Platinum Sponsor

Takeda

BSPGHAN

working for children with digestive and liver disorders

British Society of Paediatric Gastroenterology Hepatology and Nutrition

INCORPORATING JOINT STUDY DAY WITH CYSTIC FIBROSIS TRUST FRIDAY 31ST JANUARY 2020

E&OE

ANNUAL MEETING 2020

WEDNESDAY 29TH – FRIDAY 31ST JANUARY. THE DOME, CHURCH STREET, BRIGHTON

ABSTRACTS

Brighton Dome

Thanks to all of our sponsors

On behalf of BSPGHAN and the local organisers in Brighton, we are sincerely most grateful to all our partners for their contributions to the Annual Meeting.

Our commercial sponsors and charitable organisations play an integral part in our meeting to complement the independently developed scientific programme. This interaction provides the opportunity to share and acquire knowledge in partnership for the mutual benefit of patient care.

We are pleased to welcome established and new sponsors to our 2020 meeting providing some continuity but also variety with opportunity for new dialogue.

Informed dialogue and open exchange of information are of key importance; it is with this spirit, that we encourage all delegates to take time to visit the exhibitors and to thank them for their continued support of our annual BSPGHAN events.

PLATINUM SPONSOR

Takeda

SILVER SPONSOR

Mead Johnson Nutrition

rb HEALTH · HYGIENE · HOME

BRONZE SPONSORS

AMGEN

Nestlé HealthScience

ZERIA GROUP Tillotts Pharma GI-health is our passion™

Abbott

Calea the heart of homecare

POSTER IBD AWARD

Dr Falk

CHARITIES

Children's Liver Disease Foundation

CICRA

GUTS UK

SPONSORED SYMPOSIA

Takeda

NUTRICIA LIFE-TRANSFORMING NUTRITION

29TH - 31ST JAN 2020

BSPGHAN Abstract Book

Index:

Wednesday
29th January 2020

Plenary Session I
Oral Abstract Presentations

12.10 – 12.20

Nabilone for Gastrointestinal Dystonia. A single centre case series

Miss Michelle Brooks[1], Dr Joseph Symonds[2], Miss Sarah Bremner[3], Dr Victoria Merrick[1], Dr Diana Flynn[1], Miss Christina McGuckin[1], Mr Simon Fraser[4], Miss Karen Fraser[5], Mr Gregor Walker[6], Mr James Andrews[6], Mr Timothy Bradnock[6], Dr Andreas Brunklaus[2], Dr Katharine Forrest[2], Dr Valerie Orr[7] and Andrew Barclay[1].

[1]Paediatric Gastroenterology, Royal Hospital for Children, Glasgow UK; [2]Paediatric Neurology, Royal Hospital for Children, Glasgow UK;[3]Paediatric Dietetics, Royal Hospital for Children, Glasgow, UK; [4]Paediatric Pharmacy, Royal Hospital for Children, Glasgow, UK; [5]Admin Services, Royal Hospital for Children, Glasgow, UK;[6]Paediatric Surgery, Royal Hospital for Children, Glasgow, UK;[7]Paediatric Neurodisability, Royal Hospital for Children, Glasgow, UK

In severe neurodisabling conditions the clinical constellation of pain behaviour, retching, bloating, abdominal distension and constipation/pseudo-obstruction can be referred to as gastrointestinal dystonia (GID)(1).The evidence base for effective therapies are very limited (1) and symptoms can remain disabling. Nabilone a synthetic analogue of the active component tetrahydrocannabinol (THC) found in cannabis has a licence in paediatrics for the treatment of severe chemotherapy induced nausea, however, as medical cannabis has also been implicated in the treatment of pain, spasticity, gastrointestinal upset and apetite stimulation, it has been hypothesised as a potential agent to treat GID. To date we are unaware of any published work reporting the use of Nabilone in GID.

We aimed to describe our experience of using nabilone for patients with GID.

Approval was sought on a named patient basis from the clinical directorate and head pharmacist on the basis that; patients fulfilled our criteria for GID (1), patients had been trialled on medical therapies, jejunal feeding and blended diet, concomittant medication reviewed in MDT including tone management and to review potential exacerbants to GID, all members of MDT were in agreement that trial of treatment was warranted. Patients were admitted for 48hrs of documentation of symptoms using modified paediatric pain score. TPR and overnight saturations were peformed pre and post treatment for 48hrs. With the potential for pre-existing neuro-respiratory depression, Nabilone was commenced below previously described paediatric dosing and then incremented in 250ug doses to a dosage of <18kgs: 500ug bd and 18-27kg: 500ug tds. Efficacy was assessed by parents and clinicians perception, PedsQL ver 3.0 (GI questions) and sleep diary, all performed prior to treatment and one month after stable dose. Global Peds QL scores analysed by paired student's t-test ($p<0.05$).

3 patients were treated - all male, 2.4-9yrs, 10.5-18kg, 2 cerebral palsy and 1 undiagnosed condition. Parents and clinicians perceptions were that treatment had improved symptoms. All patients had an increase in their Peds QL scores post treatment. Most significant increase in median scores were in the domains 'stomach discomfort when feeding' (5 to 85 ($p<0.006$)) and 'nausea and vomiting' (6.25 to 56.25 ($P=0.02$)). The patient with sleep disturbance was awoken a median of 3 times pre to once per night on treatment.

Nabilone shows promise in treating GID , a poorly understood debilitating complication of severe neurodisability, in particular, symptoms of nausea and retching. Of note, two patients were being considered for parenteral nutrition for nutritional failure. Further evaluation of this treatment is warranted in clinical trials or on a further named patient basis. The authors advocate a wide MDT setting, with input from Gastroenterology, Surgery and Neurodisability. We currently reserve this for refractory patients for whom most known measures have been ineffective. The authors also note that the current objective tools have limitations in capturing efficacy of interventions for GID due to diffuse and complex symptoms. Future study would be aided by the existence of a specific assessment tool for symptomatology.

1. McConnell N, Beattie LM, Richards CE, Protheroe S, Barclay AR,JPGN;66 (supp 2):1002;2018

12.20 – 12.30

Children and young people with inflammatory bowel disease attend less school than their healthy peers

Claire Barnes[1], James Ashton[1], Florina Borca[2], Mick Cullen[1], Dawn Walker[3] and Robert Beattie[1].
[1]University Hospital Southampton
[2]NIHR Southampton Biomedical Research Centre; [3]University of Southampton

Aim:
Chronic diseases, such as inflammatory bowel disease (IBD), can impact negatively on education and social development. The aim of this study is to examine the extent to which IBD affects school/college attendance for children and young people (CYP) and determine contributing factors.

Methods:
We performed a cross-sectional survey to determine the school/college attendance rates, the reasons for absence related to IBD and facilitators/barriers to school/college attendance. In a subset followed-up locally, we performed a detailed review of hospital attendance data, to assess the healthcare burden.

Results:
Two-hundred-and-thirty-one questionnaires were given to CYP with IBD aged 5-17 years. The response rate was 74% (final sample 169). The median school/college attendance rate was 92.5%; significantly lower than all children in England (95.2%). 39.6% of children with IBD were persistently absent, defined nationally as missing 10% or more of school. Only five children (3%) had a 100% attendance record. Increasing age and use of monoclonal therapy were predictors of poor school attendance. Concerns about feeling unwell at school/college, access to toilets, keeping up with work, and teachers' understanding of IBD are the main issues for CYP with IBD. (This is highlighted in the word cloud - figure 1, illustrating the questionnaire free text comments). There was a significant negative correlation between number of days in hospital and school attendance.

Conclusion:
IBD has a significant impact on school/college attendance, with hospital attendance, disease burden, and school difficulties being major factors. Employing strategies to minimise healthcare burden and developing a partnership between health and education, to support children with IBD will serve to facilitate school/college attendance.

<u>Figure 1 – Word cloud of questionnaire free text comments</u>

A graphical representation of word frequency from the questionnaire free text comments. The size of the word indicates its frequency; the more often a word appears in the text the bigger and bolder it appears in the word cloud.

12.30 – 12.40

D1 biopsies increases the diagnostic yield in coeliac disease in children

Chloe Ashton[1], Jeng Cheng[1] and Veena Zamvar[1].
[1]Leeds Teaching Hospitals

Introduction/Background:

Coeliac disease is an autoimmune systemic condition, which is triggered by gluten in genetically predisposed patients. These patients are genetically predisposed to HLA markers DQ2 or DQ8. Coeliac disease can present in children with gastrointestinal symptoms, failure to thrive and dermatitis herpetiformis; and can be asymptomatic especially in high-risk individuals such as children with Down's syndrome, Type 1 diabetes and Turner's syndrome.

Children who are symptomatic, are recommended to undergo total immunoglobulin A (IgA), tissue transglutaminase (TTG) and anti-endomysial antibody testing as the first step in diagnosis. Current BSPGHAN Coeliac disease guidelines recommend that children with typical symptoms, positive TTG >10 times the upper limit of normal and positive anti-endomysial antibodies on repeat test; do not require histopathology testing and that clinical symptoms and antibody testing are sufficient for diagnosis. Duodenal biopsy is, however, still considered the gold standard for diagnosis of coeliac disease in children. Duodenal biopsies are classified according to Marsh criteria which assesses the degree of villous blunting and atrophy, crypt hyperplasia and intraepithelial lymphocytosis. The exact, optimal location of biopsy, however, remains slightly ambiguous. This is, in part, due to the patchy nature of coeliac disease and hence, the need to take multiple biopsies. BSPGHAN guidelines 2013 state 'At endoscopy, take 4 biopsies from D2 or lower and 1-2 from duodenal bulb'. There is some existing controversy that normal duodenal bulb histopathology may be mistakenly interpreted as pathological. Recent paediatric studies have highlighted the significant number of children with histology confirmative of coeliac disease seen only in D1.

Aim:

The primary aim of this study was to ascertain whether taking D1 biopsies increase diagnostic yield in coeliac disease in children in our cohort.

Subjects and Methods:

This retrospective study recruited 221 paediatric patients who were investigated for coeliac disease by upper GI endoscopy by one paediatric consultant gastroenterologist between January 2015 and April 2019. Each patient notes were reviewed and their blood markers as well as their endoscopy and histology results were recorded.

Results:

A total of 213 patients who underwent gastroscopy for suspected coeliac disease were included, 8 were excluded as the endoscopy and biopsy results were not available for review. Of those, 146 patients (71%) were diagnosed with coeliac disease. 25 patients (17.1%) had positive D1 biopsies and negative D2 biopsies. 5 (3.4%) patients had negative D1 biopsies and positive D2 biopsies. The remaining 116 patients (79.5%) had both positive D1 and D2 biopsies.

In our cohort, D1 biopsies were positive in 96.5 % of coeliac disease patients whilst D2 biopsies were positive in only 82.8% of cases. Thus, performing D1 biopsies increased the diagnostic yield by 17.1%

Summary and Conclusion:

D1 biopsies when performed with D2 biopsies improve the diagnosis and thus outcome for patients with coeliac disease. Absence of D1 biopsies would have resulted in diagnosis being missed in 17 % of the patients. Our study supports the current recommendations of multiple biopsies from duodenal bulb (D1) and distal duodenum.

12.40 – 12.50

PRedicting Outcomes For Crohn's dIsease using a moLecular biomarkEr: PROFILE trial

Nurulamin Noor[1], Biljana Brezina[1], Juan De La Revilla Negro[1], Francis Dowling[2], Leisha O'Brien[2], Simon Bond[2], Kamal Patel[3], Abigail Seward[3], Klaartje Kok[4], John Gordon[5], Lynne Whitehead[6], Sara Upponi[1], Paul Lyons[7], Eoin McKinney[7], Kenneth Smith[7], James Lee[7] and Miles Parkes[1].
[1]Addenbrooke's Hospital, Cambridge; [2]Cambridge Clinical Trials Unit, Cambridge University Hospitals NHS Foundation Trust; [3]St George's Hospital, London; [4]Barts and the Royal London Hospital, London [5]Royal Hampshire County Hospital, Winchester; [6]Clinical Trials Pharmacy, Addenbrooke's Hospital [7]Department of Medicine, University of Cambridge School of Clinical Medicine

Introduction:

The course of inflammatory bowel disease varies substantially between individuals, but the lack of reliable prognostic markers has hindered disease management because patients with aggressive disease are undertreated by conventional 'step-up' therapy while those with more indolent disease are exposed to unnecessary treatment-related toxicity if a more aggressive 'top-down' approach is indiscriminately used.

Previously, we have described a transcriptional signature detectable within peripheral blood CD8 T-cells at diagnosis, identifying two subgroups of patients, correlating with subsequent disease course. In order to translate this work to the bedside and overcome the technical challenges of separating cell populations, we sought to develop a whole-blood biomarker that could re-capitulate the prognostic CD8 subgroups and then assess whether this biomarker can improve clinical outcomes by appropriately matching therapy to disease course.

Methods:

From a training cohort of 69 newly-diagnosed patients, we simultaneously obtained a whole-blood PAXgene RNA tube and peripheral blood CD8 T-cell sample. Gene expression in both samples was measured by microarray. Statistical modelling was used to identify a transcriptional classifier in whole-blood gene expression data re-capitulating the CD8 findings and was subsequently optimised into a multi-gene qPCR assay with independent validation in a second, independent cohort of 123 newly-diagnosed patients.

The PROFILE trial has incorporated this classifier to compare relative efficacy of 'top-down' and 'accelerated step-up' therapy between biomarker-defined subgroups of 400 patients with newly diagnosed CD. Subjects within each biomarker subgroup will be randomised (1:1) to receive one of the treatment strategies until trial completion (48 weeks). The primary outcome is incidence of sustained surgery and steroid-free remission, with secondary outcomes examining a number of measures assessing burden of disease including quality-of-life.

Results:

Following the application of statistical (machine) learning methods described, a 17 gene qPCR assay was developed and optimised. In the validation cohort of 123 patient, patients could be classified into two distinct subgroups, IBDhi (high risk) and IBDlo (lower risk). Irrespective of the underlying diagnosis, IBDhi patients experienced significantly more aggressive disease than IBDlo patients, with earlier need for treatment escalation (hazard ratio=2.65 (CD), 3.12 (UC)).

Subsequently this biomarker is being used to stratify therapy in the PROFILE trial, where 42 sites have been opened and at the time of writing 200 participants have been randomised, with recruitment ongoing.

Conclusions:

We have developed, optimised and validated a prognostic transcriptional biomarker that predicts clinical outcomes from diagnosis in patients with IBD. This classifier is currently being used in the PROFILE trial – the first ever biomarker-stratified, trial in Gastroenterology. If clinical utility of a stratified treatment approach is demonstrated, this would represent a major step towards personalised therapy in IBD.

12.50 – 13.00

Scottish longitudinal home parenteral nutrition dataset shows increased prevalence and survival: a unique national survey over 16 years.

Neil McConnell[1], Christina McGuckin[2], Catherine Paxton[1], David Mitchell[1], Simon Fraser[2], David Hoole[1], Gregor Walker[2], Timothy Bradnock[2], Atul Sabharwal[2], James Andrews[2], Carl Davis[2], Kathleen Ross[3], Shyla Kishore[3], Diana Flynn[2], Paul Henderson[1] and Andrew Barclay[2].

[1]Royal Hospital for Sick Children, Edinburgh; [2]Royal Hospital for Children, Glasgow; [3]Royal Aberdeen Children's Hospital, Aberdeen

Objectives:

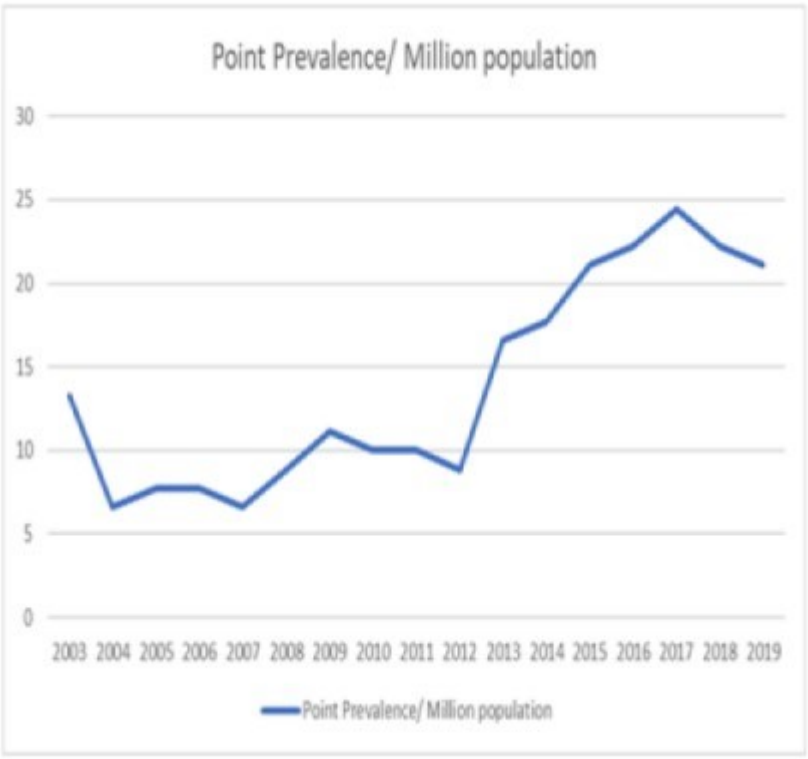
The prevalence of patients receiving home parenteral nutrition (HPN) has been rising over the last two decades in the United Kingdom. It is not clear whether this is driven by an increasing case load into HPN services, improved survival, or both. Longitudinal surveys of HPN services with reference to demographics, survival and weaning PN have not had complete ascertainment on a national level. We aim to describe the incidence, period and point prevalence, and outcomes of HPN patients within Scotland from 2003-2019, using our unique dataset collated from Scottish-wide MCN and centralised regional data, with specific reference to long term outcome and survival.

Design:

Period prevalence (calendar year), point prevalence (31st October annually) and incidence of paediatric HPN for each year from 2003-2019 were evaluated by a prospective single group cohort study. Basic demographics included age, diagnostic category (short bowel syndrome [SBS], Neuromuscular [NM]; mucosal). Outcomes were defined as: still on PN at last paediatric review; weaned PN; weaned (transplanted) and died. Population data from the National Records of Scotland provided population estimates for children aged <16 years. Trend analyses were performed between three epochs: 2003-2008 and 2009-2013 and 2014-2019 using Poisson regression. Two-year transplant-free survival and survival rates (excluding deaths not attributable to IF/HPN) were also calculated as per previous survey methodology.

Results:

A total of 76 HPN cases (58% female) with 90 HPN episodes were identified. Most recent point prevalence (31/10/19) was 21/1,000,000 Scottish population under 16 years. Incident cases across the three epochs were stable around 4/1,000,000/yr. There was no significant increase in period prevalence between 2003-2008 and 2009-2013 but a significant increase between 2003-2008 and 2014-2019 ($p<0.001$) and 2009-2013 and 2014-2019 ($p<0.001$) which suggests a more recent rise. This was driven by a significant increase in SBS ($p<0.005$) and NM ($p=0.006$) period prevalence. Diagnostic split was 43 (57%) patients with SBS, 16 (21%) with NM and 17 (22%) with a mucosal cause. There was no difference in diagnosis percentages (i.e. SBS, NM, mucosal) across the 3 epochs ($p=0.838$). Weaning was achieved in 35 (46%) of patients with 24 (31.5%) remaining on PN at last paediatric review and 17 (22.5%) having died. Two-year transplant free survival for incident cases per epoch were 13/20 (65%, 2003-2008), 18/21 (85%; 2009-2013) and 20/21 (95%; 2014-2019). Two-year transplant free survival was significantly better between 2003-2008 vs. 2014-19 ($p=0.02$). Excluding deaths non attributable to IF/HPN 2yr survival for 2014-2019 was 100%.



Discussion:

We report the first longitudinal national survey of outcomes for HPN in the UK with complete ascertainment and outcomes for cases. Our data show that incidence is stable but that prevalence is rising suggesting survival has improved significantly during the survey and our most recent data compare favourably with international centres of excellence. We postulate the use of virtual managed clinical networking with sharing of expertise to improve outcomes for small to medium sized IF centres. This dataset can provide the template and impetus for the completion of full ascertainment HPN longitudinal survey via the UK Paediatric eBANS group.

13.00 – 13.10

Paediatric e-BANS

Anthony Wiskin[1], Rachel Russell[2], Thomas Julian[3] and Akshay Batra[2].
[1]University Hospital Bristol NHS Trust; [2]University Hospitals Southampton NHS Trust;
[3]Great North Children's Hospital Newcastle

Introduction:

The Paediatric e-BANS was established in 2015 to provide an accurate contemporary database of children requiring home parenteral nutrition (HPN) in England. Prevalence data collected in September 2018 showed there were 352 patients receiving HPN in England.

Aims and Objectives:

We aimed to report the engagement of paediatric intestinal failure centers with e-BANS, by monitoring activity on the database over a 12 month period. The secondary aim was to maintain the database to provide reliable national long-term outcome data.

Subjects and Methods:

A list of paediatric centres providing HPN in England is maintained within e-BANS and all centres encouraged to enter cases regularly. This was achieved by regular contact with identified leads in each centre and site visits where required. Activity on the database between 1st September 2018 and 31st August 2019 was recorded. This included addition of new patients or updating information held for existing patients.

Results:

A total of 535 patients on home PN were notified to the database since its establishment. 352 patents were active on PN on the 1st of September 2018. 259 records were updated during the 12 month study period and 39 new patients added. Of these 17 had short bowel syndrome, 8 neuromuscular diseases, 3 congenital enterocyte disorder and 11 other disorders. 14 patients were weaned off PN, 5 transitioned and 1 died. The net increase in number of patients was 17 putting the total number of patients on Home PN up from 352 to 369.

Summary:

1. There was good engagement from all centers and 73% patient records were updated over the last 12 months.
2. The number of patients on home PN, managed by Intestinal Failure centers in England increased by 5% over the year.
3. 4% of patients on HPN were able to successfully wean off PN.

Conclusion:

A National Database of Paediatric HPN patients is achievable in England. Longitudinal data from this will be very important in service planning and in determining outcomes of care.

Thursday
30th January 2020

Plenary Session II
Oral Abstract Presentations

14.00 – 14.10

Improved neonatal outcome following implementation of NST

Femi Dhilip[1], Lauren Hill[1] and Christopher Forster[1].
[1]Leeds Teaching Hospitals

Introduction:

Recent consensus from both BAPM & ESPGHAN recommends a multidisciplinary team approach to nutrition in preterm and growth restricted neonates, ideally in the form of a 'Nutritional Support Team' (NST).

We present the outcomes from a tertiary surgical neonatal unit following the implementation of a NST .This was established within the Leeds Neonatal Service and comprised a Consultant Neonatologist, a specialist neonatal dietician and a pharmacist. Weekly ward rounds were set up with standardized nutrition proformas, along with supported implementation of new feeding guidelines, delivering substantial educational input to the medical and nursing staff, along with the daily presence of a neonatal dietician.

Objective:

To compare nutrition outcomes before and after the introduction of a dedicated NST.

Setting:

Level III Surgical NICU at Leeds Children's Hospital.

Patients:

Growth rates, Catheter Related Blood Stream Infection (CRBSI) rates and inborn NEC rates were collected prospectively for all babies seen by the NST in the 12 months following setting up the service and were compared to rates prior to the service. In addition, a cohort of thirty neonates with a birth weight of < 1.25kgborn prior to the service, were compared to an equal number of neonates born 12 months afterwards.

Design:

A service review assessed the following parameters,
Growth rates, Length of inpatient stay (days),Number of days on PN,
Time to achieve full enteral feed, Evidence of conjugated hyperbilirubinemia, Rates of: (CRBSI), (NEC)

Results:

Growth rates for preterm infants <36 weeks gestation increased by 165% - from 9g/kg/day to 14g/kg/day following implementation of the NST. In babies < 30 weeks, this degree of improvement increased from 8 to13g/kg/day.
46% of babies in the pre cohort crossed two or more centiles in the weight between birth and discharge, compared to only 25% in the post cohort.

12 months following the NST being established ,a considerable drop was found in the following :length of stay, with average admission duration dropping from 67 to 51 days, mean duration of PN, falling by 65% (26 to 9.5) with a corresponding drop in time to establish full feeds(25 days to 15 days). There were 5 cases of conjugated hyperbilirubinemia, compared to 4 babies in the post. NEC rates were 6 pre and 10 post, but CRBSI rates fell significantly from 2.85 to 0.91 per 1000 line days.

Conclusion

This service review demonstrates considerable improvements in nutritional outcomes after the introduction of a NST in a tertiary neonatal unit. Growth rates were increased by 165% with a significant drop in CRBSI, days of PN and time to full feeds , along with a drop in total length of stay, which in combination have significant financial implications. As well as these quantitative changes, there has also been a tangible change in approach to nutrition across all staff groups, with

consideration for growth and nutrition now embedded in the culture of the unit, which is likely secondary to the regular presence of the NST within the unit, standardized feeding protocols and effective and ongoing education.

14.10 – 14.20

Study of acute liver failure in children using targeted next generation sequencing technology

Robert Hegarty[1, 2], Melissa Sambrotta[2], Philippa Gibson[2], Sandra Strautnieks[1], Tassos Grammatikopoulos[1, 2] and Richard Thompson[1, 2].
[1]King's College Hospital NHS Foundation Trust ; [2]King's College London

Introduction:

Acute liver failure (ALF) in children is rare but, without treatment, a potentially fatal condition. Whilst monogenic, metabolic diseases form the largest diagnostic category, diagnosis can be difficult such that in up to 43% of cases the cause remains undetermined.

Aim:

The study's aim was to identify undiagnosed, monogenic causes of ALF using targeted, next generation sequencing (NGS) technology and report on its clinical utility. The primary objective was to identify undiagnosed monogenic diseases in children who received a diagnosis of indeterminate ALF and to describe their characteristics. The secondary objectives were to: (1) provide the research foundation to construct a diagnostic, genetic panel using targeted NGS technology for future children with indeterminate ALF; (2) identify and characterise children who have a mutation in NBAS (or any other novel gene).

Methods:

Children < 10 years of age admitted to our centre with ALF who had stored Biobank blood samples were included in the study. An NGS custom panel of 64 candidate genes, known to cause ALF and / or metabolic liver disease, was constructed using the online Agilent SureDesign tool. The Agilent SureSelect QXT kit was used for the library generation and preparation of samples.

Results:

Sequencing was performed on genomic DNA extracted from blood samples of 41 children (male = 23) with a median age, at presentation with ALF, of 2.2 years (range: 7 days to 8.6 years). The depth of coverage was > 30X in > 99.9% of targeted regions. The total number of non-synonymous variants with a minor allele frequency of < 1 % was 69 (29 children) in 40 genes: biallelic variants = 9 (8 children) in 7 genes; monoallelic = 60 (27 children) in 40 genes. Biallelic variants, including 1 child with a copy number variant, were found in the following genes (number of children): NBAS (3); TWINK (1); CPT1A (1); SUCLG1 (1); POLG (1); MPV17 (1); DLD (1), summarised in Table 1. Amongst these 8 children, 5 had received emergency liver transplantation and all patients were alive at last follow up.

Table 1. Summary of patients found to have biallelic, rare, non-synonymous variants

ID	Gene	Zygosity	Genotype	Protein	MAF	CADD / Pathogenicity
1	*NBAS*	Hom	c.4731_4733dup	p.Tyr1578dup	Not reported	*Pathogenicity likely*
2	*NBAS*	Hom	c.exons 17-19 dup	p.?	Not reported	*Pathogenicity likely*
3	*NBAS*	Het	c.1702G>A	p.Val568Ile	0.0012	15
			c.191G>A	p.Arg64His	6.30E-05	19
4	*TWINK*	Het	c.1471C>G	p.Ser491Thr	3.99E-06	24
			c.1697A>G	p.Lys566Arg	3.99E-04	22
5	*CPT1A*	Hom	c.2198A>G	p.Asn733Ser	0.0018	16
6	*SUCLG1*	Het	c.601A>G	p.Arg201Gly	1.99E-05	30
			c.236G>A	p.Gly79Asp	0.0013	27
6	*POLG*	Het	c.153_158delGCAGCA	p.Gln54_Gln55del	0.0036	*
			c.803G>C	p.Gly268Ala	0.0039	27
7	*MPV17*	Hom	c.338C>T	p.Pro98Leu	5.31E-05	25
8	*DLD*	Het	c.826A>T	p.Thr276Ser	5.42E-04	25
			c.911T>C	p.Ile304Thr	1.57E-04	26

MAF, minor allele frequency; CADD, Combined Annotation Dependent Depletion; Hom, homozygous; Het, heterozygous

* Polyglutamine tract expansion

Conclusion:

Biallelic variants of varying pathogenicity were found in 8/41 children (20%) whereby variants in NBAS and genes encoding mitochondrial proteins were the most common findings. Further studies are required to evaluate the contribution of these genetic variants to ALF. Nevertheless, decision for liver transplantation may have been influenced if these genetic results were available at the time of ALF. In the future, a rapid sequencing NGS workflow could aid in reaching a timely diagnosis and clinical decision making.

14.20 – 14.30

National study of management of acute variceal bleeding in paediatric liver centres

Suzanne Davison[1], Tassos Grammatikopoulos[2] and Jane Hartley[3].
[1]Children's Liver Unit, Leeds Children's Hospital; [2]Paediatric Liver, GI and Nutrition Centre and MowatLabs, King's College Hospital, London; [3]Liver Unit, Birmingham Women's and Children's Hospital, Steelhouse Lane, Birmingham

Background:
Evidence is lacking to guide management of oesophageal varices in children. In 2016 a retrospective audit identified differences in practice between the three National Specialist Liver Centres. In 2017 a consensus guideline was agreed and endorsed by BSPGHAN for both surveillance and treatment of varices and management of acute variceal bleeding. A prospective 12 month audit of this guideline took place at each centre in 2018, including all endoscopies for either surveillance or acute bleeding management. This study relates to those requiring management of acute variceal bleeding.

Aim:
Describe the population and management of children presenting with acute variceal bleeding who are transferred to a national liver centre during a twelve month period.

Methodology:
Proforma completed at time of procedure and discharge for consecutive children undergoing oesophagogastroduodenoscopy (OGD) for acute bleeding at each of three centres during 2018. Lead investigator at each centre collated and submitted anonymised data to co-ordinator for analysis.

Results:
Of 517 OGD in 349 children [(M=195) median age 9y8m (4m-17y10m)], 61 (12%) were for acute bleeding in 41 children [(M=26) median age 4y1m (9m-14y7)]. Underlying diagnosis was extrahepatic portal vein occlusion (EHPVO) in 21 and chronic liver disease in 20.

Table:

	all	First bleed OGD	Previous OGD
Number OGD/children	61/41	19/19	42/24
Age at OGD median(range)	3y11 (8m-14y7)	3y3 (8m-13y4)	5y1 (9m-14y7)
Interval bleed to OGD median days (range)	2 (0-11)	3 (1-11)	1 (0-7)
Rx sclerotherapy *	28 (46%)	10 (53%)	18 (43%)
Rx variceal band ligation*	26 (43%)	9 (47%)	17 (40%)
Referral from secondary care	25 (41%)	7 (37%)	18 (43%)
Referral from tertiary care	17 (28%)	10 (53%)	7 (17%)
Referral in house	19 (31%)	2 (11%)	17 (40%)

*1 patient underwent both sclerotherapy and band ligation

Initial management included infusion of blood (39%), platelets (25%), plasma derivative (11%) or cryoprecipitate (5%). Compliance with guideline for administration of IV antibiotics was 89%, octreotide 92% and PPI 100%. One child presenting in house required resuscitation and ventilation. No child had Sengstaken Blakemore tube (SBT) inserted prior to transfer, two required SBT after OGD with one having SBT re-insertion for ongoing bleeding. 13/41 children (32%) had complicated course. 12 required subsequent OGD for further bleeding: 4 had ongoing bleeding, 7 re-bleeding and 1 both ongoing and re-bleeding. Of these one with A-1-AT deficiency underwent emergency TIPSS

procedure with SBT in place preceding emergency LT, one with Alagille syndrome developed hepatic decompensation and one with Langerhan Cell Histiocytosis and SBT placement underwent early LT. One child developed aspiration pneumonia.

Summary and Conclusion: A co-ordinated national guideline and subsequent audit has been achieved. Sixty-one cases of acute variceal bleeding in 2018 were identified, with underlying EHPVO in 51%. Use of IV antibiotics and octreotide as initial management can be improved. As children presenting with acute variceal bleeding are young (median age 3y11m) sclerotherapy remains an important aspect of management. This audit provides data that will inform pre-procedural counselling.

14.30 – 14.40

Nutrition Evaluation Screening Tool (NEST): An easy to use, newly developed screening tool for hospitalised children

Kitt Dokal[1], Nadia Asmar[1], Rita Shergill-Bonner[1] and Mohamed Mutalib[1].
[1]Evelina London Children's Hospital

Introduction:

The Nutrition Evaluation Screening tool (NEST) is a nutritional screening tool that was developed at the Evelina London Children's Hospital (ELCH) and has been used in clinical practice for the last few years.

Aims:

The objectives of this study were to:

- Evaluate compliance of the NEST.
- Determine to what extent the NEST agrees with other established paediatric nutritional tools (Strongkids and STAMP) in nutrition screening.
- Determine how the NEST agrees with the Subjective Global Nutritional Assessment (SGNA) in identifying malnutrition.

Methods:

A retrospective descriptive study. 102 patient episodes were collected by randomly choosing patients from three medical and surgical wards at the ELCH.

Over 6 weeks patient episodes were collected from patient records. NEST compliance was checked to see if it was completed for patients within 24 hours of admission and once every week, while an inpatient at the ELCH.

Two medical students completed the NEST, Strongkids and STAMP screening tools, as well as the SGNA for each patient episode. Each tool uses defined criteria to assess nutrition. Criteria assessed by the NEST: weight loss, poor weight or poor food intake, diagnosis with risk of malnutrition; weight and height >2 centiles apart. The STAMP and Strongkids have their own respective criteria. A score is then produced using each screening tool, ranging from low, medium to high risk – with a recommendation of an action to be taken.

The SGNA is a nutritional assessment tool which factors in nutritional history and examination to produce an overall SGNA score ranging from normal, moderate and severe. Statistical analysis using Cohen's Kappa, was undertaken to assess agreement of the NEST with the other screening tools (Strongkids and STAMP), and with the SGNA.

Results:

89/102 (87.2%) of patient episodes were NEST tool compliant.
As shown in the table 1, there is agreement with the three other nutrition tools, for patients on initial screening at admission (N=76). The NEST is these initial cases, showed moderate agreement with the Strongkids and STAMP tools.
The NEST tool was found to have fair agreement with the SGNA in the cases at presentation.
Follow up patients (N=26) assessed with the NEST showed no statistically significant agreement compared with the three nutrition tools.

Table 1	Agreement with the NEST			
Nutrition Tool	Kappa (κ)	p-value	Standard error	Confidence Interval (95%CI)
$Strong_{kids}$	0.472	<0.005	0.082	0.311 to 0.633
STAMP	0.416	<0.005	0.084	0.251 to 0.580
SGNA	0.301	<0.005	0.083	0.138 to 0.464

Table 1: **Table showing comparison of the NEST tool against with other nutrition tools on patient with initial presentation to the Evelina London Children's Hospital** – For each nutrition tool ($Strong_{kids}$, STAMP and SGNA), kappa, p-value, standard error and confidence intervals were calculated for agreement with the NEST.

Summary:
The NEST is a user-friendly tool and practical tool at the ELCH.
For first episodes at the ELCH, the NEST has moderate agreement with both the Strongkids and STAMP. There was fair agreement between STAMP and Strongkids.
The NEST had fair agreement with the SGNA, and had the highest agreement compared with the other screening tools assessed for agreement with the SGNA. The NEST should be considered as a viable nutritional screening tool in a paediatric hospital, caring for children with complex medical problems.

14.40 – 14.50

A national study of surveillance and primary prophylaxis of oesophageal varices in paediatric liver centres

Suzanne Davison[1], Tassos Grammatikopoulos[2] and Jane Hartley[3].
[1]Children's Liver Unit, Leeds Children's Hospital; [2]Paediatric Liver, GI and Nutrition Centre and MowatLabs, King's College Hospital, London; [3]Liver Unit, Birmingham Women's and Children's Hospital, Steelhouse Lane, Birmingham

Background:
Evidence is lacking to guide management of oesophageal varices in children. In 2016 a retrospective audit identified differences in practice between Specialist Centres. In 2017 a consensus guideline was agreed for management of varices. Recommendations for surveillance included (a) platelet threshold of < 120x 109/L for initiating surveillance, (b) primary prophylaxis and re-scope interval according to grade of varices (c) routine administration of platelets not required. A prospective audit was performed at each centre in 2018: This study relates to children undergoing surveillance and primary prophylaxis.

Aim:
Describe population and management of children undergoing surveillance / primary prophylactic treatment of oesophageal varices at the three national liver centres during 2018.

Methodology:
Proforma completed at time of oesophagogastroduodenoscopy (OGD) for consecutive children undergoing surveillance and prophylactic Rx in 2018. Lead investigator at each centre collated and submitted anonymised data to co-ordinator for analysis.

Results:
Of 517 OGD in 349 children, 456 (88%) were for surveillance: 157 secondary and 299 primary prophylaxis. Only those in whom primary prophylaxis was initiated during the audit period [n=121, median age 10.2y (4m-17y 7)] are included. Diagnosis was chronic liver disease (112) and extrahepatic portal vein occlusion (9). Indication for OGD was platelets < 120x109/L in 43, and in 78 with platelets > 120x109/L was opportunistic during other procedure (44) splenomegaly (16), varices on imaging/ history of possible bleed (8), management planning / transplant assessment (8), other (2.) Two with only fundal varices are excluded.

Table:

Grade of varices and Rx	n	Platelet count $x10^9$/L median (range)	Platelets < $120x10^9$/L n (%)	Platelets > $120x10^9$/L n (%)
All	119	148 (34-592)	42	77
Grade 0	67	204 (38-504)	11 (16%)	56 (84%)
Grade 1 and 2 no Rx	31	135 (34-592)	14 (45%)	17 (55%)
Grade 2 Rx and 3	21	77 (40-316)	17 (81%)	4 (19%)

Platelet threshold of 120x109/L had sensitivity 40% and specificity 95% for presence of varices requiring Rx.

Primary prophylaxis was banding in 19/21, sclerotherapy (1) and both (1). One child (indication varices on CT, platelets 166 x109/L) had unsuccessful banding resulting in trauma to varices and acute bleed. He developed aspiration pneumonia and subsequently underwent embolization of varices. One child required observation overnight for respiratory symptoms, and one received platelets post OGD. Of 67 children undergoing OGD only no other child received blood products. Discharge six hours post OGD occurred in 28/67. Reason for delayed discharge included : other management / assessment 15, distance home in 22, respiratory symptoms (1) difficult OGD (1).

Planned follow up interval in those commencing surveillance for platelets < 120 was concordant with guideline in 17/36 (6 no plan stated), being longer in 18 and shorter in one.

Summary and Conclusion:
National consensus guideline and audit of surveillance endoscopy has been achieved. Surveillance OGD and Rx may be successfully performed as day case procedure. Platelet threshold 120x109/L has good specificity for predicting varices that require Rx. Interval to next OGD was according to operator decision rather than compliant with guideline.

14.50 – 15.00

A single-centre experience of outcome of Kasai Portoenterostomy with adjuvant protocol

Kavitha Jayaprakash[1], Suzanne Davison[1], Sanjay Rajwal[1] and Naved Alizai[1].
[1]Leeds Children's Hospital, Leeds Teaching Hospitals NHS Trust

Background:

Biliary atresia (BA) is a progressive fibro-obliterative disorder of the intra and extrahepatic biliary tree in infants. Kasai hepatico-portoenterostomy (KP) is preferred initial management with aim of improving bile flow and preserving native liver. Published European national rates of jaundice clearance after KP ranged from 40-60%, with 5-year-survival of native liver of 53%. However data from national studies reflect multicentre experience, combining differing surgical approaches and varying regimes for post KP adjuvant therapy. Evidence supporting adjuvant therapy remains lacking.

Since our centre became a designated Specialist Liver Centre in 2000, all KP operations are performed by a designated paediatric hepatobiliary surgeon, with postoperative management guided by a single protocol including corticosteroids, antibiotics and choleretic adjuvant therapy.

