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ANNUAL MEETING 2018

WEDNESDAY 24TH – FRIDAY 26TH JANUARY. THE QUEENS HOTEL, CITY SQUARE, LEEDS

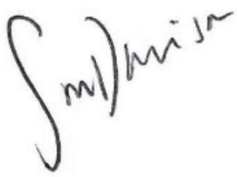
ABSTRACTS

County Arcade, Leeds

Thanks to all of our sponsors

On behalf of BSPGHAN and the local organising committee for Leeds 2018, I would like to express sincere gratitude to all who contribute to the Annual Meeting. In particular, to our commercial sponsors who not only provide financial support, but through their participation provide opportunities for us to share and acquire knowledge, provide a platform to stimulate research and future development, and enrich the meeting through their single topic symposia.

I also wish to acknowledge the valuable role that Charitable Organisations fulfil. Their partnership with the Society and the Annual Meeting provides a vital interface with families and the wider community, updates us on current issues and priorities, and guides us in maintaining a strategy that is child and family focussed.



Suzanne Davison
Local Organiser, BSPGHAN Leeds 2018

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24TH - 26TH JAN 2018

Plenary Abstracts
Wednesday 24th January 2018

Group sessions are a good medium for educating patients with coeliac disease

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Introduction: Prior to March 2016 the newly diagnosed coeliacs were seen in the dietetic gastroenterology clinic for a 30 minute slot. Finding extra clinic slots for the increasing number of children being referred was difficult so group sessions were introduced in March 2016. Until October 2017 6 families/month were invited to the group session and from November 2017 the number was increased to 10 families/month.

Aims and Objectives: To assess the impact of educating children and their families on the gluten free diet in a group session.

Subjects and Methods: Children with behavioural or learning difficulties or with parents who need an interpreter are excluded. These patients are offered a standard clinic appointment. The group session invitation advises parents that they are attending a group session and asks them to contact us if they do not consent to the group session. These patients will also be offered a standard clinic appointment.

Group sessions are held monthly and last for 1.5 hours. They are run by a dietitian and dietetic assistant.

A room is used that can hold at least 30 people so children can bring additional members of their family who may be involved in food provision such as grandparents.

A standardised presentation devised from the Dr Schär Institute presentation teaching pack. This includes endoscopy pictures and useful apps to demonstrate in the session. Parents are asked to add their own practical knowledge and active discussions are encouraged between families.

At each session the dietetic assistant supervises the younger children and gives them a picture specially designed by our medical illustrations department that contains gluten and non-gluten containing food for them to colour in during the session. She also works with them to create a plate of their favourite food and discusses alternatives

Results: Until November 2017, 83 children have attended the sessions. The practical session allows the children to learn in a fun way so their parents can focus on the dietitian's presentation. Parents can exchange contact details with other parents for support and the children can meet others with their condition.
We have had very positive feedback such as "we wish we'd had this when she was diagnosed", they felt the dietitian was very knowledgeable and they enjoyed the session and will continue to offer this as service.

Summary and Conclusion: The group sessions are an effective medium for educating children and their families on the gluten free diet and the experience of attending the session is a much better experience for the child rather than in a clinic room. There are health economics benefits of the group session because they have a 1.5 hour session rather than 30 minutes session. Offering 6 places per month increased our capacity by 100% and 10 places increased our capacity by 233% without increasing the frequency of our clinic sessions

An open label pilot study to determine whether a diet low in short chain carbohydrates reduces symptoms in children with functional gastrointestinal disorders.

