working group

Reducing unwarranted variation in investigations of children with early development impairment in East of England

Introduction:

Early development impairment (EDI) is when a child's development falls 2 SD below the mean for their age in two or more developmental domains before 5 years age. It is also called global developmental delay (GDD) or intellectual disability (ID) in school age population. It affects 1-3% of the children.

Aims:

Primary: To analyse the reasons for variation in practices and improve equity in investigations for children with early development impairment in East of England.

Secondary: Creating a regional project for paediatric trainees through CHEERI which is a paediatric trainee research network of East of England that will allow them to meet their QI/research curriculum requirements.

Methods:

Phase 1 – to establish the extent of variation in the practices:
Between July 2023 and September 2023, community paediatricians from different community trusts in East of England were contacted for an informal survey. In this survey we enquired about any guidelines (local/regional/published) that clinicians followed during evaluation of children with EDI. We contacted few community paediatrics consultants who were happy to champion our project amongst the wider community paediatricians.

Phase 2 – to analyse the reasons for variation:
After analysing the initial responses, an EDI working group was formed within the CHEERI which consisted of nine paediatric trainees who were enthusiastic and motivated to be part of the project.

Between October 2023 to March 2024, the group analysed the guidelines collected from the initial survey. A scoping review of existing literature was performed to understand the current recommendations. Based on the
initial analysis, a second literature analysis was performed by smaller group from within EDI working group, focusing only on genetic investigations in EDI. We liaised with lead clinical biochemist to obtain their feedback. The findings from existing guidelines and both the literature analyses were further presented in a joint meeting between CHEERI and representatives of community paediatricians and East genomics. Based on the recommendations from the meeting, a formal survey was sent to all the community paediatricians in the east of England region between March 2024- April 2024. The results from this survey and draft regional guideline for

investigations in EDI was further presented in the regional community paediatrics meeting which was also attended by few members of East genomics.

Results:

First informal survey: We received responses from 9 out of 14 community trusts. 3 followed regional PHG foundation guideline published in 2014, 3 followed individual expertise, 2 had local guidelines and 1 followed Mithyanta et al guidelines from Alder Hey published in Archives in diseases of childhood in 2017.

On liaison with clinical biochemist, UK National Metabolic Biochemistry Network Best Practice Guideline published in 2020 for investigations of GDD was obtained. Analysis of all the guidelines showed huge variation in investigations of EDI.

First literature analysis: A scoping review was performed for articles published between 2013-2023. We attempted to find answers to the following questions- In the investigation of EDI 1) What tests are recommended? 2) What tests are not recommended? 3) Should tests be done in a particular order? 4) What is the yield of the tests? 5) What is the evidence for and costs of the tests? There was lot of variation in the literature. However, the best quality evidence was for genetics. There was very low yield for metabolic tests but they were potentially treatable. MRI was not recommended as first line due to risks associated with general anaesthesia.

Second literature analysis: A focused second literature analysis on genetic investigations gave the following provisional conclusions- Even though yield of whole exome sequencing (WES)/ whole genome sequencing (WGS) was higher than aCGH (microarray based comparative genomic hybridization), it was more cost effective to do them as second line.

Second survey (formal): A formal survey sent to all community paediatricians in the East of England prior to the regional meeting received 26 responses. The barriers identified in the survey along with potential solutions and draft guideline were discussed in the meeting. There was consensus in favour of (re)- developing a regional framework to guide EDI investigations.

Trainees: Poster presentation of literature analysis and initial survey results was done in ARU Chelmsford conference in March 2024.

Future steps:

Currently, the work is ongoing for creating the final version of regional guideline in liaison with paediatric neurologist. Based on the findings of survey in April, further larger survey focused on solutions to the barriers to genetic investigations in children with EDI has been created in collaboration with East genomics and extended to all community and acute paediatricians in East Midlands along with East of England.

Varma R, Kayani R

A retrospective audit for pre-hospital RESPECT documentation for children with complex life limiting neuro-disability admitted to paediatric intensive care unit.

Introduction:
As per NICE guidelines [NG61], discussion of end-of-life care in children with complex life limiting neuro-disabilities or conditions, should be taking place long before they need PICU admission. If the discussions have taken place prior to admission, parents are more mentally prepared and accepting of the final outcome.

Objectives:
To assess, all children with complex life limiting neuro-disabilities or conditions, who were admitted and have passed away in the paediatric intensive care unit in East of
England, in the last 8 years, if they were a) known to any other team/hospice care or b) had documented RESPECT form or end of life care discussion.

Methods:
Retrospective audit of medical records between 2014 to 2022 was done. Electronic notes were scanned for evidence of advance care plans or their discussion prior to admission to paediatric intensive care unit.

Exclusion criteria: Children who died in PICU but did not have any pre-existing life limiting neuro-disability or conditions; children who died in PICU with cancer.

Results:
Total number of children who passed away during this period in the unit was 149. Only 2 out of eligible 33 children (6%) had documented advance care plans or referral to palliative care prior to PICU admission.

Discussion:
The emotional distress and other pressures inherent in situations in which patients are approaching the end of their life sometimes lead to misunderstandings and conflict between doctors and patients and those close to them, or between members of the healthcare team. However, this can usually be avoided through early, sensitive discussion and planning about how best to manage the patient's care.

Together for Short Lives outlines four types of illness trajectory which will require children's palliative care provision. This categorisation is important for the purposes of planning and needs assessment.

A child's best interests are not always limited to clinical considerations and it's important to take account of any other factors relevant to the circumstances of each
child in partnership with parents when considering decisions about their child's treatment.

Conclusion:
It is important to start discussions of advance care planning with families of children with complex neuro-disabilities prior to their PICU admissions wherever possible as it will allow parents and children to make informed decisions and have more say about their care away from stressful atmosphere of Paediatric Intensive Care.

Identifying and categorisation of trajectory of illness can guide to where and when these conversations should be initiated. Establishing who and where these conversations should take place and adequate training for healthcare professionals in this regard is essential. Documentation of these discussions and sharing with wider disciplinary team is of paramount importance.