Aim:

Assess outcome (minimum of two years post KP) of consecutive infants undergoing KP managed according to a single protocol. Outcome includes rate of clearance of jaundice, mortality, survival with / without native liver, and development of portal hypertension.

Method:

Retrospective study of all infants with BA identified through an electronic database that underwent KP at our centre between 01/01/2000 and 31/12/2016. Data collection included age at admission, age at KP, presence of Biliary Atresia Splenic Malformation (BASM), clearance of jaundice (bilirubin < 2200μmol/L at 6 months), date of death, date of transplant, presence of portal hypertension, date of last follow-up.

Results:

164 children were identified of whom 23 (14%) had BASM. KP was performed by one of two surgeons. Median age at admission was 43 (4-135) days and at operation was 48 (11-143) days). Clearance of jaundice occurred in 104/164 (63%). In successive quarters of the cohort, median age at Kasai in days/jaundice clearance were 50/56%, 51/49%, 45/68% and 38/80% respectively.

Of 164, 17 with follow up data < two years (due to transfer elsewhere) are excluded. Of remaining 147, median duration of FU is 8 (2.1-17.2) years. Overall survival is 138/147 (94%). Of nine children who died, deaths occurred > 30 days post KP. Causes included pneumococcal sepsis (1), end stage liver disease awaiting transplant (3), post-transplant (2) and unrelated causes (3). Survival with native liver is 73/147 (50%). Of those with BASM, overall survival is 20/22 (91%) (one no FU data) and survival with native liver 16/22 (73%).

In 73 surviving with native liver [median FU 6.2 (2.1-17.2) y], portal hypertension defined as splenomegaly, thrombocytopenia, presence of varices or ascites developed in 26 (36%).

Summary and Conclusions:

Outcome at a single centre with uniform protocol for adjuvant therapy is at least compatible with national outcome data, with jaundice clearance rate of 63%, overall survival 94% and survival with native liver 50%. Improving trend in jaundice clearance warrants further investigation. Of those surviving with native liver, 64% have no evidence of portal hypertension after 6.2 y follow up. Children with BASM did not have an inferior outcome. This data provides supporting evidence to continue the current protocol, guide counselling and informed consent.

The authors acknowledge the contributions of colleagues past and present who have contributed to the data.

Friday 31st January 2020

Plenary Session III
Oral Abstract Presentations

9.00 -9.10

How many children with severe neurodisability are being sent home on parenteral nutrition?

Elena Cernat[1] and John Puntis[1].
[1]Leeds Children's Hospital, Leeds General Infirmary, Leeds, UK

Introduction/Background:

Up to 85% of children with severe neurodisability have eating problems, many requiring tube feeding. Rarely, due to severe gastrointestinal dysmotility and feed intolerance, parenteral nutrition (PN) may need to be considered.

Aim:

The aim of the study was to identify which intestinal failure (IF) centres in England offer home PN (HPN) for this group, how frequently this occurs, and how decisions are made.

Subjects and Methods:

A questionnaire was sent to the IF centres in England identified via the eBANS HPN registry after approval from the eBANS committee. Questions included: numbers of patients currently on HPN; the proportion with severe neurodisability; the number of these over a five year period; underlying diagnosis; who made the decision to initiate PN; if there was a written policy regarding selection for HPN; how many regained enteral autonomy; was there routine involvement of the palliative care team in the decision making?

Results:

18 IF centres offering HPN were contacted and 13 (72.2%) replied. In 12/13 (92.3%), the questionnaire was completed by a doctor and in one by a dietician. 12/13 centres accepted patients with severe neurodisability for HPN although some indicated reluctance to do so, emphasising the need for careful consideration on a case-by-case basis. The 13 centres were managing a total of 256 HPN patients; among these, 19/256 (7.4%) had severe neurodisability. Over the last 5 years, HPN was provided for 36 patients with severe neurodisability; 3 centres had never had such a patient on HPN but 2 of these would offer PN for inpatients. In 1 case, PN was withdrawn in hospital and the outcome was death. In the majority of cases, the nutrition team initiated the PN, in 2 cases it was the referring gastroenterology team from another hospital and in another 2 cases the neonatal/surgical team. 9 centres provided information on underlying diagnosis – the most common being various genetic conditions (n=11) followed closely by cerebral palsy (n=10); in 4 cases the diagnosis was unknown although is likely to be a genetic disorder. None of the centres had a written policy regarding which patients would be accepted for HPN and which excluded, but 8 centres mentioned that the decision to initiate HPN was only taken after each case had been discussed in a multi-disciplinary nutritional team meeting; only 2 centres involved a clinical ethical committee in the decision making. 11 centres included the palliative care team in the management of patients; 1 had twice withdrawn PN as part of a palliative care plan, both patients subsequently dying. Over five years 7/36 (19.4%) patients regained full enteral autonomy.

Summary and Conclusions:

Despite difficult ethical issues relating to judging quality of life and use of resources, the majority of IF centres in England offer HPN to patients with severe neurodisabilty. This usually happens following careful MDT discussion and with involvement of a specialist in palliative care. Only 1 in 5 children were weaned from HPN after spontaneous improvement in gut motility.

9.10 – 9.20

EMERGENCi: A UK national prospective survey of severe GI bleeding (requiring upper GI endoscopy) and emergency endoscopy in under 16s

Nicholas Croft[1, 2], Ramiya Kirupananthan[3], Polychronis Kemos[2, 4], Michael Stanton[5, 6] and Emergenci Collaborators[1, 5].

[1]BSPGHAN; [2]Barts and the London School of Medicine, Queen Mary University of London; [3]Children CRF, Royal London Children's Hospital, Barts Health NHS Trust; [4]Centre for Immunobiology, Blizard Institute, Barts and the London School of Medicine, Queen Mary University of London

[5]BAPS; [6]University Hospital Southampton

Objectives and Study:

EMERGENCi is a prospective national cohort study of emergency endoscopy and severe upper GI bleeds in children. Objectives were to identify information about emergency paediatric endoscopy services available across the UK, plus produce national data of the incidence and indications for emergency endoscopy.

Methods:

Units were identified through the UK Paediatric Gastroenterology and Paediatric Surgical Societies (BSPGHAN & BAPS). Denominator data was collected (using Redcap) to identify existing facilities, staffing and rotas and the geographical area and total population covered. Once registered a fortnightly email was sent asking for reports of severe upper GI bleeds and/or emergency upper endoscopies over a 6 month period. Inclusion criteria were < 16 years AND either Upper GI bleed requiring endoscopy (i.e. with significant haematemesis/melaena/other suspicion of major upper GI bleed) OR urgent upper GI endoscopy performed for other reasons (e.g. foreign body). Any cases reported required a short case report form to be completed.

Results:

26 units responded and provided denominator data (covering 83.2% of the UK population of <16 years= 10 million). 17/26 were children's hospitals/units co-located with adult hospital, 5/26 were standalone children's hospitals and 4/26 were in a district general setting. 62.5% have an on call medical paediatric gastroenterology rota of which at least half do not include return to hospital out of hours, i.e. for telephone advice only. 64% of units reported availability of out of hours emergency endoscopy rota.

62.5% of centres had a protocol for severe upper GI bleeds and 40% for management of other indications.

20/26 registered centres (covering 61% of the UK population) then provided prospective cases by responding to fortnightly emails. 91 cases have been reported: 33 for severe upper GI bleeds (7 variceal), 35 for ingestion of foreign body (4 button & 2 AA batteries) and 12 for 'other' reasons (e.g. food bolus, caustic/acid ingestion, less severe bleed). The mean age of the patients was 6.2 years with 28/91 patients <=12 months old at the time of the endoscopy. Of the GI bleeds 17 required endoscopic therapy (6/7 variceal) including haemospray, clips and injection. Bleeds were mostly managed by the medical teams, FB and 'other' by surgeons.

Preliminary calculations suggest an incidence for all procedures of 2.9 (95% CI: 2.4 – 3.6) per 100,000 children <16 annually. This means that approximately 350 of these cases occur across the UK every year. For GI bleeds alone, the incidence was 1 (95% C.I. 0.7 – 1.5) and of these those that required therapeutic intervention was 0.7/100,000 children <16 per year.

Conclusion:

This is the first National prospective study of its kind examining the most urgent and severe endoscopy cases in under 16s. Initial incidence data confirms these are rare events. These data can support planning for national emergency endoscopy services to provide the highest quality in the

best location for patients. Further work will examine the case reports in more detail including initial presentation, co-morbid status, severity of the bleeds, waiting times and outcomes.

9.20 – 9.30

Blended Diet: an adjunct to weaning home parenteral nutrition?

Diana Flynn[1], Christina McGuckin[1], Tracey Cardigan[1], Hazel Duncan[1], Andrew Barclay[2], Elaine Buchanan[1] and Simon Fraser[1].

[1]Royal Hospital for Children, Glasgow; [2]NHS Greater Glasgow and Clyde

Objectives and Study:

Weaning from home parenteral nutrition (HPN) in children can often be challenging due to limited tolerance of enteral feed. This can be further exacerbated by food aversion and limited oral intake. Here we describe our initial experience with blended diet (BD) in weaning children from HPN, where oral intake was limited and enteral milk formula feeds were not able to be advanced due to threshold of tolerance to feed being reached.

Methods:

Retrospective notes review of all patients on HPN since introduction of blended diet at RHC (Feb - Nov 2019). All HPN patients commenced on blended diet were included.

Results:

Records of all 14 HPN patients in a large regional unit were reviewed over the last 10 months. Three patients received blended diet for a period of 5-8months, age range 4 to 9 years, 2 female and one male. All had been on parenteral nutrition since birth. Diagnosis was short bowel syndrome in all three. Prior to commencement of BD all patients required at least 5 nights of HPN per week and had reached their threshold of formula tolerance with enteral feeding via gastrostomy, and all had poor oral intake of solids. BD was commenced slowly and volumes and food variety in blends increased as tolerated under senior dietetic supervision, commencing with 10-20ml boluses via gastrostomy. Duration of BD is 8 months in two patients and 5 months in one patient. BD volumes achieved are 200-600ml/day, and none have reached their threshold of tolerance as yet. BD was well tolerated, and has resulted in improved stool consistency in all patients. In two of the three patients stool frequency improved significantly from around 5 per day to 1-2 per day and in two of three patients night time stooling stopped completely. No allergic reactions were encountered. All have weaned one night per week from their PN. Complications include one episode of gram negative central venous access device (CVAD) sepsis 2 months after starting BD and difficulty in administering BD at school. All three remain on BD to date.

Conclusion:

These preliminary results suggest that blended diet should be considered in children on HPN where threshold of tolerance has been achieved using enteral feeds and/or where there is poor oral intake due to food aversion/limited appetite. This is a cost effective potential alternative treatment to teduglutide, as BD may allow further weaning from HPN when thresholds of tolerance with enteral feed has been reached. BD can also improve stool output/consistency which may improve quality of life. Further studies should be performed to determine effectiveness of BD in weaning from HPN. It is possible that CVAD infection whilst commencing BD may have been due to bacterial translocation, hence caution should be used in rate of introduction of BD in this patient group.

9.30 -9.40

Oesophageal mucosa innervation in paediatric Eosinophilic Oesophagitis.

Kornilia Nikaki[1], Ahsen Ustaoglu[1], Chung Lee[1], Philip Woodland[1] and Daniel Sifrim[1].
[1]Wingate Institute of Neurogastroenterology, Blizard Institute, QMUL

Introduction:

The mechanism for symptom generation in eosinophilic oesophagitis (EoE) is not completely clear. Together with mechanical obstruction due to decreased oesophageal wall distensibility, oesophageal hypersensitivity has been noted. Hypersensitivity can be due to impairment of mucosal integrity and/or sensitization of mucosal nerves. The aim of our study was to delineate the mucosal innervation in the proximal and distal oesophagus in paediatric EoE and control subjects and correlate this with the histological findings.

Methods:

We prospectively recruited children undergoing an upper GI endoscopy for clinical reasons and identified a subgroup with normal histology +/- normal impedance pH-metry that served as controls and a patient group with histologically confirmed EoE. We obtained oesophageal biopsies from 3-5cm above the lower oesophageal sphincter and from the proximal/upper third. The biopsies were immunohistochemically stained for CGRP and TRPV1. CGRP positive nerve fibres were identified and their position relative to the lumen was determined (expressed as median number of cell layers from the lumen for all sections examined). The peak eosinophilic count per high power field (HPF) was used as proxy of disease activity in EoE.

Results:

Nineteen children were included in the control group (12M:7F, median age: 11 years) and 12 in the EoE group (9M:2F, median age: 12 years). In the control group, CGRP positive nerve fibres were identified at a median of 19.5 cell layers from the lumen in the proximal and 19 cell layers in the distal oesophagus (Image 1). In the EoE group, CGRP positive nerve fibres were identified at a median of 14 cell layers in the proximal and 12.6 cell layers in the distal oesophagus ($p=0.0009$ and 0.01 respectively). There is a weak correlation between the number of eosinophils per HPF and the position of the nerve fibres in the oesophageal mucosa ($r=-0.46$). None of the nerve fibres identified expressed the TRPV1 receptor.

Conclusion:

The oesophageal mucosa innervation in children with EoE is similar in the proximal and distal oesophagus with more superficial lying nerve fibres compared to controls. The superficiality of the nerve fibres may be driven by the underlying inflammation. These findings may contribute to the oesophageal hypersensitivity noted in EoE despite histological remission.

9.40 – 9.50

Wilson's disease is associated with good long term outcome but neurological symptoms and signs may be under-recognised in children

Meranthi Fernando[1], Carla Lloyd[1], Indra Vanmourik[1], Mona Abdelhady[1], Deirdre Kelly[1], Jane Hartley[1], Evangeline Wassmer[1] and Girish Gupte[1].

[1]Birmingham Women's and Children's Hospital NHS Trust

Introduction:

Wilson disease (WD) is a recessively inherited disorder of copper metabolism mainly affecting liver and brain. It has a pre-dominant hepatic phenotype with low awareness for WD associated neuropsychiatric signs/symptoms in children

Aim:

To identify the presentations, treatment and outcome of a cohort of paediatric WD.

Subjects and methods:

Retrospective analysis of medical records was carried out from 1987 to 2018 to identify children with a diagnosis of WD, and children with Wilson's diagnostic score of 4 or more as per ESPGHAN position paper 2018 were included for further analysis. King's Wilson index (KWI) was calculated for patients who required Liver transplant (LTx).

Results:

Table 1: Demographics (n=67)

Age (yrs) at diagnosis (median(range))	12 (3 to 17)	
Sex	M=33; F=34	
Ethnicity (E)	Caucasian = 33(49%) Asian = 30(45%) Other = 4(6%)	
Presentations	Symptomatic = 39 (58%) Asymptomatic = 28 (42%) 10 siblings	
<u>Index cases – presentations (n=57)</u> Hepatic	Asymptomatic Acute liver failure Acute on chronic liver disease (CLD) Hepatitis & stable CLD	18(35%) 6(11%) 23(44%) 5(10%)
Neurological Other	3 (4%) 2 (3%)	
Outcome	LTx – 24 (36%) Died without LTx – 1 (1%) Chelators – 42 (63%)	all had KWI ≥8

Demographics are presented in table 1

3/67 presented with neurological features at 9, 12 and 15 years. 2/3 had only minor changes in the liver biopsy and one had evidence of chronic liver disease. In those presenting with liver disease a myriad of neuropsychiatric features was reported including poor scholastic performance (9), behavioural disorders (8), headache (4), movement disorders (7) speech abnormalities (6) and encephalopathy (11).

Neuroimaging was available in 9 with 6 having normal MRI findings and 3 showing typical basal ganglia involvement.

Kayser-Fleisher rings were observed in 23/67 children.

Chelators used in 42 patients, three required LTx within a year, the remaining 39 were alive with a median follow up of 4.5 years (range 1 to 8.5). First line medications were Zinc (n= 22), Penicillamine and Zinc (n=19), Trientine (n=1). Second line therapy with Penicillamine was used in 6, due to lack of control and 3 were started on Trientine due to adverse effects (AE) related to Penicillamine. Gastro intestinal symptoms were noted in 10/37 received Zinc. Neutropenia (4/30), nephrotic range proteinuria (2/30) and perforating collagenosis (1/30) were among AE noted in Penicillamine group. No significant AE were noted in Trientine group.

Summary:
This cohort highlights the heterogenous clinical phenotype of WD and interestingly neurological signs and symptoms were observed on follow-up. Chelation therapy is safe and effective in asymptomatic and children with chronic liver disease, when started early, whilst acute liver failure/acute on chronic liver failure presentation could be rescued with liver transplant. Limitations of our study: single centre retrospective, no information about current outcome after transition to adolescent; brain MRI imaging available in limited number of children, detailed neurological assessment was not possible in all children.

Conclusion:
Children with WD have a good long term outcome and input from paediatric neurologist/psychologist with modern neuroimaging techniques may help in characterisation and early identification of neurological/neuropsychiatric involvement.

9.50 – 10-.00

Gene expression profiling identifies risk factors for progressive liver graft fibrosis

Nicola Ruth[1], Carla Lloyd[1], Deirdre Kelly[1], Stefan Hubscher[2] and Alberto Sanchez-Fueyo[3].
[1]Birmingham Children's Hospital; [2]University of Birmingham; [3] King's College, London

Background:

Long term outcome following paediatric liver transplantation (LTx) continues to improve despite the development of asymptomatic graft hepatitis and fibrosis in >75% of survivors. The underlying mechanisms for this graft dysfunction remains unclear, but it may be a form of chronic rejection. We have analysed gene expression profiles in protocol liver tissue samples and correlated the results with clinical and histological outcomes in transplant recipients.

Aim:

• Identify gene expression in liver graft tissue
• Define the transcript sets associated with histological damage
• Define cross-platform multi-parameter classifiers predicting either stability of progression of histological damage.

Subjects and Methods:

Patients who had undergone protocol liver allograft biopsy at 5 and 10 years between 2011 and 2015 were identified. Patients were assigned to one of 3 groups based on their histology grading: (1) Acute rejection (AR); (2) Minimal fibrosis at 5y with no progression at 10y (Non-progressors); (3) Moderate fibrosis at 5y with progression of fibrosis at 10y (Progressors). Baseline demographics and biochemical analyses were collected. mRNA was obtained from paraffin block samples. For gene expression profiling studies Nanostring platform and the PanCancer Immune Profiling Panel were employed, which contain probes for the multiplex quantification of transcript levels of 800 genes from a single sample (770 pre-defined genes and 30 custom genes). The selected custom genes: ABMR/TCMR/Stellate cell/Fibrosis were identified. Data obtained was correlated with PCR and histological features.

Results:

35 cases were identified. AR (n=15) included patients at first rejection median (range) 1.20m (0.2 – 13). Non-Progressors and Progressors (n=10 each). 51% were male. Biliary atresia was the main indication for LTx (49%). Mean age at LTx was 3.7y (range 0.1-16.7). In groups 2 and 3, mean age at 10y protocol biopsy was 12.3y (range 9.6-13.6) and the mean interval from transplant to biopsy was 10.4y (range 9.5-11.3y). Nanostring analysis revealed the transcriptome of AR was significantly different compared to the Non-Progressors and Progressors. Progressors differed from AR in specific transcriptional pathways associated with rejection e.g. Interferon-gamma (IFNG) and MHC-II antigen presentation while Non-Progressors showed intermediate changes.

Conclusion:

AR is transcriptionally different to sub-clinical damage with fibrosis, but functional pathway analysis revealed that the liver tissue transcriptome of patients with progressive fibrosis was enriched in proinflammatory pathways sharing some similarities with rejection. Based on this, it may be possible to identify patients at risk of fibrotic change and tailor their immunosuppression accordingly.

Posters
Selected for BSPGHAN Best Poster Award

P6

Reducing bed days whilst training for Home Parenteral Nutrition

Linsay Rajfeld[1].
[1]Leeds Teaching Hospitals NHS Trust

Parenteral Nutrition (PN) is the artificial method of providing nutrition intravenously by bypassing the normal digestion in the stomach and bowel (Aspen 2016). Children who have intestinal failure require long term Home PN and therefore parents must be trained in all of the complex procedures required to care for their child at home.

Previously this process has been extremely lengthy, taking months to complete. This was due to lack of structure and the availability of Nutrition Nurses. This has an obvious effect on the Trust's resources and the patients' hospital experience.
In producing a new training programme and involving the ward nurses, we aimed to reduce this time (from start of training to day of discharge) to 4 weeks.

An audit was conducted of the previous 4 parents to be taught Home PN, which concluded that their bed stays from starting the training to discharge varied between 50 and 127 days, costing the trust between £16,250 and £41,275 (£325 per bed day).

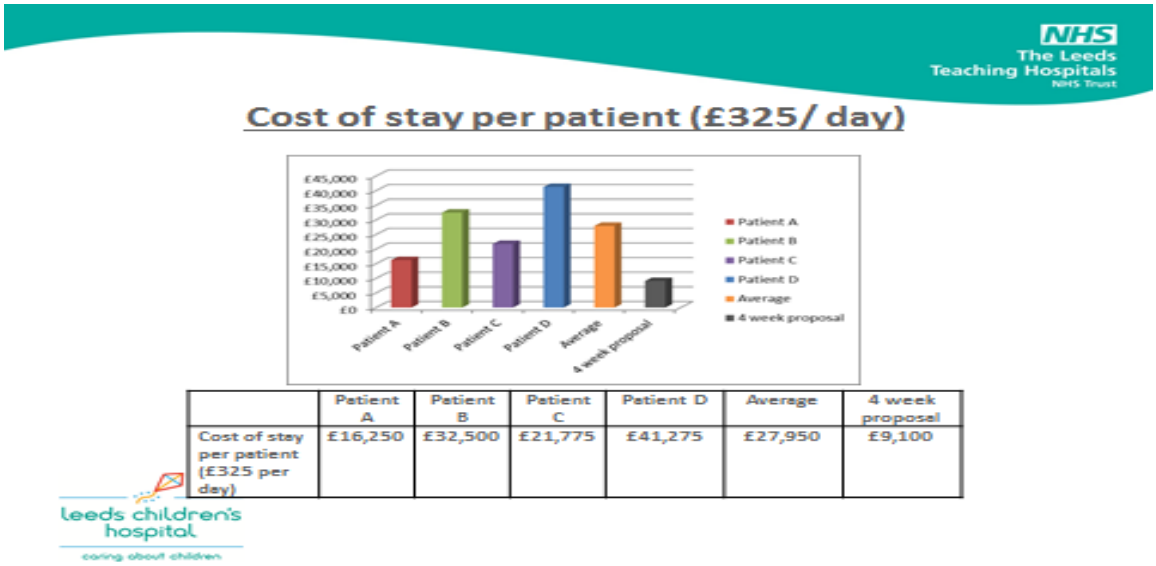
Our practice was benchmarked against other centres, whose training varies between 2 and 6 weeks.

We then devised a 4-week training plan, covering all aspects of care required to keep the child safe at home. We involved the ward staff to continue the training on the weekends (when the Nutrition Nurses are not available) using our step-by-step guides.

We have been using the 4-week training plan for twelve months now, with 3 children successfully discharged home within 4 weeks. The cost of a 28 day stay in hospital whilst training is £9,100, which is evidently a huge saving.

Parental satisfaction has also improved with them having a clear plan and expected date of discharge. Reduced hospital admission improves child development and strengthens family bond.

Ward nurses and Nutrition Nurses have also been working more collaboratively, with the ward nurses feeling more empowered and skilled in their teaching of parents.



	Patient A	Patient B	Patient C	Patient D	Average	4 week proposal
Cost of stay per patient (£325 per day)	£16,250	£32,500	£21,775	£41,275	£27,950	£9,100

leeds children's hospital
caring about children

P10

The microbiota is a key factor influencing gut rehabilitation in intestinal failure due to early onset short bowel syndrome

Jemma Cleminson[1], David Derry[1], Maureen Lawson[1], Carly Martin[1], Monica Chiu[1], Karen Hartley[1] and Julian Thomas[1].

[1]GNCH, Newcastle upon Tyne

Introduction:

Short bowel syndrome (SBS) is the most common cause of paediatric intestinal failure (IF) and its prevalence is rising. Home parenteral nutrition (HPN) is life-saving but can be associated with serious complications such as intestinal failure-associated liver disease (IFALD) and catheter related blood stream infections (CRBSI), therefore the ultimate aim is intestinal autonomy. Current understanding of predictive factors for achieving intestinal autonomy is limited, but include remaining bowel length, type and quality of bowel, and earlier age of onset of SBS. Significant dysbiosis also occurs in SBS and may impact on the likelihood of successful intestinal rehabilitation (IR).

Aim:

To phenotype patients with SBS, comparing children who achieved intestinal autonomy to those who remained on HPN or required intestinal transplant, to identify possible factors that impact upon the likelihood of successful IR.

Subjects and methods:

Retrospective analysis of all North East Paediatric SBS patients who have been cared for by the HPN team during the past 11 years, using anonymised routinely collected data. Comparative statistical analyses were performed, using parametric or non-parametric tests as appropriate, to evaluate the significance of differences in predictive factors between children who achieved intestinal autonomy and those who have not yet done so.

Results:

70 children who had received HPN were identified. 35 had SBS, 35 had non-SBS diagnoses. 33 had neonatal onset, 2 had childhood onset. 2 died (from illnesses not relating to SBS). Of the remaining 31 in the neonatal onset group, 15 have achieved successful IR to date. The underlying causes of SBS were similar between the two groups. Bowel length was significantly longer in the successful IR group (48.3cm vs 24.5cm, $p<0.01$), the number of central lines was significantly higher in the unsuccessful IR group (3 vs 6, $p=0.01$), and the number of line infections post-discharge was significantly higher in the unsuccessful IR group (1 vs 10, $p=0.005$).

Summary and Conclusion:

We confirmed that the initial length of bowel is important in determining outcome amongst children with early onset intestinal failure. We further show that the number of central lines and the number of courses of antibiotics given may be even more important in predicting outcome, which suggests that intestinal microbiota play a key role in intestinal rehabilitation.

P11

Is faecal immunochemical test a suitable alternative to faecal calprotectin in children?

Kirn Sandhu[1], Ruth Ayling[2] and Sandhia Naik[1].
[1]Royal London Hospital; [2]Barts Health NHS Trust

Background:
Faecal calprotectin (FCP) has widely been used as a non-invasive marker for intestinal inflammation in children. Faecal immunochemical test (FIT) is well established in bowel cancer screening programmes in adults. FIT is cost-effective and easier to handle in comparison to FCP.

Aim:
We aimed to evaluate the performance of FIT in the paediatric population and compare it with FCP.

Subject and Methods:
Clinicians in paediatric gastroenterology clinic who requested FCP for further investigation to assess bowel symptoms. These patients provided a sample of FIT from the same stool. These samples were collected over a 10-month period from November 2018 to September 2019. FIT samples were taken into a proprietary tube (Eiken Chemical, Tokyo, Japan) and stored at 4 degrees until analysis. FCP was measured using Liason Calprotectin (Diasorin, Italy). FIT was measured using the OC-sensor (Eiken, Tokyo, Japan).

Results:
131 samples were returned; 131 FIT and 102 FCP of which 7 calprotectin samples were insufficient for analysis. In 95 patients we had paired samples for FIT and FCP. Normal range for FCP was 0-200 µg/g and for FIT was 0-4 µg/g. 23 of 95 patients did not have a diagnosis of inflammatory bowel disease (IBD) (24.2%). In the IBD group; 42 had Crohn's, 27 ulcerative colitis and 3 indeterminate colitis. 15 were new diagnosis of IBD.

In the 95 patients; FIT was normal (<4 µg/g) in 50 patients and abnormal (>4 µg/g) in 45 patients. FCP was normal (<200 µg/g) in 45 patients and abnormal (>200 µg/g) in 50 patients.

FIT positively correlated with calprotectin with a Spearman's rank coefficient 0.653, $p<0.001$. There were 32 patients with FIT >20µg/g and in 29 of these patients FCP was >200µg/g.

In 60 patients with FIT and in 35 patients with FCP underwent colonoscopies. Table 1 shows the diagnostic value of FIT in comparison to histological inflammation.

Table 1: Diagnostic value of FIT in comparison to histological inflammation (n=60).

	FIT ≥ 4 µg/g
Sensitivity	81.3% (95%CI 68.1-89.8)
Specificity	91.7% (95%CI 64.6-98.5)
Positive predictive value	97.5% (95% CI 85.6-99.6)
Negative predictive value	55% (95%CI 39.8-69.3)

There were 25 FIT samples in which the histology showed normal and mild inflammation. All patients with normal histology had a FIT of <4 µg/g. There were 35 FIT samples in which the histology depicted moderate and severe inflammation. In 91.4% of patients with moderate to severe inflammation on histology had a FIT of >5 µg/g.

Summary and Conclusion:
Our study is the first to compare FIT and FCP in the paediatric population. Our results suggest that FIT correlates well with FCP and can be used to differentiate between functional bowel disease and inflammation in children. A FIT of > 20 µg/g was consistent with the finding of severe inflammation.

P14

Variation in coeliac disease management in specialist paediatric gastroenterology centres across the United Kingdom - a 2019 service snapshot

Siba Paul[1], Varathagini Balakumar[2] and Peter Gillett[3].
[1]Torbay Hospital, Torquay; [2]Cardiff University; [3]Royal Hospital for Sick Children, Edinburgh

Background:

ESPGHAN 2012 guidelines on coeliac disease (CD) recommended a no-biopsy pathway [NBP] for symptomatic children with IgA anti-tissue transglutaminase titres (IgA-TGA) >10X the upper limit of normal (ULN) and with positive IgA anti-endomysial antibody (IgA-EMA) and HLA-DQ2/8 positivity. A survey in Southwest England highlighted the need for greater awareness of the NBP and the utility of HLA-DQ2/8 amongst paediatricians (1).

Aims:

1) To assess the implementation of 2012 ESPGHAN guidelines and
2) To understand variation in practice in CD management across the UK in specialist GI centres

Methods:

We sent all specialist gastroenterology centres (n=29) providing endoscopy services for CD diagnosis across the UK an email survey in August 2019. This included 20 questions under 4 main headings:

- Pre-diagnosis
- Post-diagnosis
- Annual review
- Transition

Results:

All 29 (100%) centres responded. NBP was fully implemented in 28/29 centres, 22/28 centres started in the 2 years post publication of the ESPGHAN guidelines. 5/29 centres had stopped HLA-DQ2/8 testing for NBP diagnosis. 2/29 centres had started offering NBP diagnosis in asymptomatic children from high-risk groups. IgG-TGA or IgG-EMA were mainly used in IgA-deficient children as initial screening. Diagnosis of CD was initially discussed by a doctor in 65% centres [n=19]. Most centres [n=21] waited at least 6-months post-diagnosis to start gluten-free oats. DEXA is offered as routine in 4/29 centres. Routine vitamin-D supplementation was suggested by 25% centres, and all centres would repeat IgA-TGA to assess normalisation. Annual follow-up was with a combination of doctors/dietitians [n=26]. Transition was mainly to primary care between 16-18 years, with 3/29 exclusively transferring to adult gastroenterology services.

Summary and Conclusions:

Our survey highlighted excellent uptake of NBP. HLA-DQ2/8 remained an important test in 83% centres and there remain other considerable differences in ongoing management. Most do not use the NBP for asymptomatic children. The new ESPGHAN guidelines (2) will help confirm and harmonise recommendations from previous iterations, allow a secure NBP diagnosis for asymptomatic children and remove HLA-DQ2/8 testing from routine diagnosis.

Reference:
(1) Paul SP, Adams HL, Basude D; Collaborators. Indian J Gastroenterol. 2019;38(3):203-210.
(2) Husby S, Koletzko S, Korponay-Szabó I, et al. J Pediatr Gastroenterol Nutr. 2019 Oct 17. doi: 10.1097/MPG.0000000000002497. [Epub ahead of print]

P19

The use of extensively hydrolysed and amino acid formulas outside of cow's milk allergy – when, what and why?

Rosan Meyer[1], Chris Smith[2], Laura Sealy[3], Sara Mancell[4] and Luise Marino[5].
[1]Imperial College, London; [2]Royal Alexandra Childrens Hospital, Brighton; [3]Bristol Royal Hospital for Children; [4]King's College Hospital NHS Foundation Trust; [5]University Hospital Southampton NHS Foundation Trust

Background:

Where breastmilk is not available, extensively hydrolysed formulas (EHF) and amino acid formulas (AAF) are used for infants/children with a proven cow's milk allergy (CMA). However, these feeds are often used in other medical conditions where tolerance and absorption are affected. This practice survey assessed the use of these feeds in paediatric conditions outside of CMA; aiming to describe the population, growth parameters and micronutrient status.

Methodology:

Four National Health Service tertiary paediatric centres participated in this practice survey. Inclusion: children between 0-18 years, consuming >25% of their estimated energy requirements of an EHF/AAF for any condition other than allergic disease. Anonymised data was collected: (i) descriptive information (ii) indications (iii) type and route of feeding (iv) growth status and nutritional deficiencies (v) medication and vitamin and mineral supplementation.

Results:

One hundred-and-ninety-one children were included with a median age of 19 months [IQR: 4 to 63]. Seventeen percent (33/191) were on AAF and 83% (158/191) on EHF. The feeds were commonly used in critical illness (31%), cancer (26%) and GI disease (18%). When assessing the indications for using an EHF or AAF, 32% responded that this was standard practice in their unit and 29% used these feeds when children were deemed not to tolerate standard whole protein paediatric feeds. Indications for use included: vomiting 12%, congenital/acquired gastrointestinal pathology 7%, malabsorption 5%, diarrhoea 3%, reflux 1% and constipation 1%. In addition, 10% marked "other" reasons for using an EHF or AAF. The diagnostic categories and indications for use of AAF and EHF were combined into a heat map (figure 1). The majority (73%) of children had enteral feeds via a nasogastric tube. Nutritional blood markers were performed in 29% of children and 83% were on a vitamin or mineral supplement.

Figure 1: Heatmap of primary indications for use of formula.

Diagnostic Category	Grand Total	Standard clinical Practice	Not tolerating Standard feeds	Vomiting	Other	Congenital/Acquired GI pathology	Malabsorption	Diarrhoea	Constipation	Reflux
PICU ALL	60	85%	3%	0%	10%	2%	0%	0%	0%	0%
PICU / Other	45	80%	4%	0%	13%	2%	0%	0%	0%	0%
PICU / Cardiac	11	100%	0%	0%	0%	0%	0%	0%	0%	0%
PICU / Prematurity	4	100%	0%	0%	0%	0%	0%	0%	0%	0%
Oncology	49	12%	57%	29%	2%	0%	0%	0%	0%	0%
GI disease	34	3%	47%	3%	6%	24%	6%	9%	3%	0%
Liver	18	11%	0%	0%	50%	6%	33%	0%	0%	0%
Other	12	8%	25%	42%	0%	0%	8%	0%	8%	8%
Prematurity	10	0%	10%	0%	10%	40%	10%	20%	0%	10%
Cardiac disease	8	0%	63%	25%	0%	0%	0%	13%	0%	0%

Conclusions:

This practice survey found that EHFs and AAFs are used in a variety of medical conditions. Indications for feed choice varied with disease but GI symptoms were common. Evidence-based research supporting the use of these feeds outside cow's milk allergy is scarce. Whilst awaiting further research, patients on these types of feeds should have regular nutritional monitoring.

P24

2 Years Outcomes of Percutaneous Endoscopic Gastrostomy insertion in a Large Tertiary Centre.

Jeng Haw Cheng[1], Hanin Tawfik[1], Elizabeth Renji[1], Marcus Auth[1], Manjula Nair[1], Brenda Hill[1], Claire Mckie[1], Raj Parmar[1], Fiona Cameron[1] and Sarang Tamhne[1].
[1]Alder Hey Children's Hospital

Introduction:
Gastrostomy tube feeding plays a vital role in patient who required long term support in enteral nutrition. It serves to deliver nutrition (feeds), fluid and medication straight into gastric lumen and helps overcome limitations due to problems like unsafe swallow, feed/food aversion, severe GOR and also augments calorie supplementation to improve growth faltering. In our centre, percutaneous endoscopic gastrostomy (PEG) insertions are performed by both paediatric gastroenterologist and paediatric surgeon. We take pride for the fact that we are the only tertiary paediatric gastroenterology centre in the country that use a single operator technique in PEG insertion. 3 safety checks are performed to identify a safe insertion site such as endoscopic trans-illumination, finger indentation and rule out any bowel entrapment through a local anaesthetic needle insertion and aspiration technique. Once safe site is identified, the same operator who performs the endoscopy will perform the puncture through the abdominal wall with trocar, insert guidewire and pull through the PEG tube. Post procedure, patients will be reviewed by our MDT team and observed overnight in hospital.

Aim:
To assess the outcome and complication rate of PEG insertions with single operator pull through technique in our centre.

Method:
Detailed review of MDT assessment letters prior to PEG insertion, operative and post procedure notes from the electronic medical records of patients who underwent PEG insertion by paediatric gastroenterologists from January 2017 till December 2018.

Results:
A total of 110 patients had PEG insertion during this period. 2 patients were excluded due to lack of clinical details. Out of 108 patients, 55 are females and 53 males. The median age at insertion is 14 months (23 days to 16 years) and the median weight is 9.4kg (2.44kg - 55.4kg).107 patients had a 12 Fr Corflo PEG inserted and 1 patient had 15 Fr Freka device (to facilitate a jejunal extension). The common co-morbidities for this cohort are cardiac, genetics, neurology, respiratory and a few with renal. Indications for PEG insertions were unsafe swallow (24%); vomiting (22.2%); failure to thrive (15.7%); oral aversion (14.8%); feeding difficulties (12.9%);delivery of medication and fluid (9.2%) and others (0.9%). 98% of patients had both symptom and growth improvement with their PEG.
15 (14%) patients required a jejunal extension due to intolerance of gastric feeds.
2 out of these 15 patients required jejunal extension within 2 months.

Only 1 out of the 108 patients had an immediate complication of PEG site infection. (managed successfully with systemic antibiotics). 8 out of 108(7.4%) patients developed late onset complication such as episodes of PEG site infection, leakage, prolapsed mucosa and wound healing problem.

Summary:
98% of patients had a positive outcome from their PEG insertion. Only 0.9% of patients developed early onset complication and 7.4% developed late onset complications. Based on the fact that in our centre, PEG insertion is performed by a single operator technique and patients have a short inpatient stay, these results have shown that not only our approaches in PEG insertion are safe but also cost effective.

P25

To evaluate the acceptability, gastrointestinal tolerance and compliance of a low calorie peptide based paediatric tube feed formula

Clare Thornton-Wood[1], Sharan Saduera[1], Michelle Burke[2], Liesl Silbernagl[3] and Hayley Kuter[4].
[1]Nestle Health Science; [2]Brighton & Sussex University Hospitals NHS Trust;[3]Lewisham and Greenwich NHS Trust; [4]Manchester University NHS Foundation Trust

Rationale:

The prevalence of cerebral palsy (CP) children who require a low calorie feed is between 8-15% (1,2). The ESPGHAN working group recommends using a low-fat, low-calorie, high fibre, micronutrient replete formula for maintenance of enteral tube feeding after nutritional rehabilitation in immobile Neurological Impaired children (1). To address the nutritional needs of cerebral palsy children who have low energy needs, a nutritionally complete, low calorie tube feed formula has been developed.

Method:

Participants were recruited from the National Health Service, mainly community settings. Children were all exclusively tube fed (n = 9) under the care of a dietitian or a multidisciplinary team. Informed consent was obtained prior to the start of the study. Participants were given the new low-calorie formula with fibre (Peptamen Junior 0.6 kcal/ml, Nestlé Health Science)for 7 days. Formula intake and gastro-intestinal tolerance was recorded by parents or caregivers throughout the study period.

Results:

All participants (aged 1-11 years) had a neurological issue; majority had CP with a gross motor function IV, one child had Aicardi –Goutieres Syndrome, another had hypoxic ischaemic encephalopathy.

Children with CP have a complex medical history and recruitment can be challenging. The recruitment numbers for this study are similar to other clinical trials including children with severe developmental delay or CP (3,4).

Nine children were recruited, 7 completed the 7-day trial. One child did not complete the study; they withdrew from the study as they noticed a slight increase in flatulence, this child was previously on a non-fibre containing feed. The other child suspended the feed on day 3 due to user error. The average daily formula intake was 1032mls (600-1300ml) for those completing the trial. All those who completed the trial tolerated 100% of the low calorie formula.