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Introduction: Short chain fermentable carbohydrates are widespread in the diet, comprising of monosaccharaides (fructose), disaccharides (lactose) oligosaccharides (fructans and galactans) and polyols termed FODMAPS. FODMAPS are incompletely absorbed and lead to physiological effects in the gastrointestinal tract that exacerbate symptoms in susceptible individuals. Clinical demonstrations have shown that FODMAPS induce gastrointestinal symptoms. The low FODMAP diet is a nutritionally balanced elimination diet of short chain carbohydrates, which has been used to resolve functional bowel disorders (FGIDs) in adults with Irritable bowel syndrome (IBS) for more than 10 years. Most patients with FGIDs try many elimination diets which often prove to be nutritionally imbalanced due to omitting major food groups. Current NICE and BDA (British Dietetic Association) guidelines for adults with IBS recommend the low FODMAP diet after general lifestyle and dietary advice.

Aims and Objectives:

1) To determine whether the implementation of a low FODMAP diet in children with functional gastrointestinal disorders reduces symptoms in a group of patients after a 2 week period.
2) Whether the low FODMAP diet provides participants with satisfactory relief of their gut symptoms after a 2 week period.

Subjects and Methods: Children aged 5-16 were identified by screening referrals and preliminary assessments in a dietetic outpatient's clinic from August 2016 - November 2017 for suitability of the low FODMAP diet. Twelve participants/carers with either Rome criteria IV-defined IBS or functional abdominal pain – not otherwise specified alongside clinical judgement were invited to complete an arbitrary 2 week trial of the low FODMAP diet. Symptoms were captured using a subjective visual aid scale (VAS) measuring symptom severity 0-10cm. Participants/carers completed the VAS pre and post the low FODMAP dietary intervention. Symptoms measured were categorised into primary and secondary outcomes. Primary outcomes included; abdominal pain (discomfort/distention), wind, belching, stomach gurgling and satisfactory control over gut symptoms. Secondary outcomes captured urgency to open bowels, incomplete evacuation, nausea, acid regurgitation and lethargy/ tiredness. Stool type was not assessed however all participants had either loose stools (types 5, 6, 7) or a mixed stool (type 2 and 6). Low FODMAP dietary advice was provided and a 2- 3 week future clinic appointment was made for re- introduction advice. A further dietetic assessment was offered during and at completion of the reintroduction diet at which point a probiotic was recommended.

Results: Twelve participants agreed to be part of the study, 9 of which completed the elimination diet and returned to clinic for reintroduction advice. One participant moved out of area, 1 participant unknowingly did not follow the low FODMAP diet and 1 participants' follow up appointment was not timely enough for his results to be included in this study. Five boys (56%) mean age: 9.8yrs and 4 girls (44%) mean age: 11.25yrs completed the diet. The pre and post dietary intervention VAS evaluations were measured, and the difference between them was calculated as the percentage improvement. Symptoms improved in both primary and secondary outcomes for all participants. (Fig 1)

Figure 1

Percentage improvement difference pre and post symptom evaluation										
Abdo. pain	Abdo. distention	Wind/ flatulence	Belching/ burping	Stomach gurgling	Urgency to open bowels	Incomplete evacuation	Nausea	Acid regurg.	Tired lethargic	Satisfaction over gut symptoms
73.8%	80.6%	48%	80.5%	92.8%	70.2%	73.5	87.5%	87.4%	67.3%	83.9%

Summary and Conclusion: This study demonstrates that in child/adolescent IBS and functional abdominal pain – not otherwise specified alongside clinical judgement, a low FODMAP diet decreases symptoms and increases satisfactory relief over gut symptoms. The rates of efficacy in the low FODMAP diet in adults has consistently been approximately 75% for clinical reduction in symptoms, results from the present study indicate similar results. Current research into this area is limited; however this study begins to throw light upon dietary factors and the implication of a dietary intervention in pursuit of future research for paediatric FGIDs.

Targeting Biological treatment in children using drug levels improves patient outcomes

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Background: There remains no current paediatric consensus whether therapeutic drug monitoring for biological therapy should be part of standard practice. Increasing evidence suggest in the long term use of targeted treatment using drug levels is ultimately cost effective and improves quality of care and reduces disease burden. [(1)]

Aims and Objectives: To determine whether use of drug levels significantly changed patient management and improved patient outcomes in children with IBD. Anti-TNF drug levels have been available in our institution since November 2014.