One child saw increase in stool frequency (usually type 6) from 2 to 4 times per day and a slight increase in flatulence and bloating; it was noted this child was also previously on a non-fibre containing feed and suggest a slow introduction of fibre containing feed in children who are sensitive to fibre could be beneficial.

Conclusion:

The new low-calorie formula was well tolerated by the majority of participants. This formula could be useful to ensure full nutritional goals are met for those children with low energy needs.

References:
1. Romano C, et al.(2017) European Society for Paediatric Gastroenterology, Hepatology and Nutrition Guidelines for the Evaluation and Treatment of Gastrointestinal and Nutritional Complications in Children With Neurological Impairment. J Pediatr Gastroenterol Nutr. Aug;65(2):242-6.
2. Marchand et al.(2006). Nutrition Support for Neurologically Impaired Children: A Clinical Report of the North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition. Journal of Pediatric Gastroenterology and Nutrition (43):123Y135.
3. Fried MD et al.(1992).Decrease in gastric emptying time and episodes of regurgitation in children with spastic quadriplegia fed a whey-based formula. The Journal of paediatrics, 12(4):569-72.
4. Khoshoo et al.(1996). Incidence of gastroesophageal reflux with whey- and casein-based formulas in infants and in children with severe neurological impairment. J Pediatr Gastroenterol Nutr. 22(1):48-55.

P26

To Evaluate the Acceptability (Including Gastrointestinal Tolerance and Compliance) of a Paediatric Enteral Formula with Ingredients Derived from Real Food for Children Over 12 Months of Age.

Clare Thornton-Wood[1], Sharan Saduera[2], Jackie Falconer[3], Rita Shergill-Bonner[4], Haidee Norton[5], Marie Watson[6] and Jayne Lewis[7].

[1]Freelance Dietitian; [2]Nestle Health Science; [3]Chelsea and Westminster Hospital NHS Foundation Trust; [4]Evelina Children's Hospital, London; [5]Birmingham Women's and Children's NHS Foundation Trust; [6]Sheffield Children's Hospital, Sheffield; [7]Oxford Health NHS Foundation Trust, Oxford, United Kingdom

Rationale:

Blenderised foods being given to children requiring tube feeding is increasing. The previous position statement did not recommend the use of blended diets as it may increase the risk of tube occlusion and nutritional inadequacies (1,2,3). The current position statement has been recently updated and allows the option of blended diets if appropriate (4).

Methods:

A tube feed formula has been developed to address safety and nutritional inadequacies. Study design followed the UK Advisory Committee on Borderline Substances (ACBS) criteria to support submission for prescription usage in the National Health Service (NHS). All participants (n=19) were tube fed, recruited from NHS settings and under the care of a dietitian/doctor. All participants were given the new formula for 7 days, Isosource Junior Mix, Nestlé Health Science. Demographic, medical data was obtained and gastrointestinal (GI) tolerance recorded. Stool type was measured using the Bristol Stool Chart.

Results:

Participants (1-14 years) had a range of medical conditions; global developmental delay, epilepsy, cerebral palsy and Down's syndrome; 16/19 completed the 7-day trial and average daily formula intake was 730mls (480-1400ml) for those completing. A number of participants reported positive changes in stool consistency; becoming firmer and decreasing in frequency. One child saw improved mood, eye contact and concentration. Resolution of reflux and gradual decrease in retching were observed in 2 participants. One child experienced bloating and flatulence; they were previously on a tube feed without fibre which may have caused symptoms. There were no weight changes during the study.

Conclusion:

The new tube feed was well tolerated by majority of participants, with a decrease in GI symptoms and beneficial changes in stool type.

References:

1.BDA Practice tool kit. Liquidised food via Gastrostomy tube. May 2015.
2.Sullivan MM et al. Nutritional analysis of blenderized enteral diets in the Philippines. Asia Pac J Clin Nutr. 2004; 13(4):385-912.
3.Hurt RT et al. Blenderized tube feeding use in adult home enteral nutrition patients: a cross-sectional study. Nutr Clin Pract.2015;30(6):824-829
4. BDA Policy Statement. The Use of Blended Diet with Enteral Feeding Tubes. Nov 2019

P27

Huge Hiatal Hernia: A case report.

Hermione Race[1], R Gowda[1], B Bhaduri[1] and A Desai[2].
[1]Maidstone Tunbridge Wells NHS Trust; [2]Kings College Hospital London

Introduction/Background:

Congenital para-oesophageal hiatal hernia (CPOH) is rare in children. We present an interesting case of a large type IV hiatus hernia with organo-axial volvulus and its management.

Aims and objectives:

A two-year-old girl presented with persistent, effortless, non-bilious vomiting since weaning. She had no dysphagia or weight loss and no neurological symptoms (MRI brain normal). Routine bloods, a coeliac screen and immunoglobulins were normal and urine microscopy and culture was negative. Trials of milk, wheat and gluten free diets did not improve symptoms. Neither did treatment for reflux with omeprazole.

Initial investigation with upper gastrointestinal endoscopy showed a normal appearance of the oesophagus and stomach. However, the operator was unable to pass the scope beyond the first part of the duodenum. Subsequent imaging with a barium contrast study showed a huge central and right-sided type IV hiatus hernia containing the entire stomach. The stomach was abnormally dilated with the fundus rotated posteriorly suggesting a chronic organo-axial gastric volvulus. The position of the gastroduodenal junction appeared as a distinct transition point at the expected level of the oesophageal hiatus. Only small quantities of contrast passed through into the duodenum into normally positioned left-sided jejunal loops. The patient underwent successful laparoscopic reduction of the hiatus hernia and gastropexy. Post-operatively, patient developed gastroparesis leading to lacto-bezoar. She needed OGD to dissolve the same. She was treated with erythromycin which resolved the gastroparesis.

Whilst CPOH is rare it should be considered within the differential diagnosis of patients presenting with persistent vomiting.

Summary:

We present an interesting case of huge congenital diaphragmatic hiatal hernia. Barium contrast study is the best investigation for diagnosing CPOH and should be included in the work up of a chronically vomiting child as first line of investigation.

P28

Enteral feeding tube changes causing procedural anxiety in children: a case review

Lindsey Knight[1] and Jenny Goldthorpe[1].
[1]The Leeds Teaching Hospitals

Enteral feeding tubes are used in children for short and long term use, for various reasons including unsafe swallow.

Children experience difficult enteral tube changes, this can increase anxieties of the enteral feeding tube been replaced routinely or in an emergency. We reported a number of cases in children, in which this has happened. Following a sample of case reviews, the current practice was reassessed and a new pathway was developed to provide better patient experience.

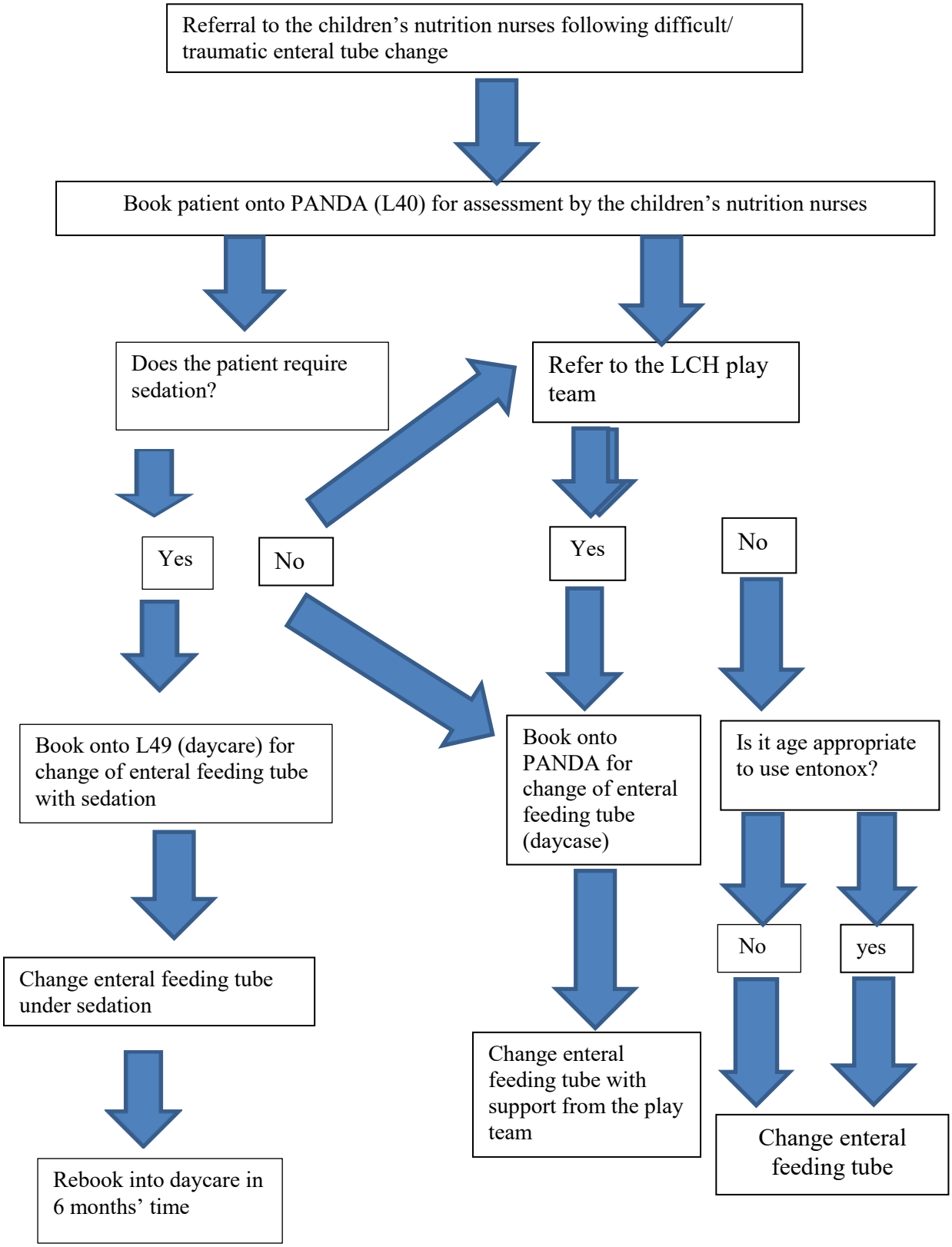
Case 1: Child with gastrostomy balloon button in situ for oral aversion and hyperglycaemia. Referral from Children's Community Nurse (CCN) team reluctant to perform further changes, due to increasing anxieties. On assessment, child presented as guarded and afraid. Decision to perform the 1st planned gastrostomy change under sedation (midazolam), in hospital, unsuccessful and resulted in the child being held to change gastrostomy. 2nd gastrostomy change: Child had pain relief prior to the procedure, despite dad assisting with the procedure, led to unsuccessful gastrostomy change. On the 3rd attempt, one nurse worked alongside Dad on how to change the gastrostomy on a manikin. Simultaneously, the child was engaged in distraction on a doll. Successful gastrostomy change performed by Dad, with the child becoming less anxious, in comparison to the 2 other gastrostomy changes. Child has since been handed back to the local team, in supporting family to change gastrostomy independently.

Case 2: Child with complex medical needs. On long term Parenteral Nutrition (PN). Nasogastric feeding tube (NGT) in situ for drainage. Previous NGT changes under general anaesthetic (GA). Acute displacement of the NGT, led to the child been in pain and discomfort. NGT inserted with Entonox, with play input, uneventful. Referral made to the play team for distraction and next NGT change performed under sedation. Remains under the play team for ongoing input and requires routine NGT changes under sedation

Case 3: Child with gastrostomy balloon button in situ, post nissens fundoplication. Presented to Accident and Emergency (A&E), due to device being pulled out, balloon intact. Gastrostomy reinserted, after slowly dilating the tract. Previous traumatic button change. Referral made to the play team, by the children's nutrition nurses and for next planned gastrostomy change with sedation, on the ward.

Enteral feeding tubes can be difficult to replace especially when the child has heightened anxieties and fears. We must take into consideration the individual needs and previous traumatic enteral tube changes, making appropriate adjustments. We recommend that the enteral feeding tube is changed routinely alongside the play team, with the use of sedation. Regular assessments of the child must be made in order to make reasonable adjustments of the most appropriate intervention to provide a positive experience for the child. This includes a weaning sedation programme at each of the routine tube changes and by including the play team early on the pathway will relieve anxiety and be able to discharge back to the local team earlier on which is better for the child and family.

Pathway for difficult enteral feeding tube changes



P29

Improving the Nutrition of Long-Stay Paediatric Surgical Inpatients: A Quality Improvement Project

Harry Donkin Everton[1], Gabrielle Prager[2], Natalie Howard[2], Mohamed Mutalib[2] and Manasvi Upadhyaya[2].

[1]Kings College London, GKT School of Medical Education, London, UK; [2]Evelina London Children's Hospital, London, UK

Introduction:

Paediatric surgical inpatients are at increased risk of failure to thrive (1). Measurement of patient weights and centiles aids in monitoring the nutritional status, identifying those at risk of failing to thrive, and aiding in recovery post-surgery. Additionally, long-stay paediatric surgical patients are often complex and undernourished.

An audit assessing documentation of weight and follow-up of long-stay surgical paediatric inpatients was carried out in 2017; which showed room for improvement. Work was undertaken to highlight the importance of documentation in aiding the identification of at-risk patients, including presenting previous findings to the team and facilitating involvement of the dietician team in weekly surgical MDTs. An e-noting tool, NEST (Nutritional Evaluation Screening Tool) was incidentally introduced in April 2018; aiming to improve the nutritional assessment of patients. A re-audit of current practice was then undertaken.

Aim:

To evaluate weight assessment documentation and identification of at-risk long-stay paediatric surgical inpatients, aiming to improve their nutritional management.

Methods:

A retrospective audit of paediatric surgical inpatients was carried out. Inclusion criteria included inpatient stay greater than two weeks, between April and December 2018. 14 eligible patients were identified. Their electronic notes were accessed for data including: age, sex, weight and centile on admission, discharge, and throughout inpatient stay, and referral to dietitians or gastroenterology. This was compared to data collected between May 2017 and January 2018, where the same data points and inclusion criteria were used.

Results:

Following the interventions 43% of patients had their weights recorded twice weekly; improved compared to 14% previously. 93% (n=13) of patients had a discharge weight documented in comparison to 57% previously (n=12). Only 29% (n=4) of patients had centiles calculated on admission, and 14% (n=2) had centiles calculated on discharge. There was a decrease in patients referred for gastroenterology follow up from 38.1% to 28.6% (p=0.721) and dietician follow up from 90.5% to 71.4% (p=0191).

Table 1: Weight documentation pre and post-intervention

	May 2017 to January 2018	April 2018 to December 2018	P value
Number of patients	21	14	
Admission weight	95% (n=20)	79% (n=11)	p=0.279
Twice weekly weights	14% (n=3)	43% (n=6)	p=0.112
Discharge weight	57% (n=12)	93% (n=13)	p=0.028

Summary and Conclusion:

All long-stay paediatric surgical inpatients should have regular weight assessment and follow up. Unfortunately, weight documentation remains inadequate. While there was an overall improvement

in documentation following interventions; there was a decrease in those referred onwards. Four patients had significant weight loss on discharge, however only two had dietician follow-up.

Centiles were not regularly calculated; these are useful in assessing trends. E-noting has functionality for these to be calculated and graphed, we will work to promote this. The re-audit was likely too soon after NEST implementation; it however may have helped raise awareness of the importance of patient nutrition.
Further work is required to improve the nutritional assessment of paediatric surgical inpatients, which will require long-term efforts from multiple angles with multi-disciplinary teamwork. Work to improve e-noting functionality and to highlight at-risk patients at weekly MDTs is being undertaken. A re-audit is planned.

Reference: (1) Cole SZ, Lanham JS (2011). Failure to thrive: an update. American Family Physician. Apr 1;83(7): 829-834.

P30

High Vitamin B12 in Paediatric long-term parenteral nutrition patients

Anne Willmott[1] and Natasha Patel[1].
[1]UHL

Introduction/Background:

Vitamin B12 is a water-soluble vitamin, essential for normal erythropoiesis and neurological function. Approximately half of enteral B12 is absorbed via the ileum in those with normal GI tract, linked with Intrinsic factor. There is an enterohepatic circulation of B12 and body stores (2-4mg) are in the liver.

Current Parenteral Nutrition recommended (generous) daily allowance is 0.3mcg/kg/day in infants and 1mcg/ day in children[1], which exceeds the daily requirement and will lead to rapid build-up of stores[1].

Aims

1) We assessed all B12 results over several years, from a cohort of stable paediatric home parenteral nutrition patients.
2) Water soluble vitamins are given in "Solivito" added to PN. We reviewed our recommended Solivito dosage, and that of other centres. This was compared to the recommended daily dose of vitamin B12 and other vitamins (ESPGHAN)[1].

Subjects and methods:

1) We looked at 88 Vitamin B12 measurements on 9 stable home PN pts over up to 7 years. Their diagnoses include bowel aganglionosis / Hirschprungs, gastroschisis, multiple atresias and dysmotility
2) We then compared the doses of vitamins in Solivito to recommended daily doses

Results:

1) All measurements except one were above normal (one was 697 ie near top of N range) and 27 (30.7%) were >2000ng/L (table 1)

Table 1

Vitamin B12 levels readings (mcg/l)			
697-1000	1001-1499	1500-2000	>2000
4 (4.5%)	29 (32.9%)	29(32.9%)	27 (30.7%)

2) Our recommended dose for Solivito (as for a number of other centres) is 1ml/kg up to max of 10ml. This gives significantly more than the daily recommended Vitamin B12 dose. Our recommended Solivito dose is compared to the recommended dose of its component according to ESPGHAN (table 2)

Table 2

	Daily requirement in TPN[1]	Solivito N (10ml vial)
Vitamin B1	1.2mg	2.5mg
Vitamin B2	1.4mg	3.6mg
Nicotinamide	17mg	40mg
Vitamin B6	1mg	4mg
Pantothenic	5mg	15 mg
Biotin	20 micrograms	60 micrograms
Folic Acid	140 micrograms	400 micrograms
Vitamin B12	1 microgram	5 micrograms
Vitamin C	80mg	100mg

Summary and Conclusion:

1) Whilst no B12 toxicity is reported in the literature, we report extremely high levels over the long term (30.7% >2000)

2) The usual dose of Solivito gives significantly more B12 than is needed, and also more of all other vitamins, giving scope to consider reducing the maximum dose given

3) Whilst current dose of Solivito continues, we would suggest to consider stopping routine measurement of Vitamin B12, since it is always raised, and never results in any alteration of PN. It is not an expensive assay but it does require quite a high volume of blood and we would advocate it is not currently necessary. If recommendations change and Solivito and B12 doses are reduced then this will need further monitoring

4) In adults high B12 can be a first marker of more significant disease such as malignancy and whilst this is not generally an issue in children, any pathological cause of high B12 is masked by the very high amounts given to PN patients.

5) It is very important to monitor B12 once PN is weaned, as deficiency can develop more quickly in those with significant bowel resection than in others such as vegans/vegetarians, and supplementation is quite often needed.

1) ESPGHAN/ESPEN/ESPR/CSPEN guidelines on pediatric parenteral nutrition: Vitamins
Bronsky, J.Braegger, Christian et al.
Clinical Nutrition, Volume 37, Issue 6, 2366 – 2378

P31

Successful Dietary Management of Eosinophilic Oesophagitis in children with Endoscopic Surveillance.

Carin Swart[1], Rajesh Rawlani[1], Bim Bhaduri[1] and Rohit Gowda[1].
[1]Maidstone and Tunbridge Wells Hospital NHS Trust

Introduction and background:

The role of dietary management in Eosinophilic Oesophagitis (EoE) has been extensively studied in children. Empiric eliminations diets in the form of six food exclusion diet (SFED), four food exclusion diet (FFED) and two food exclusion diet (TFED) has been shown to be successful and practical in children. However, patient compliance with continued avoidance of food allergens and sustained individualised dietetic support to ensure nutritional adequacy, is key to successful dietary management. As symptom reporting in paediatrics cannot be relied upon, objective assessment with repeated endoscopic review is required to ascertain the efficacy of dietary treatment.

Aim:

To show that successful clinical and histological remission can be achieved through empirical elimination diet and the causative food allergen/s identified through systematic reintroduction and timely endoscopic surveillance.

Subjects and Methods:

A retrospective audit looking at all newly diagnosed paediatric patients (0-16 years) with EoE in Maidstone Hospital from 2014 – 2019, was performed. For the purpose of the review, patients who received steroids (Budesonide gel) were excluded. Steroids were considered in those patients who either became non-compliant on dietary therapy or had severe disease.

The elimination diet were categorised as SFED (cow's milk, egg, wheat, soy, fish and all nuts), FFED (cow's milk, egg, wheat and soy) or TFED (milk and wheat only). Patients were given dietary education, patient resources and were in regular contact with the dietitian. All patients had clinical and endoscopic review at 12 weeks after the initial food elimination and then 1-2 foods were re-introduced.

Results:

A total of 15 patients were diagnosed to have EoE. Four patients were excluded as they were on steroids. Two patients were lost to follow up. Nine patients with a mean age of 12 years, had exclusive dietary therapy. Of these 6/9 were started on a SFED, 1/9 patient on a FFED and 1/9 patient on a TFED. One patient excluded Milk only due to difficulties with compliance. All 9 patients had full clinical remission and had endoscopy at 12 weeks. Full histological remission (<15 eosinophils /hpf) was achieved in 7/ 9 patients: 1 patient reintroduced all foods prior to the endoscopy and 1 patient didn't show full histological remission. They were excluded from further analysis. Food reintroduction was started 1-2 foods at a time and this was followed by three monthly endoscopies until the causative food was identified. A further two patients moved out of area. Causative foods were identified in the remaining 4/5. The causative foods were cow's milk in 3 patients, cow's milk & wheat in 1 and 1 patient remained on a FFED (parental choice).

Summary and Conclusion:

Our review showed that with good dietetic support high levels of compliance was achieved. Full clinical and histological remission (78%) and identification of causative agent (55%) can be achieved on empirical elimination diet. We recognise that the patients did have multiple endoscopies for surveillance and are currently considering 2 food exclusions (step up approach).

P32

Case Report of a Benign Squamous Papilloma of Oesophagus in an Adolescent

Anne Vijaykumar[1] and Nandhini Kumaraguru[1].
[1]Watford General Hospital

Introduction:

Squamous papilloma of oesophagus is a benign epithelial tumour and it is very rare in paediatric population.

Aim:

To discuss a case of Benign Squamous Papilloma of Oesophagus diagnosed at endoscopy.

Subjects and methods:

Following identification of this interesting and rare case, detailed case notes review of both paper and electronic records was done.

A fifteen-year-old girl was referred to the Paediatric Gastroenterology clinic by her GP for persistent and worsening epigastric pain. Blood tests including full blood count, C-reactive protein, urea, electrolytes, HbA1c, haematinics, liver function tests and coeliac screen were normal. GP had commenced her on Gaviscon and Ranitidine to no effect and also trialled Omeprazole which unfortunately was not tolerated due to nausea. The epigastric pain was affecting her school performance and quality of life, therefore specialist input was sought.

At the Paediatric Gastroenterology clinic, the patient reported no exacerbating or relieving factors to her symptoms like positional variations, association with food or drinks or there were no systemic symptoms of note. On examination, she had epigastric tenderness and was commenced on Lansoprazole to which she had an initial clinical response. Subsequently she became refractory to the treatment and she was booked for an Upper Gastrointestinal Endoscopy.

Results:

During gastroscopy, a small oval white polypoid lesion less than 0.5cm diameter was found in the mid-oesophagus. During biopsy, the whole lesion was excised leaving nearly a normal looking mucosa behind. Histology of this biopsy was reported as squamous papilloma of the oesophagus. Rest of the Upper Gastrointestinal mucosa looked normal.

After three months, her symptoms were fairly under control in the absence of medications during the follow- up.

Summary:

This case reports a fifteen-year-old girl with epigastric pain who has shown an improvement after resection of a small oesophageal squamous papilloma.

Conclusion:

Squamous papilloma of the oesophagus is usually an incidental finding and only found in 0.4% of endoscopies1.It is very rare in the paediatric population and gastro-oesophageal reflux symptoms resistant to medical therapy and also non-progressive intermittent dysphagia have been reported as symptoms with this condition. There is a paucity of literature of squamous papillomas and most reported cases have excellent prognosis, treated by excision only, with very low rates of recurrence2. Although it is generally considered as a benign condition, literature also reports that there could be a malignant association rarely3. Hence complete excision of the lesions is recommended.

References:
1. Yoo SS, Lee WH, Ha J, et al. The prevalence of esophageal disorders in the subjects examined for health screening]. The Korean Journal of Gastroenterology. 2007 Nov;50(5):306-312
2. Szántó I, Szentirmay Z, Banai J, et al. Squamous papilloma of the esophagus. Clinical and pathological observations based on 172 papillomas in 155 patients. Orvosi Hetilap. 2005 Mar;146(12):547-552.
3. d'Huart MC, Chevaux JB, Bressenot AM, et al. Prevalence of esophageal squamous papilloma (ESP) and associated cancer in northeastern France. Endosc Int Open. 2015;3(2):E101–E106. doi:10.1055/s-0034-1390976

P35

Serological tests and duodenal biopsy results in Insulin-Dependant Diabetes Mellitus (IDDM) with suspected Coeliac Disease (CD); validating ESPGHAN's updated guideline

Hina Rizvi[1] and Ronald Bremner[2].
[1]Birmingham Heartlands Hospital; [2]Birmingham Children's Hospital

Background:

Clinical practice guidelines support the diagnosis of CD without duodenal biopsies, when anti-tissue transglutaminase (TTG) levels are persistently ten times the upper limit of normal in symptomatic children[1] . CD in IDDM has prevalence of around 4.5%[2]. Understanding the prevalence of pCD (abnormal serology with normal biopsy) in this group is an important consideration when deciding the suitability of serology-only diagnosis, to avoid unnecessary invasive testing and over-diagnosis. Aims: To determine the prevalence of potential coeliac disease in children with IDDM referred for investigation for endoscopy and its relationship with serology results at the time of duodenal biopsies

Methods:

A retrospective review of endoscopy reports, histology and serology results of children with IDDM attending for upper gastrointestinal endoscopy for all indications at a regional referral centre from January 2014 to December 2018. Diagnosis was determined by the treating clinician. Biopsies (median 5 per case, range 2-9) were reported by paediatric pathologist and findings categorised per the modified Marsh grading system. IgA anti-tissue transglutaminase (TTG) testing was with the Thermo Fisher EliA Celikey. Raised TTG results were categorised: up to ten times the upper limit of normal (1-10xULN); and, more than ten times the upper limit of normal (>10xULN). Anti-endomysial antibody (EMA) tests were tested on the same serum sample. EMA were categorised: strongly positive, weakly positive & negative.

Results:

Review of endoscopy electronic records identified 84 children with IDDM undergoing first diagnostic endoscopy, and five were excluded from analysis because TTG results from samples taken on the same date were not available. The median age was 10 years (range 2 – 16 years); 39 were male. Coeliac disease was confirmed in 43 (54%). All 26 cases with TTG >10xULN had enteropathy (Marsh grade 3a-c), and in 23 cases EMA was positive (not tested in 3). In the 24 cases with TTG 1-10xULN, 15 had CD (Marsh 2 in two) and normal biopsies in 9. Two cases had negative TTG (2.5%) at the time of endoscopy, and both had raised TTG titres on previous samples. EMA results were available for all 24 cases when TTG 1-10xULN, and was strongly positive in 17 (10 had CD), weakly positive in 3 (2 had CD) and negative in 3 (none had CD).

Conclusions:

These data lend further support for biopsy-free diagnosis for children with IDDM and coeliac-associated symptoms, when TTG >10xULN on serial testing, using this assay. No case with TTG>10xULN had pCD. When TTG 1-10xULN, 9 (37.5%) had pCD. Positive EMA did not exclude pCD when TTG 1-10xULN. The likelihood of coeliac disease (CD) in children with insulin-dependent diabetes mellitus (IDDM) is predicted by TTG level >10xULN. When TTG 1-10xULN, EMA result did not predict either CD or pCD, supporting the recommendation for biopsy based on TTG result.

References:
1. ESPGHAN Guidelines for Diagnosing Coeliac Disease 2020
2. Holmes GKTScreening for coeliac disease in type 1 diabetes Archives of Disease in Childhood 2002;87:495-498

P36

Pancreatitis as a complication of a displaced gastrostomy.

Maria Van Veelen[1], Helen Yorke[2], Anna Pigott[2] and Nadir Khan[2].
[1]Keele University; [2]University Hospital of North Midlands

Background:

Since its initial implementation in 1980, percutaneous endoscopic gastrostomy (PEG) insertion has been a popular means of providing nutrition to children in need of enteral feeding. A rare complication of gastrostomy placement, particularly with re-insertion after incidental removal, is acute pancreatitis. Acute pancreatitis due to migration of a temporary device is not found frequently within the paediatric literature. A temporary device commonly involves the use of a Foley catheter to maintain the patency of the gastrocutaneous tract if attempts at placing a new gastrostomy tube have failed. There appears to be very few protocols both in the literature as well as among hospital policies regarding placement of temporary devices. Here we present the case of a four year old child who had a temporary device placed after dislodgment of her PEG tube, which subsequently migrated into her duodenum, leading to the discovery of acute necrotising pancreatitis. The importance of creating a standardised protocol for managing displaced gastrostomies, as well as proper documentation with imaging requests to radiology, are discussed.

Aim:

To highlight the potential complication of pancreatitis when using a Foley catheter to maintain a gastrostomy tract secondary to migration of the catheter into the duodenum.

Method:

We report a case of a child whose balloon gastrostomy device became displaced (the 6th occasion the device had been displaced). Staff were unable to replace the device and instead placed a Foley catheter to maintain the tract. The catheter balloon was inflated. The patient subsequently experienced bilious vomiting and deteriorated with a rigid, tender abdomen. The patient developed respiratory distress and shock requiring transfer to PICU. An amylase was markedly raised at over 4000. Unfortunately there was a delay in recognising the Foley catheter was causing obstruction to both the bowel and the ampulla. A CT abdomen showed the catheter in the duodenum, but as the presence of a gastrostomy was not included in the CT request, the presence of the inflated catheter balloon in the duodenum only became apparent when a paediatric radiology consultant reported the scan 72 hours later.

Results:

Inflated Foley catheters used either as permanent gastrostomy devices, or to temporarily maintain a gastrostomy tract when a device has been displaced can cause obstruction to the duodenum and ampulla resulting in necrotising pancreatitis.

Conclusion:

Guidance should be made available to staff managing displaced gastrostomy devices to ensure that if it is necessary to inflate the balloon of a catheter to prevent displacement, the staff monitor the external length of the catheter. Staff must also be aware of the possibility of displacement should discomfort on feeding or vomiting occur. When requesting imaging all lines and devices should be indicated on the request so radiologists can comment on the inappropriate placement of devices.

P37

Evaluation of Current Growth and Fracture Risk in Paediatric Patients with Intestinal Failure Receiving Long-Term Parenteral Nutrition

Jason Jia Shyan Ong[1], Christina McGuckin[1], Andrew Barclay[1], Tracey Cardigan[1], Hazel Duncan[1], Simon Fraser[1] and Diana M Flynn[1].
[1]NHS Greater Glasgow and Clyde

Patients who have enteropathy or intestinal failure are not only at risk of poor growth but are deficient in micro- and macronutrients which negatively impacts bone metabolism. This association is made worse when these children are placed on long-term home parenteral nutrition (HPN). This study reviews our current cohort of HPN patients.

All patients on HPN on 1st September 2019 were identified using our West of Scotland HPN database. We performed a retrospective case note review examining patients' primary diagnosis, age of starting HPN, weight and height SDS, current calcium/phosphate and vitamin D levels, history of fracture and bone mineral density, where performed.

14 patients were included. 11 were female, 3 were male, receiving HPN for median 67 months (3 to 108 months). There was a diagnosis of short bowel syndrome in 8 patients, 2 patients had enteropathy and 4 patients had motility disorder. Weight SDS was less than –2 in 2 patients and Height SDS was less than –2 in 6 patients. Mean weight SDS score was –0.80 and mean height SDS score was –1.49. A DEXA scan was done in seven patients which showed a mean BMD Total Body SDS score of –1.10 (-2.4 to -0.5) and mean BMD Lumbar Spine SDS score of –0.18 (-1.1 to 0.6). A total of 4 patients were diagnosed with a fracture. Of these, 3 were incidental X-ray or DEXA findings. One child had osteopaenia and one had clinical rickets, thought to be part of her underlying disease – Total Intestinal Aganglionosis. Average calcium and phosphate provision from PN were 0.3 mmol/kg/day and 0.64 mmol/kg/day. Vitamin D levels were checked 6 monthly, and results from most recent bloods showed 5 patients having inadequate levels (30-50 nmol/L) and the rest having sufficient amount (>50 nmol/L). None of the patients were Vitamin D deficient (<30 nmol/L).

Studies have previously shown a correlation between bone health and children with intestinal failure on long term PN. We have demonstrated a similar result to those published with 4 of 14 patients (29%) sustaining non-pathological fractures. Approximately 43% of patients had short stature, which may result from the heterogenous sample and be a consequence of the underlying intestinal disorder as opposed to being on HPN. This study confirms high fracture rate in this complex patient group, as well as short stature. Further optimization of HPN composition and vitamin D and enteral calcium supplementation, may improve bone health and reduce fracture risk. Endocrine assessment is recommended for all patients on HPN.

P40

A qualitative review: The psychosocial and emotional experiences of children and adolescents with coeliac disease

Maya Harvey[1], Dr Sophie Velleman[1], Alison Dinning[1] and Lauren Mcveigh[1].
[1]University Hospitals Bristol NHS Foundation Trust

Background:

Coeliac disease (CD) is diagnosed in around 1.4% of the global population, and prevalence is higher in children than in adults. Currently the only effective treatment for managing CD is strict adherence to a lifelong gluten-free diet (GFD), but compliance to the GFD can be challenging and has been associated with a number of social and practical burdens. Furthermore, a recently published quantitative review suggests that young people with CD have increased symptoms of anxiety and depression and a lower quality of life. Despite these findings, there is limited knowledge about the subjective experiences of young people with CD and the impact of having CD.

Aim:

To identify and synthesise the themes across qualitative studies to develop an integrated understanding of the psychosocial and emotional experiences of children and adolescents with CD.

Methods:

EMBASE, Medline and PsycINFO databases were searched for studies using exclusively qualitative methodology up to September 2019. Thematic synthesis was used to analyse the findings from across studies.

Results:

Seven studies from four countries were included. An overarching theme emerged: A journey of adjustment, and two subordinate themes, each with three subcategories: Adaptive coping (practical strategies, cognitive strategies, identity) and Struggle to cope (shame of being different, "food-handicapped", identity) (see Figure 1). Identity was found to be an influential pattern across experiences of young people with CD and therefore fits under both of the subordinate themes.

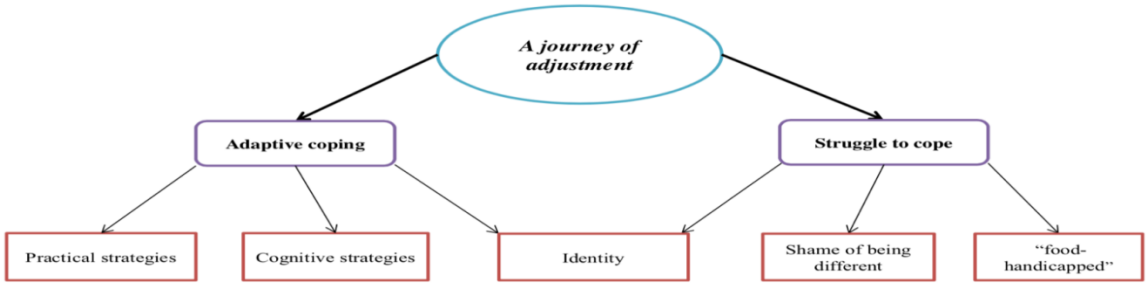


Figure 1: The themes identified through synthesis of findings across qualitative studies, capturing the psychosocial and emotional experiences of children and adolescents with CD.

Summary and conclusions:

Children and adolescents with CD have a diverse range of psychosocial and emotional experiences. Some young people make use of coping strategies and personal identity positively influences adjustment and adherence to the GFD, whereas others struggle to integrate the diagnosis into their identity. Young people with CD also experience shame around being different and feel limited by the need to follow a GFD, particularly in social situations. These findings highlight the need to consider the young person's individual situation and personal outlook when providing support with managing their condition. Additionally, strategies which aim to increase a sense of belonging and reduce feelings of constraint may be useful for this population. Further research is required to understand the experiences of children and adolescents with CD within different cultures and healthcare services, for example, this review found no qualitative studies from the UK.

P42

The Importance of MDT working in a Paediatric Intestinal failure service

Rachel Wood[1] and Andrew Fagbemi[1].
[1]Manchester Foundation Trust

Introduction:

We are a large tertiary children's hospital and gastroenterology service, with 37 Home Parenteral Nutrition (HPN) patients with intestinal failure (IF) and 56 non parenteral nutrition IF patients. Historically the surgical IF patients (see table 1) were managed under the surgeons and the non-surgical IF patients under gastroenterology.

Table 1: Breakdown of intestinal failure patients on Home PN			
Surgical IF		Non-Surgical IF	
Gastroschisis	10	pseudo obstructions	3
Malrotation/volvulus	4	tufting enteropathy	2
necrotising enterocolitis	8	villous atrophy	1
Hirschsprungs	2	congenital and syndromic diarrhoea	2
		cystinosis	1
		sodium losing enteropathy	1
		dysmotility	1

Previously there was no consistency of care; patients were seen by different teams at different times. Many of the non PN IF patients were not seen by professionals specialising in Intestinal failure. There was poor communication between the surgical, gastroenterology and neonates teams.

Aim:

18 months ago we started work on a different method of working, a team approach with the aim to become one of the best IF services in the UK, streamlining care and working together to improve the care of our patients. A patient satisfaction questionnaire was sent to all our HPN patients for feedback on our service.

Methods:

We agreed a weekly MDT meeting to discuss all of our inpatients and outpatients on PN would be a start. This was set up for 8am once a week for 90 minutes and it is a forum for open discussions regarding PN changes/prescriptions, feeding, medications, blood results, social issues and moving forward with each patient. This meeting has expanded and now includes all the consultant paediatric gastroenterologists, specialist nutrition nurses, pharmacist, junior doctors, and specialist dietitian and play specialist.

We reviewed the number of intestinal failure patients on and off PN. ESPGHAN guidelines (2018) on Home PN advise the management of these children should be in a specialist centre with a multidisciplinary team with expertise in PN. Recommendations are reviews are done between 1-3 months post discharge and more for infants with at least 4 visits per year for older children. This was unachievable with 1 clinic per month which has gradually increased to a weekly MDT clinic and a weekly specialist nurse and dietitian clinic.

Results:

We have found that since setting up our MDT service, we have much stronger relationships with our patients and families as they get to know the team in hospital prior to discharge and they trust us with the safe care of their child, with 90% of our satisfaction responses stating the care as excellent

over the last 18 months, with 100% feeling confident in the team and seeing all the professionals at appointments. Recommendations from ESPGHAN highlight that early referral to an expert centre may reduce PN related complications, which is our aim as a team to improve the outcomes for our entire cohort of IF patients.

Table 2: Summary of satisfaction survey of IF service over the last 18 months
93% satisfied with the team
90% satisfied with frequency of review
95% satisfied with child's care
88% confidence in the team
100% satisfied with the current team/service
Comments 1 response not satisfied that all the professionals needed were at the appointments 1 response requested that the PN prescriptions were changed more frequently 1 response stated that we don't always know the answers 1 response states team very quick to respond when unwell

Conclusion:
This is just our experience at RMCH and future research would be to look at all tertiary centres and compare the type of MDT service they provide alongside patient outcomes and patient experiences. All people in the community having parenteral nutrition should be supported by a co-ordinated multidisciplinary team, (NICE 2017).