Subjects and Methods: Demographic data was obtained from Cerner Millenium.
Clinical outcomes were collated from electronic records and weekly virtual biologics rounds.
Infliximab levels and antibodies were measured using Theradiag assay.
Indications for measuring levels were poor, no response, or suspected loss of response based on activity score, biochemical markers, faecal calprotectin and reported symptoms.

Results: November 2014 - November 2017 102 children received Infliximab

Male:Female 54%:46%.
Data expressed median (range)
Age median 13 years (7 -17)
Crohn's disease (CD) n=79
Ulcerative Colitis (UC) n=19
IBDU n=4

Current patients on Infliximab 42, length of treatment 13 months (1-54)
Current total infusions 280 infusions in last 12 months (median per patient 5)
Paediatric drug levels n =59

14% developed antibodies and switched treatment with 100% response
29/59 escalated their treatment - 66% responded (n=19)
8% switched due to secondary loss of response- 50% subsequent response
7% of patients discontinued treatment due to adverse effects:
Anaphylaxis (n=6) psoriasis (n=2)
3% had subtotal colectomies for UC after failing medical treatment
2 children switched to Vedolizumab - ongoing

Conclusion: Targeted treatment of children with IBD using drug levels leads to an improved response and remission rate. Cost benefit in the longer term can be assessed by objective measures such as admission rates, complications, reduction in number of investigations for example MRI or colonoscopies.

Therapeutic drug monitoring for biological and biosimilars should be part of standard paediatric IBD care.

[1]Inflamm Bowel Dis 2017 Dec; 23(12):2083-2088 Selinger et al

Long term safety and efficacy of single dose parenteral iron in children with inflammatory bowel disease (IBD) in one of the largest tertiary centres in North England.

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Introduction: Iron-deficiency anaemia is a common complication amongst children with inflammatory bowel disease and it can have a significant impact on the quality of their lives. Although single-dose parenteral iron preparations are an easily available treatment for children, there are still concerns surrounding its adverse reactions. There are not many longitudinal studies showing its sustained efficacy and effects.

Aims and Objectives: The primary aim of this study was to evaluate the safety, side effects and efficacy of IV iron maltoside 1000 (Monofer®) at 6 weeks, 3 months, 6 months and 1 year after treatment in children with IBD. The secondary aim was to identify any evidence of iron overload.

Subject and Methods: A comprehensive search was performed using the hospital's IBD database to identify patients who have been given parenteral iron from 2012 to 2016. Primary indication, underlying diagnosis, dose of iron (mg), adverse reactions and laboratory values before and after treatment were among the parameters recorded. Dose calculations were based on the Ganzoni formula. Parenteral iron was used only if oral iron therapy was ineffective (<20g/L rise in 3 months), not tolerated, not advisable (IBD unstable) or iron-deficiency anaemia with haemoglobin levels of <100 g/L. Repeated measures ANOVA was conducted for statistical analysis.

Results: A total of 27 patients were identified. The median age was 15 and median weight was 33.9 kg. Repeated-measures ANOVA conducted on 27 patients showed that mean haemoglobin differed significantly between time points [$F(4, 104) = 29.416$, $p < .001$]. Normality checks were carried out and were approximately normally distributed. Post-hoc tests using the Bonferroni correction revealed that mean haemoglobin increased significantly by 6 weeks and remained stable thereafter ($p < .001$).

Only one patient had an acute type 1 allergic reactions but did not fulfil the criteria for anaphylaxis. Two patients had evidence of hair loss at 3 months post-infusion. This reversed in the first child with the use of biotin for 3 months and in the second child, the hair loss reversed with the stoppage of azathioprine. Hence, these are unlikely to be secondary to iron overload. None of the patients had evidence of dysmetabolic iron overload syndrome (DIOS). All children had normal LFTs and GGTs on follow up biochemistry with no evidence of diabetes, chronic fatigue or hepatosplenomegaly within the notes of their follow up consultations.