P45

Taurolock® Effectivity in Reducing Central Venous Catheter-Related Blood Stream Infection in Patients Receiving Home Parenteral Nutrition

Natalia Iglesias[1] and Elena Cernat[1].
[1]Leeds General Infirmary, Great George Street, Leeds Teaching Hospitals

Introduction:

Central venous catheter related blood stream infection (CRBSI) is the most frequent serious complication on long term parenteral nutrition (PN). Evidence suggests that use of Taurolidine-citrate lock solution (Taurolock®) reduces CRBSI compared to heparin lock in this group of patients.

Aim:

To compare incidence of CRBSI in home PN patients when using Taurolock® vs patients using heparin locks over a 2 year period.

Methods:

In our Trust it is policy that central venous catheter (CVC) of patients receiving HPN are locked with heparin 10 IU/mL as first line and patients are changed to Taurolock® line locks when they have suffered one CRBSI.
A retrospective case note review identified the number of CRBSI/1000 catheter days and antibiotics usage pre and post Taurolock® in 19 HPN patients (3 patients previously excluded as PN infused over 24 hours and line locks were not required) over a period of 2 years, from September 2017 to September 2019.

Results:

Of the 19 patients, 5 received heparin initially and 14 received Taurolock® as they already had an episode of CRBSI prior to the period reviewed.
12,905 catheter days (1,203 heparin and 11,702 Taurolock®) were evaluated during that period. CRBSI/1000 catheter days for patients on heparin locks were 3.3, compared to 0.42 for patients on Taurolock®. The total number of antibiotic days for the heparin group was 60, which corresponds to 49.9 antibiotic days/1000 catheter days compared to 108 antibiotic days on the Taurolock® group which equals to 9.2 antibiotic days/1000 catheter days. No adverse effects related to use of Taurolock® were seen.

Conclusions:

In our experience, the use of Taurolock® during cyclical HPN in our cohort of patients with intestinal failure has been associated with a reduction in CRBSI. This has also extended the potential for maintaining long term venous access, reducing hospital admissions for suspected CRBSI and also reducing overall antimicrobial use.

P48

***Single Paediatric Centre Experience of Vitamin D Deficiency and Metabolic bone disease in the Home Parenteral Nutrition Program*.**

Jeng Cheng[1], Kate Simpson[1], Sarang Tamhne[1], Emma Jones[1], Claire McKie[1], Chloe Dempsey[1], Marcus Auth[1], Manjula Nair[1], Raj Parmar[1], Fiona Cameron[1] and Elizabeth Renji[1].
[1]Alder Hey Children's Hospital

Introduction:

Vitamin D is vital for maintenance of bone health and pivotal for calcium and phosphate regulations. Suboptimal 25-OH vitamin D levels have been reported in children on long-term Home Parenteral Nutrition (HPN) formulations constituted with recommended Fat-soluble vitamins. HPN patients are at higher risk due to poor intake, absorption and chronic illness. The ESPGHAN guidelines state that the actual amount of vitamins delivered to the patient may be much lower than the intended dose as vitamins degrade over time in the HPN bag. In addition amino acid, glucose, sodium and calcium infusion from parenteral nutrition (PN) increase the urinary calcium excretion. All these factors contribute to vitamin D deficiency and metabolic bone disease (MBD) in HPN patients.

Aims:

To document the vitamin D status and assess MBD in paediatric HPN patients.

Methods:

Retrospective review of biochemistry, health records and medications between September 2017 till September 2018 for HPN patients in our tertiary paediatric gastroenterology centre. Each patient had 3 measurements of Vitamin D level and parathyroid hormone (PTH).

Results:

From the 20 patients,13 (65%) had short bowel syndrome; 4 (20%) had intestinal dysmotility; 2 (10%) had intestinal pseudo-obstruction; and 1 (5%) has microcytic megacolon. 2 (10%) of HPN patients had vitamin D deficiency (Vitamin D <25nmol/L); 7 (35%) of HPN patients had vitamin D insufficiency (Vitamin D 25 -50nmol/L) and 11(55%) had sufficient vitamin D. Out of 9 HPN patients with Vitamin D deficiency or insufficiency, 5 (56%) had short bowel syndrome; 2 (22%) had intestinal dysmotility and 2 (22%) had intestinal pseudo-obstruction. 1 out of 9 required less than 50% of PN; 4 of 9 patients required more than 50% of PN but not 100%; and 5 of 9 patients required full 100% of PN.

20 patients should have had a total of 60 measurements of both Vitamin D and PTH but only 51 measurements were performed. 25 measurements showed high PTH suggestive of secondary hyperparathyroidism. 15 had normal Vitamin D and 10 low Vitamin D level. 3 (15%) patients had fractures with MBD seen on DEXA scan. 16 (80%) of patients were on Vitamin D supplement.

Conclusion:

Prevalence of vitamin D insufficiency or deficiency in paediatric HPN patients is 45%, similar to other studies. The percentage of fractures is similar to other published studies and was seen in children on HPN for longer duration. High PTH levels are seen despite normal vitamin D. This indicates vitamin D deficiency can be identified biochemically with high PTH level prior to vitamin D level dropping (Fischer's exact test 0.026, $p<0.05$). Although short bowel patients made up 56% of the Vitamin D insufficient/deficient patients, this was not significant (Fischer's exact test 0.6424, $p>0.05$). Regular surveillance of vitamin D and PTH and reactive replacement of Vitamin D are not sufficient in this complex cohort. Therefore, our suggestion is implementation of standard Vitamin D supplement for all home PN patients regardless of PN volume as prophylaxis for Vitamin D deficiency rather than only treating when there is abnormal Vitamin D or PTH

P50

Comparing the quality of life between children and young people with functional constipation, inflammatory bowel disease and functional abdominal pain – not otherwise specified.

Michael Cornish[1], Georgina Knott[2], Jochen Kammermeier[1], Rakesh Vora[2] and Mohamed Mutalib[2].
[1]Guy's & St Thomas' NHS Foundation Trust; [2]Evelina London Children's Hospital

Introduction/background:

Gastrointestinal symptoms, including pain, discomfort, toileting difficulties, can lead to significant stress for children and young people. They have to deal with feelings of embarrassment and shame; and as a result of their symptoms are more likely to have difficulties with concentration, poorer attendance at school and can feel unable to participate in social and recreational activities.

Aim:

To investigate the quality of life of patients treated for either functional constipation (FC), inflammatory bowel disease (IBD) or functional abdominal pain - not otherwise specified (FAP-NOS).

Subject & Methods:

Patients who were referred to the paediatric gastroenterology psychology service were asked to complete the Pediatric Quality of Life Inventory (PedsQLTM) generic module at their first appointment, or early on in treatment, as part of routine care. Their parents were asked to complete a parent/carer proxy report.

Between April 2017 and November 2019, 45 eligible patients aged 5-17 years, and 46 of their parents completed the PedsQLTM. This group was made up of 9 patients (20%) and 12 parents (26%) with FC, 12 patients (27%) and 10 parents (22%) with IBD and 24 patients (53%) and 24 parents (52%) with FAP-NOS.

Results:

Young people with FAP-NOS had a lower self-reported scale score (mean = 54.70, SD=18.05) than either the FC group (mean=60.51, SD=17.14) or the IBD group (mean=65.76, SD=11.72), indicating that they perceive themselves as having a worse quality of life. This finding was also found in the parent/carer proxy report although the scores were less variable (FAP-NOS mean=52.28, SD=19.78; FC mean=52.90, SD=14.53 and IBD mean=54.01, SD=16.01).

These results were compared to a healthy UK sample (Upton et al, 2005). The patients and parents proxies in this sample, across all three diagnostic groups scored lower than the healthy sample, (mean=83.89, SD=11.84) indicating they have worse quality of life.

Summary and conclusion:

Young people with gastrointestinal disorders have lower quality of life than a healthy sample. Across the gastroenterology cohorts in this study, individual's with FAP-NOS has lower quality of life than individuals with constipation or IBD. These results indicate the need for psychological support in the paediatric gastroenterology population.

Reference: Upton, P., Eiser, C., Cheung, I., Hutchings, H. A., Jenney, M., Maddocks, A., Russell, T. & William, J. G. (2005). Measurement properties of the UK-English version of the Pediatric Quality of Life InventoryTM 4.0 (PedsQLTM) generic core scales. Health and Quality of Life Outcomes, 3(22).

P54

Neurodisabled children have a higher incidence of abnormal findings at endoscopy when scoped for upper gastrointestinal symptoms than non neurodisabled children

Huda Atta[1], Fahad Siddiqui[2] and Lisa Whyte[1].
[1]Birmingham Women's and Children's NHS Foundation Trust;[2]West Midlands Deanery

Introduction and background:

It is wildly believed that some of the gastrointestinal disorders are more frequent in children with neurological disabilities, which interfere with both quality of life and nutritional status (1).

Gastro oesophageal reflux disease (GORD) is one of the most frequently (up to 70%–75%) reported disorders (2,3) and is multifactorial in aetiology. Making a clinical diagnosis of GORD in children with neurological disability is challenging as symptoms are often lacking (communication deficits), non-specific (unexplained irritability, food rejection, hyper salivation) or atypical (anaemia, increased dystonia, seizures, laryngospasm, or recurrent pulmonary infections), and can be associated with other complications (inhalation, swallowing difficulties) in this high-risk group. (4)

Aim of the study:

To review endoscopic findings in chronically, neurodisabled patients who undergo upper GI endoscopy for suspected GORD. Furthermore, to compare this group of patients with a control group of patients, who are not neuro-disabled and undergo upper GI endoscopy for suspected GORD.

Method:

• Neurodisabled - identified prospectively from referrals to the lead consultant, age 2-16 years and non-verbal (< 30 words). These children were scoped in Birmingham Children's Hospital (BCH) between September 2018 and September 2019.
• Non neurodisabled - retrospective review of data for all the endoscopies done in the last quarter of 2018 in BCH for complains including vomiting, abdominal pain, feeding difficulties and nausea.

Results:

In total 78 patients' endoscopic findings were evaluated, of which 16 (20%) were neurodisabled. The mean age at the time of endoscopy was 9.3 (2-16) years in the neurodisabled-group and 8.3 (11 months-16) years in the control group with no statistically significant differences ($p>0.05$).

Endoscopic diagnosis of GORD and reflux esophagitis were the commonest findings in the neurodisabled-group (11 and 8 respectively), which has high statistically significant differences from the control-group ($p<0.00$).

Table 1 shows comparison between the two groups regarding the demographic data, endoscopic findings and the final diagnosis. An incidental finding was the increased prevalence of eosinophilic esophagitis in the neurodisabled-group (18.7% vs 5%) which was statistically non-significant ($P<0.06$).

Parameter	neurodisabled-group No=16	Control-group No=62	P Value
Age mean(±SD, years)	9.3±4.6	8.3±4.5	.06
Gender (male/female)	10/6	32/30	.4
PPI at time of the endoscope	10 (62%)	14(22%)	.002*
Endoscopic Findings: 1. Esophagitis	8	6	.000*

2. Reflux 3. Gastritis 4. Gastric erosions 5. Gastric ulcers 6. Duodenitis 7. Duodenal Ulcers	11 2 4 0 1 1	1 11 3 3 3 3	.000* .6 .012* .37 .82 .82
Main Final Diagnosis • Normal • GORD • Peptic Ulcer • Functional • Eosinophilic esophagitis • H pylori • Intestinal dysmotility • Constipation • Benign polyps	 4 7 0 0 3 0 1 0 0	 20 2 4 11 3 5 0 7 4	 .004*

Summary and Conclusion:
Children with neurodisability were more likely to have an endoscopic diagnosis of GORD than non-neurodisabled children when scoped for symptoms suggestive of GORD. Other conditions such as eosinophilic esophagitis could be present, thus the endoscopic and the biopsies' findings are of significant value to the patients' plan of care.

P56

Supporting patients and their families in a busy supra regional liver specialist unit out-patient setting

Rita Duru[1], Julie Taylor[1], Carla Lloyd[1] and Lindsay Hogg[1].
[1]Liver Unit, Birmingham Women's & Children's Hospital

Introduction:

RCPCH and BSPGHAN published Quality Standards (QS) for PGHAN in January 2017 that apply to children receiving GHAN in secondary and tertiary care settings across the UK[1].

Established in 1989 the Liver Unit at Birmingham Women's & Children's Hospital has provided supra regional treatment for children with acute and chronic liver disease & those requiring liver +/- intestinal transplantation. The unit appointed a dedicated specialist clinic co-ordinator in the early 1990s and to date is the only liver specialist unit offering this service.

The clinic co-ordinator is the first point of contact in clinic offering different levels of support and information to patients and families reducing anxiety at their visits to a specialist unit.

Aim:

Review out-patient clinical co-ordinator service offered to families and patients to determine whether we meet some or all of the The Future Vision for PEGHAN as set out in the QS.

Methods:

To compare clinic co-ordinator role support in matching QS as set out on page 35

Results:

We can identify areas that meet the QS.

• Clinic Co-ordinator has appropriate knowledge and skills to provide support in an outpatient setting.
• Ensuring the clinic runs in a co-ordinated to reduce anxiety, stress and possible miscommunications.
• Communicate with patients and their families who require specialised drugs i.e. immunosuppression, on importance of adherence to treatment providing accurate results.
• Inform MDT of patients who are infection risks and appropriate precautions are adhered to within the out-patient settings and those families who require extra support/information regarding their medical condition.
• Signposting to other departments/specialities.
• Ensure all non-English patients have support of an interpreter to ensure concise information is provided from an independent source
• Signposting to Patient Parent Support Groups
• Work in partnership with the transition team to encourage young people 15+ years to see doctors without parental support
• Weekly reviews pre and post clinics with consultants and MDT staff
o highlighting issues for upcoming clinics o raising outstanding issues from previous clinic

Summary:

The clinic co-ordinator in the liver unit outpatient setting delivers a unique service to provide continuity of care working in partnership with the MDT, to provide a coherent approach within the out-patient setting currently matching some of the Future Vision for QS.

Conclusion:

Communication between the clinical co-ordinator and MDT team is essential to ensure patient journey is consistent and QS are met. Ensuring families receive high standards of care within the out-patient setting is fundamental to enable the unit to work in partnership to develop trust.

References

1. Quality standards for Paediatric Gastroenterology, Hepatology & Nutrition, RCPCH: January 2017

P57

A rare case of microscopic colitis in a child with coeliac disease

Akshatha Mallikarjuna[1] and Christine Spray[2].
[1]Yeovil District Hospital; [2]University Hospital Bristol

Introduction:

Microscopic colitis is a common cause of chronic watery diarrhoea that becomes more common as we age.[1] It is an inflammatory disorder of colonic mucosa that causes chronic non-bloody diarrhoea, characterised using distinct microscopic findings on colonic tissue biopsy in an otherwise endoscopically and radiologically normal colon.[2] Several studies have demonstrated that microscopic colitis is among the causes of persistent or recurrent diarrhea in patients with coeliac disease, despite adherence to the diet.[3-5]

Aim:

Investigate for an alternative explanation for the symptoms in a patient with coeliac disease showing sub optimal response to gluten free diet.

Subject and Methods:

15 year old girl presented with dizziness, fatigue since 8-9 months, loose stools (6-7 times/day, type 6) since 2-3 months. Blood tests result showed IgA TTG>100mIU/L, endomysial antibody positive. She was diagnosed with coeliac disease on a non-biopsy pathway as per ESPGHAN guidelines and started on gluten free diet. No significant improvement in her symptoms. 2-3 months later she had severe loose stools, nocturnal stools, further weight loss and severely impaired quality of life. Compliance with gluten free diet was confirmed. No other associated symptoms. Repeat blood test showed IgA TTG of 92mIU/L. Fecal calprotectin was 285mcg/gm.

Result:

She underwent upper and lower GI endoscopy, as she continued to be bed ridden with no improvement in her symptoms. Duodenal biopsy showed focal villous atrophy and focal intraepithelial lymphocyte infiltration fulfilling Marsh III criteria for coeliac disease. Biopsy of the colon showed intraepithelial lymphocyte infiltration and erosion of surface epithelium in most part suggesting microscopic colitis.

Summary:

Coeliac disease is an immune-mediated disorder affecting genetically predisposed subjects and is caused by the ingestion of gluten present in cereals.[6]

The etiology of microscopic colitis however is unknown, but leading models of pathogenesis include autoimmunity, an immune or inflammatory response to luminal factors, and, for collagenous colitis, myofibroblast dysfunction.[7] Collagenous colitis (CC) and lymphocytic colitis (LC) are the two recognised forms of microscopic colitis, each with characteristic histopathological features.[8,9]

Coeliac syndrome is associated with HLA-DR3-DQ2 haplotype, which also correlates with microscopic colitis: the inflammatory process is similar between the two syndromes.[10] One of the study suggests that in cases of non-response or symptom relapse in coeliac disease additional pathology should be considered and colonoscopy be part of the follow-up in patients who present with chronic watery diarrhea, even if initial tests indicate only CD.[11]

Conclusion: CD can co-exist with other gastrointestinal diseases of an auto immune origin. Although microscopic colitis is common in elderly females, it should be considered especially in children not showing response to gluten free diet. Chronic watery diarrhoea is the clue warranting colonoscopy in these patients. Further studies looking for incidence and characteristics of microscopic colitis in children is essential.

References.

1. Guagnozzi D, Arias A, Lucendo AJ. Systematic review with metaanalysis: diagnostic overlap of microscopic colitis and functional bowel disorders. Aliment.Pharmacol.Ther.2016 Apr; 43(8): 851-862. [Pubmed: 26913568]
2. Stewart M, Andrews CN, Urbanski S et al. The association of coeliac disease and microscopic colitis: a large population-based study.Aliment Pharmacol Ther.2011 Apr; 33: 1340-1349.
3. Fine KD, Meyer RL, Lee EL. The prevalence and causes of chronic diarrhea in patients with celiac sprue treated with a gluten-free diet. Gastroenterology 1997;112:1830–1838.
4. Abdulkarim AS, Burgart LJ, See J, et al. Etiology of nonresponsive celiac disease: results of a systematic approach. Am J Gastroenterol 2002;97:2016–2021.
5. Leffler DA, Dennis M, Hyett B, et al. Etiologies and predictors of diagnosis in nonresponsive celiac disease. Clin Gastroenterol Hepatol 2007;5:445–450.
6.Di sabatino, Corazza GR. Coeliac disease. Lancet 2009;373:1480-93.
7. Darrell SP, Ciaran PK. Reviews in basic and clinical Gastroenterology and Hepatology. Gastroenterology 2011;140:1155–1165.
8. Bohr J, Olesen M, Tysk C, et al. Collagenous and lymphocytic colitis: a clinical and histopathological review. Can J Gastroenterol 2000; 14: 943–7.
9. Williams JJ, Beck PL, Andrews CN, et al. Microscopic colitis – a common cause of diarrhoea in older adults. Age Ageing 2010; 39: 162–8.
10.Wickbom A, Nyhlin N,Montgomery SM et al.Family history, comorbity, smoking and other risk factors in microscopic colitis:a case control study. Eur J Gastroenterol Hepatol.2017 May;29(5):587-594
11.Green PH, Yang J, Cheng J, Lee AR, Harper JW, Bhagat G. An association between microscopic colitis and celiac disease. Clin Gastroenterol Hepatol2009;7:1210–1216.

P59

From status to stricture – risks of PR paraldehyde

Tine Alliet[1], Robert Heuschkel[1] and Rishi Arora[2].
[1]Addenbrookes Hospital - Cambridge University Hospitals NHS Trust; [2]Bedford Hospital NHS Trust

Introduction/Background:

A 16-year old boy with a background of developmental delay, behavioural issues and refractory epilepsy was admitted after a status epilepticus requiring intubation and ventilation for which he was given paraldehyde PR twice. Following this he developed severe perianal lesions, rectal bleeding, fever and constipation. He was initially treated with IV antibiotics, oral laxatives and local wound care. Promethazine was commenced for deterioration of challenging behaviour. He underwent an endoscopy under GA which revealed a colonic stricture and severe rectal inflammation. The stricture with a length of 4.5cm was confirmed on abdominal CT scan. Due to the critical nature of the stricture, a defunctioning colostomy was performed. Unfortunately, there is no certainty that a join up will be possible in the future.

Aim:

To highlight the side effects and risks of paraldehyde and to summarize previous reports regarding corrosive damage following rectal paraldehyde in paediatric patients.

Subjects and Methods:

A systematic literature search was performed in the MEDLINE database. A combination of the following MeSH terms was used: [rectal paraldehyde], [paraldehyde AND side effect], [paraldehyde AND injuries], [paraldehyde AND adverse effect], [paraldehyde AND toxicity], [paraldehyde AND burn(s)], [paraldehyde AND stricture], [paraldehyde AND proctitis]. Abstracts were screened for relevance. All paediatric reports of adverse events due to rectal paraldehyde were included.

Results:

We found 6 relevant articles regarding 772 paediatric cases who received rectal paraldehyde. Age varied from 1 month to 24 years. The dose administered was between 0.3ml/kg and 0.6ml/kg, in 3 of the cases this dose was divided over multiple doses in 1-2 hours. The amount of paraldehyde was mixed in an equal amount of oil in 67 of the cases and in +/- 11.5 times the amount of saline in 654 cases. There were no specifics about the solution in 51 cases. Another article reported 677 paediatric patients who were sedated with either chloral hydrate alone or a combination with rectal paraldehyde. Unfortunately, the number of patients treated with both was not specified. They used a premixed solution of 1:1 paraldehyde liquid with olive oil.

In 4 of all the cases above respiratory depression was named as a side effect but as in those 4 patients a combination with phenytoin was used, it is unclear what has caused the respiratory depression. All others didn't report any side effects following rectal paraldehyde.

Summary and conclusion:

Even though the corrosive and irritating characteristics of paraldehyde are well prescribed in pharmacological reports, there is very little literature available about this topic. Medical staff should be aware of the possible serious outcomes following paraldehyde administration, especially because this drug is up until today part of the UK's APLS algorithm for the management of status epilepticus.

P61

Case study: An unusual side effect of blended diet via gastrostomy

Julia Callan[1] and Salvestrini Camilla[1].
[1]Cambridge University NHS Foundation Trust

Background:

The use of blended food provided via gastrostomies is an increasingly popular choice for parents and carers of tube fed children in the UK. Recent British Dietetic Association guidance recommends dietitian-led indications of blended diet as an option, based on potentially physiological, social or emotional benefits to the tube fed individual. Importantly, the perceived risks of blended diet including potential nutritional deficiency have not been widely researched. Specifically, risks of nutritional excesses due to blended diet has not been identified as a concern.

We present a case of beta-carotenaemia in a four-year-old male who was fully blended diet fed via gastrostomy.

Case report:

The patient had a background diagnosis of Transposition of Great Arteries, vomiting and food aversion and been on full blended diet via gastrostomy for 11 months. During this time, he had demonstrated a marked improvement in volume tolerance with cessation of vomiting and notably no longer required jejunal feeding. Weighed food diaries had been analysed at commencement of diet, four and ten months in. These had not indicated any nutritional imbalances. On review in clinic it was noted that the patient had orange pigment of his face and palms. Biochemical analyses of serum beta carotene revealed excessive levels at 2373µg/L (ref: 90-310). On detailed history taking, the patient's family denied the nutritional intake to be different to the reported regimen as well as the use of other nutritional supplements or powders.

Discussion:

A number of concerns over nutritional adequacy of full blended diet for gastrostomy fed children have made its use a controversial issue. However, this discussion is mainly focussed around nutritional deficiencies In contrast, this case raises the important question of bioavailability of nutrients in blended feeds.

Conclusion:

Gastrostomy fed children who receive blended diet may be at risk of nutritional deficiencies or micronutrient intoxication. Hence, the frequency of dietary analyses, clinical assessments and biochemical monitoring in this vulnerable group of patients must be carefully indicated.

P62

The pathophysiology of Non-Erosive Reflux Disease (NERD) is different in children compared to adults in regards to oesophageal mucosa integrity and innervation.

Kornilia Nikaki[1], Chung Lee[1], Ahsen Ustaoglu[1], Philip Woodland[1] and Daniel Sifrim[1].
[1]Wingate Institute of Neurogastroenterology, Blizard Institute, QMUL

Introduction:

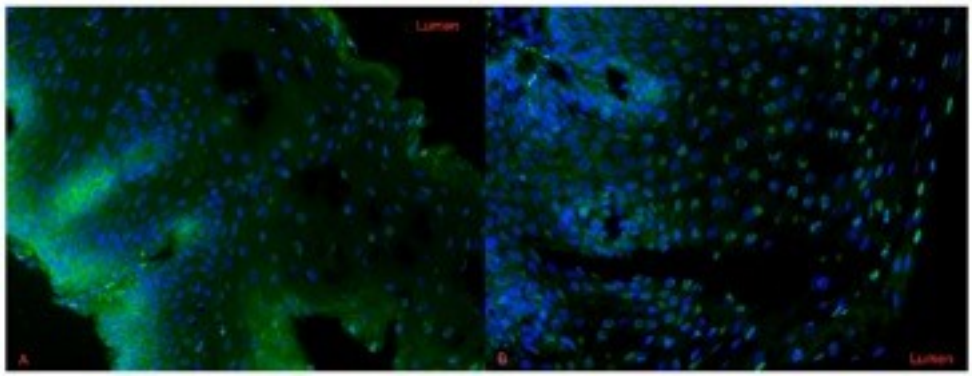
The mucosa of adult patients with Non-Erosive Reflux Disease (NERD) shows increased vulnerability "in vitro" when exposed to refluxate-like solutions. Moreover, the normal oesophageal mucosa has deep lying afferent nerves in the distal oesophagus and more superficial in the proximal oesophagus. In contrast, NERD patients have superficial nerves along the oesophageal length. Our study aimed to assess oesophageal mucosal integrity and innervation in children with NERD and controls.

Methods:

We prospectively recruited children undergoing an upper GI endoscopy for clinical reasons and identified a subgroup with normal histology +/- normal impedance pH-metry that served as controls and a patient group with increased acid exposure on impedance pH-metry. We obtained oesophageal biopsies from 3-5cm above the lower oesophageal sphincter and from the proximal/upper third. Fresh distal biopsies were placed in Ussing chambers (surface area:0.011cm2) and exposed to pH2 and pH5 solutions containing bile acids and pepsin. The baseline trans-epithelial resistance (TER) and its % of drop when exposed to the refluxate-like solutions were recorded. Distal and proximal biopsies were immunohistochemically stained with CGRP. CGRP positive nerve fibres were identified and their position relative to the lumen was determined (expressed as median number of cell layers from the lumen for all sections examined).

Results:

For the mucosal integrity studies, 19 children were included (12M:7F, median age: 11 years, range: 0.83-15 years) in the control group and 9 in the NERD group (4M:5F, median age: 6 years, range: 1-15 years). The baseline TER were comparable between the two groups (179 vs 175 Ohms). The drop of the TER when mucosa was exposed to refluxate-like solutions with pH2 and pH5 was -44% and 0% in the control group and -65% and +10% in the NERD group (non-significant differences; Image 1). For the innervation studies, 19 children were included in the control group (13M:6F , median age: 11 years, range 0.83-15 years) and 15 children in the NERD group (8M:7F, median age: 10 years, range 1-16 years). In the control group, CGRP positive nerve fibres were identified at a median of 19.5 cell layers from the lumen in the proximal and 19 cell layers in the distal oesophagus. In the NERD group, CGRP positive nerve fibres were identified at a median of 17.5 cell layers in the proximal and 17 cell layers in the distal oesophagus (non-significant differences; Image 2). There is no correlation between the age and the position of the nerve fibres identified.



Images 1 and 2

Conclusion:
Unlike adults with NERD, the oesophageal mucosa in children with NERD does not show increased vulnerability to acid exposure. The oesophageal mucosa innervation in children is similar in the proximal and distal oesophagus with deep lying nerve fibres both in controls and NERD patients; in contrast to adult healthy volunteers who have superficial nerves in the proximal oesophagus and NERD patients who have superficial nerves in the distal oesophagus too. These differences may be due to different composition/pH of the refluxate, duration of exposure to reflux and subsequent inflammation and/or repair mechanisms.

P65

High Prevalence of Autism Spectrum Disorder in Paediatric Patients with Eosinophilic Oesophagitis: Experience of a single tertiary centre in UK

Chai Leng Lee[1], Alexandra Patrick[1], Adepoju Akinlolu[1], Emma Jones[1], Christopher Greaves[1], Sharon Watters[1], Helen Garrett[1], Sarang Tamhne[1], Elizabeth Renji[1], Manjula Velayudhan Nair[1], Raj Parmar[1], Taimur Babar[1], Fiona Cameron[1], Laura Grey[1], Alexandra Brown[1] and Marcus Karl-Heinz Auth[1].
[1]Alder Hey Children's NHS Foundation Trust, Eaton Road, Liverpool, UK

Background and Aims:

Eosinophilic oesophagitis (EoE) is a chronic immune/antigen mediated oesophageal inflammatory disease associated with oesophageal dysfunction resulting from severe eosinophil-predominant inflammation. Children with EoE present with feeding difficulties, including feed refusal, vomiting, regurgitation, dysphagia and food impaction, often resulting in changes of their food intake. Autism spectrum disorder (ASD) refers to a group of complex neurodevelopment disorders characterised by repetitive and characteristic patterns of behaviour and difficulties with social communication and interaction. They typically display different eating and feeding problems such as foods refusal, ritualistic eating behaviours, picky eating behaviour, and food rejection due to their texture.

Both EoE and ASD can present with food selectivity and feeding disorders. We noticed co-existence of both conditions in our patients and were interested in pattern of food restriction behaviours, food allergies and atopy in these patients. No previous studies have explored the prevalence of ASD in patients with EoE. We aim to evaluate if there is an association of EoE and ASD.

Subject and Methods:

This is a retrospective case-cohort study. A proforma was designed for data collection. Consecutive n=30 patients with EoE were selected from the EoE registry 2018-2019 to identify those with ASD. Consecutive n=30 age, gender and time-matched subjects were selected as control group from children listed for OGD. Indications included abdominal pain, gastro-oesophageal reflux, possible coeliac disease, haematemesis and failure to thrive. All control group subjects did not have EoE as proven by histology report. Exclusion criteria for both groups include patients with severe neurological impairment and patients who were exclusively enterally fed. Significance was calculated with chi square statistic test ($p<0.05$).

Results:

From 30 consecutive EoE patients, ASD was detected in n=7 (23.4%) (p=0.023), whereas matched control patients had ASD within the expected prevalence of our regions (n=1, 3.3%). As reported before, patients with EoE were predominantly male. There was no significant difference in the prevalence of reported restrictive food intake, food allergy and atopy.

Results

	EoE Group n=30	**Control Group** n=30
Male	25	20
female	5	10
Age (years)	9.7 (2.7- 17.4)	10.2 (5.1- 16.7)
ASD	**7 (23.4%) (p<0.05)**	**ASD n=1 (3.3%)**
Restrictive food intake	3	8
Food allergy	7	3
Atopy	9	5

Conclusions:
For the first time, this retrospective study demonstrated a high prevalence of ASD in patients with EoE. Prospective, larger studies are planned to elucidate if restrictive food intake is the cause or result of EoE. Unrecognised GI disorders in patients with ASD may contribute to their behavioural and feeding difficulties. This finding suggests that feeding disorders in ASD should not be assumed to be purely behavioural and should be evaluated further with upper GI endoscopy with a low threshold to rule out EoE.

P66

The role of endoscopic retrograde cholangiopancreatography in children with Hereditary Pancreatitis.

Sophia Kanwar[1], Deepak Joshi[2], Anil Dhawan[1] and Tassos Grammatikopoulos[1].
[1]Paediatric Liver, GI & Nutrition Centre and MowatLabs, King's College Hospital, London, UK
[2]Institute of Liver Studies, King's College Hospital, London, UK

Hereditary pancreatitis (HP) is a rare, debilitating condition of recurrent episodes of pancreatitis and intermittent pain. Treatment for HP involves a combination of pain management, endoscopic interventions, monitoring of pancreatic function and lastly pancreaticojejunostomy. There are no clear management guidelines for paediatric HP and we investigated the role of ERCP (endoscopic retrograde cholangio-pancreatography) in those.

Methods:

We performed a retrospective, single centre study in patients with chronic pancreatitis between 2006-2019. Those with genetically confirmed HP were identified and demographic, biochemical, radiological and endoscopic data were collected.

Results:

We identified 39 (11 male) children with HP with a median age of 8.5 years (range 2-15) at diagnosis. Genetic variants were found in SPINK1 51% (n=20), PRSS1 41% (n=17) and CFTR 10% (n=4). Median values at presentation were 102 IU/L (27-1290) for serum amylase, 6 µmol/L (<3-23) for bilirubin, 12 IU/L (6-183) for GGT and 164 µg/g (1->500) for faecal elastase.

Indications for the first ERCP are shown in Table 1 and interventions performed were stent insertion (n=15), sphincterotomy (n=14) and complications recorded were (pancreatitis n=1 and prolonged pain n=2).

Indication	**PRSS1**	**SPINK1**	**CFTR**
Pancreatic duct (PD) changes/ stricture	5/2	2/1	
Pseudocyst drainage	1	3	
Pancreatic leak	1	2	
Pancreaticolithiasis	1	2	
Pancreas divisum		1	1
Delineation of pancreaticobiliary system/ biopsy	2/1		
Intervention	13	11	1
PD stent insertion	6	8	1
Sphincterotomy	9	6	1

Table 3

Median follow up was 5 years (0-14). Overall, during the follow-up period management included no ERCP, 1 ERCP, 2-3 ERCPs and 4-5 ERCPs before symptoms were appropriately managed or they were referred for surgery in 15, 7, 14 and 3 patients respectively. Of the 24 patients who underwent 1st ERCP, 25% (6) had resolutions of their symptoms, 4% (n=1) required surgery due to complete PD blockage from lithiasis and 71% (n=17) required a 2nd ERCP. Median time between ERCPs was 8 weeks (range 1-156). Following ERCP, 41% (7) had no more interventions, 6% (1) had surgery and 53% (9) required further ERCPs: 5 had stent insertion and 3 had stent removal. Following 3rd ERCP, 44% (4) had no more interventions, 1 patient had thoraco-splanchnic nerve surgery, 1 patient had surgery and 3 patients required another ERCP, all requiring stent insertion. The three patients

required a 5th ERCP for persistent PD changes or pancreaticolithiasis, following which 2 had no interventions at last follow up and one was referred for surgery.

Five patients underwent pancreaticojejunostomy and one patient underwent a total pancreatectomy and auto-islet transplantation. Average time to surgery was 1.75 years (range 0.4-5 yrs). Median values at last clinic visit were 37 mmol/mol (31-72) for HbA1c and 238 µg/g (23->500) for faecal elastase for all 39 patients.

Conclusion:
ERCP has an important role in the management of HP with 61% of our cohort requiring one. PD changes are the commonest indication and stenting the primary intervention with resolution of PD changes in 80% of patients after 3rd ERCP.

P68

Nutritional Blood Monitoring in Blenderised Diet; food for thought

Miss Michelle Brooks[1], Miss Hazel Duncan[2], Miss Jacqueline Hay[2], Miss Sarah Bremner[2], Miss Gillian Smith[3], Miss Cardigan Tracey[2], Mr Timothy Bradnock[4], Mr James Andrews[4], Miss Christina McGuckin[1], Dr Victoria Merrick[1], Mr Simon Fraser[5], Miss Elaine Buchanan[2], Dr Konstantinos Gerasimidis[6], Dr Richard Hansen[1], Mr Gregor Walker[4], Dr Diana Flynn[1] and Dr Andrew Barclay[1].
[1]Paediatric Gastroenterology, Royal Hospital for Children, Glasgow UK; [2]Paediatric Dietetics, Royal Hospital for Children, Glasgow, UK; [3]Paediatric Dietetics, Wishaw University Hospital, UK
[4]Paediatric Surgery, Royal Hospital for Children, Glasgow, UK; [5]Paediatric Pharmacy, Royal Hospital for Children, Glasgow, UK; [6]University of Glasgow

Objectives and Study:

The practice of administering blended diet (BD) as opposed to enteral feeds is increasingly prevalent amongst the paediatric population, either in conjunction with health professionals or by parents and families independently. Limited data exist on the long-term efficacy and safety of BD (1) and to our knowledge there are no data on nutritional blood monitoring (NTBM) in prolonged BD.

We aimed to describe our experience of NTBM in BD patients in a large regional service.

Methods:

Patients commenced on BD between 1/1/17-1/6/19 were identified prospectively from our complex enteral feeding clinic and regional dietetic database. Patients were established on BD by local process; dietetic assessment of nutritional requirements; risk assessment of patient and family circumstances; educational package including discussion of nutritional adequacy, safe preparation, storage and administration of BD; BD volumes were prescribed in combination with enteral feeds and titrated upwards with tolerance; individualised meal plans were reviewed for nutritional adequacy and alterations were advised in a tailored manner. After stabilisation of enteral regimen patients were defined as either full or partial (where an enteral formula is used as substantial component of the BD meal or enteral feeds were still administered as separate part of a feed regimen) BD. Local guidance recommends NBTM at baseline and 6, 12/12 thereafter for; U+E, LFT, Bone Profile, PTH Vits A, B12, D, E, zinc, copper, selenium, magnesium, manganese, FBC, ferritin and folate. Patients were screened for acute inflammatory response with a combination of clinical assessment, serum albumin and C-reactive protein. Alterations to dietetic intake and supplementation were actioned via MDT forum.

Results:

33 patients (12F) commenced BD with our support. 29 patients had moderate/major neurodisability as a primary diagnosis with 21 fulfilling our criteria for gastro-intestinal dystonia (2), 6 patients had short bowel syndrome (SBS). 10 (30%) patients received full BD and 23 (70%) partial BD. 20 (8 full 12 partial) patients had NTBM as per local standards. 7/20 all results within normal limits. Abnormalities (Table 1). 17 (85%) patients required no intervention, 2 patients had zinc supplementation and 1 had major modifications to BD profile as it was found to be markedly iron deficient.

Blood Indice	**Abnormality (no pts)**	**Action**
Vit E	Mild elevation (4)	None
Selenium	Mild elevation (4)	None
Vit A	Mild elevation (3)	None
B Vits	Mild elevation (3)	None
Zinc	Moderate deficiency (2)	Supplementation
Ferritin	severe deficiency (1)	Dietary modification

Table: NTBM abnormalities in 20 patients receiving BD

Conclusion:
We describe BD practice across our regional service, and report that the majority of patients receiving BD, known to health professionals, in our region are in the context of severe neuro-disability and/or short gut using partial BD. The frequency and severity of NTBM abnormalities compare favourably with previous review of enteral or orally fed patients with neuro-disability (3). We postulate that these favourable outcomes in terms of NTBM reflect careful patient selection and intensive dietetic support for patients. Whether the use of adjuvant enteral formula as part of a BD diet is a stabilising factor for micronutrients warrants further study.

1. Armstrong J, Buchanan E, Duncan H et al Arch Dis Child 2017;102:152-8
2.McConnell N, Beattie LM, Richards CE, Prothero S, Barclay AR, JPGN;66 (supp2): 1002;2018
3.Brooks M, Paxton CE, Cardigan C, Wilson DC, Barclay AR. JPGN 2014(58) Supp 1; S4

P72

High chronicity of Eosinophilic Esophagitis (EoE) and need for treatment modifications in children with EoE - a prospective tertiary hospital experience

Raj Singh Parmar[1], Alex Patrick[1], Adepoju Akinlolu[1], Emma Jones[1], Christopher Greaves[1], Sharon Watters[1], Sarang Tamhne[1], Elizabeth Renji[1], Manjula Velayudhan Nair[1], Helen Garrett[1], Taimur Babar[1], Laura Grey[1], Fiona Cameron[1] and Marcus K H Auth[1].
[1]Alder Hey Children's Hospital

Introduction:
Eosinophilic esophagitis (EoE) constitutes the most prevalent cause of chronic esophagitis is associated with reduced quality of life in children. As major UK contributor site to the prospective ESPGHAN Pediatric EoE Registry (PEER) we are collecting prospectively data on disease characteristics, progression, and treatment response. According to ESPGHAN guidelines, treatment consist of monotherapy with oral viscous budesonide (OVB), six food elimination diet (SFED) or proton pump inhibitors (PPI)

Aim:
To understand the proportion of chronic disease activity and treatment response in children with EoE.

Methods and subjects:
Prospective review of all patients newly registered in our centre from 2017-2019 (also on PEER registry). Follow up endoscopy period range from 3 months to 47 months with median of 16 months. Out of 42 cases 3 did not book in for follow up endoscopy.

Results:
N=42 patients were newly registered. As reported before, male preponderance with male:female ratio 5:1 existed. Age at diagnosis ranged from 1 yr to 17 years (mean age 10 years). In n=4 (9.5%) of children EoE was diagnosed incidentally.
OVB was prescribed as the most common initial treatment (n=24; 57.1%) followed by elimination diet of six food (n=14; 33.3%) or targeted (n=12; 28%) based on patient preference and compliance. N=2 patients required combination treatments (4.7%) for co-existent IBD. PPI was prescribed in n=26 (61%) at initial diagnosis in combination with other treatments for signs of concurrent GORD. At follow up endoscopies were macroscopically normal in 19/39 cases, followed by furrows (18/39) and exudate (4/39) with one trachealisation.

Initial gastroscopy showed normal macroscopic findings in 13 (30%) cases. Most common macroscopic endoscopic finding were linear furrows 59% followed by exudate 21% and trachealisation in 11%. We found no correlation between severity of macroscopic finding and number of eosinophils on histopathology.

Out of 39 cases only n=20 (51%) cases were in remission on follow up, the mean endoscopic reference (EREFS) score dropped from 1 to 0.6. At last follow up there was significant reduction in prescription of OVB from 57% to 21% and 20/42 (47%) children were still on PPI.

Summary and Conclusion:
After mean follow-up period of 16 months only half of children were in remission of EoE. Notably, in two thirds of children, maintenance treatment was changed, which deserves further analysis to better understand and address motivation, compliance, side-effects, health related and treatment related quality of life in children with EoE.

There was no correlation between symptoms, macroscopic finding and histopathology report, which makes it difficult to predict the outcome and indicated the need for validated tools of disease activity such as Paediatric Eosinophilic Esophagitis Symptom Score (PEESS) 2.0 (which is now part of our surveillance plans). Effective dietetic input is crucial as evident from shift from SFED to TED. The launch of BSPGHAN EoE working group with national quality standards will provide an essential milestone to improve outcome and quality of life in children with EoE.

P73

Role of Azathioprine as a novel therapy for Refractory Paediatric Eosinophilic Esophagitis

Raj Singh Parmar[1], Elizabeth Renji[1], Sharon Watters[1], Emma Jones[1], Manjula Velayudhan Nair[1], Fiona Cameron[1], Marcus K H Auth[1], Jeng Haw Cheng[1] and Sarang Tamhne[1].
[1]Alder Hey Children's Hospital

Introduction/Background:
Eosinophilic esophagitis (EoE) constitutes one of the most prevalent cause of chronic esophagitis. Untreated EoE is usually associated with persistent symptoms and inflammation resulting in stricture formation and functional abnormalities. PPIs, elimination diet six food (SFED), targeted (TED) and topical steroids are offered as first line therapy. Although systemic and topical corticosteroids are effective in treating EoE, some patients develop corticosteroid dependency. Alternative therapeutic approaches to avoid corticosteroids are scarce and this makes management of refractory EoE very challenging.

Aims and objectives:
To assess whether Azathioprine as an immunosuppressant therapy can be effective to bring EoE in remission. We present our experience of management of refractory EOE that has failed all available commonly used treatments.

Methods and subject:
Retrospective review of a refractory case of EoE.

A 7 yr old boy with past history of partial colectomy secondary to appendicectomy presented with symptoms of blood and mucus in stools with no upper GI symptoms. Upper GI endoscopy done in Jan 2014 showed incidentally EoE and normal colon. He was started on oral Omeprazole. Repeat endoscopy in June 2014 showed persistent eosinophilic esophagitis and developed symptoms of dysphagia. He was prescribed oral viscous Budesonide (OVB) 8 wk course. Endoscopy in October 2014 showed persistent eosinophilic oesophagitis with new finding of eosinophilic enteritis. All his bloods were unremarkable with no allergy to common foods. From Jan 2014 till March 2019 child had 16 upper GI endoscopies (Table1). He had very marginal effect of available first line treatments. He received wide range of treatments including PPI, topical and oral steroids, Montelukast, Sodium cromoglycate, SFED, TED, elemental feeds (Neocate, Elemental 028).

S/N	Date of endoscopy	Histopathology findings	EoE treatment started / changed
1	Jan 2014	EoE, normal colon	Oral Omeprazole (PPI)
2	June 2014	Persistent EoE	Oral viscous budesonide (OVB)
3	Oct 2014	Persistent EoE + Eosinophilc enteritis	Six food elimination diet (SFED) Neocate (as more symptomatic)
4	Jan 2015	EoE in remission Enteritis persist	Stay on neocate + Hypoallergenic food introduced. Symptoms worse so oral Budesonide started in June 2015
5	Aug 2015	Improvement in EoE and enteritis	PPI + Montelukast + Sodium cromoglycate
6	October 2016	Recurrence of symptoms and evidence of EoE	Oral budesonide
7	Jan 2017	EoE persistent	OVB represcribed
8	July 2017	EoE persistent	OVB continued + Elemental diet

9	Nov 2017	EoE persistent (eosinophils upto 40/hpf)	Exclusive enteral nutrition (EEN) via nasogastric tube
10	Feb 2018	EoE persistent but improvement in eosinophil count	Elemental diet finished recently + OVB represcribed
11	March 2018	EoE more worse (eosinophils upto 120/hpf	SFED + Oral steroid (Prednisolone 40 mg)
12	June 2018	Mild EoE (eosinophils upto 25/hpf)	Omeprazole + Elemental 028 feed
13	Sept 2018	EoE improvement	TED 2 (egg and soya free) + Omeprazole
14	Nov 2018	Significant EoE	Oral Prednisolone + Omeprazole high dose
15	March 2019	EoE some improvement, (eosinophils upto 70/hpf)	Azathioprine 1mg/kg + Elemental diet to bridge
16	Aug 2019	EoE in remission	Azathioprine to continue

Despite all above treatments endoscopy in March 2019 showed significant EoE disease activity. Child become more symptomatic. So far child had already received 3 courses of oral steroid (Budesonide or prednisolone) with no long term EoE remission. At this stage we planned to try Azathioprine 1mg/kg as rescue therapy with elemental diet as bridging strategy. Interestingly last endoscopy done in August 2019 showed significant improvement. At last EoE is in long term remission histologically and symptomatically. No side effects of Azathioprine noted on clinical review and on blood monitoring.

Results:
Immunosuppression with Azathioprine in case of refractory EoE has shown significant improvement to achieve possible long term remission.

Summary and conclusions:
EoE is increasingly recognised as a very challenging condition that has huge negative effect on health quality of life. To date there is very limited published data about efficacy of azathioprine or 6-mercaptopurine for EoE in paediatric age group. Our experience of effective use of Azathioprine brings new hopes to manage this debilitating disease and inspire more research and clinical trials in future.

P74

Strategies developed to support families facing intestinal transplantation

Lindsay Hogg[1], Velma Wright[1], Julie Taylor[1], Jane Hartley[1] and Girish Gupte[1].
[1]Birmingham Women's & Children's Hospital

Introduction:
A child/young person facing an Intestinal Transplant can be extremely difficult for families. It involves meeting a new team and hospital at a time when medical care of the child is challenging. An MDT approach has been demonstrated to benefit the family and help to ensure they are adherent to treatment and care. Working in partnership is essential to ensure children and young people receive the best care.
Our aim is to describe the strategies we have developed to meet the needs of our intestinal transplant families, children and young people.

Methods:
Introducing families to our transplant MDT at the time of transplant assessment, ensuring they understand team members' roles and responsibilities.

Giving families regular opportunities to meet with different members of the transplant team to gather information and ask questions. Provide families with information both written and verbal to empower and encourage them to participate in the discussions with healthcare professionals. Daily communication with the family is done in their own language via interpreters.

Establish links with the local support network early and ensure they are kept up to date with treatment and care plans. Eg Shared care consultant, community nursing teams

Support the parents to communicate with the nursery/school as appropriate. Discuss return to school plans and educational care plans to ensure the child/ young person can resume their education.

Behavioural contracts to establish families understand their roles and responsibilities.

Signpost families to support services e.g. Children's Liver Disease Foundation, Multi Organ Transplant Support.

Family support team advice parents/carers on welfare benefits, housing issues and financial support in the form of grants available.

Investigate and discuss opportunities for respite and other support services available in the local area.

Summary:
Feedback from families is that they feel part of the team looking after their child. They are able to confidently voice their opinions and concerns. Fostering a culture of parent and healthcare professionals working together is required to ensure children and young people's needs are met.

Future Development:
Further development of unit website, links for families to use prior to admission to orientate them to the unit and staff and development of apps.

P75

Setting up a transition service for intestinal transplant patients

Lindsay Hogg[1], Monica Smith[1], Heather Howe[2], Lisa Vokes[2], Philip Allen[2], Jane Hartley[1], Lisa Sharkey[2], Julie Taylor[1] and Girish Gupte[1].
[1]Birmingham Women's & Children's Hospital; [2]Oxford University Hospital NHS Foundation Trust
[3]Addenbrookes Hospital, University of Cambridge;

Introduction:

Improving survival of intestinal transplantation resulted in an increase in number of young people surviving into adulthood. The prospect of transition is exciting but "scary" in the words of our young people requiring the development of a service which meets their unique needs. It is vitally important that a robust supportive transition process is established for young people and families.
Method: Transition is discussed from 12 years with the family at their annual anniversary admission, introducing the concept and describing the process. Young people meet with the transition nurse, youth worker and psychologist. We use a transition programme called Ready, Steady Go and the HEADSS document to support transition.

Families first have the opportunity to meet the adult team at our biannual family day. The adult team consisting of the Lead Consultant, Specialist Nurse and Dietician contribute to the information session during the day. These can be a talk or information boards/posters. This allows families to gather information to make informed choices about their young person's future care.

The adult team join us in the outpatient clinic at the children's hospital to meet the young person and their parents/carer. It is the start of the formal transition. The Paediatric Consultant and Specialist Transition Nurse attend the adult clinic. The adult consultant will lead the consultation with the Paediatric Consultant contributing.

Results:

Ten young people have been transitioned and care handed over to the adult multidisciplinary team. The emphasis is very much on the young person and the time scale will be driven by their needs.

Retrospective review of the medical notes shows that young people are seen once in the paediatric clinic with the adult team, seen two/three times in the adult clinic with the paediatric team in attendance prior to formal handover. Formal handover can take two/three years.Six young people are in the process of being transitioned.

Conclusion:

Feedback from families has shaped the service. Communication between paediatric and adult teams has highlighted differences in protocol processes. This has helped us prepare the young people to cope with the differences. Healthcare professionals need to therefore work collaboratively with young people to ensure they have all the information and resources to engage with the service. This will help to ensure a good long-term outcome in young people undergoing transition.

P77

Audit of complications of jejunal tube feeding- a single centre experience: Following the introduction of individualised patient pathways.

Michelle Brooks[1], Christina McGuckin[1] Karen Fraser[1], Susie Goodwin[1], Victoria Merrick[1], Diana Flynn[1], Atul Sabharwal[1], James Andrews[1], Tim Bradnock[1], Gregor Walker[1] and Andrew Barclay[1].
[1]NHSGGC

Objectives and study:

Jejunal feeding is becoming a common strategy for the provision of nutritional support in children with Gastrointestinal Dystonia (GID). Complications of jejunal tube feeding are well reported, however there is little documented around the methods to prevent complications in the first instance.

We aimed to examine the frequency and types of complications associated with jejunal tubes within our Complex Enteral Nutrition (CEN) Service since the introduction of de-escalation plans which provide a feeding regimen ie gastric feeds or intravenous fluids in the event of a complication and a standardised troubleshooting guide.

Methods:

Patients managed within the CEN service in the Royal Hospital for Children were included; patients were identified using the CEN database, jejunal feeding commenced between 01/01/2016 and 30/08/2019. Demographics including age, gender, underlying medical condition, reason for jejunal feeding, device type, complications and outcome of jejunal feeding were collected.

Results:

62 patients were identified, 36/62 (59%) male. The commonest underlying condition was neurological impairment/Cerebral Palsy in 21/62 (34%), followed by GI Dystonia in 13/62 (21%) other conditions included underlying neurological syndromes, reflux (GOR) and cardiac disease. The most common indication for jejunal feeding was poor enteral feed tolerance in 19/62 (30%) followed by GOR in 12/62 (19%) and GI dystonia in 7/62 (11%); other reasons include poor weight gain/failure to thrive.

17/62 patients had a Corflo (Halyard) with jejunal extension,29/62 had a gastrojejunal (GJ) button,4/57 had a GJ tube (Halyard),14/57 had a freka (Fresenius Kabi) with jejunal extension. 4 Patients had a witzel jejunostomy, 9 children required placement of more than 1 type of device due to multiple complications. 205 devices were placed in 62 patients; 41/205 were elective changes and 77/205 initial placements therefore discounted, leaving 54 unplanned replacements. The most common indication for replacement was migration of the jejunal tube into the stomach 19/54 (35%), blockage 7/54, (13%), with 4 of these occurring in the 6Fr Corflo extension, and dislodgement 14/54 (26%). We previously reported blockage of the Corflo device as our main complication (54%), since the introduction of our troubleshooting guide this has reduced dramatically. Other complications included broken ports and snapped/cracked tubes.

By the end of the study period, 29/62 (47%) patients were de-escalated to gastric feeds using a de-escalation plan following a complication with their jejunal tube.5 children died due to unrelated reasons.22 children remain jejunally fed, 4 children progressed to formal roux-en-y jejunostomy. 2 children are nil by mouth on home parenteral nutrition.

Conclusion:

Jejunal feeding can be a beneficial method of providing nutrition to children with GID and other complex medical conditions either in the short term, or longer term. However, the complications of

jejunal feeding tubes can be burdensome to patients, families and the MDT caring for these children. Effective training for staff and parents is required to minimise risks of complications where possible. The implementation of individualised de-escalation plans and a standardised trouble shooting guide for jejunally fed children have allowed for successful de-escalation of jejunal feeding in the majority of our cohort, and reduced the number of device complications substantially.

P82

Case report of Hereditary angioneurotic oedema associated acute pancreatitis

Sunita Amar Rajani[1], Ilhan Omar[1], Fiona Shackley[1], Catherine Waruiru[1], Prithviraj Rao[1] and Priya Narula[1].
[1]Sheffield Childrens Hospital

Objectives and study:

We report a case of a teenage boy with hereditary angioedema (HAE) type 1 presenting with recurrent abdominal pain. Abdominal pain is a common gastrointestinal symptom in HAE due to intestinal oedema, however pancreatic involvement is rare. To our knowledge this is the first paediatric case reported in the UK.

Methods:

A retrospective review of clinical notes, clinic letters, and investigations was undertaken. Review of literature revealed there were 13 cases of HAE associated with pancreatitis reported worldwide including one paediatric patient.

Results:

A 14-year-old athlete had multiple hospital admissions with recurrent abdominal pain. He was diagnosed with HAE at four years of age with an uneventful early childhood. His first few episodes of abdominal pain were secondary to constipation which was adequately treated. As he continued to have episodes of abdominal pain further investigations revealed H.pylori gastritis which was resistant to first line therapy. He also had three episodes of abdominal angioedema with bowel wall thickening and ascites on ultrasound, which responded to C1 esterase inhibitor (C1INH).

In 2019 he had two admissions with severe abdominal pain and vomiting with raised pancreatic enzymes consistent with diagnosis of acute pancreatitis. Amylase checked on previous presentations had been normal. He responded with conservative management and a second dose of C1INH. He had a complete diagnostic work up to rule out other aetiologies of acute and recurrent pancreatitis including viral, metabolic, immunologic and genetic screens and imaging to rule out structural anomalies.

Conclusion:

HAE patients can have recurrent episodes of subcutaneous or submucosal oedema and can present with varying gastrointestinal involvement. This case highlights the association of pancreatitis and HAE. The proposed mechanism for pancreatitis in HAE is postulated to be oedema or swelling of the pancreas that interferes with normal pancreatic drainage or obstruction of pancreatic duct/ampulla of vater due to intestinal oedema.

References:

1. Berger TD, Garty B-Z. Hereditary Angioedema Presenting as Recurrent Acute Pancreatitis. Pediatrics. 2016;137(2):e20150620-e20150620.
2. Veronez, Camila Lopes et al. “Hereditary Angioedema-Associated Acute Pancreatitis in C1-Inhibitor Deficient and Normal C1-Inhibitor Patients: Case Reports and Literature Review.” Frontiers in medicine vol. 6 80. 17 Apr. 2019, doi:10.3389/fmed.2019.00080
3. Visy B., Fust G., Bygum A., Bork K., Longhurst H., Bucher C., Bouillet L., Farkas H. Helicobacter pylori infection as a triggering factor of attacks in patients with hereditary angioedema (2007) Helicobacter, 12 (3) , pp. 251-257.

P85

The use of bone densitometry scan in the evaluation and management of bone health in paediatric patients with coeliac disease

Huey Miin Lee[1], Andrew Hubbert[1], Bukunola Kukoyi[1], Pamela Allen[1], Manuela Vadrucci[1] and Betina Lorentzen[1].

[1]King's College Hospital NHS Foundation Trust

Introduction/Background:

Low bone mineral density (BMD) can affect up to 75% of patients newly diagnosed with coeliac disease (CD).

Aim:

To review our single centre's experience in the use of bone densitometry (DEXA) scan in the evaluation and management of bone health in paediatric patients with CD.

Subjects and Methods:

Data of all patients diagnosed with CD in a tertiary paediatric gastroenterology unit from March 2000 to June 2017 were retrospectively reviewed. Information including patient demographics, date of diagnosis, results of initial and follow-up DEXA scans, subsequent management and Vitamin D levels were collected.

Results:

Over the 17-year period, 104 patients were diagnosed with CD. Median age at diagnosis was 6.8 years (range 11 months–16.4 years). 83/104 patients had DEXA scans. Median age of first DEXA scan was 9 years (range 4.7–17.8 years). 37/83 patients had a second DEXA scan. Median duration between the first and second DEXA scans was 3.1 years (range 1.0–5.4 years). 15/83 patients had a third DEXA scan and 3/83 patients had a fourth DEXA scan.

56/83 patients had lumbar spine (LS) BMD and total body (TB) BMD Z-scores ≥ -1.0.

19/83 patients had LS BMD Z-scores and/or TB BMD Z-scores between -2.0 to -1.0.
6/83 patients had LS BMD Z-scores and/or TB BMD Z-scores ≤ -2.0.
Median (range nmol/L) Vitamin D levels for the 3 group of patients above were 62.8 (31 – 113), 57 (23.7 – 98.1) and 53.7 (11.2 – 64.9), respectively.

2/83 patients were 4.7 years and 4.9 years old. As there were no normative data for patients at this age, their LS and TB BMD were reported as 0.530 and 0.681 (+/- 0.01 g/cm2) for the first patient, and 0.705 and 0.842 (+/- 0.01 g/cm2) for the second patient.

24 out of the 56 patients with initial LS and TB BMD Z-scores ≥ -1.0 had follow-up DEXA scans. Of these, 21 patients continued to have LS and TB BMD Z-scores ≥ -1.0, and 3 patients had LS and/or TB BMD Z-scores between -2.0 to -1.0 at their second DEXA scans.

8 out of the 19 patients with initial LS and/or TB BMD Z-scores between -2.0 to -1.0 had follow-up DEXA scans. Of these, 3 patients had improvement of Z-scores to ≥ -1.0, 3 patients continued to have Z-scores between -2.0 to -1.0, and 2 patients had worsening of Z-scores to ≤ -2.0 at their second DEXA scans.

5 out of the 6 patients with initial LS and/or TB BMD Z-scores ≤ -2.0 had follow-up DEXA scans. Of these, 1 patient had improvement of Z-scores to ≥ -1.0, 1 patient had improvement of Z-scores to -2.0 to -1.0, and 3 patients continued to have Z-scores ≤ -2.0 at their second DEXA scans.

Interventions to improve BMD include commencement of Vitamin D and calcium supplements, ensuring compliance with gluten-free diet and lifestyle changes.

Summary and Conclusion:

We share our centre's experience in using DEXA scans to monitor the bone health of children with CD which may be useful for development of future guidelines.

P87

Does blended diet improve symptoms in Intestinal Failure children on Home Parenteral Nutrition?

Rachel Wood[1].
[1]Manchester Foundation Trust

Home Parenteral Nutrition (HPN) can be very debilitating for our patients and families, they have high stool losses and it is very challenging to increase their enteral nutrition and reduce their parenteral nutrition without impacting on their growth, development and quality of life.

The aim of the trial was to introduce blended diet through the gastrostomies of our short bowel HPN dependent patients to see if any tolerance was established compared to standard feeds.

Methods:

Five of our patients on HPN for 5 and 6 nights a week over the last few months have introduced blended diet at least 3 times a day, daily via their gastrostomy. All families had wanted to start this and our BDA guidance (2019) is now as dietitians we can and should support families introducing this. Each patient was reviewed in clinic to check they had the correct tube in situ and risk assessment completed.

Each patient introduced one blended feed per day at a time at a mealtime, avoiding fruits and foods that can increase stool output. The quantity was increased to twice then three times a day. Family foods were encouraged, focusing on starches, protein and fats. The patients were reviewed between 2-4 weeks to check tolerance and advice on quantities and types of food was given to the parents to ensure the foods were high in calories and protein and as balanced as possible. Their stool output, bloods including Liver function, growth and volume of PN was closely monitored.

Results:

The biggest improvement that was seen is the change in stools when giving blended diet. These reduced in quantity and generally improved in consistency. The 2nd improvement was with parental autonomy and quality of life for these children around meal times, as the children felt more included in meal times and the parents had some control over their care.

Patient No	No of nights on PN	Stools pre blended diet	Stools on blended diet	Improved consistency in stools	Reduction in PN nights	Change in LFTS
1	6	4 x large volume undigested Vomits daily	2x large stools much thicker	√	no	Sig ↓in ALT and GGT
2	6	3 x day 2 x O/N watery loose	1 x day 1 x overnight and soft stools	√	no	↓GGT, ALT and Bili
3	5	BO x 4 overnight BO x 3 day, watery loose	BO x 1-2 overnight BO 2 day, thickening in consistency	√	no	LFTS already within range
4	5	4/ 24hours loose and watery	BO x 1 thicker and formed consistency	√	no	No change
5	5	BO x 3 overnight BO x 2-3 day large and loose	BO x 3/24hours thicker in consistency	√	no	No change

Conclusions:
Larger studies are required over a longer period of time to assess the real impact of blended diets on reducing dependence on HPN. In the future it would be helpful to look at multicentre data to be able to draw some significant conclusions. Overall blended diet has been really positive for the families that have started it.

P88

Bone health in coeliac disease patients- the role of DXA scan as a routine screening tool

Nastasia Hadjichristou[1], Jochen Kammermeier[1], Rakesh Vora[1] and Mohamed Mutalib[1].
[1]GSTT

Background:

Children with coeliac disease (Cd) are prone to develop impaired bone health. Dual energy x-ray absorptiometry (DXA) is considered gold standard in assessing the bone health. However, the optimal time, the necessity and clear recommendations of its use are still elusive. The aims of this study are to report the outcome of bone mineral density (BMD) in children with Cd by using DXA scan and to assess the duration of disease on skeletal health.

Methods:

Retrospectively review all paediatric Cd patients who had DXA scan between March 2013 and August 2019. As facilities to perform DXA scan became routinely available in our institution over the last 12 months we aimed to screen all children with Cd. Patients' demographics, BMD Z- scores (± SD) in lumbar spine (LS) and in whole body less head (WBLS), body mass indices (BMI), 25-OH vitamin D (25-OH-D) level and tissue transglutaminase antibody (tTG) at the time of DXA scan were analysed.

Results:

49 Cd patients (female: male ratio, 1.5: 1) were included. Median age at diagnosis was 6.6 years (IQR: 4.8- 9.2). Median time interval between diagnosis and DXA scan was 22± 24 months. Mean Z-score in LS was -0.2± 0.9 and WBLS -0.2 ± 1.1. Mean Z- score of 25-OH-D was 61 nmol/L± 21, BMI 18kg/m2± 3.3 and tTG 25± 38 at the time of DXA.

We further divided the cohort into three groups based on the time elapsed between diagnosis and DXA scan (Group A= 0- 12 months, Group B= 13- 24 months, Group C: > 24 months). In group A, Z-score in LS was -0.1± 0.9, WBLS: -0.1± 0.9, BMI: 18± 3.8 and 25-OH-D: 65± 21 and tTG: 30± 32. In group B, Z- score in LS was 0.1± 0.7, WBLS: 0.1± 1.2, BMI: 18± 4 kg/m2, 25-OH-D: 56nmol/L ± 16 and tTG 17± 38. In group C, Z- score in LS was -0.3± 0.9, WBLS: -0.5± 1.1, BMI: 17± 2.3 kg/m2, 25-OH-D: 62nmol/L ± 23 and tTG: 26± 51.

In one patient (2%) the BMD Z- score in LS was less than -2.3SD. Out of the total, 7/49 Cd patients (14%) were at risk for low BMD with Z- score in LS:-1.3± 0.1, WBLS:-1.2± 0.4, BMI 16.3kg/m2 ± 1.8, 25-OH-D: 44nmol/L ± 18 and tTG: 38± 49. One out of 49 patients (2%) presented with compression fractures after diagnosis.

Conclusions:

In paediatric patients with Cd the outlook of bone status as measured by DXA scan appear to be negative and did not improve despite gluten free diet and normal vitamin D level. 2% of Cd patients had low BMD and one patient had symptomatic vertebral fracture requiring treatment. DXA scan, is a useful, non-invasive and low radiation risk tool to risk stratify bone health in children with Cd.

P89

Assessment of factors predictive of abnormal gastric emptying in a large cohort of children

Maryam Hussein[1], Esme Poole[1], Vinod Kolimarala[1], Mich Lajeunesse[2], Efrem Eren[3], Francis Sundram[4] and Nadeem Ahmad Afzal[1].

[1]Department of Paediatrics, Southampton Children's Hospital; [2]Department of Paediatric Allergy and Immunology, Southampton Children's Hospital; [3]Department of Allergy and Immunology, University Hospital Southampton; [4]Department of Nuclear Medicine, University Hospital Southampton

Introduction:

Slow gastric emptying is a recognised cause of paediatric Gastroesophageal reflux often diagnosed with a gastric emptying scan.

Objectives:

To better understand the role of the gastric emptying test in the paediatric population, we set out to assess demographics and factors predictive of abnormal gastric emptying in a large cohort of children at Southampton Children's Hospital.

Subjects and Methods:

99mTc-DTPA is a non-absorbable compound used for assessing gastric emptying time. This was administered orally or via a Nasogastric/Gastrostomy tube mixed with their milk/feed. The radioactivity exposure is reduced in paediatrics compared to adults in conjunction with the Administration of Radioactive Substances Advisory Committee (ARSAC) recommendations with the minimum dose in children being 10MBq. Children aged 0-18 years who had the gastric emptying scan between 2009 and 2018 at Southampton Children´s Hospital were included in the study. Patient´s demographics, underlying diagnosis, and symptoms at the time of the scan were recorded.

Results:

A large cohort of 285 children who had gastric emptying scan were identified and recruited in this study. 182/285 (63.9%) children were reported to have an abnormal gastric emptying scan. The mean gastric emptying time was 62.84 minutes (SD=65.83). The mean age was 6.3 years (SD=5.5) with 55.4% males. Patients had a mean height of z=-1.14 (SD=2.1) and weight of z=-1.02 (SD=1.82). At the time of the scan, 90.8% (n=258) had symptoms of vomiting and acid reflux, 26.6% had constipation, 21.7% abdominal pain and 3.9% had diarrhoea. History of nausea was reported by 'older children' accounting for 11% of the subjects recruited in the study.

With regards to underlying diagnoses, 41.2% had an underlying neurological diagnosis, 29.9% had developmental delay and 21.6% had an underlying genetic condition. 22.7% had allergies with the top 3 identified allergies being milk (96.9%), soya (34.4%) and wheat/gluten (21.9%). 15.5% were born premature and 3 patients were diabetic. 38.5% had some form of surgery by the time they had the scan. Of the type of surgeries, 23.9% had a gastrostomy and 6.4% had cardiac surgery. 42.6% had a pH study and 55.2% had a gastroscopy. Of the total patients having a gastroscopy, about 1/4th (27.5%) had oesophagitis.

Univariate regression analysis shows having an underlying genetic diagnosis to be predictive of slow gastric emptying whereas multivariate regression analysis shows allergies especially milk and the presence of diarrhoea to be predictive of slow gastric emptying.

Conclusion:

This study reports the use of a gastric emptying scan in one of the largest reported cohorts in paediatrics. Chronic reflux in a child with an underlying genetic diagnosis should alert the clinician about the possibility of delayed gastric emptying. This is the first large scale study to show a link between delayed gastric emptying and milk allergy.

P94

A rare combination of Wilson's disease and Hyperornithinaemia-Hyperammonaemia-Homocitrullinuria (HHH) syndrome in a child; lessons learned!

Meranthi Fernando[1], Suresh Vijay[1], Sai Santra[1], Mary-Anne Preece[1], Astor Rodrigues[2] and Girish Gupte[1].

[1]Birmingham Women's and Children's Hospital NHS Trust; [2]John Radcliffe hospital Oxford

Introduction:

Wilson disease (WD) is a recessively inherited disorder of copper metabolism due to mutations of the ATP7B gene with predominantly hepatic presentation during childhood, but can have diverse clinical presentations.

Hyperornithinaemia-Hyperammonaemia-Homocitrullinuria syndrome (HHHs) is a recessively inherited, rare type of urea cycle disorder caused by mutations of SLC25A15 gene. Clinical manifestations range from mild hyperammonaemia to encephalopathy whilst liver involvement could range from mildly raised liver enzymes to acute liver failure (ALF).

Case report:

We report a 6-year-old boy who was diagnosed to have WD soon after birth by mutational analysis as parents were known heterozygotes. His WD was managed with zinc, commenced at 6 months of age due to transaminitis. Developmental delay and seizures were noted at 3 years. Extended investigations revealed hyperammonaemia with raised ornithine, orotic acid and homocitrulline in urine compatible with HHHs, confirmed by SLC25A15 mutational analysis.

At 6 years, he presented with hyperammonaemia and decompensated liver disease preceded by a viral illness and had encephalopathy. He made a full recovery from this episode with conservative management. The signs of chronic liver disease became gradually apparent afterwards with coarse liver on ultrasonography and new onset splenomegaly. At 6 yrs 3 month she presented with an episode of acute fulminant liver failure secondary to an infection and required liver transplant.

His transplant was uneventful However, he was found to be unsettled and gradually became encephalopathic during immediate post-transplant period with a normal ammonia. Subsequently he developed respiratory distress, progressive encephalopathy and succumbed to death 23 days after his transplant.

Discussion:

We report on a rare association of a combination of WD and HHHs patient who was adequately controlled with medications for both conditions. Since WD was diagnosed at birth and early treatment started, he would have expected to have a good clinical outcome. This child was diagnosed with a less severe form of HHH as this was manifested at 3 years of age, unlike the severe form which is manifested during infancy.

Explanted liver showed established biliary cirrhosis which would be unusual in his age group with clinically good control of both conditions from an early period. The combination of WD and HHH probably contributed to the histopathological progression due to the combined genetic load and possibly presence of modifier genes. The encephalopathy seen in post-transplant period could not be fully explained as liver transplantation would have resulted in a cure of both conditions. We postulate that encephalopathy was secondary to infection that contributed to ALF in pre-transplant period and was probably exacerbated in context of immunosuppression in post-transplant period.

In conclusion we would like to raise awareness about the histopathological progression of liver disease where two conditions may co-exist. Timely investigations to establish the severity of liver disease may have to be done to decide about the appropriate timing of transplantation.

P102

Review of value of annual liver ultrasound scan in Chronic Liver Disease

Rachel Pybus[1] and Palaniswamy Karthikeyan[1].
[1]Leeds Teaching Hospital NHS trust

Introduction:

Chronic Liver Disease (CLD) in children contributes to a significant proportion of outpatient appointments in Paediatric Liver service. Current monitoring of CLD is based on limited evidence and involves regular imaging by ultrasound (US) and biochemical monitoring by blood tests. At our centre it has been our practice to for children with CLD to undergo annual review with US.

Aims:

1. To assess changes detected by annual US in children with CLD and consider whether they have contributed to management decisions
2. To review the need for routine annual US.

Methods;

Retrospective data was collected from children aged 7 years or above who have undergone annual ultrasound imaging at our centre for at least two years and have CLD due to Alagille syndrome, Biliary Atresia, Autoimmune Hepatitis or Alpha-1-Antitrypsin deficiency. The local computer records were used to review all US reports, blood results and clinical findings. Two groups were defined according to whether US at 5 years of age was (A) normal or (B) abnormal. Abnormal liver architecture, splenomegaly, polysplenia, bile duct abnormalities or any vascular abnormality were classified as abnormal ultrasound findings.

Results:

37 patients were identified (8 Alagille syndrome, 9 alpha-1-antitrypsin deficiency, 20 biliary atresia.

Group A: Clinical course of patients with Normal liver ultrasound at five years of age:
Seven children (18.9%) had normal US and platelet count median 350 x109/L (316-471). Age at latest FU was median 12.72 (10.16-15.11) years. Median number of annual scans per patient was 3 (2-5). All continue to have normal US and have no clinical signs of portal hypertension or any other complication of chronic liver disease.

Group B Clinical course of patients with abnormal liver ultrasound:
Thirty patients were included in this group. The median age was 12.5 years (9.28 - 16.62) years and the median platelet count was 269 x109/L (52-476). Splenomegaly was seen in 66.7% of patients. Three patients had polysplenia. Median number of annual scans per patient was 5 (1-11).
Of these, three developed suspicious nodules on annual US requiring further axial imaging. All these nodules were benign and did not require any further treatment. No other changes on US led to clinical intervention.

Summary and conclusion:

Routine annual US led to additional significant findings warranting intervention only in children already known to have established liver disease and abnormal US appearances. In those with CLD but no US changes by 5 years of age, annual US did not identify any significant abnormalities and did not affect management.
We conclude therefore that reducing frequency of US monitoring in children with normal liver appearance at 5 years, from yearly to two yearly, may be appropriate.

P104

A case report of a child presenting with a gastrointestinal manifestation of Henoch Schonlein Purpura indistinguishable from Crohn's disease

Kirn Sandhu[1], Kirsteen Macdonald[2], Irene Scheimberg[2], Esin Karaa[2], Louise Langmead[2] and Protima Amon[1].

[1]Royal London Hospital, Barts Health NHS Trust; [2]Barts Health NHS Trust

Introduction:

Henoch Schonlein purpura (HSP) is an IgA small vessel vasculitis which is seen in 10-20 cases per 100,000 children per year. HSP is characterised by palpable purpura, arthralgia, abdominal pain and renal disease. Gastrointestinal complications associated with HSP such as intussusception, gastrointestinal haemorrhage, ileal perforation, stricture and protein losing enteropathy are encountered in 5% of patients.

History and presentation:

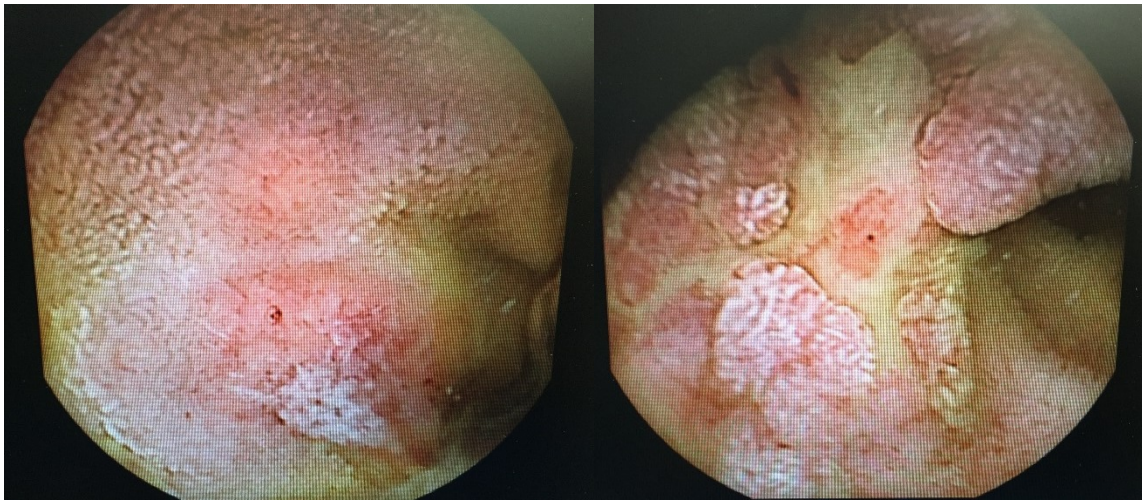
An 8-year-old girl was referred to the paediatric surgical team with a 4-day history of intermittent colicky abdominal pain and 1-day onset of bilious vomiting with no fevers, diarrhoea or weight loss. She had a viral upper respiratory tract illness two weeks earlier. She was otherwise well with no significant family history. Her abdominal pain was severe requiring gut rest and opiates.

Investigations:

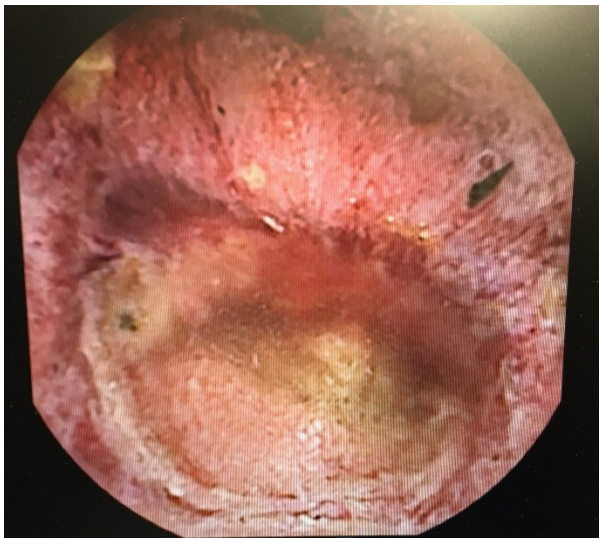
Investigations revealed raised inflammatory markers and hypoalbuminaemia. Ultrasound of the abdomen showed thickening of terminal ileum and MRI abdomen demonstrated jejunal thickening suggestive of Crohn's disease. She underwent endoscopic examination revealing gastritis and duodenitis, colonoscopy was normal. Video capsule endoscopy was performed which revealed ulcers throughout the jejunum and terminal ileum (figure 1), indistinguishable from Crohn's disease.

Figure 1: Video capsule endoscopy images depicting ulceration in the jejunum (a) and ileum (b).

(a) Jejunum showing mucosal oedema, inflammation and ulceration



(b) Ileum depicting inflammation and deep ulcers.



Clinical progress:
On day 8 of her admission she developed a symmetrical purpuric rash over both ankles leading to the diagnosis of Henoch Schonlein related ileitis. She was commenced on intravenous steroids followed by an oral tapering course. She made a good recovery with no sequelae. She was discharged from the paediatric gastroenterology team and remains well on follow up.

Summary and Conclusion:
In 10-15% cases of HSP the gastrointestinal manifestation prefaces the skin presentation as demonstrated in this case which leads to a significant diagnostic challenge to distinguish this from IBD. However, the history was short and atypical for Crohn's disease. Ileitis is a rare complication of HSP and only 2 paediatric cases have previously been reported. In both cases, children underwent exploratory laparotomy.

MDT discussion is crucial to ensure patient is appropriately managed and unnecessary surgical intervention avoided. In this case paediatric surgeons, gastroenterologist, radiologists, histopathologists, and general paediatricians were all involved. HSP should be considered as a differential diagnosis in children presenting with an acute abdomen with terminal ileitis.

P105

1 year clinical outcome of a new tertiary joint paediatric GI allergy clinic in Wessex, UK

Esme Poole[1], Maryam Hussein[1], Vinod Kolimarala[1], Sally-Ann Denton[1], Joan Gavin[1], Mary Halsey[1], Emma Grainger-Allen[1], Mich Lajeunesse[1], Efrem Eren[2] and Nadeem Ahmad Afzal[1].
[1]Southampton Children's Hospital, Paediatrics, Southampton, United Kingdom
[2]Department of Allergy and Immunology, University Hospital Southampton

Background:
Children with multiple allergies, especially those with considerable gut symptoms despite being on restricted diets, remain a challenge to manage. This is quite often due to the paediatrician trying to impact on general symptoms, functional gut symptoms and food exclusions all at the same time. To manage an increasing number of regional referrals of such children, we formulated a new joint paediatric allergy and GI clinic in Wessex. This clinic is supported by two consultants, GI/Allergy dieticians, specialist nurses and a clinical psychologist.