Summary: Parenteral iron appears to have sustained efficacy in the treatment of iron deficiency anaemia in children with IBD. Iron status increased significantly at 6 weeks and sustained till 1-year post-infusion. The immediate reaction rate was 3.7% and none of the remaining 26 patients had any long-term or short-term side effects including any evidence of DIOS.

Conclusion: Parenteral iron appears to be safe and has sustained efficacy in the treatment of iron-deficiency anaemia in children with IBD.

Case report: "Skin lesions in Paediatric Crohn's disease: thinking beyond Erythema Nodosum."

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Introduction: Extraintestinal conditions associated with Inflammatory Bowel Disease (IBD) are identified in about 5-7% of children at presentation, and up to 30% within the first few years after diagnosis.[1,2] Erythema nodosum and pyoderma gangrenosum are well recognized manifestations of paediatric IBD.[1,2] We describe a rarer cutaneous manifestation of IBD in a 12 year old boy with newly diagnosed Crohn's disease.

Aims and Objectives: To raise awareness of a rare cutaneous manifestation of Crohn's disease in a child.

Subjects and Methods: A 12 year old boy presented with 24 months of intermittent abdominal pain, faltering growth and altered stooling. Based on positive serological and genetic markers, a diagnosis of coeliac disease was made. Despite a gluten free diet his symptoms persisted. He presented acutely with a rash to his lower limbs, initially treated as cellultis and later described as erythema nodosum. He was referred for investigation for IBD. The endoscopic findings and biopsy results were typical for Crohn's disease.

Results: The skin rash worsened and spread despite topical steroid treatment. Dermatological review suggested erythema nodosum but it was also noted that he had subcutaneous nodules on his knees, elbows and forearms which raised suspicion of histiocytoid sweet syndrome (H-SS). A skin biopsy confirmed this diagnosis.

Due to failure to induce gastrointestinal remission with exclusive enteral nutrition (EEN) the patient was commenced on biological therapy with Infliximab and azathioprine. His skin lesions responded to this in combination with a further course of more potent topical steroids.

Summary: Sweet syndrome (SS) is characterised by acute onset of painful nodules or erythematous/violaceous plaques on the limbs, face and neck, usually accompanied by fever. Histopathological diagnosis is based on dermal neutrophilic infiltration. Histiocytoid Sweet syndrome (H-SS) is a histological variant, mainly composed of lymphocytes and histiocytoid myeloperoxidase-positive cells.

SS is associated with infection, malignancies, autoimmune disease, inflammatory bowel disease, pregnancy and drugs. Paediatric SS is uncommon, most frequently associated with infectious diseases. The histiocytoid type (H-SS) is even rarer and to date, only six cases of H-SS have been reported in children worldwide[3,4], only one of which was associated with IBD[4].

Conclusion: Knowledge of skin lesions associated with inflammatory bowel disease is important in Paediatric Gastroenterology as it may be the first manifestation of the underlying disease process. The chronology of the skin lesions tends to follow that of the underlying bowel disease with healing occurring in tandem with disease remission.

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HLA-DQ2/DQ8 typing for non-biopsy diagnosis – is it necessary?

Paul, SP[1], Consultant Paediatrician; Chopra, J[2], 5th Year Medical Student; Vaina, C[3], Consultant Paediatrician; Basude, D[4], Consultant Paediatric Gastroenterologist;
[1]Torbay Hospital, Torquay; [2]Medical School, University of Bristol; [3]Yeovil District Hospital; [4]Bristol Royal Hospital for Children;

Introduction: Non-biopsy pathway of diagnosis of coeliac disease (CD) was implemented in the SW England in May 2013. This requires anti-tissue transglutaminase (anti-tTG) titre greater than 10 times the upper limit of normal (ULN), positive anti Endomysial antibody (EMA) and positive HLA-DQ2/DQ8 haplotype. While the HLA-DQ2/DQ8 positive status in symptomatic children with anti-tTG >10xULN is considered necessary for non-biopsy diagnosis of CD, it has caused potential confusion and risk of misdiagnosis of CD if used inappropriately.

Aims and Objectives: This clinical study was set with the following objectives:

(1) Identify the symptomatic paediatric patients in SW England diagnosed with CD via the non-biopsy pathway since May 2013.
(2) Determine HLA-DQ2/DQ8 status in these patients
(3) Final diagnosis when HLA DQ2/DQ8 was negative in these patients.
(4) Feasibility of withdrawing HLA-DQ2/DQ8 testing from the non-biopsy pathway.

Subjects and Methods: Cases were identified from the electronic non-biopsy pathway register kept at the Bristol Royal Hospital for Children (BRHC) which was updated based on voluntary reporting of cases diagnosed serologically in BRHC and DGHs. The endoscopy register from BRHC was cross-checked for symptomatic cases with anti-tTG>10xULN but had negative HLA-DQ2/8.

Results: HLA-DQ2/DQ8 results were available for 96/110 patients. 95/96 patients (99.0%) were positive for HLA-DQ2/DQ8 (figure 5). Of these, 90/95 (94.7%) were HLA-DQ2 positive, 18/95 (18.9%) were HLA-DQ8 positive and 13/95 (13.7%) carried both haplotypes. For the remaining 14/110 patients, HLADQ2/DQ8 typing was not requested or not reported, but a diagnosis of CD was confirmed serologically nonetheless. One patient was negative for HLA-DQ2/DQ8, however subsequent small-bowel biopsy confirmed CD histologically of Marsh classification 3b.

Conclusion: We conclude that identification of the HLA DQ2/DQ8 status did not contribute towards confirming the CD diagnoses. Provided the high prevalence of HLA-DQ2/DQ8 in the general population, dangers of misuse and misinterpretations, alongside the significantly higher costs, we suggest consideration towards the removal of HLA-DQ2/DQ8 testing from the non-biopsy serological diagnostic criteria for CD. To clarify this further we propose a prospective national survey through the BSPGHAN to report HLA-DQ2/DQ8 negative cases in symptomatic children who are anti-tTG >10xULN and EMA positive.

Plenary Abstracts
Thursday
25th January 2018

Validation of Direct Observation of Procedural Skills (DOPS) for Paediatric Gastroscopy

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Introduction: Direct observation of procedural skills (DOPS) are tools designed by the Joint Advisory Group (JAG) to assess competence in endoscopy. These were expanded in July 2016 (new DOPS) to include those specific to paediatric gastroscopy (OGD). However, paediatric OGDDOPS assessments have not been validated.

Aims and Objectives: To correlate overall trainee competence with components of the paediatric OGD DOPS.

Subjects and Methods: We performed a prospective UK-wide analysis of formative paediatric OGDDOPS submitted to the JETS e-Portfolio over one-year (August 2016-2017). Scores were averaged across procedural domains (pre-procedural, procedural, post-procedural and endoscopic non-technical skills – ENTS). Each DOPS item, except for ENTS, were grouped into cognitive and technical skillsets by two independent investigators, and correlated with the overall performance score.Correlation analyses were performed using Spearman's test (rho >0.70 indicating high positive correlation).