Aim:
We aim to describe the demographics and symptoms of children seen in the last two years in this clinic. We also aim to assess clinical outcome at 1 year follow up.

Methods: All children referred to the joint clinic between December 2016 and October 2019 were included in the study. Details of demographics, presenting symptoms and growth centiles were collected. Clinical outcome at 1 year follow up was also assessed.

Results:
43 children were recruited to the study. Referrals came from 12 hospitals, with 65.1% of referrals from regional hospitals in Wessex. The patients had previously been seen by general paediatricians, paediatric allergy and GI consultants with little clinical progress before being referred to this new joint clinic for multi-professional team input.

At first appointment, the mean age was 6.9 yrs (SD: 4.48) with 27/43 (62.8%) males. The mean Z score for height was -0.47 (SD 1.09) and for weight - 0.30 (SD 1.4). Patients generally had considerably restricted diets with a median of 3 food exclusions (IQR: 2-5); milk, soya and wheat/gluten being the top 3. Despite these food exclusions, patients had significant GI symptoms with the most common symptom being constipation (62.8%), followed by abdominal pain (58.1%) and vomiting/reflux (48.8%). Other diagnoses included anaphylaxis (18.6%), Eosinophilic Oesophagitis (20.9%), allergic rhinitis (16.3%), asthma (14.0%) and eczema (20.9%).

During the course of follow up, 15 children had x-rays confirming stool loading in 86.7%; treated as functional constipation with laxatives. 13/17 showed mucosal abnormality on endoscopy of which 9 were diagnosed to have Eosinophilic Oesophagitis.

At 1.01 years (SD 0.81) follow up (n=27), there was no significant change in height (z=-0.48 to z=-0.71) and weight (z=-0.38 to z=-0.59). The team was able to impact on improving the diets with a significant drop in number of food exclusions from 3 to 2 (p=0.019). In addition, there was a significant reduction in constipation and reflux symptoms. There was, however, no impact on abdominal pain.

Conclusion:
This study highlights the benefits of a specialised MDT positively impacting on reduction of symptoms and decreasing burden of food exclusions in multi-allergic children. This is where paediatricians and GI/Allergy specialists had previously failed to impact on clinical management in their individual clinics. The improvement in symptoms despite dietary relaxation alludes to a more functional cause in this group of patients. Despite effective management strategies, surprisingly little impact was made on abdominal pain. We believe management of abdominal pain in this group of children remains complex and multi-factorial, making it a management challenge.

P106

A rare case of Multiple Intestinal atresia with Combined Immuno-Deficiency

Sripriya Eachempati[1].
[1]West Suffolk Hospital

Introduction:

Hereditary multiple intestinal atresia (HMIA), the rarest form of recurrent multiple atresia, was first reported by Winter and Zeltzer in 1956. HMIA involves multiple atretic lesions along with homogenous intraluminal calcifications. The immune deficiency affects T- and B-cell functions, with lymphopenia, agammaglobulinemia, and impaired mitogen responses. Death occurs before 2 years of age in most patients. Clinicians face a difficult situation as the severity of the intestinal presentation and risk of severe and opportunistic infections precludes long-term survival.

Case report:

We report a rare case of Multiple Intestinal atresia with Severe combined Immunodeficiency in a term baby who was antenatally diagnosed with small bowel atresia. Her postnatal abdominal ultrasound on Day 0 revealed biliary dilatation and small bowel obstruction. Her contrast study on Day 0 showed obstructed first part of duodenum, dilated loop reaching proximal jejunum and collapsed distal bowel. She was transferred to quarternary centre for ongoing liver and surgical management. In the quarternary centre her Ultrasound was suggestive of duodenal atresia with possible antenatal perforation. On day 2 of life she had an exploratory laparotomy which featured multiple atresias at pylorus and small bowel including ascending colon. Colonic lumen was absent.

Surgery: She had small bowel enterotomy, Jejunostomy, Pyloroplasty, Tube jejunostomy on day 2.

She is now left with ultra short gut, high stoma, multiple atresias and liver disease, needing lifelong parenteral nutrition or Intestinal transplant. She is on parenteral nutrition with a very small amount of enteral feeds. She is not for live vaccines or BCG. She is eligible for Paluvizumab. She is nursed in isolation with strict infection control measures. This obviously affects her development as she has never seen any faces. She is on prophylactic antibiotics and antifungals. Her best interests are in receiving PN at home, distancing her from antibiotic resistant organisms that she can acquire nosocomially.

Her sister who was born premature at 36+1 weeks had malrotation with Ladd's bands and complete gastric outlet obstruction. She had sudden loss of cardiac output during laparotomy and despite active CPR sadly passed away on the operating table.

Investigations:

Suspicion of MIACID was raised and TTC7A gene was sent and was discussed with quarternary centre Immunology as SCID. This was confirmed on day 8 of life. She had absent CD8 and some CD4 cells. She was deemed to have severe end of SCID. The Immunologist has discussed with parents and started the process of tissue typing for bone marrow transplant which can take several weeks.

Conclusion:

Hereditary multiple intestinal atresia (HMIA), a presumed autosomal recessive disorder, is an unusual and rare form of recurrent intestinal atresia which can be associated with severe combined immunodeficiency (SCID). The combination of HMIA and SCID is invariably lethal. This obviously has implications for any further siblings and would necessitate genetic counselling. This disease has a high burden in terms of infection control, lifelong parenteral nutrition, bone marrow transplant and/or intestinal transplant.

P107

Clinical outcomes of abnormal gastric emptying in 182 children at 2 years follow up

Maryam Hussein[1], Esme Poole[1], Vinod Kolimarala[1], Mich Lajeunesse[2], Efrem Eren[3], Francis Sundram[4] and Nadeem Ahmad Afzal[1].

[1]Department of Paediatrics, Southampton Children's Hospital; [2]Department of Paediatric Allergy and Immunology, Southampton Children's Hospital; [3]Department of Allergy and Immunology, University Hospital Southampton; [4]Department of Nuclear Medicine, University Hospital Southampton

Introduction:

Although gastric emptying is well documented as a cause of gastroesophageal reflux in children, little is known about the outcome of children with delayed gastric emptying. We report a 2-year outcome of delayed gastric emptying in a large cohort of children in Southampton.

Aims:

To assess clinical outcomes of children with abnormal gastric emptying scan after 2 years at Southampton Children's Hospital.

Methods:

Children aged 0-18 years with gastrointestinal symptoms (90.8% vomiting/reflux, 26.6% constipation, 11% nausea, 21.7% abdominal pain & 3.9% diarrhoea) who had gastric emptying scan between 2009 and 2018 at Southampton Children's Hospital were identified. Those with reported abnormal gastric emptying scan were included in the study. Patient's demographics, underlying diagnosis, symptoms, growth data and medications taken at the time of the scan were recorded. Clinical symptoms, medications and growth data were re-checked at 2 years follow up.

Results:

A total number of 285 children underwent gastric emptying scan of which 182/285 (63.9%) children were reported to have an abnormal gastric emptying scan and were recruited to the study. The mean gastric emptying time was 82.9 minutes (SD=75.3).

In this cohort of patients with an abnormal gastric emptying scan, 90.6% (n=258) had symptoms of vomiting and acid reflux, 31.3% constipation, 22.5% abdominal pain, 10.6% nausea and 4.5% had diarrhoea at presentation. A review of treatment charts shows 65.9% to be on anti-reflux therapies. Of the total number, 54.9% were treated with proton pump inhibitors, 14.5% with Ranitidine, 8.7% with Gaviscon, 35.8% with Domperidone and 3.5% with Erythromycin. 38.2% of the patients were on concomitant laxatives.

Patients were followed up for a median of 22 months (IQR 14-25 months). At the final follow up appointment although there was an increase in height, the change was insignificant (z=-1.19 to z=-1.09). A similar non-significant increase was seen in weight (z=-1.02 to z=-0.85).

However, a significant reduction in symptoms of vomiting/gastroesophageal reflux (p=0.000), nausea (p= 0.02) and abdominal pain (p=0.013) were seen with no significant change in symptoms of diarrhoea (p=ns). Parallel to the significant clinical improvement there was a significant reduction in the prescription of medications which included Domperidone (p=0.001), Proton pump inhibitors (p=0.002) and Gaviscon (p=0.001) but not Ranitidine, Erythromycin or Laxatives (p=ns).

Using improvement in reflux symptoms as a proxy for improved gastric emptying; multivariate regression analysis of symptoms shows gastric emptying to improve with the presence of underlying allergies (p=0.026). Additional diagnoses such as developmental delay, underlying neurological diagnosis, presence of a genetic condition, presence of oesophagitis at endoscopy and history of

previous surgery failed to reach significance (p=ns).

Conclusion:
This is the first longitudinal study looking at clinical outcome in a large cohort of children with delayed gastric emptying. Although symptoms of vomiting/reflux did not improve in children with genetic and neurological conditions or those who had previous surgery, the outcome remains good in children with underlying allergies. The paediatrician's clinical decision in reducing prescriptions of reflux medications based on improvement in symptoms at 2 years reflects good practice.

P108

Anorectal Manometry in Children with Defaecation Disorders BSPGHAN Motility Working Group Consensus Statement

Eleni Athanasakos[1], Stewart Cleeve[1], Nikhil Thapar[2,3], Keith Lindley[2], Steve Perring[4], Hannah Cronin[2], Osvaldo Borrelli[2] and Mohamed Mutalib[5].

[1]The Royal London Hospital, Barts Health NHS; [2]Great Ormond Street Hospital; [3]UCL Great Ormond Street Institute of Child Health; [4]Poole General Hospital;[5]Evelina London Children' Hospital

Background:

Chronic Constipation and Functional/Structural Faecal Incontinence (CCFSFI) is common (0.5% - 30%), debilitating, the pathophysiology is poorly understood, investigations and treatment are limited and the prognosis is poor. Anorectal manometry (ARM) and high resolution manometry (HRAM) are the gold standard investigations in adult patients with proven diagnostic and therapeutic benefits. Unfortunately early standardisation in adult ARM/HRAM did not occur and has resulted in significant variations in practice and reporting, hampering progress in research and collaboration. The BSPGHAN Motility Working Group (MWG) aims to encourage standardisation by producing a detailed consensus on the use of ARM/HRAM in children. It is anticipated that there will be incremental refinements to the consensus over time.

Methods:

The authors conducted a literature search using PubMed between 2004 -2018 for publications in English using all possible combinations of the following keywords: (1) 'anorectal', 'rectoanal', 'malformat*', 'Hirschsprung*', dyssynerg*', 'constipat*'. (2) 'manometr*', 'physiolog*', and (3) 'paediatric*', 'pediatric*', 'child*', 'neonat*'. The literature search formed the basis of the following questions, which were discussed and answered, by members of the MWG that attended the consensus meetings:

1. What are the indications for performing ARM in children?
2. Should ARM in children be performed awake or under sedation?
3. How should children be prepared for ARM?
4. What catheter should be used to perform ARM in children?
5. How to perform ARM in children?
6. How should ARM in children be analysed and reported?

Results:

Consensus was reached if more than 80% of the working group members voted (>5 on a Likert scale 0 to 9). A consensus was reached for all of the questions 1 to 6. Below is a synopsis of the outcomes:

1. Indications: CCFSFI unresponsive to medical management, diagnosis (including anal sphincter injury), to guide treatment efficacy, assessment of congenital anomalies (e.g. anorectal malformations), assessment of defaecation dynamics (qualitative) and screening for Hirschsprung disease.
2. All patients should be offered ARM/HRAM awake rather than under sedation.
3. A pre-test screening interview should be carried out, informed consent is mandatory, bowel preparation is recommended.
4. Water perfused or solid state catheters, ARM or HRAM can be used.
5. A detailed, stereotyped protocol should be used in all patients.
6. A detailed report of quantitative values and qualitative interpretation should be provided in all patients. Normative values in children remain extremely sparse.

Conclusion:

BSPGHAN MWG has reached consensus on the use of ARM/HRAM in children with CCFSFI. The group looks forward to reviewing the usefulness of the consensus (national survey or audit) and demonstrating its utility in collaboration and research.

P110

IgE mediated food allergy to gastrostomy administrated blended diet in children with neurodisability

Jamie Motion[1], Iain Chalmers[1], Kathleen Ross[1], Tracy Cameron[1], Michelle Brown[2], Shyla Kishore[1], Gillian Vance[3] and Susan Bunn[1].
[1]NHS Grampian; [2]NHS Highland; [3]Great North Children's Hospital, Newcastle

Objectives and Study:

In children with complex neurodisability and foregut dysmotility gastrostomy administered blended diet is often better tolerated than commercial liquid nutritional therapy(1) and has recently been recognised and supported by the British Dietetic Association(2). We present 3 children with IgE-mediated allergy to blended diet as a new, unexpected complication.

Methods:

Retrospective review of clinical notes and further interview of parents/carers.

Results:

In a defined geographic area of Scotland there are currently 22 children with neurodisability receiving blended diet of which 3 have presented with an IgE mediated allergy to blended food administration.

Case 1: 14 yr boy with cerebral palsy (CP) secondary to hypoxic ischaemic encephalopathy (HIE). GMFCS (Gross Motor Function Classification System) level V and severe bowel dysmotility requiring fundoplication, gastrostomy, jejunal feeding and, subsequent to colonic distension, ileostomy formation. Blended diet initiated with good clinical effect but first exposure to cow´s milk and subsequently to wheat both induced anaphylaxis; that to wheat requiring 3 doses of IM adrenaline. Testing suggested egg allergy as well as confirming milk and wheat sensitisation (table 1).

Case 2: 6 yr girl with epileptic encephalopathy, severe neurodisability and feed intolerance. Liquid ketogenic diet achieved good epilepsy control but increased vomiting so she was switched to blended ketogenic diet with improved tolerance. First exposure to egg resulted in anaphylaxis requiring IM adrenaline. Testing confirmed egg allergy (table 1).

Case 3: 6 yr girl with CP secondary to HIE, GMFCS level V with severe feed intolerance and malnutrition despite jejunal feeding. On initiating blended diet her parent recalled an urticarial rash and facial swelling when egg white was whisked nearby. Allergy testing confirmed egg allergy and likely peanut allergy (table 1).

A retrospective allergy focused history was taken from each family (table 1). All children had received standard infant formula early in first year of life and then liquid nutritional therapy - exclusive amino-acid based feed (case 2 & 3) and peptide-based feed (case 1), until initiation of blended diet.

Table 1 – Allergy focused history, IgE and skin prick test results for 3 cases

Conclusion:
Blended diet can offer significant benefits to feed tolerance in children with complex neurodisability (1) and its use has increased markedly over the last 2 years (3). We highlight an important complication which is preventable by recognition prior to exposure. We hypothesise that these children are at increased risk of food allergy due to their lack of usual early life enteral exposure to foods necessary for promoting oral tolerance. Risk of a major allergic reaction is increased by direct administration of allergen into the stomach, bypassing the protective oral recognition of allergy and commonly a fundoplication preventing vomiting. Additionally, risk of adverse outcome is higher due to their fragility and comorbidities. In all 3 cases an allergy focused history would have identified risk factors.

	Retrospective allergy focused history	Relevant specific IgE (kU/L) (<0.35 kU/L negative)	Skin prick test (≥3mm positive)
Case 1	Personal hay fever First degree relative – fruit-pollen syndrome	Cow's milk 4.66 Egg white 9.16 Egg yolk 6.76 Wheat 90.8	
Case 2	Mild eczema Half sibling – egg and peanut allergy	Total egg 51.8	
Case 3	Personal hay fever Skin sensitivity to grass	Egg white 2.21 Nut screen 8.36	Egg 6mm Peanut 4mm Cashew 2mm

We recommend a careful allergy focused history in all children initiating blended diet and, if positive, allergy testing prior to starting high allergenic foods.

References:
(1) Nutr Clin Pract. 2019 Sep 24. doi: 10.1002/ncp.10406
(2) www.bda.uk.com/
(3) N-P-035 ESPGHAN 2019

P112

Severe erosive gastritis due to ingestion of liquid from chemiluminescent plastic rods (glow sticks)

Shehriyar Khan[1], Rohit Gowda[1] and Bimal Bhaduri[1].
[1]Maidstone and Tunbridge Wells NHS Trust

Introduction/Background:

We present a case report of a 4-year-old child who presented to our gastroenterology team with ongoing episodes of vomiting and abdominal pain after ingesting liquid from chemiluminescent plastic rods (glow sticks).

Aim:

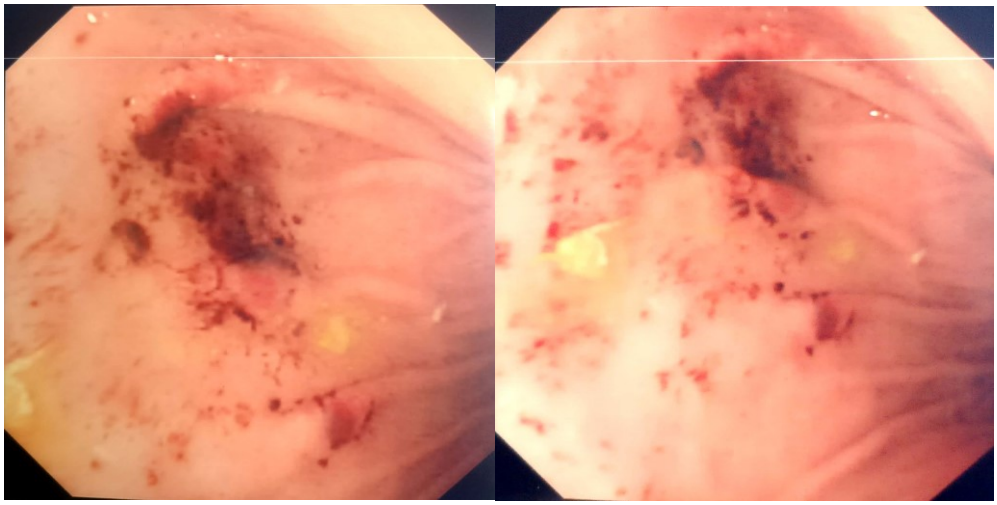
To raise awareness about the severity of accidental ingestion of liquid from these chemiluminescent rods. There is only one published case series reporting no significant symptoms and only transient symptoms to site of exposure. Our patient presented with significant ongoing symptoms and severe erosive gastritis on gastroscopy.

Subject and Methods:

A 4-year-old female child presented to the emergency department with intermittent episodes of vomiting and abdominal pain after accidental ingestion of liquid from two glow sticks which she was using as a bracelet. The ingested amount was estimated to be about 10-15 mls and such chemiluminescent glow sticks contain hydrogen peroxide, oxalate ester (e.g. diphenyl oxalate) and a fluorescent dye. Glow sticks previously contained dibutyl phthalate (luminescer) and dimethyl phthalate (activator). There was no significant past medical or medication history.
She bit on the cap at the end of the stick, ingested the liquid and drank water with it. Mother found her with the empty sticks and she later on started vomiting which prompted her presentation to the emergency department. Baseline bloods were done which showed haemoglobin of 124 g/L. She was sent home after a period of observation and successful fluid challenge. She represented after a week with ongoing episodes of vomiting at home. An urgent oesophagogastroduodenoscopy (OGD) was booked and she was commenced on omeprazole for symptomatic relief. In the meantime, a barium swallow was performed which did not show any evidence of oesophageal stricture, holdup or gastric outlet obstruction.

Results:

The OGD showed severe erosive gastritis in the body of the stomach. The patch of gastritis was not well demarcated and showed some minor ulceration on the edges. The oesophagus and duodenum appeared normal. Haemoglobin was repeated which dropped to 103 g/L over a period of 8 days from the previous one.



Summary and Conclusion:

There is no case report (paediatric or adult) of erosive gastritis on an OGD leading to severe ongoing symptoms after ingestion of material from such sticks. This material is very irritating to the oral mucosa therefore it is not normally ingested in significant amounts. Our patient ingested about 10-15 mls because she took water with it, which helped it to go down to the stomach and cause the significant gastritis. Her abdominal pain and vomiting improved with omeprazole therefore we would suggest treatment with Proton pump inhibitors or an H2 receptor antagonist. In severe/ongoing symptoms, we would suggest an urgent OGD to rule out oesophageal or gastric pathology. From the public health perspective, such items should have a warning label about the chemicals inside it and not to be ingested or exposure to skin and eyes.

P114

Thiopurine drug monitoring in paediatric patients with autoimmune liver disease

Zuzana Londt[1], Jonathan Hind[1], Anil Dhawan[1], Dino Hadzic[1] and Marianne Samyn[1].
[1]King's College Hospital

Introduction:

Autoimmune liver disease (AILD) is a chronic hepatitis of unknown aetiology, characterized by presence of high immunoglobulin levels, circulating autoantibodies and particular changes on liver histology. Induction of treatment is with prednisolone however azathioprine is commonly introduced as a second line agent and titrated up in order to achieve remission. Routine monitoring of the active metabolites of azathioprine, Thioguanine nucleotides including 6-thioguanine nucleotide (TGN) and 6-methylmercaptopurine (MMP) is routinely done in patients with inflammatory bowel disease however experience in children and young people (YP) with AILD is limited.

Aim and Objectives:

The aim of the study was to evaluate TGN levels in a group of patients with AILD attending a multidisciplinary clinic.

Subjects and Methods:

A retrospective, single centre review of a cohort of YP diagnosed with AILD between 2012-2017 that had TGN and MMP levels measured. TGN levels between 235-450 pmol/ 8x10^8 cells are considered to be therapeutic in our laboratory and MMP/TGN ratio ≤ 11 recommended. All patients were treated with a combination of prednisolone and azathioprine. Those with associated inflammatory bowel disease were excluded. Demographic and clinical data including laboratory values (white cell count (WCC), platelets, Aspartate aminotransferase (AST) and Alanine aminotransferase (ALT) at time of TGN levels) were collected. Statistical analysis was carried out using descriptive statistics, Mann-Whitney U and Spearman correlation with p value <0.05 considered to be significant.

Results:

Forty-one TGN levels were measured in 17 patients (9 female) diagnosed with AILD at a median age of 15 (range 12-17) years. Fifteen (88%) patients had autoimmune hepatitis and the remainder had autoimmune sclerosis cholangitis. Treatment was induced with prednisolone and azathioprine treatment was added at the median of 2.5 (range 1-19) months after the initial diagnosis. The median dose of prednisolone was 12.5 (0.25mg/kg, range 5-30) mg when azathioprine was introduced. TGN levels were collected between 0.5-48 (median 8) months after the start of treatment. Median (IQR) values were as follows: AST 45 (24,72) IU/l, ALT 43 (21,98) IU/l, TGN 265 (178,369) pmol/ 8x10^8 cells, MMP 511 (264,1043) pmol/ 8x10^8 cells, MMP/TGN 2 (1,4) and azathioprine dose/kg bodyweight 1.7 (1.3,2.0) mg/kg. No significant correlation was found between AST, ALT and TGN levels respectively. TGN levels were therapeutic in 46% and below and above therapeutic range in 39% and 15% respectively. Interestingly, median azathioprine dose/kg was similar for those with sub-therapeutic and therapeutic levels (1.7 mg/kg) and could suggest suboptimal adherence to treatment in the group with low TGN levels. Self-reported adherence of < 80% in keeping with non-adherence was recorded for 20% (n=8) samples taken with abnormal AST in 75% and therapeutic TGN levels only in 25%.

Summary and Conclusion:

Although our study could not find the reason of treatment failure based on TGN levels. The TGN level monitoring helped in selected patients to tailor their treatment and also identified non-adherence in some of the individuals.

P116

Finger-lickin' good? : An evaluation of a pilot intensive messy play programme for severe oral sensory sensitive children

Amanda Harvey[1], Kate Jones[1], Chris Smith[1], Michael Hii[1] and Assad Butt[1].
[1]Royal Alexandra Childrens Hospital, Brighton

Introduction:

Oral Sensory Sensitivity (OSS) leading to significant feeding issues is rare, but complex medical conditions and negative disease-based experiences can increase the risk. This can negatively impact on many areas including emotional and social development and growth. Severity can vary but use of feeding tubes may become necessary further exacerbating issues. Progress to a more normal intake can be slow and families are often frustrated with the impression of little or no progress over long periods of time. Our centre developed a short intensive intervention messy play course for children with significant OSS.

Aim:

To evaluate the impact of an intensive intervention course using a detailed scoring system investigating 7 key areas.

Method:

Children with recognised established OSS were identified through dietetic and Speech and Language therapist (SALT) caseloads. Parents were invited to attend a series of weekly sessions over a 6-week block. The programme developed had a progressive hierarchy structure with graded sensory experiences involving desensitisation to food. It followed typical sequential feeding development using a range of tastes and textures. Parents completed a pre-course sensory sensitivity questionnaire covering 7 key categories (Food approach/Behaviours at meal, Food approach/Time, Range of foods, Sensory issues, Tactile/messy response, Ability to taste new foods, and Behavioural reaction to foods). Each of these categories had a scale score ranging from 1 (normal) to 5 (profound problems). On completion of the course, families repeated the questionnaire to identify any significant or subtle change in the 7 areas.

Results:

Complete data was available for 7 children (4 male 3 female, age range: 3 years 3 months to 8 years 7 months) who completed the course. Pre-intervention results showed highest scoring (worse) areas of sensitivity as food approach behaviour, range of foods and behavioural reactions.

As a group, using the total score from each category, an overall improvement was only seen in 1 category (Food approach/time), no difference was seen in 4 and worse in 2. However, looking at the results individually, the range of changes within all the categories shows a more varied picture (Table1). Improvements were seen in tactile/messy response, food approach/time and ability to taste new foods. A negative impact was seen in behavioural reaction to foods (worse score in 4/7 children). On an individual level, every child showed at least 1 improvement in 1 category.

	Behaviour category						
	Food approach /Behaviours at meal	Food approach /Time	Range of foods	Sensory issues	Tactile/ messy response	Ability to taste new foods	Behavioural reaction to foods
Number of pts who's score showed improvement	0	3	0	1	4	3	2
Number of pts who's score showed no difference	5	2	7	3	1	3	4
Number of pts who's score got worse	2	2	0	3	2	1	1

Table 1

Discussion:
Improvements in children with long established OSS will often be slow and subtle. However, our short-term intervention showed a positive impact on at least two areas of behaviour and identified areas in which the interventions were unhelpful. Worsening scores could be explained by the potential negative experience of the intervention, which is likely challenging for such a group. We speculate that longer term, regular interventions would likely yield better outcomes.

Conclusion:
Detailed intervention outcome questionnaires are a useful tool to identify small, subtle but important positive steps for the child, family and health professional in OSS. Information yielded can help tailor future interventions according to individual patient responses in this challenging group of children.

P117

Disaccharidase Deficiency: Clinical Characteristics, Treatment and Outcomes

Eve Moreland[1], Jennifer Morris[1], David Wands[2], Richard Russell[2]
[1]University of Glasgow, Medical School; [2]Royal Hospital for Children, Glasgow

Objectives and Study:

Disaccharidase deficiency, and the resulting carbohydrate malabsorption, is considered a potential cause of many presenting gastrointestinal symptoms in children. Small bowel biopsies assayed for disaccharidase activity remains the gold standard for confirming diagnosis. The clinical significance and interpretation of abnormal results has not yet been fully characterised while data analysis on patient response to treatment, particularly enzyme replacement, is lacking. The aim of this study was to examine our patient cohort and evaluate their clinical characteristics, results of investigations and response to treatment.

Methods:

This study examined all patients in our centre who underwent upper GI endoscopy with biopsies assayed for disaccharidase activity over a 9-year period. Data was collected retrospectively from patient's electronic medical records. The data set included patient demographics, presenting features, clinical characteristics and biopsy results. The patient's clinical course and response to treatment was then examined. Emphasis was put on those who received specific treatment as current data lacks a thorough review of its efficacy. Invertase is an industrial enzyme derived from yeast and hydrolyses sucrose into glucose and fructose.

Results:

A total of 245 paediatric patients were included. 70 (29%) patients presented with diarrhoea, 41 (17%) with abdominal pain, 21 (8%) with bloating, 16 (7%) with vomiting and 76 (31%) with various other symptoms. Overall, 154/245 patients (63%) had normal disaccharidase levels while 91/245 (37%) had abnormal levels; 61 of which (67%) were male. Out of 91 abnormal biopsies, 62 (68%) had normal small bowel histology. The majority of patients had pan-disaccharidase, lactase or sucrase-isomaltase deficient while a variety of other combinations existed. Those with lactase deficiency were often managed by a dairy free diet or lactase supplementation, some were self-resolving with no treatment. 15 patients with either sucrase-isomaltase, lactase-maltase or a pan-disaccharidase deficiency were managed with Invertase, with a mean effective dose of 0.4mls/kg/day. Invertase outcomes included 5 (33%) patients with resolved symptoms, 5 (33%) patients with improved symptoms, 3 (20%) patients showing no response, 1 (7%) with worsening symptoms and 1 (1%) patient with an unknown outcome to treatment. In total, there were 4 patients with congenital sucrase-isomaltase deficiency identified and symptoms fully managed with Invertase.

Conclusion:

Carbohydrate malabsorption should be considered in the differential diagnosis of children presenting with a range of gastrointestinal complaints. In this study, while the majority had normal disaccharidase levels a significant proportion had a disaccharidase deficiency. In those with abnormal results there is an important subpopulation that respond to enzyme supplementation. In this tertiary paediatric hospital Invertase was successfully used providing a significant cost-saving compared to other enzyme replacements available.

Dr Falk
Award
IBD Posters

P1

Alopecia areata in children with ulcerative colitis: extra gastrointestinal manifestation or medicine side effect?

Hermione Race[1], Akshatha Mallikarjuna[1], Rohit Gowda[1] and Bim Bhaduri[1]
[1]Maidstone and Tunbridge Wells NHS Trust

Introduction:
The hair loss associated with inflammatory bowel disease is of three varieties i) telogen effluvium, ii) alopecia areata (AA) and iii) primary cicatricial alopecia. Telogen effluvium normally occurs on the top of scalp and is exacerbated by nutritional deficiencies, stress and adverse effects of treatment. However, AA is predominantly a T-lymphocyte-mediated autoimmune disease resulting in either patchy scalp hair loss or universal alopecia.

Aims and objectives:
To highlight the possible aetiology of hair loss in children with ulcerative colitis and review the current literature.

Subjects and methods:
A 13-year-old boy presented with bloody stool for six months. Investigations showed raised fecal calprotectin and mild anemia. Endoscopy showed mucosal erythema, granularity and superficial ulcerations involving the right side of the colon. Histopathology showed active chronic pancolitis with cryptitis and crypt abscesses. The child was diagnosed with ulcerative colitis and started on mesalazine. After five months of treatment, he presented with patchy hair loss in right parieto-occipital scalp. His mesalazine was stopped and dermatology referral made.

Results:
Patches of hair loss showed balding with inverted exclamation mark hairs typical of AA. There was no ophiasis, other body hair or nail involvement. He was commenced on treatment with intralesional corticosteroids and mesalazine restarted.

Summary:
Historically, hair loss is commonly reported in patients with inflammatory bowel disease, with recent prospective studies of adult patients reporting a much higher prevalence than previously suggested of around 33%. The causes of hair loss have been linked via case reports to autoimmune disease, stress, and nutritional deficiencies including iron and B12, as well as medications used to treat IBD such as Mesalazine. As such, Mesalazine was stopped in this patient pending dermatological review. However, recent evidence suggests that AA is less common in patients with UC treated with Mesalazine or Infliximab. Furthermore, Mesalazine has been reported by dermatology journals as a relatively new therapeutic option for steroid resistant AA. Following dermatological review, the patient was commenced on treatment with intra-lesional corticosteroids and Mesalazine was restarted.

Conclusion:
There are multiple reasons for alopecia in IBD patients. In UC patients, the prevalence of AA is significantly higher than in general population and logically can be linked via autoimmune disease. Recent evidence suggests Mesalazine may be a treatment rather than a cause of hair loss in AA and therefore early dermatological reference should be sought to correctly diagnose and treat it swiftly. Further studies are required to fully elicit the prevalence, pathogenesis and treatment of AA in children with UC.

P5

Evaluation of a managing exam stress and IBD group in a pediatric gastroenterology service

Sarah Densham[1], Dr Rachel Tyler[1] and Dr Sophie Velleman[1].
[1]Bristol Royal Hospital for Children

Introduction/background:

An increasing number of young people with IBD were reporting difficulties in managing stress in school, particularly around revision and exams, to the pediatric gastroenterology team.

Aim:

To address this need, the service designed and provided a brief intervention for young people aimed at increasing their confidence in managing their IBD in school, being more prepared for exams and managing stress during exams.

Method:

The group was facilitated by a Clinical Psychologist and a Trainee Clinical psychologist over 4 hours. The group drew from cognitive-behavioural principles using a combination of didactic and interactive exercises, including: psycho-education about stress, introduction to CBT model, relaxation and mindfulness techniques practiced in vivo, behavioural strategies, managing unhelpful thoughts. Handouts were provided as an aide memoir. Participants completed idiosyncratic measures at pre-group and post-group time points to evaluate the group. Ratings were from 0 - 10 where higher scores reflect higher levels of stress and confidence. Young people (N=34) in year 10, 11, 12, 13 at school were invited to attend the group. Females between the ages of 15 and 17 attended the pilot group (N=3).

Results:

Results demonstrate the group effectively increased confidence in managing stress during exams, feeling more prepared for exams and managing IBD at school. Results show that attending the group led to reductions in feeling stressed about exams. Overall, participants rated the group average 9 for helpfulness, 9 for relevance, 9 for 'group fit', 8 for goal attainment. Qualitative feedback from participants included: "The most helpful aspect was talking about how we can look after ourselves, sometimes I forget that" and "meeting other people who have IBD was really good because it makes you realise that you aren't the only one struggling". Please see attachment for tabular depiction of results.

	Pre-group	Post-group
How prepared do you feel for your exams?	4	7
How confident do you feel about managing stress during exams?	3	8
How stressed do you feel about exams at the moment?	5.5	3
How confident do you feel about managing IBD at school?	4	7

Table 1. *Pre-group and post group mean scores*

Summary/Conclusions:

The service aim was to provide a brief intervention for managing exam stress and IBD. Despite the low number of participants, the results demonstrate that the group effectively increased confidence in managing stress during exams, managing IBD at school, being more prepared for exams and reduced exam stress. Participants found the group helpful, relevant and to be a good fit. These experiences were corroborated by qualitative feedback from participants. Potential Particular identified reasons for low group attendance related to the timeliness and timings of the group. A leaflet describing the group will be provided in greater advance for age-appropriate patients, to facilitate a more timely intervention and alternative times to run the group (e.g. evenings) will be considered.

P9

Optimising anti-TNF through treatment drug monitoring (TDM) in children with an early diagnosis of inflammatory bowel disease

Marco Gasparetto[1], Natasha Burgess[1], Qamar Hussein[1], Sandhia Naik[1], Ian Sanderson[1] and Protima Amon[1].

[1]Barts Health NHS Trust, The Royal London Children's Hospital, Department of Paediatric Gastroenterology

Background:

Anti-TNF agents are recommended for induction and maintenance of remission in children with active inflammatory bowel disease (IBD) despite optimal use of other recognised treatments. In the recent years, therapeutic drug monitoring (TDM) has allowed early measurement of drug and anti-drug antibody concentrations helping to rationalise patient management. We hypothesized that in very young children the trough level may deviate from that expected using doses calculated according to body weight.

Aim:

We reviewed the dose effectiveness of anti-TNF in a population of children with IBD diagnosed at 10 years of age or below, by looking at their need for dose escalation based on trough level (TDM) and/or clinical and biochemical response. We examined if the infliximab (IFX) dose of 5 mg/Kg were calculated per surface area, to what extent would this childhood dose differ from the dose per surface area given to an adult (receiving 5 mg/Kg).

Subjects and Methods:

Retrospective review of prospectively collected data available from the hospital electronic patient records. Body surface area was calculated using the Du Bois formula. Adult standard body surface area used as reference was 1.9 m2 for men (Wt 70.5 Kg, Ht 175.3 cm) and 1.6 m2 for women (Wt 57.6 Kg, Ht 161.6 cm).

Results:

A total of 38 children (M=24) were diagnosed with IBD at the age of 10 years or below in our Department over the past 10 years. Twenty-five had Crohn's disease (CD), 12 ulcerative colitis (UC) and 1 IBD-unclassified. Age at diagnosis was 7 years +/- SD 2 (2-10). All children were started on 5 mg/Kg/dose of IFX, which corresponded to 116.1 mg/m2 +/- SD 25.86 (78.6 – 184.3) for males and to 106.23 mg/m2 +/- SD 22.69 (76.6 – 150.3) for females. This resulted to provide only 50-75% of the dose requirement by body surface area based on 18-year old UK men and women population data (185.5 mg/m2 and 180 mg/m2 respectively).

Twenty-six children needed escalation to 10mg/Kg of IFX. Indication for escalation was based on low level only (n=9), symptoms only (n=3) or a combination of low level and symptoms (n=14). The time to dose escalation from start of IFX was 8.62 months +/- SD 11.42 (2-50).

Four patients were primary non-responders to IFX, 1 had anaphylactic reaction. Twenty-two patients were escalated to other biologics, including 12 of those on escalated IFX regime.

Eight patients had IBD related surgery (4 CD and 4 UC) at 31.25 months +/- 19.6 (17-66) from diagnosis.

Overall, the number of patients who maintained remission on IFX (standard or escalated dose) was 13.

Summary and conclusion:
Our findings suggest that a standard IFX dose of 5 mg/Kg in children diagnosed under 10 years of age may not meet their requirements if they had been based on body surface area. TDM is therefore particularly recommended in this age group for early proactive dose optimization. Consideration could be given to prescribing biologics according to surface area in young children as happens with a number of other drugs.

P23

Inflammatory Bowel Disease and Takayasu Arteritis: A Case Report.

Rachel Pybus[1], Veena Zamvar[2], Joanne Sims[1], Nasim Tahir[2] and Mark Wood[2].
[1]Bradford NHS Teaching Hospitals Trust; [2]Leeds NHS Teaching Hospitals Trust

Background:

Takayasu arteritis (TA) is extremely rare, especially in childhood, and its occurrence with inflammatory bowel disease (IBD) is even rarer. There have been suspected associations between TA and IBD in adult literature, but only a few paediatric case reports. We describe an Asian female (X) with indeterminate colitis diagnosed at age 4 after endoscopy showed pancolitis. She remained steroid dependant and further endoscopy 2 years later revealed ulcerative colitis which has since been well controlled with Sulfasalazine and Azathioprine. At 12-years-old X presented with 3-months of right sided neck pain, pain and fatigue in her lower limbs, intermittent fevers, night sweats and weight loss. There had been no changes in her bowel symptoms.

On examination X was found to be pale, tachycardic at 135 bpm and had a blood pressure of 101/61 with no discrepancy between limbs. She had normal heart sounds and a clear chest. There were some small tender submandibular lymph nodes and a right sided torticollis.

X was admitted for pain management and noted to have raised inflammatory markers, anaemia and thrombocytosis. She had a neck ultrasound which revealed evidence of large vessel arteritis. A CT angiogram was performed showing arteritis of the ascending, arch and descending thoracic aorta with aneurysmal change. There was involvement of the proximal arch vessels and proximal superior mesenteric artery (SMA) with aneurysmal changes in the distal SMA, as well as involvement of the right femoral and popliteal arteries. This was consistent with a diagnosis of Takayasu arteritis. She was referred to the paediatric rheumatologists and started on 30mg/kg of pulsed intravenous Methylprednisolone for 3 days. After a brief period of hospitalisation, she was discharged on a weaning dose of oral Prednisolone, weekly subcutaneous Methotrexate and low dose Aspirin. Unfortunately, she had inadequate response to Methotrexate, this was discontinued and Mycophenolate Mofetil and intravenous Tocilizumab were commenced.