Results: 157 DOPS assessments were completed by 20 unique trainers for 17 trainees. Overall performance score comprised: 1: Maximal supervision (4.5%), 2: Significant supervision (14.0%), 3: Minimal supervision (24.8%) and 4: Competent (56.7%). By domain, overall competence correlated most with mean scores for the 'Insertion and Withdrawal' domain (rho: 0.884, p<0.001), followed by 'Management' (rho 0.834, p<0.001), 'Visualisation' (rho 0.819, p<0.001), ENTS (0.773, p<0.001), 'Post-procedural' (rho 0.611, p<0.001) and 'Pre-procedural' (rho 0.575, p<0.001) domains.By skillset, overall score correlated most with performance in 'Technical'(rho 0.860, p<0.001), followed by ENTS and 'Cognitive' domains (rho 0.788, p<0.001) domains. There was strong correlation between cognitive and ENTS skillsets (rho 0.852, p<0.001). In terms of DOPS items, overall competence score correlated most with 'Management of Complications' (rho 0.852, p<0.001) and 'Angulation and Tip Control' (rho 0.834, p<0.001), and least with 'Confirms Consent' (rho 0.396, p<0.001) and 'Equipment Check' (rho 0.528, p<0.001).

Summary: Our data identify the aspects of OGD which assessors relate most closely with overall competence. In OGD, performance in the 'Insertion and Withdrawal' domain, 'Management of Complications' items, and ''Technical' skillsets had greatest correlation with overall procedural competence.

Conclusion: Competencies in paediatric OGD, as assessed within DOPS, vary in their correlation with overall competence. As assessors are completing the new DOPS in a consistent manner, this provides novel validity evidence for the new paediatric OGD DOPS.

Electrolyte Abnormalities in Preterm Neonates Receiving Electrolyte-Free Versus Standard Parenteral Nutrition Solutions: A Two-Centre Retrospective Study

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Introduction: There is current debate as to whether "electrolyte-free" parenteral nutrition (PN) solutions should be used for preterm infants during the first days of PN provision. Electrolyte-free PN does not contain Na+, K+, Cl-, Ca2+, PO4, or Mg2+ and is preferred by some to restrict early Na+ intakes during the postnatal transitional period of extracellular fluid contraction and physiological weight loss. Yet sufficient early electrolyte supplementation is needed for adequate protein accretion when delivering higher amounts of amino acids. In the East of England region some centres routinely use electrolyte-free PN while others use electrolyte-supplemented PN from birth.

Aims and Objectives: We compared the incidence of electrolyte disturbances of Na+, K+, Ca2+ and PO4 in preterm infants according to whether they received electrolyte-free PN or standard (electrolyte-supplemented) PN. Our hypothesis was that there would be no difference in incidences of hypernatraemia or hyperkalaemia during the first postnatal week.

Subjects and Methods: Retrospective cohort study done at two UK tertiary-level neonatal units. Centre 1 routinely uses electrolyte-free PN in the first 48-72h after birth, while centre 2 routinely uses standard PN. We included preterm neonates <36 weeks' gestational age who started on PN within 24 hours of birth within two discrete 6-month epochs.

Outcome measures were first-week peak and nadir serum Na+, K+, Ca2+, and PO4 concentrations; and proportions with hypernatraemia (Na+ >150 mmol/L), hyponatraemia (Na+ <130 mmol/L), hyperkalaemia (K+ >7.0 mmol/L), hypokalaemia (K+ <3.5 mmol/L), hypercalcaemia (Ca2+ >3.0 mmol/L), and hypophosphatemia (PO4 <1.5 mmol/L).

Results: A total of 81 patients (n=43 centre 1; n=38 centre 2) were included. Median (IQR) gestational age was 27.1 (25.7-29.2) weeks and mean (SD) birth weight was 986 (321) g, with no significant demographic differences between centres. More babies who received electrolyte-free PN in the first week (centre 1) had hyperkalaemia (12% vs 0%, p=0.03), hyponatraemia (28% vs 3%, p=0.001), hypokalaemia, (77% vs 40%, p=0.001), and hypophosphataemia (81% vs 37%, p=0.002) compared with standard PN babies (centre 2). Nadir Na+, K+, and PO4 concentrations were also significantly lower in babies who received electrolyte-free PN.

Comparative rates of hypernatraemia (16% vs 34%, p=0.06), hypercalcaemia (18% vs 21%, p=0.8), and median peak Na+ (147 mmol/L vs 148 mmol/L, p=0.6) and Ca2+ (2.9 mmol/L vs 2.8 mmol/L, p=0.3) concentrations were similar.