Discussion:

Although rare, simultaneous occurrence of both diseases cannot be put down to coincidence alone; with the frequency of IBD in patients with TA reported between 5.8-9.3%. The exact causative mechanism for both IBD and TA is unclear, however there may be genetic markers which link both diseases. A significantly higher proportion of patients with IBD and TA were HLA-B52 positive compared to TA alone. Other genetic markers including HLA-DR2 and IL-12B have also been shown to be present in both diseases. It is also interesting to note that Infliximab has been effective in treating combined IBD and TA. This could support the theory that TNF-α may have a role in both conditions leading to mucosal and vessel inflammation.

Conclusion:

TA in IBD is extremely rare and requires a high degree of suspicion. Ongoing symptoms of pain, weight loss, fever and high inflammatory markers, in patients with otherwise well controlled IBD, should raise suspicion of alternative pathologies such as Takayasu arteritis. Early diagnosis and prompt treatment are likely to reduce morbidity and mortality in a group of patients who already suffer a high disease burden.

P33

An Unusual Case of Spontaneous Bowel Perforation as First Presentation of Crohn's Disease

Michael Hii[1], Georgina Yan[1], Chris Smith[1], Assad Butt[1], Joanne Parker[1] and Ruth Hallows[1].
[1]Royal Alexandra Children's Hospital

Introduction:
We describe the case of spontaneous bowel perforation in a 14-year old boy, preceding a diagnosis of Crohn's disease.

Background:
Outside of the neonatal population, spontaneous bowel perforation (SBP) in children is exceedingly rare. It tends to occur in children with pre-existing conditions such as infection, Ehler-Danlos and established Crohn's disease (CD).

Case Presentation:
The patient presented to the paediatric emergency department with sudden onset left upper quadrant abdominal pain radiating to his left shoulder. He described the pain as “stabbing”, worse on inspiration and when lying down. He had a 1-year history of vague intermittent abdominal pain. There was no associated vomiting, diarrhoea, fever or weight loss. Chest and abdominal examinations were unremarkable apart from generalised abdominal tenderness without peritonism.

Investigations and Management:
A chest XR showed extensive free air under the diaphragm and subsequent CT abdomen confirmed pneumoperitoneum although the site of perforation could not be identified. As he was haemodynamically stable, the surgeons treated him conservatively with IV antibiotics. It was thought that the perforation was most likely due to Helicobacter Pylori so he was discharged with appropriate eradication therapy. He was referred to gastroenterology for outpatient follow -up.

Despite resolution of the pneumoperitoneum on follow-up imaging, his abdominal pain persisted. He also developed constipation and reported weight loss. Faecal calprotectin was significantly elevated. Upper GI endoscopy showed generalised erythema in the stomach and terminal ileum but no obvious ulcers were observed. MRI of the small bowel showed a slightly thick-walled terminal ileum. The rest of the small bowel and large bowel appeared normal. Histology showed terminal ileitis and non-specific chronic gastritis. There were no granulomas observed. A subsequent video capsule endoscopy revealed multiple small but deep punctuate ulcers throughout the small bowel and right-side of the colon, confirming diagnosis of CD.

Discussion:
SBP is a rare entity in children but carries potentially significant morbidity and mortality (1-2). To date, there have been 22 reported cases of SBP in children with CD, of which 18 percent were first presentation of CD. All of the patients required surgical intervention (3).

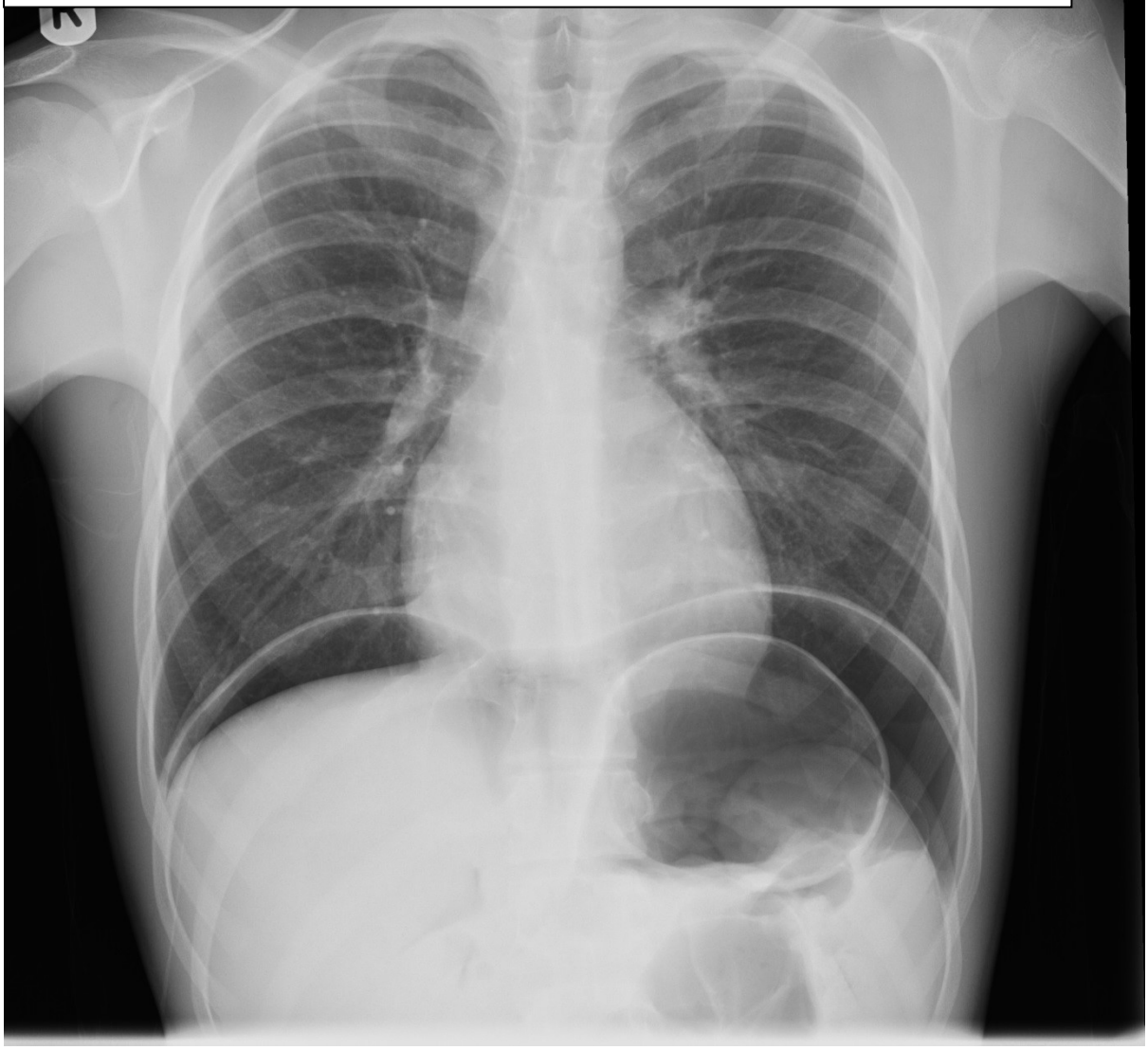
Conclusion:
Although rare, CD should be considered in children presenting with SBP. It is regarded as a surgical emergency however conservative treatment can be successful as demonstrated by this case and reported in the literature (4).

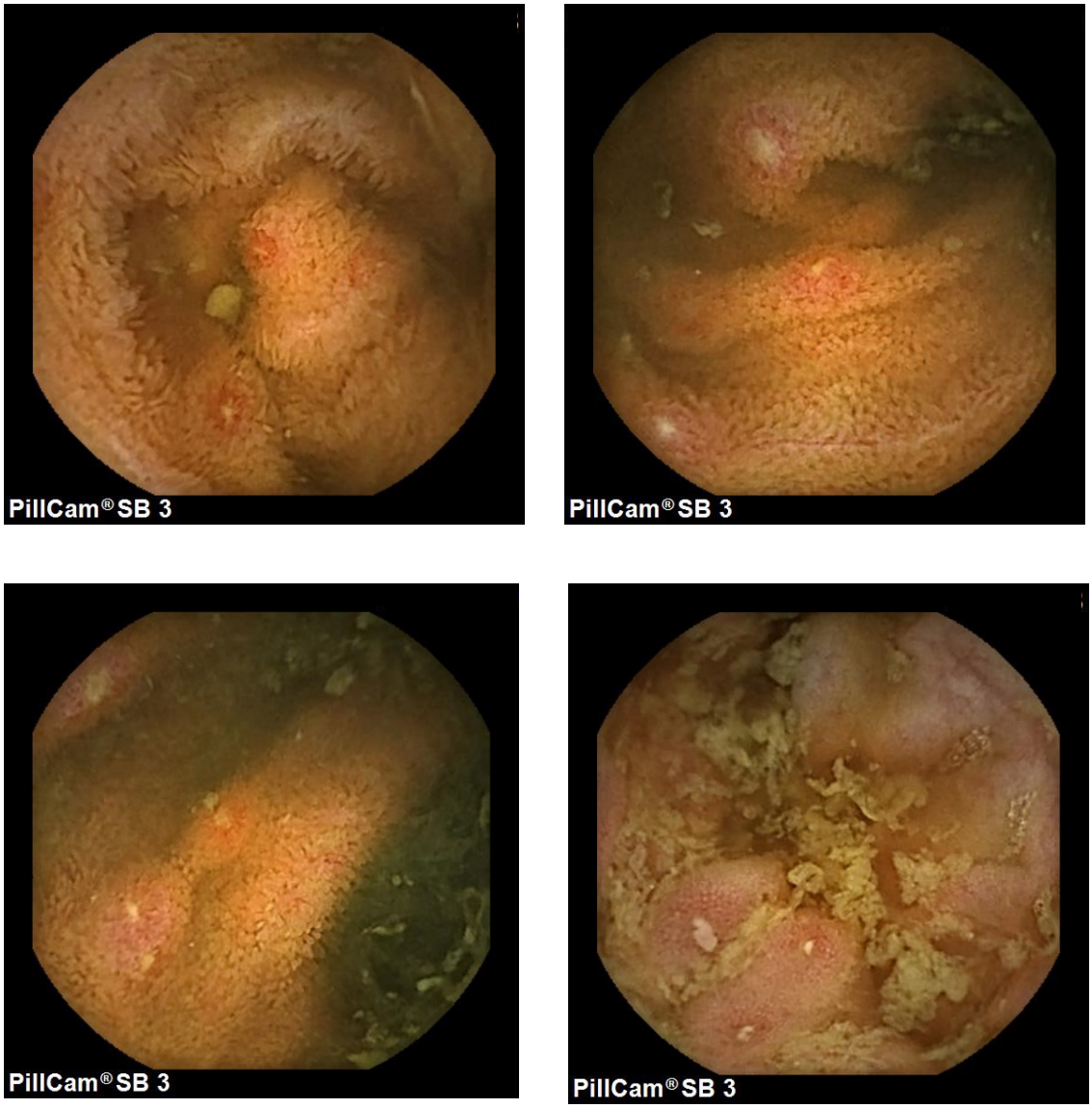
References

1. Chen JC, Chen CC, Liang JT et al. Spontaneous Bowel Perforation in Infants and Young Children: A Clinicopathologic Analysis of Pathogenesis. J. Pediatr. Gastroenterol. Nutr. 2000;4;432-435
2. Choi YJ, Cho YH, Kim SH et al. Clinical implication of spontaneous gastrointestinal perforation in

pediatric patients: its difference according to age group. Ann Surg Paed Res. 2018;95(3);141-146
3. Kolar M, Pilkington M, Winthrop A et al. Free intestinal perforation in children with Crohn's disease. Journal of Pediatric Surgery Case Reports. 2018;32:5-10
4. Gasparetto M, Giorgi B, Kleon W et al. Colonic Perforation in a Child with Crohn's Disease: Successful Medical Treatment Rescues from Colectomy. Case Reports in Gastrointestinal Medicine. 2012;Article ID 152414;4 pages

Chest XR: extensive free air under the diaphragm.





Video Capsule Endoscopy: small but deep punctuate ulcers were seen throughout the small bowel and right side of the colon.

P39

Analysis of 61 exclusive enteral nutrition formulas used for induction of remission in Crohn's disease - new insights on dietary disease triggers

Michael Logan[1], Konstantinos Gkikas[1], Vaios Svolos[1], Ben Nichols[1], Simon Milling[1], Umer Z Ijaz[1], Richard Hansen[2], Richard K Russell[2] and Konstantinos Gerasimidis[1].
[1]University of Glasgow; [2]Royal Hospital for Children, Greater Glasgow & Clyde NHS

Objectives and Study:

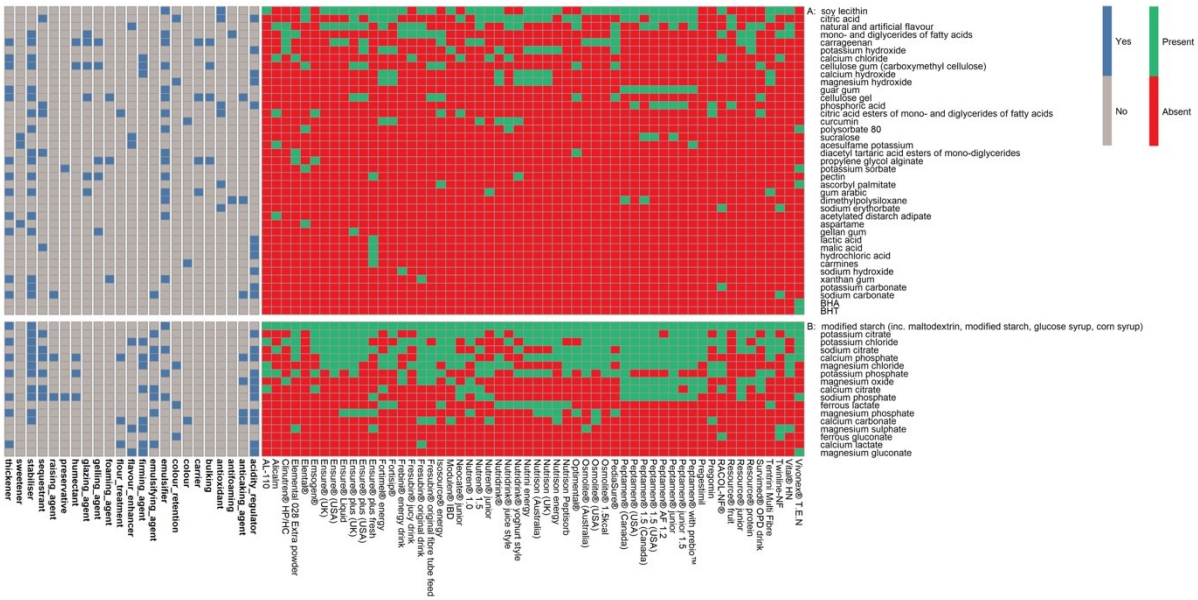
Exclusive enteral nutrition (EEN) is an effective treatment of Crohn's disease (CD). We hypothesise that food ingredients which are included in EEN formulas are less likely to initiate a disease flare and that their dietary elimination is not essential for disease amelioration.

Methods:

We performed a compositional analysis of EEN formulas with evidence of clinical efficacy for the induction of remission in active CD. Macronutrient content was compared against the dietary reference values (DRVs), the UK National Diet and Nutrition Survey (NDNS), and children with CD. Food additives included with EEN formulas were cross-referenced against the FAO/WHO database.

Results:

Sixty-one formulas were identified with variable composition [carbohydrates (23-89%), protein (8-30%), fat (0-50%)]. Modified starches (e.g. maltodextrin), milk protein and vegetable/plant oils were the commonest sources of carbohydrate, protein and fat, respectively. The n-6:n-3 fatty acid ratio varied from 0.3 to 46.5. Fifty six unique food additives were identified contained within EEN formulas (median per formula: 11), figure 1. All formulas were lactose, gluten free. 80% lacked fibre. The five commonest food additive functional classes were emulsifiers, stabilisers, antioxidants, acidity regulators, and thickeners. Food additives, implicated in CD aetiology, were present in formulas [modified starches (100%), carrageenan (23%), carboxymethyl cellulose (14%), polysorbate 80 (5%)]. EEN formulas derived 7.9% less energy from saturated fat than participants from the NDNS (p=0.001). CD children consumed more sugars, total/saturated fat than the EEN content.



Conclusion:

We provide a list of food ingredients which are unlikely, in the amount provided within EEN, to trigger CD activity. Current perceptions about the role of these ingredients in CD management are challenged.

P41

Juvenile polyps in patients with suspected VEOIBD –a case series.

Nastasia Hadjichristou[1], Mohamed Mutalib[1], Rakesh Vora[1] and Jochen Kammermeier[1].
[1]GSTT

Objectives:

Hamartomatous polyps, solidary or syndromic, are commonly observed in the paediatric population. Clinical presentation is often non-specific and diagnostic biomarkers are not available. Presentation in early childhood, rectal bleeding and raised faecal inflammatory markers may also be indicative of inflammatory bowel disease (IBD). The aim of this study is to characterise a cohort of patients with juvenile polyps and further evaluate the role of faecal biomarkers as a diagnostic tool.

Methods:

We retrospectively reviewed all paediatric patients referred with suspected very early-onset IBD (VEOIBD) between January 2014 to November 2019 who were subsequently diagnosed with hamartomatous polyp/s. Patients' demographics, clinical presentation, polyp's size, location, number of polyps, blood tests and level of faecal calprotectin (FC) were analysed.

Results:

A total of 11 patients (female: male ratio 1.2:1) with histologically confirmed hamartomatous gastrointestinal polyp/s were included. Median age at diagnosis was 5.4 years (IQR: 3.6 -7.3) and median duration of symptoms prior to the diagnosis was 8 months. All 11 children presented with rectal bleeding, 3/11 (27%) had diarrhoea, 3/11 (27%) experienced abdominal pain and 2/11 (18%) suffered from chronic constipation. One out of 11 patients (9%) presented with rectal prolapse.

In 8/11 cases (73%), a solitary juvenile polyp was identified. Three out of 11 children (27%) had polyposis syndrome (two patients with juvenile polyposis and one with Peutz-Jeghers syndrome (PJS)). Polyps were found in the recto-sigmoid in 9/11 patients (82%), in the caecum in 1/11 (9%) and transverse colon in 1/11 children (9%). The patient with PJS had additional polyps in the stomach.

FC levels (median FC: 733ug/g) were determined in all 11 patients within 2 months prior to endoscopy. Eight out of 11 children (73%) had a single colonic polyp with a median size of 12mm. Four out of eight solitary polyps were larger than 15 mm and the remainder 4/8 solitary polyps were smaller than 10mm. The median FC levels for both groups were 1453ug/g and 244ug/g respectively. Patients with polyps larger than 15mm were diagnosed at an earlier stage (median age: 3.4 years [IQR: 3.3- 3.6]) compared to those with polyps smaller than 10mm (median age: 5.9 years [IQR: 5.4-6.5]). Histological findings were similar, highlighting cystically dilated glands, elongated and branching crypts with superficial ulcers and inflammation in the adjacent lamina propria with no evidence of malignancy. None of the patients had a family history of gastrointestinal polyps. No complications were noted and all subjects report complete resolution of symptoms within 48 hours post polypectomy.

Conclusions:

Juvenile polyps frequently present with raised faecal calprotectin and should be considered as a differential diagnosis for patients with suspected VEOIBD. Our data suggests that there is an association between the level of faecal calprotectin, age at presentation and polyp size.

P47

Dietary triggers of colonic inflammation following treatment with exclusive enteral nutrition in children with Crohn's disease

Konstantinos Gkikas[1], Michael Logan[1], Ben Nichols[1], Clare Clark[1], Umer Z. Ijaz[1], Lisa Gervais[2], Hazel Duncan[2], Victoria Garrick[2], Lee Curtis[2], Elaine Buchanan[2], Tracey Cardigan[2], Lawrence Armstrong[3], Caroline Delahunty[4], Diana Flynn[2], Andrew R. Barclay[2], Rachel Tayler[2], Richard Hansen[2], Richard K. Russell[2] and Konstantinos Gerasimidis[1].

[1]University of Glasgow; [2]NHS Greater Glasgow & Clyde; [3]NHS Ayrshire and Arran; [4]NHS Lanarkshire

Background:

Exclusive enteral nutrition (EEN) ameliorates gut inflammation in children with Crohn's disease (CD). We have previously described the rapid rise in faecal calprotectin levels (FC) when children with CD return to their habitual diet after EEN treatment (1).

Aim:

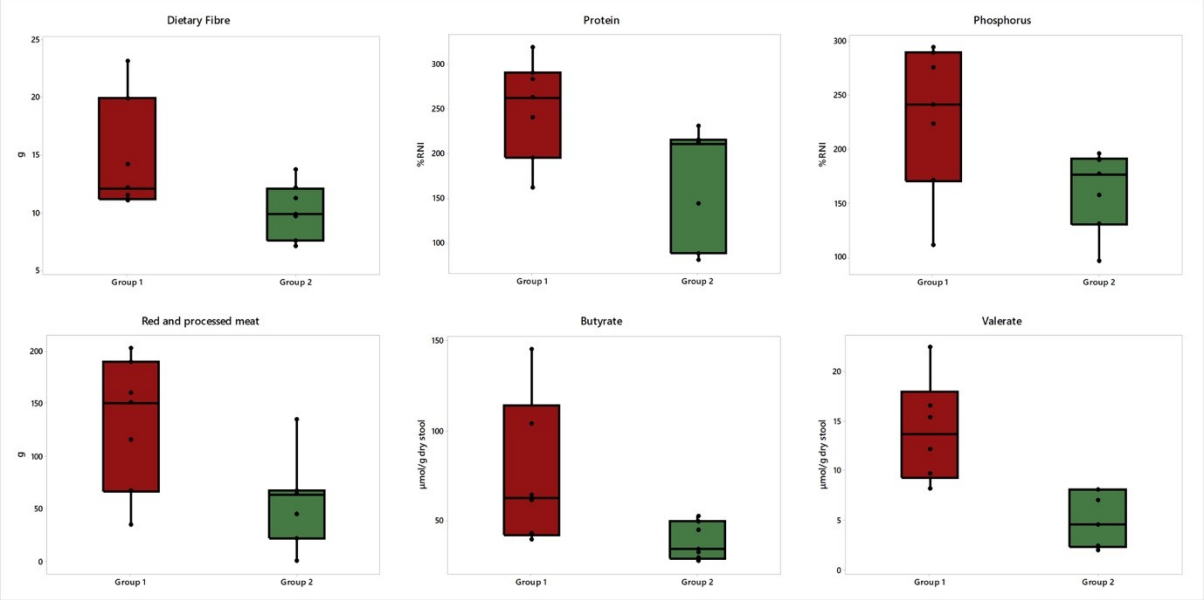
We aimed to explore dietary triggers of CD relapse by performing dietary assessment and measuring biomarkers of food consumption in faeces of children with CD during early food reintroduction.

Subjects and methods:

A composite outcome of clinical remission post EEN (weighted Paediatric Crohn's Disease Activity Index <12.5) and a significant drop in FC (FC decrease: >500 mg/kg / >35%), was used to define the patient group. All patients completed 3-day estimated weight food diaries and provided a faecal sample. Patients were divided equally for statistical analysis purposes in two groups; above (Group 1) and below (Group 2) the median FC concentration at food reintroduction [900 mg/kg (341, 1,243)]. Nutrient analysis was performed with WinDiets. Short chain fatty acids, the gluten immunogenic peptide (GIP) and starch were measured in stool, as proxy of fibre, gluten and malabsorbed/resistant starch respectively. Food groups were assigned using the National Diet and Nutrition Survey approach and statistics employing taxonomy were utilised in the food group analysis. All data are displayed using medians (Q1, Q3).

Results:

14 children provided a FC sample within 21 (15, 51) days post EEN. Classification of patients in the two groups resulted in significantly different FC values; Group 1: 1181 mg/kg (1024, 1781) vs Group 2: 411 mg/kg (130, 651) ($p<0.001$). Age, weight and height were similar between the two groups ($p=0.40$, $p=0.37$, $p=0.79$). Total energy intake did not differ between the groups ($p=0.37$). Patients in Group 1 consumed more fibre than Group 2 [12.1 g (11.3, 19.9) vs 9.9 g (7.6, 12.1), $p=0.04$]. Protein and phosphorus intakes, expressed as a percentage of reference nutrient intakes, were also higher in Group 1 than Group 2 [Protein (%): 262 (195, 291) vs 211 (88, 215), $p=0.02$; phosphorus (%): 241 (171, 290) vs 177 (131, 1901), $p=0.04$]. Butyrate and valerate levels were higher in Group 1 than Group 2 [Butyrate: 62.5 (41.8, 114) vs 34.1 (29.2, 49.6), $p=0.02$; valerate: 13.7 (9.3, 17.9) vs 4.5 (2.3, 8.1), $p<0.01$]. There were no differences in GIP and faecal starch levels between the two groups ($p=0.18$, $p=0.12$). Red & processed meat intake was higher in Group 1 than Group 2 [151 g (66.7, 190) vs: 63.3 g (21.7, 67), $p=0.02$)]. Cereals intake was non-significantly higher in Group 1 [389 (207, 405) vs 231 (141, 279), $p=0.08$]. Overall diet diversity did not differ between the two groups ($p=0.47$).



Summary and Conclusion:

The current analysis suggests that fibre, protein, phosphorus and red & processed meat may be associated with recurrence of colonic inflammation in children with CD during early food reintroduction. These findings should be confirmed in larger studies.

References

1. Logan, M. et al. The reduction of faecal calprotectin during exclusive enteral nutrition is lost rapidly after food re-introduction. AP&T 50, 664–674 (2019).

P49

What do teenagers want from their Inflammatory Bowel Disease service? Findings from a focus group.

Michael Cornish[1], Caron Lawson[2], Georgina Knott[1], Rita Shergill-Bonner[1], Gemma Lee[1], Jochen Kammermeier[1], Rakesh Vora[1] and Mohamed Mutalib[1].
[1]Evelina London Children's Hospital; [2]Kings College London

Introduction/background:

Following the introduction of clinical psychology, to the paediatric gastroenterology team at the Evelina London Children's Hospital, we held a focus group to explore what are the needs of young people with Inflammatory Bowel Disease (IBD). This focus group aimed to find out what young people want from their gastroenterology service, and particularly the new gastroenterology psychology service.

Aims:

1. To explore young people's experience of having IBD and find out what they want from their IBD service.
2. To think about how the new paediatric gastroenterology psychologist could best support young people with IBD.
3. To help young people meet others with the same medical diagnosis and learn from each other's experiences.

Subject & Methods:

This qualitative study invited young people between the ages of 15 to 17 who were patients with IBD under the care of the Evelina London Children's Hospital to attend a focus group. Four patients attended. One female and three males. Two were 15 years old and two were 16 years old. Focus group members were from diverse ethnic backgrounds.

The focus group were asked about the challenges they face living with IBD. They were also asked what might help them to manage their IBD better and what they might want from the paediatric gastroenterology psychologist.

A psychology assistant supported the running of the group and documented the group discussions. From this several themes were identified.

Results:

The group all reported feelings of frustration, particularly with GP services, around how long it took to get diagnosed with IBD. Once diagnosed, the main challenges of living with IBD that emerged were dealing with fatigue and managing the early treatment after diagnosis. Most of the young people also reported it being challenging talking about their IBD to other people.

The focus group members made several suggestions that might improve their care. This included, as they get older they wanted some individual time to talk to their doctor without their parents. They would like more access to psychological/emotional support. They reported wanting more support during the transition into adult services. They also wanted quick and easy access to the medical team. They also gave ideas about improving their inpatient experience, including more privacy, entertainment and age appropriate ward space.

Summary and conclusion:

The young people in the focus group highlighted some of the difficulties and challenges from having IBD and gave some suggestions that would improve their experience of living with IBD.

P64

Assessment of Nutritional Status in Paediatric Inflammatory Bowel Disease

Burcu Dadi[1], Mohamed Mutalib[1], Gemma Lee[1] and Rita Shergill-Bonner[1].
[1]Evelina London Children's Hospital-Guy's and St Thomas' NHS Foundation Trust

Introduction/Background:

Inflammatory bowel disease (IBD) is a chronic inflammatory condition of unknown aetiology that is thought to result from a combination of genetic, immunologic and environmental factors. Malnutrition, protein-energy, and micronutrient deficiencies are common among patients with IBD. 10%-56% of Crohn's disease and up to 10% of UC patients had growth failure at the time of diagnosis. Nutritional deficiencies may be the result of reduced oral intake, active GI inflammation and occasionally from drug nutrient interactions.

The incidence of obesity in IBD patients is also increasing as in the normal population which may suggest a common pathophysiologic link. Obesity plays a major role in the IBD burden, associated comorbidities and reduced quality of life. 10% of children with Crohn's disease are shown to be overweight or at risk of being overweight at the time of diagnosis. The prevalence of overweight and obesity in paediatric crohn's disease is around 20%.

Aim:

To assess nutritional status in children with IBD as measured by z scores and BMI at diagnosis, 3, 6 and 12 months after diagnosis.

Subjects and Methods:

This is a single centred, retrospective, observational study. Patients' gender, weight, height, BMI and z-scores were recorded at prediagnosis, at 3 months, 6 months and 1 year.

Results:

46 children were included 29 Crohns, mean± SD age at diagnosis 11.5 ± 3.3 (1-16years). at diagnosis weight 39.3 ± 14.8 BMI 17.3 ± 3.5 z score -0.3 ± 1.1. At 3 months wt 41.2±14.7, BMI 18.2±3.2, zscore -0.2±1.2, at 6 months wt 43.5±16.3 BMI18.7±3.4 zscore -0.05±1.1 and at 1 year wt 44.9±14.8 BMI 19.2±3.6 zscore 0±1.

Summary and Conclusions:

Z scores and BMI at diagnosis were low and appears to be slowly corrected over the course of one year. Children below the age of 2 years are the least affected nutritionally with positive z scores pre and post diagnosis. Adolecents over the age of 15 years are at high risk with persistently low z score up to one year after optimum medical treatment.

P76

Bone health in IBD patients- the role of DXA scan as a routine screening tool

Nastasia Hadjichristou[1], Jochen Kammermeier[1], Rakesh Vora[1] and Mohamed Mutalib[1].
[1]GSTT

Introduction:

Children with chronic gastrointestinal (GI) conditions are prone to develop impaired bone health. Dual emission x-ray absorptiometry (DXA) is considered gold standard in assessing the skeletal health. However, the optimal time, the necessity and clear recommendations of its use are still elusive. The aims of this study are to report the outcome of bone health in children with inflammatory bowel disease (IBD) by using DXA scan and to assess the duration of disease on skeletal health.

Methods:

Retrospective review of all paediatric IBD patients who have had a DXA scan between January 2015 and October 2019. As facilities to perform DXA scan became routinely available in our institution over the last 12 months we aimed to screen all children with IBD. Patients' demographics, bone mineral density (BMD) Z-score in lumbar spine (LS) (± SD) and in whole body less head (WBLS) (± SD), body mass indices (BMI) and 25-OH vitamin D (25-OH-D) level at the time of DXA scan were analysed.

Results:

61 IBD patients (Crohn's disease [CD]: 44, Ulcerative colitis [UC]: 17) were included. Male to female ratio (1.4:1). Median age at diagnosis was 12.8 years (IQR: 10.3- 14.4). Mean time interval between diagnosis and DXA scan was 15± 17months. Mean Z-score in LS was -0.67± 1.0 and WBLS -0.68 ± 1.1. Mean 25-OH-D level and BMI was 48 nmol/L± 23 and 19 kg/m2± 3.7 at the time of DXA scan.

We divided the cohort into three groups based on the time elapsed between diagnosis and DXA scan (Group A= 0- 6 months, Group B= 7- 24 months, Group C: > 24 months). In group A, Z- score in LS was -0.7± 0.9, WBLS: -0.7± 1.0, BMI: 18± 3.8 and 25-OH-D: 42± 21. In group B, Z- score in LS was -0.5 ± 0.7, WBLS: -0.5± 1.1, BMI: 19.6± 4 kg/m2 and 25-OH-D: 54nmol/L ± 24. In group C, Z- score in LS was -0.9 ± 1.3, WBLS: -0.8± 1.2, BMI: 19.9± 3 kg/m2 and 25-OH-D: 49nmol/L ± 22.
Z-scores were similar in children with CD and UC, Z- score in LS: -0.7± 1.1 vs -0.7± 0.75, p=0.69 and WBLH: -0.7± 1.2 vs -0.8 ± 1, p=0.51 respectively.

In 7/61 IBD patients (11.5%), (CD: 6, UC: 1), mean Z-score in LS was -2.6± 0.7, WBLS: -2.5± 1.1, BMI: 19± 2.3 kg/m2 and 25-OH-D: 46nmol/L± 20. Median age at diagnosis was 12.6 years (IQR: 11.9- 14.5) and mean interval between diagnosis and DXA was 26± 35 months. One patient (1.6%) presented with multiple spinal compression fractures within two months of diagnosis.

Conclusions:

In paediatric patients with IBD the outlook of bone status as measured by DXA scan appears to be negative and did not improve despite optimal medical therapy.11.5% of IBD patients had low BMD and one patient had symptomatic vertebral fractures. DXA scan, is a useful, non-invasive and low radiation risk tool to risk stratify bone health in children with IBD.

P81

Large Vessel Vasculitis as extraintestinal manifestation (EIM) of Inflammatory Bowel Disease (IBD)

Sunita Amar Rajani[1], Rachel Tattersall[1], Sarah Louise Maltby[1], Daniel Hawley[1] and Arun Urs[1].
[1]Sheffield Children's Hospital

Objectives and study:

Extraintestinal manifestations (EIM) are relatively common in paediatric patients with inflammatory bowel disease (IBD). We describe a rare extraintestinal complication of large vessel vasculitis (LVV - Takayasu's arteritis) in 16-year-old young man. There is only sparse literature LVV in patients with IBD and treatment is challenging.

Methods:

A retrospective review of clinical notes and investigations was undertaken with review of literature.

Results:

A 16-year-old young man of Eastern European descent was diagnosed with very early onset IBD, managed as Ulcerative colitis age 2. Initial treatment comprised Sulfasalazine and Azathioprine with frequent relapses responsive to steroids. Treatment was escalated to Infliximab(combination therapy) following an endoscopy which showed pancolitis. He lost immunologic response to Infliximab and was switched to Adalimumab with to which he also experienced secondary treatment failure.

Symptoms persisted with abdominal pain, bloody stools, nocturnal symptoms and poor quality of life.Endoscopic assessment was suggestive of Crohn's disease with ulcerations in duodenum, jejunum, ileum (deep seated) with patchy involvement of colon with histological evidence only limited to active pancolitis. MRI small bowel showed thickening of terminal ileum. He continued to be steroid dependent. Serum inflammatory markers were persistently raised along with raised faecal calprotectin: Erythrocyte sedimentation rate > 60 mm/hr, CRP >70 mg/L, Platelets >500 x 109/L and IgA elevated >2.9 g/L. Extensive immunological work up (anticardiolipin antibodies, angiotensin converting enzyme, rheumatoid factor, anti dsDNA antibodies, complement and antinuclear antibodies) was negative. Serum amyloid A was raised at 478 mg/L.

Treatment was escalated to Vedolizumab with improvement in bowel symptoms but inflammatory markers remained persistently elevated and he complained of pain in left mandibular region. Carotid ultrasound suggested stenotic disease of the carotid arteries and a widespread large vessel arteritis (LVV) involving the aorta, subclavian, carotids was confirmed on PET scan with a mixed picture of damage and active inflammation.

Since LVV is life-threatening, high dose, pulsed intravenous steroids were started, vedolizumab stopped and Tocilizumab (treatment of choice for LVV) started with joint rheumatology management. Aspirin and methotrexate were also added. Inflammatory markers and systemic symptoms have improved and the patient reports improved quality of life.

Conclusion:

IBD patients are at a risk of LVV although this remains a rare association. Clinicians need to have high index of suspicion where patients with IBD have significantly elevated inflammatory markers or unusual symptoms and screen for vasculitis.

LVV is reported to have been induced by anti-TNF treatment but it is not clear this is the pathogenesis here. In adult patients vedolizumab is reported to be associated with of significant flaring of arthritis and asthma and it is plausible this was the cause of worsening symptoms in this case but this also remains unproven. We have prioritised treatment of the LVV here but tocilizumab has no effect on IBD and may predispose bowel perforation so continued therapy is challenging. There may be a need to consider combining biologic therapies eg vedolizumab with tocilizumab with theoretical risk of significant immunosuppression and associated morbidity. Further research into LVV and IBD is needed.

P83

Intravenous ferric carboxymaltose in a paediatric population with iron-deficiency anaemia; current practice in a tertiary centre

Sahan Samaraweera[1], Zachary Green[1], James Ashton[1], Tracy Coelho[1], Nadeem Afzal[1], Akshay Batra[1] and R Mark Beattie[1].

[1]Department of Paediatric Gastroenterology, Southampton Children's Hospital, Southampton

Objectives and Study:

Gastrointestinal diseases contribute a significant proportion of causes for iron deficiency anaemia (IDA) in children. Studies have found that around 70% of children with inflammatory bowel disease (IBD) are anaemic at some point during their illness. BSPGHAN guidelines recommend iron replacement in this population if Haemoglobin (Hb) is ≤110g/L and intravenous therapy if there is an inadequate response or intolerance to oral supplementation. Patients with elevated inflammatory markers and a ferritin less than 100 mcg/L may also be considered for treatment. Ferric carboxymaltose (FCM) is an effective, and relatively safe, dextran-free intravenous iron preparation with the preferred product being Ferinject®, although it is unlicensed for those under 14 years. At Southampton Children's Hospital (SCH), Ferinject® is routinely administered as a day case procedure. Recent studies have shown a robust safety profile in the paediatric population with an estimated intravenous iron treatment dose of 15mg/kg. Current practice is variable with initial dose calculation, dosing schedule and monitoring of haemoglobin and ferritin proving inconsistent. No common guidance exists for those under 18 years old or less than 35kg in weight. Current practice at SCH was evaluated in this study.

Methods:

Retrospective data was collected from electronic patient records over a 3-year period from 2016-2019. The inclusion criteria comprised of children (≤17 years old) with gastrointestinal causes of IDA, including IBD and intestinal failure. Ideal Ferinject® doses were calculated based on weight (15mg/kg). Initial Ferinject® dose and any subsequent doses were then recorded, along with ferritin and Hb pre and post-infusion. C-Reactive Protein (CRP) values were noted to contextualise the ferritin result.

Results:

The mean age of the cases was 12.6 years, with the youngest child being 3 years old. Mean weight was 46.42kg (range 15-66.2kg). Mean pre-dose Hb was 104.69 (range 72-127g/L), which rose to 121.97 (range 89-147g/L) post-dose. Post-dose blood tests were taken a mean of 3 weeks after receiving an infusion.

Mean pre-dose ferritin was 66.31, which rose to 160.79 post dose. CRP was measured in 2/3 of the cases following infusion (mean 11, range 1-99mg/L). Post-dose Hb and ferritin were not checked in 4 and 11 of the cases respectively.

Ferinject® doses were rounded to the nearest 100mg, with no more than 1000mg given per dose to any patient. The mean dose difference between what was prescribed versus the 15mg/kg recommendation was 139.44mg (corrected for patients prescribed more than 1000mg).

No adverse reactions were recorded in any cases. 26% of the patients required a second transfusion, and of these patients 71% initially received an inadequate dose of Ferinject® (less than 15mg/kg).

Conclusion:

There was significant variation in the administration and indication for Ferinject® prescription. Intravenous iron was safe and resulted in an increase in Hb for all patients, typically within 3 weeks, although ¼ required a second infusion potentially related to an inadequate initial dose. A consensus guideline for Ferinject® prescription, monitoring and follow-up is needed to help clinician's prescribe appropriate doses and optimise treatment for children with IDA caused by gastrointestinal diseases.

P95

Real life outcomes of Adalimumab in biologic naive and experienced children with inflammatory bowel disease, achieving remission over a 4 year period: a tertiary centre review

Vaia Zouzo[1], Manjula Nair[1], Sarang Tamhne[1], Marcus Auth[1], Fiona Cameron[1], Edwarda Baretto-White[1] and Elizabeth Renji[1].
[1]Alder Hey Childrens' Hospital

Introduction:
Adalimumab is widely used in paediatric inflammatory bowel disease (pIBD) either as first- or second-line treatment.

Aims and Objectives:
Comparing the efficiency of Adalimumab in pIBD between biologic naïve and experienced patients. The primary outcomes were remission (PUCAI<10, or wPCDAI<10, or reduction >15 from initial) at 6 months and 1 year. Secondary outcomes were steroid free remission and optimal therapeutic drug monitoring (TDM) levels, Serum level (SL) associated with remission.

Subjects and Methods:
Retrospective review of electronic records of pIBD patients treated with Adalimumab from January 2015 until November 2019 and document outcomes.

Results:
55 patients (Male: 36) were commenced on Adalimumab over a period of 4 years. In our cohort, the biological naïve patients who were in remission at 6 and 12 months, were 21% and 6.6 % respectively. In the biological experienced group, the percentage was 19.3% and 21.4% respectively. Steroid free remission in the biologic naïve group at 6 months and 12 months were 95% and 93% respectively, compared to 84% and 57% in the biologic experienced group. SL > 5mg/L (with intensification of regime in 20/55) was associated with better outcomes, despite positive antibodies. 21% of naïve and 45.7% of experienced patients required intensification of regime within maintenance phase. Concomitant Immunosuppressants (CI) did not prevent loss of response.

P 98

Normalisation of Stool Calprotectin- Is it practical in Children with Inflammatory Bowel Disease in real-life setting?

Meranthi Fernando[1], Wolfram Haller[1] and Rafeeq Muhammed[1].
[1]Birmingham Children's Hospital

Background:

Endoscopic healing is associated with better outcome in patients with Inflammatory bowel disease (IBD). Normal stool calprotectin values in patients with IBD correlates well with endoscopic healing. Serial monitoring and documentation of stool calprotectin is particularly important in children with IBD, because of the practical difficulties of reassessing with colonoscopy.

Aim:

To identify the proportion of patients who achieve normalisation of stool calprotectin values 12 months after diagnosis and correlate with clinical remission and CRP normalisation.

Methods:

Retrospective chart review of children diagnosed with Crohn's disease (CD) and ulcerative colitis (UC) from 1/1/2017 to 30/06/2018. Stool calprotectin was measured in the hospital by Buhlmann ELISA method and value less than 250 microgram/gram is considered as normal. Normal range of CRP was 1-10 mg/l. Clinical remission was assessed by physician global assessment (PGA). Endoscopic healing was defined as lack of ulceration in the colon. Histology was considered normal if no abnormalities were reported by the pathologist.

Results:

59 (32 CD & 27 UC) out of 119 children (71 CD & 48 UC) diagnosed with IBD in the study period had stool calprotectin documented at diagnosis. Median age of children diagnosed with CD was 12.5 years (range 3-15 years) with B1 phenotype in 91% and B2 phenotype in 9% (L1 16%, L2 21%, L3 63% with perianal disease in 47%). Median age of children with UC was 13 years (range 3-16 years) with disease extent E1 11%, E2 3%, E3 15%, E4 71%. Data on stool calprotectin and CRP at diagnosis and treatment in the first 12 months are summarised in Table 1.

Table1
Results of stool calprotectin and CRP at diagnosis and treatment received in the first 12 months after diagnosis

	Crohn's Disease	**Ulcerative Colitis**
Number of patients (%) with abnormal CRP >10 mg/L at diagnosis	19/31 (61%)	8/27 (30%)
Number of patients (%) with abnormal stool calprotectin (>250 microgram/gram) at diagnosis	30/32 (93%)	23/27 (85%)
Median (range) CRP mg/L at diagnosis	16 (1-133)	6 (1-86)
Median (range) Calprotectin microgram/gram at diagnosis	1800 (34-1800)	1800 (39-1800)
Number of patients (%) on treatment with biologics in the first 12 months after diagnosis **Infliximab** **Adalimumab**	24/32 (75%) 11 13	13/27 (48%) 13 0
Number of patients (%) on Azathioprine mono therapy	4/32 (12.5 %)	5/27 (18.5%)
Other treatment/No treatment	4/32 (12.5 %)	
Number of patients (%) on Mesalazine mono therapy		9/27 (33.5)

In patients with CD, only 63% (20/32) children had stool calprotectin result available at 12 months after diagnosis (median 179 range 30-1800 microgram/gram). CRP was available at 12 months in all patients (median CRP 1 range 1-9 mg/l). In patients with UC, only 67% (18/27) children had stool calprotectin result available 12 months after diagnosis (median 291 range 30-1800 microgram/gram). 24/27 (89%) children had CRP results available at 12 months (median 1 range 1-22 mg/l). Data on clinical remission, normalisation of CRP, stool calprotectin, endoscopic healing and histologic normalisation at 12 months after diagnosis for patients with CD and UC are summarised in Table 2 and 3.

Table 2
Outcome of Crohn's disease patients 12 months after diagnosis

	Clinical remission	**Normal CRP**	**Normal Stool Calprotectin**	**Normal CRP & stool calprotectin**	**Endoscopic remission**	**Histologic remission**
Number of patients data available	32	30	20	20	14	14
Yes	27 (84%)	30 (100%)	11 (55%)	11 (55%)	9 (64%)	9 (64%)
No	5 (16%)	0	9 (45%)	9 (45%)	5 (36%)	5 (36%)

Table 3
Outcome of ulcerative colitis patients 12 months after diagnosis

	Clinical remission	**Normal CRP**	**Normal Stool Calprotectin**	**Normal CRP & stool calprotectin**	**Endoscopic remission**	**Histologic remission**
Number of patients data available	27	24	18	18	17	17
Yes	21 (78%)	23 (96%)	8 (44%)	8 (44%)	9 (53%)	9 (53%)
No	6 (22%)	1 (4%)	10 (56%)	10 (56%)	8 (47%)	8 (47%)

Conclusion:
A high proportion (~80%) children with CD and UC achieved clinical remission by 12 months after diagnosis with about 50% children achieving normalisation of CRP and stool calprotectin. Stool calprotectin monitoring and documentation in the first 12 months of treatment was possible only in about 65% of children with IBD. Home based stool calprotectin assay may be more acceptable to some children with IBD resulting in better adherence to stool calprotectin monitoring.

P100

Are we giving up too soon on Vedolizumab?

Chloe Corlett[1], Astor Rodrigues[1], Lucy Howarth[1], Esther Quinn[1], Tejuswi Patel[1] and Holm Uhlig[1].
[1]Oxford University Hospitals Trust

Introduction:

Vedolizumab is an anti-integrin monoclonal antibody, licensed and approved by NICE for the treatment of Ulcerative Colitis (UC) and Crohn's disease (CD) in adults. Its efficacy to induce and maintain remission and its safety profile is well described in the adult population. It is usually commenced in patients with moderate to severe inflammatory bowel disease (IBD) who have failed conventional and anti-TNF therapy and NICE recommends that its continued use should be carefully reconsidered if no evidence of therapeutic benefit is observed by week 10. Ledder et al. (2017) reported a multi-centre retrospective study describing its efficacy and safety in the paediatric population; however its use in this population is still under evaluation. Recent ECCO/ESPGHAN guidelines published in JPGN (Turner et al. 2018) state that after commencing Vedolizumab, a response can be seen by week 6, although may not be evident until week 14 of therapy. In this study we look at whether a longer duration of treatment (than 10-14 weeks) is potentially beneficial.

Aim:

To assess response to Vedolizumab in paediatric patients with UC; in particular to look at the duration between commencing Vedolizumab and remission of disease and whether persistence with treatment is worthwhile if an initial response at the expected 14 weeks is not achieved.

Subjects and Methods:

All paediatric patients from our tertiary centre with UC who had been on Vedolizumab for more than 14 weeks were included in this study. Data was collected regarding biometrics, previous treatments, dose and indications for commencing Vedolizumab, duration until remission (defined by faecal calprotectin (FC) levels and/or colonic histology findings), concurrent medications, and need for surgery.

Results:

13 patients were identified with a mean age of 14.5 yrs (range 12 to 18 years) at the time of Vedolizumab initiation. There were 5 male and 8 female patients. All patients had been on prior immunomodulation and had failed anti-TNF therapy. The dose of Vedolizumab was 5 mg/kg up to 300 mg at 0, 2, 6 weeks and 4-8 weekly thereafter depending on response.

8 patients (62%) achieved deep remission (normal FC and/or colonic histology) with a mean duration of 37 weeks (range 13 – 78 weeks) of which 2 patients took more than a year to achieve this target, all 8 patients were responding clinically and did not require additional rescue treatment; 3 patients (23%) showed partial response (reduction in FC and/or persisting abnormalities on colonic histology) having been on Vedolizumab for an average of 48 weeks and are still continuing on it; and 2 (13%) required colectomy.

Summary and Conclusion:

Although a small-scale single-centre review, 62% of patients achieved deep remission when treated with Vedolizumab. Duration of time between commencing therapy and entering remission varied widely from 13 to 78 weeks, with a mean of 37 weeks. In fact, 2 patients achieved deep remission having persevered with treatment for more than 52 weeks. This suggests that persistence with Vedolizumab for longer may lead to improved outcomes and potentially higher chance of remission.

P101

Infliximab Treatment Failure in Children with Crohn's disease- is it avoidable?

Rulla Al-Araji[1], Wolfram Haller[1] and Rafeeq Muhammed[1].
[1]Birmingham Children's Hospital

Background:

Infliximab is an effective treatment for patients with Crohn's disease, however experience in adult IBD patients show treatment failure occurring in many patients in the first year of treatment.
Aim: We aimed to identify proportion of children with Crohn's disease failing Infliximab therapy in the first 12 months after treatment initiation with Infliximab. We have analysed the relationship between Infliximab levels, anti-Infliximab antibodies and non-response at week 14 and week 54.

Methods:

Retrospective chart review of 100 consecutive children with Crohn's disease commenced on Infliximab as their primary biologic therapy. Serum infliximab levels were measured using an in-house ELISA method. Anti-infliximab antibodies (both free and drug bound) were measured using a commercial kit.

Results:

100 consecutive children (male: female 2:1 median age 12.4 years range 2.2-16 years) with Crohn's disease (L1 16%, L2 29%, L3 52% Perianal disease 3%) received Infliximab as their primary biologic treatment. 89 were on Infliximab combination therapy with Azathioprine. 2 patients had severe infusion reaction during induction, hence their treatment with Infliximab was discontinued. 69/98 (70%) patients were in steroid-free clinical remission at week 14. 61/98 (62%) children had Infliximab levels measured at week 14 (median 4.8 microgram/ml range 0.4-10). By week 54, 92 patients remained on Infliximab treatment. 69/92 (75%) had Infliximab level measured at week 54 (median 5.15 microgram/ml range 0.4-10). 77/98 (79%) patients were in steroid-free clinical remission at week 54. Rates of steroid-free clinical remission, therapeutic levels of Infliximab, anti-Infliximab antibody positivity at week 14 and 54 are summarised in Table 1.

Table 1
Infliximab levels, aniti-infliximab antibodies and steroid-free clinical remission at week 14 and week 54

	Patients (%) with therapeutic levels of Infliximab	Patients (%) with anti Infliximab antibody positivity	Patients (%) with steroid-free clinical remission and therapeutic levels of Infliximab	Patients (%) with steroid-free clinical remission and sub-therapeutic levels of Infliximab
Week 14	46/61 (75%)	24/61 (40%)	33/41 (80%)*	5/41 (12%)*
Week 54	63/69 (90%)	33/69 (48%)	54/57 (95%)*	3/57 (5%)

* statistically significant

22/29 (76%) patients not in clinical remission at week 14 continued to remain on Infliximab at week 54 with 16/22 (73%) achieving steroid-free clinical remission. 19/22 (86%) patients received Infliximab with dose and/frequency escalation and 15 of these 19 patients (75%) patients achieved steroid-free clinical remission at week 54. Of the 69 patients who achieved steroid-free clinical remission at week 14, data on clinical remission at week 54 was documented only in 62 patients. 57 of these 62 patients (92%) remained in steroid-free clinical remission at week 54 on treatment with Infliximab, with 32 patients (52%) receiving Infliximab on increased dose and/frequency.

Conclusions:

Approximately 80% children with Crohn's disease commenced on Infliximab therapy remained in steroid-free clinical remission at week 54. Infliximab levels in therapeutic range at week 14 and 54 showed statistically significant association with clinical remission in children with Crohn's disease. Anti-Infliximab antibodies did not tend to affect the rate of clinical remission, however a large proportion of patients were treated with increased dose and/frequency of Infliximab. Standard dose of Infliximab induction regime did not result in therapeutic levels of Infliximab at week 14 in 25% patients. Majority of patients not in clinical remission at 14 responded to Infliximab treatment with increased dose and/frequency and achieved clinical remission by week 54.

P103

Therapeutic Monitoring of Paediatric Patients with Inflammatory Bowel Disease on Thiopurines: Does It Make a Difference?

Judwin Ndzo[1], Ladan Yusuf[1], David Devadason[1], Sabarinathan Loganathan[1], Sian Kirkham[1], Heidi Pike[1] and Michalis Papadopoulos[1].
[1]Nottingham University Hospitals NHS Foundation Trust

Introduction:
Azathioprine is an immunosuppressant antimetabolite drug commonly used in the management of Inflammatory Bowel Disease (IBD). Its side effects include bone marrow toxicity and myelosuppression, potentially causing reactivation of latent infections. Azathioprine is metabolised to 6-thioguanine metabolites (6-TGN) by the enzyme Thiopurine Methyltransferase (TPMT). 1/300 people have a deficiency of TPMT, putting them at further risk of toxicity.

Aim:
To assess the current practice of prescribing and monitoring Azathioprine at a tertiary paediatric IBD centre. Secondly, to look at correlation between TPMT activity, 6-TGN levels, dosing and blood results.

Methods:
A retrospective study of all children <18 years old with newly diagnosed IBD who had been initiated on Azathioprine during a 2-year period from January 2017 to December 2018 and followed up for at least 11 months. Clinic letters and data on blood results were obtained from an electronic reporting system. Data were collected using Google forms and analysed using SPSS version 20.

Results:
51 children were included (26 female and 25 male). Mean age at diagnosis was 12.7years (SD 3.4years). 32(62.7%) had Crohn`s Disease; 11(21.6%) Ulcerative Colitis; 8(15.7%) had IBD-Unclassified. 48(94%) had TPMT levels tested prior to initiation of Azathioprine, 2 had levels done post initiation, and one was never tested. 90%(45) had normal TPMT levels, the remainder had low levels. Median time from diagnosis to initiation of Azathioprine was 62days (IQR 26,167). For those with normal TPMT levels, the mean starting dose of Azathioprine was 0.6-2.5mg/kg (SD 0.7mg/kg); for those with low levels the mean starting dose was 0.5-1.8mg/kg (SD 0.5mg/kg).

43(84.3%) had 6-TGN levels tested after initiation of Azathioprine; 8 did not have their levels tested at all. Median time from initiation of Azathioprine to first testing of 6-TGN levels was 4months (IQR 2.5, 8). In 20patients (39.2%) the first 6TGN level was normal; 16(31.4%) had low levels and 7(13.7%) had high levels. Of those with high levels, 5 patients had raised lipase levels, 5 had raised ALT, 3 had leukopenia, 1 had developed neutropenia and 3 had lymphopenia. The only patient who did not have TPMT levels checked developed acute pancreatitis. Following an abnormal TGN, 17 had their Azathioprine doses reviewed (stopped(2), reduced(4), increased(7) or changed to an alternative medication (4)). For those with abnormal 6-TGN levels, only 15(48%) had their levels rechecked at some point.

Prior to initiation of Azathioprine, EBV was tested in 46(90.2%) of patients. 24 had evidence of past infection; 18 were EBV naive and 3 had equivocal results. After initiation of Azathioprine, 13 were retested. 3 of these had seroconverted.

Conclusion:
Whilst the recommended practice of measuring TPMT is adhered to, there is variability in the dosing schedule and in the frequency of the monitoring of thiopurine metabolite levels.
High thiopurine metabolite levels appears to be associated with abnormalities in liver function tests, bone marrow function and pancreatic enzymes.
EBV status of the patient currently does not seem to influence the choice of immunosuppressant used. It may be useful to look at EBV seroconversion during the course of treatment.

P111

Proactive therapeutic drug monitoring following biosimilar infliximab switch in paediatric inflammatory bowel disease (PIBD)

Kathryn Allan[1], Heidi Pike[1], Sian Kirkham[1], Sabarinathan Loganathan[1], Michalis Papadopoulos[1] and David Devadason[1].

[1]Nottingham University Hospitals NHS Trust

Introduction:

Therapeutic drug monitoring (TDM) and anti-drug antibody (ADA) concentrations now an established strategy to maintain response in patients with Inflammatory Bowel disease (IBD) on biologic agents. Biosimilar infliximab is now the preferred modality across the UK. Data on TDM in patients on biosimilar Infliximab over a prolonged period is lacking, and the benefits, if any, of proactive TDM are unknown.

Aim:

(1) To study the impact of proactive therapeutic drug monitoring on dosing and response to changes; (2) to investigate loss of response as a result of ADA on patients on Remsima (biosimilar Infliximab), (3) to study the impact on therapeutic drug monitoring on PIBD patients who switched from originator Infliximab to Remsima.

Methods:

PIBD patients who were proactively managed with measurement of TDM and ADA on Remsima for a period of at least 12 months were included. Two cohorts were studied (A) those who switched from Remicade and (B) those who were Remicade naïve.

Results:

54 patients (29M/25F) were on Remsima for a period of at least 12 months 37 patients had CD, 7 had UC and 10 had IBDU). 25 patients commenced biological treatment on Remsima. 29 switched from Remicade having been on Remicade for a mean period of 23.9mo (3 – 58 months).

Sub-therapeutic trough levels (<3ug/ml) were detected in 24 patients within 12 months of commencing / switching to Remsima.

In the patient group that was Remicade naïve, the starting dose on all patients was 5mg/kg and the induction regime was at 0w, 2w and 6w. 16 patients had subtherapeutic levels (<3ug/ml) during course of treatment, and 14 resulted in a clinician led decision to alter dose/ interval (56%). Post induction trough levels (at a median of 15w; 6-31w) were available on all patients). 8 patients had dose/interval changed for following the results of the first sub therapeutic post induction levels.

29 patients switched from Remicade. 13/29 were on standard regime (5mg/kg 8w) at the point of switch. 28 patients had levels in the preceding 3 months and also within 6 months of switching to the biosimilar. None of these patients had pre-existing anti TNF antibodies (ADA) at the point of switching. 22 patients had levels within the desired range both before and after the switch. 6 patients had subtherapeutic levels on Remicade that subsequently remained subtherapeutic in 4 patients on Remsima.

7 out of 54 patients patients had at least one measured level of <1ug/ml. 2 of these continued on Remsima and achieved therapeutic drug levels. 1 patient discontinued treatment because of non-attendance. 4 stopped Remsima because of loss of effect within 6 months. 3 had detectable antibodies and 1 did not.

Conclusion:

1. Proactive TDM results in dose/interval alteration for the majority of patients on Remsima.
2. Levels of less than 1ug/ml accurately predict the development of antiTNF antibodies and may be the harbinger of ensuing loss of response.
3. Restricting measurement of antibodies to patients with a measured drug level of <3ug/ml may be cost saving without changing the yield.

P118

Genital Crohn's Disease: a case series in a tertiary Paediatric Gastroenterology Centre

Matilde Pescarin[1], Aruna Sethuraman[1], Edward Gaynor[1], Abraham Cherian[1], Tom Watson[1] and Fevronia Kiparissi[1].
[1]Great Ormond Street Hospital

Background:

Metastatic Crohn's Disease (MCD) affecting the genitalia is a rare condition, characterized by granulomatous inflammation of the genital tract. Only few paediatric cases has been described in the literature with a female preponderance.

Aim:

The aim of the study was to describe the clinical presentation, associated features and response to treatment in a cohort of patients diagnosed in a tertiary Paediatric Gastroenterology centre.

Methods:

All cases of genital MCD were retrospectively collected over a 10-year period from January 2009 to November 2019. Medical history, symptoms at presentation, histological characteristic, results of investigations and response to treatment were reviewed.

Results:

A total of 6 patients, 4 males, were diagnosed with genital MCD. The mean age at presentation of symptoms was 10.6 years of age (8-14 years). 5 patients presented initially with genital disease without any gastrointestinal symptoms and were found on soft tissue biopsies to have non-infectious and non-caseous granulomata. Only one patient received a diagnosis of penile MCD 7 years after the diagnosis of Crohn's Disease. As gastrointestinal Crohn Disease was suspected in all patients, they underwent upper and lower endoscopies with biopsies. 4 patients were found to have gastrointestinal disease compatible with Crohn Disease even if they were asymptomatic. 2 patients are still awaiting for endoscopies. 4 patients were treated with Infliximab infusions, all of them improved the MCD pathology. One patient developed resistance to IFX with antibodies so was switched to Adalimumab injections and one patient did not respond so the treatment was escalated to Adalimumab and Methotrexate.

Conclusions:

MCD involving the genitalia is a rare and challenging diagnosis, usually patients present to Urology or Dermatology who then refer to gastroenterology, this process can take months. Awareness for MCD needs to be raised, referral pathways shortened with an MDT approach; all patients should receive extensive investigations to assess the gastrointestinal tract and start appropriate treatment.

Danone Sponsored Symposium

Abstract

Infant regurgitation: how to manage?

Yvan Vandenplas
KidZ Health Castle, UZ Brussel,Vrije Universiteit Brussel, Brussels, Belgium
yvan.vandenplas@uzbrussel.be

Abstract

Safety and tolerance of a novel anti-regurgitation formula combining carob bean gum, pre- and postbiotics: a double-blind, randomized, controlled trial.

Y. Vandenplas; T. Ludwig; M. Arciszewska; A. Bongers; A. Świat; F. Dakhlia; A. Piollet; R. Oozeer; M. Bellaiche

Background: Regurgitation is the most frequent functional gastrointestinal (GI) disorder in the first year after birth and has been reported to be commonly associated with other GI symptoms. International recommendations emphasize parental reassurance and nutritional advice as the preferred management for uncomplicated regurgitation. This includes optional usage of thickened anti-regurgitation (AR) formulas for non-exclusively breastfed children. Postbiotics derived from the Lactofidus™ fermentation process, and a specific mixture of short chain galacto-oligosaccharides and long chain fructo-oligosaccharides (scGOS/lcFOS, ratio 9:1) have been shown to positively modulate gut microbiota function and alleviate gastrointestinal symptoms such as hard stools.

Objectives and study: To investigate in a double-blind, randomized, controlled trial the GI tolerance and safety of a new AR formula. The novel AR infant formula containing 0.4 g/100 ml carob bean gum (CBG) as thickener, postbiotics (derived from the Lactofidus™ fermentation process) and 0.4 g/100 ml scGOS/lcFOS, was compared to a control AR formula with 0.4 g/100 ml CBG and postbiotics, but without scGOS/lcFOS. The latter has a history of safe usage in multiple countries.

Methods: 182 fully formula fed healthy term infants aged between 3-13 weeks at inclusion and diagnosed with uncomplicated regurgitation based on adapted Rome IV criteria were included after independent ethical committee approval and registration of the study (ClinicalTrials.gov Identifier NCT03371615). The intervention duration was 4 weeks with optional 4 weeks extension. Outcome parameters included Infant Gastrointestinal Symptom Questionnaire (IGSQ) sum score at 4 weeks of intervention, weekly IGSQ score, stool characteristics, growth parameters, (Serious) Adverse Events ((S)AE) and regurgitation severity.

Results: IGSQ sum scores improved for both groups from baseline within one week of intervention. At four weeks of intervention, preliminary uncorrected analysis (n=166, per protocol population) showed equivalence of the IGSQ sum scores of the novel AR formula and control AR formula. Stool characteristics were comparable between both groups with low incidences of diarrhea or hard stools. The (S)AE pattern was not clinically significantly different between the two study groups. Both formulas supported adequate growth. Infants fed the novel AR formula had a decrease in regurgitation severity comparable to the infants in the control group.

Conclusion: The newly-developed infant milk formula combining CBG with scGOS/lcFOS and postbiotics is well-tolerated, safe and supports adequate growth

BSPGHAN

Annual Meeting Awards

Alex Mowat Memorial Prize

The Alex Mowat Memorial Prize was established by his widow Ann and was presented at RCPCH Spring Meeting till 2008 when, with agreement with Ann, the prize was then presented at the BSPGHAN Annual Meeting.

Obituary from The Independent 21st November 1995
https://www.independent.co.uk/news/people/obituary-professor-alex-mowat-1583000.html
Alexander Parker Mowat, paediatrician, hepatologist: born Cullen, Banffshire 5 April 1935; Consultant Paediatrician and Paediatric Hepatologist, King's College Hospital, London 1970-95, Head, Department of Child Health, King's College Hospital 1993-95; Clinical Teacher, London University 1970-95; Professor of Paediatric Hepatology, London University 1990-95, Senior Examiner in Paediatrics 1993-95; married 1961 Ann Hunter (two sons); died Santiago, Chile 11 November 1995.
Alex Mowat had been Professor of Paediatric Hepatology at King's College Hospital, London, and his death represented a great loss to British paediatrics and to the many young patients he helped, both in Britain and throughout the world; he died while on a lecture tour in Chile.

Mowat was proud of his Scottish ancestry and his medical education in Aberdeen. The seeds of his brilliant academic career were sown during clinical appointments in the 1960s in Aberdeen, Hong Kong, and New York and matured in a research post in the Enzymology Department of the Rowett Research Institute, Aberdeen, and during a two-year Training Fellowship with Dr Irwin M. Arias in the Department of Medicine at the Albert Einstein College of Medicine, Yeshiva, New York. These posts gave Mowat an expertise in biochemistry, enzymology and hepatology which formed the basis of great clinical contributions to his chosen specialty of paediatric liver disease and in the care of children in general paediatric medicine. At the very early steps of his career, Alex Mowat met and married Ann Hunter, a continuous source of inspiration, support and love.

In 1970 Mowat was appointed to King's as Consultant Paediatrician and Paediatric Hepatologist, a post which was unique and a timely recognition of a completely new specialty. Although there had previously been no sustained academic interest in liver disorders in children in Britain, Mowat developed a first-class clinical unit for children who suffered with these rare conditions. The clinical work of the unit was backed up at all levels by research into causes and treatment; it needed staff from many disciplines and Mowat forged a team of hepatologists, paediatric and transplant surgeons, radiologists, pathologists, nurse specialists, dieticians and other specialists which had no equal at that time.

In 1986 the unit received official government recognition and funding, thus becoming the first supra-regional centre for the treatment of children with liver disorders from all over Britain. The concentration of the children into one unit increased the knowledge and expertise in management and this was reflected in the improved results which formed the basis of more than 200 publications. Biliary atresia, portal hypertension and liver tumours were some of the conditions which were treated with results which were not surpassed in any centre in the world.

Mowat was supportive of the introduction of new techniques of treatment and this included the development of liver transplantation in children. His unit pioneered the development of auxiliary transplants and the successful introduction of the living-related programme - in which one of the parents gives part of their liver to be transplanted into the child - which has helped to ease the shortage of available organs in transplantation. Last year more than 560 children were admitted with life-threatening liver disorders and over 30 received liver transplants.

The international standing of the unit is remarkable and many of the research projects have been carried out in collaboration with university departments abroad. An example of the value of this work was the discovery of the key role of dietary copper in the causation of Indian Childhood cirrhosis, a finding which has led to the disappearance of the disease in parts of India in which this information has been made known.

The experience from King's was distilled by Mowat into his textbook Liver Disorders in Childhood (1979). The book reached its third edition in 1994 and is generally regarded as the reference book on the subject. Mowat has also been credited with raising the general awareness of his subject by introducing liver medicine into gastroenterological and general paediatric meetings. However his work was not restricted to the confines of the medical profession. In 1980 he encouraged parents of children attending the liver service at King's to develop an association which has become the Children's Liver Disease Foundation, a national charity. This organisation is dedicated to making the problems of children's liver disease more widely known, to improving outcome by funding research and to providing support for affected families. It has raised over pounds 3m.

Academic and clinical work produced other responsibilities for Mowat which he handled with skill. He was Head of the Academic Department of Child Health within the hospital and an examiner for London University and the Royal College of Physicians. He was also Honorary Consultant in Paediatrics to the Royal Air Force and Chairman of the Hospital Consultants' Committee.

Alex Mowat also had a full life outside his work. He loved his golf and taught many friends the art of whisky tasting.
Edward R. Howard and Giorgina Mieli-Vergani

Sean Devane Memorial

The Sean Devane Memorial Prize was established in 2008 with agreement from his widow to acknowledge Sean's contribution to the field of Gastroenterology and to BSPGHAN
In Memoriam Sean Devane, 1956–2008
Greenough, Anne

Journal of Pediatric Gastroenterology and Nutrition: August 2008 - Volume 47 - Issue 2 - p e1

Sean Devane was one of the last of a generation who had interests and expertise in a wide range of specialities. Although his main commitment was to the intensive care of babies on a tertiary neonatal intensive care unit, his other interests included paediatric gastroenterology, the education of medical students, and the administration and monitoring of postgraduate training. Sean's major contribution to the European Society for Paediatric Gastroenterology, Hepatology, and Nutrition was the Web site of the society that he managed and maintained to extremely high standards.

His thoughtful and intelligent advice and support were greatly appreciated by many aspiring paediatric consultants. Sean gained his medical training at the University of Dublin, where he obtained honours in 6 subjects, qualifying in 1980. Over the next 6 years he undertook house officer and senior house officer posts and obtained broad experience in a wide range of paediatrics in Ireland, before coming to the UK as a senior house officer at Cambridge University. He then undertook a research fellowship at the Institute of Child Health under Professor Dame June Lloyd and Dr Peter Milla, which led to his MD thesis. In 1989 he was appointed lecturer in Child Health at King's College School of Medicine and Dentistry at King's College Hospital, and in 1993 consultant to the tertiary neonatal intensive care unit (Frederic Still Ward) at King's College Hospital with a particular interest in developmental gastroenterology.

Sean's numerous strengths as a consultant included his abilities to relate to all levels of staff and provide efficient administrative structures to teaching and training programmes (where many others had failed), but above all, to bring calm and sense to troubled situations. Sean coped with his prolonged and painful illness with a bravery that humbled all of us. He is survived by his wife Stephanie and his 2 sons, Eoin and Aidan, his family, his many friends and colleagues, particularly on "Fred Still," and the babies and families whom he looked after.

Previous Prize winners

2008 Southampton

Alex Mowat Prize:	Dr Andrew Barclay
Best Abstract Presentation:	Ms Elaine Buchanan
Best Presentation:	Dr Sherina Ross

2009 Sheffield

Alex Mowat Prize:	Dr Johann van Limbergen
Sean Devane Memorial:	Dr Jenny Epstein
Best Allied Health Professional:	Ms Jackie Falconer

2010 Liverpool

Alex Mowat Prize:	Dr Emer Fitzpatrick
Sean Devane Memorial:	Dr Rachael Taylor
Best Poster Presentation:	Dr Paul Henderson

2011 Edinburgh

Alex Mowat Prize:	Dr Paul Henderson
Sean Devane Memorial:	Dr Emer Fitzpatrick
Best Poster Prize:	Ms Helen French

2012 Nottingham

Alex Mowat Prize:	Mark Goddard
Sean Devane Memorial:	Anna Gregory
Challenging Case:	Lisa Whyte
Best Poster:	Ms Hannah Williamson

2013 Manchester

Alex Mowat Prize:	Dr Protima Amon
Sean Devane Memorial:	Dr Lisa Whyte
Best Poster Prize:	Dr Rana Bitar

2014 London

Alex Mowat Prize:	Dr Vandana Jain
Sean Devane Memorial:	Dr Ed Giles
Best Poster Prize:	Dr Bradley Keller

2015 Stratford upon Avon

Alex Mowat Prize:	Dr Mona Abdel-Hady
Sean Devane Memorial:	Dr Kelsey Jones
Best Poster Prize:	Sarah Macdonald and Katherine Fawbert
Best PICO Presentation:	Dr Huey Miin Lee

2016 Bristol

Alex Mowat Prize:	Dr Nicola Ruth
Sean Devane Memorial:	Y Koh
Best Poster Prize:	Martin Lister

2017 Glasgow

Alex Mowat Prize:	Dr Suzanne Davison
Sean Devane Memoria:	Dr Neil Chanchlani
Best Poster:	Chris Smith

2018 Leeds

Alex Mowat Prize:	David Wands
Sean Devane Memorial:	Natasha Burgess
Best Poster:	Dr Ozan Hanci

2019 Oxford:

Alex Mowat Prize:	Joost van Haasteeren
Sean Devane Memorial:	Chris Bakewell
Best Poster:	Dr Eleni Athanasakos

Dr Falk Pharma Award established in 2018

£200 voucher and free registration to one of their European Symposium, with registration, travel and accommodation expenses

Leeds 2018	Dr Sik Yong
Oxford 2019	Annette Mulcahy

Thanks to everyone who helped score and select abstracts for presentation at the meeting and those who judged abstracts at the meeting

Abstract Selection Panel:
Dr Loveday Jago, Chair BSPGHAN Education Group
Dr David Campbell, Chair BSPGHAN Research Group
Dr Jeng Cheng, BSPGHAN Education Trainee Representative
Ms Nicky Heather, Chair of BSPGHAN Associate Members' Group
Dr Michael Hii, Consultant Paediatric Gastroenterologist, Brighton
Dr Hina Rizvi, BSPGHAN Education Trainee Representatives
Mr Chris Smith, Education Rep Associate Members' Group
Dr Indra van Mourik, Chair of BSPGHAN Hepatology Group

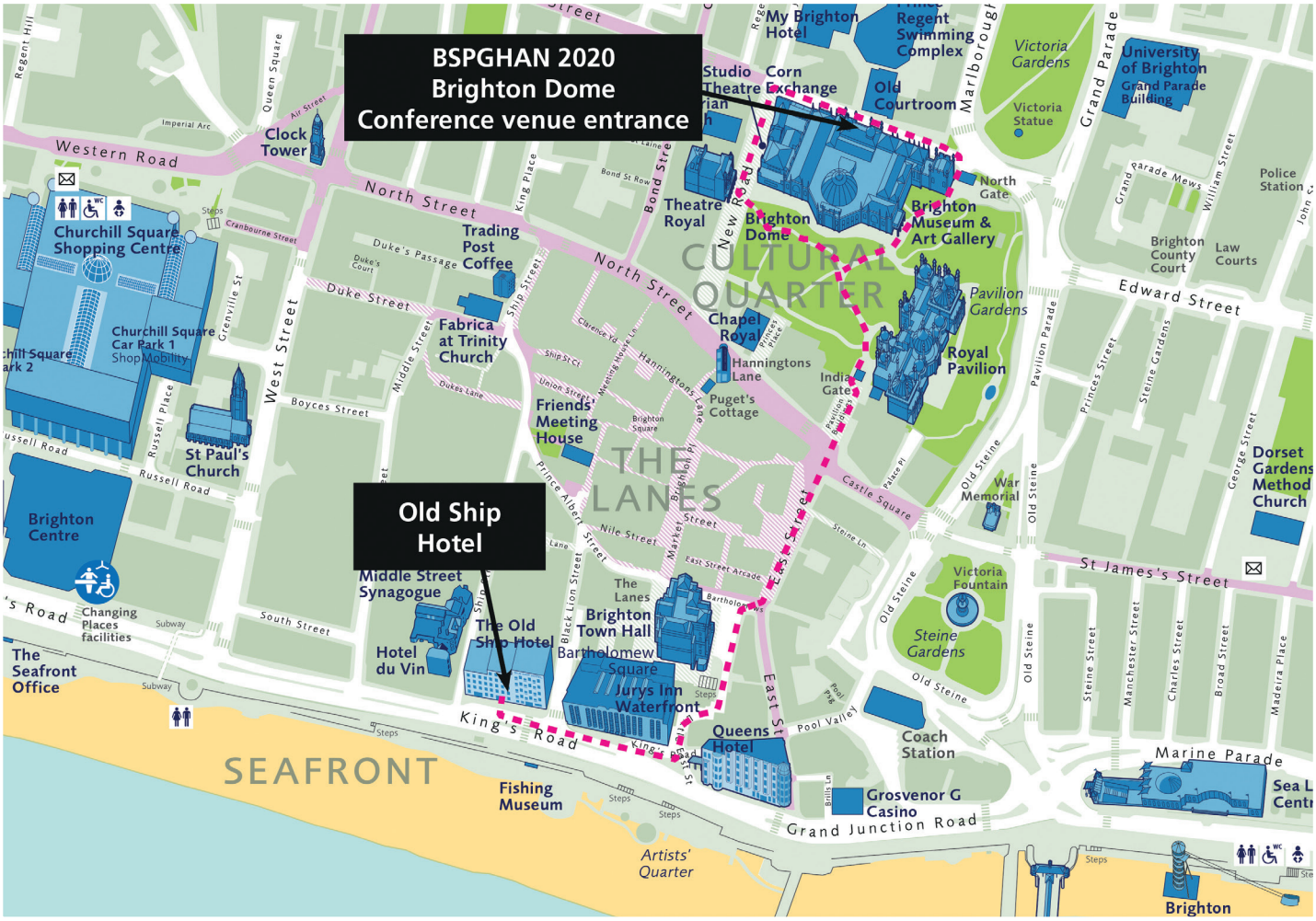
Plenary Judges:
Dr Akshay Batra, BSPGHAN Nutrition Working Group Chair, Consultant Paediatric Gastroenterologist, Southampton
Dr Jeng Cheng, BSPGHAN Education Trainee Representative
Ms Nicky Heather, Chair of BSPGHAN Associate Members' Group, Dietitian, Southampton
Dr Kwang Yang Lee, Specialist Grid Trainee, BSPGHAN Webmaster,Birmingham
Ms Minal Patel, Paediatric Dietitian, Royal London Hospital
Dr Ros Rabone, Chair BSPGHAN Trainee Members' Group, SpR, Sheffield
Dr Astor Rodrigues, BSPGHAN Treasurer, Consultant Paediatric Gastroenterologist, Oxford
Dr Indra van Mourik, Chair of BSPGHAN Hepatology Group, Consultant Paediatric Hepatologist, Birmingham,
Mr Chris Smith, Education Representative Associate Members' Group, Dietitian, Brighton

BSPGHAN Poster Judges

Dr Marcus Auth, Consultant Paediatric Gastroenterologist, Liverpool
Dr Andrew Barclay, Consultant Paediatric Gastroenterologist, Glasgow
Dr Ronald Bremner, Consultant Paediatric Gastroenterologist, Birmingham
Dr Su Bunn, Consultant Paediatric Gastroenterologist, Aberdeen
Dr David Campbell, Consultant Paediatric Gastroenterologist, Sheffield
Dr Protima Deb, Consultant Paediatric Gastroenterologist, London
Dr Lucy Howarth, Consultant Paediatric Gastroenterologist, Oxford
Dr Sally Mitton, Consultant Paediatric Gastroenterologist, London
Dr Sandhia Naik, Consultant Paediatric Gastroenterologist, London
Dr Siba Paul, Consultant Paediatrician, Exeter
Dr Anna Pigott, Consultant Paediatrician, North Staffordshire
Mrs Catherine Richards, Consultant Paediatric Surgeon, London
Dr Astor Rodrigues, Consultant Paediatric Gastroenterologist, Oxford
Dr Chris Spray, Consultant Paediatric Gastroenterologist, Bristol
Dr Sophie Velleman, Consultant Psychologist, Bristol
Dr Adrian Thomas, Consultant Paediatric Gastroenterologist, Manchester
Dr Anthony Wiskin, Consultant Paediatric Gastroenterologist, Bristol
Dr Anne Willmott, Consultant Paediatric Gastroenterologist, Leicester

Dr Falk IBD Poster Awards

Dr Rafeeq Muhammed, Chair, Consultant Paediatric Gastroenterologist, Birmingham, Chair of BSPGHAN IBD Working Group
Dr Jochen Kammermeier, Consultant Paediatric Gastroenterologist, London
Dr Amar Wahid, Consultant Paediatric Gastroenterologist, Cardiff
Dr Mashood Ayaz, Consultant Paediatrician, Surrey
Ms Janis Maginnis, IBD Nurse, Staffs



Exhibitors:

Laborie Medical Technologies UK
www.laborie.com

Ferring
www.ferring.com

Pfizer
www.pfizer.co.uk

Vertex
www.vrtx.com

Nutricia Danone
www.nutricia.com

Special Charity Exhibitor:

tofs.uk