Summary: Use of electrolyte-free PN within the first 2-3 days after birth was not associated with significantly lower rates of first-week hypernatraemia, but was instead associated with higher rates of hyponatraemia, hyperkalaemia, hypokalaemia and hypophosphataemia.

Conclusion: Routine use of electrolyte-free PN solutions may not be optimal for preterm neonates in the first week.

Paediatric Crohn's disease patients in remission have a reduced skeletal muscle protein balance after feeding.

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Introduction: Sarcopenia is common in active Crohn's disease (CD) & still prevalent in remission. This can lead to fatigue, physical inactivity and poor quality of life. The aetiology is unclear. Low levels of physical activity, inability to respond to anabolic stimuli such as food (anabolic resistance, AR), & insulin resistance (IR) could all be implicated in the failure of CD patients in remission to re-build muscle mass.

Aims and Objectives: To investigate the association between sarcopenia and AR & IR and the role of physical activity in age, gender matched children with CD.

Subjects and Methods: 18 fasted, male & female CD patients (on thiopurines ± anti-TNFα) in deep remission (16y, BMI = 21) and 9 matched controls (Con) (16y, BMI = 21) drank a liquid meal (Ensure plus, 44g CHO, 14g PRO, 11g fat) at t = 0. Arterialised hand & venous forearm blood samples were collected concurrently and brachial artery blood flow measured at baseline & every 20 mins for 2 hrs. Net balance of branched chain amino acids (BCAA) and glucose were derived, providing indices of skeletal muscle protein balance and IR. Participants also had a DEXA scan & handgrip dynamometer test on the day, and wore a pedometer & completed a food diary (each for 3 days), to assess physical activity & food intake. Patient-related outcome measure questionnaires, including IBD-fatigue, were completed.

Results: CD and Con exhibited an initial response to feeding by increasing BCAA flux: Con from 0.3 ± 0.5 µmol/min at t = 0 to 1.1 ± 0.7 µmol/min at t = 20, and CD from -0.8 ± 0.4 µmol/min at t = 0 to 0.8 ± 0.3 µmol/min at t = 20. This positive response was only sustained beyond t = 60 in Con, such that net BCAA balance across the 2 hrs was lower in CD (0.6 ± 0.3 vs -0.1 ± 0.2 µmol/min, respectively, p=0.05). IBD-fatigue scores indicated CD suffer from moderate fatigue (6.2), which had a moderate effect on daily activities (16.7). Handgrip dynamometer testing showed a trend towards greater fatigue in CD vs Con (+8 %points) in the dominant arm (p=0.061). A trend towards lower total body lean mass in CD (-15%, p=0.084) was found. No differences were detected in strength, physical activity, diet or IS.

Summary: Despite not exhibiting AR, as they initially responded to the meal stimulus, CD could not maintain a positive protein balance post feeding compared to Con. This was associated with reduced muscle mass and function.

Conclusion: The inability to sustain a positive protein balance post-prandially could provide an explanation for the reduced muscle mass seen in CD patients in remission. This could be contributing to fatigue and poor muscle function. Pharmacological interventions to reduce protein breakdown and a high protein diet to improve the anabolic response to food could both be investigated as potential treatments.

Investigating CD8+ T-cell gene expression signatures as potential prognostic biomarker in paediatric inflammatory bowel disease.

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Introduction: Inflammatory Bowel Diseases (IBD) are characterized by a relapsing-remitting disease course, with severity varying substantially amongst patients. At present, no reliable prognostic biomarkers are available for clinical practice. Previous work on adult patients identified CD8 positive T-cell gene expression as a promising prognostic biomarker in IBD.

Aims and Objectives: To investigate CD8+ T-cell gene expression as a prognostic biomarker in children newly diagnosed with IBD.

Subjects and Methods: 112 children (age 5-16) were prospectively recruited at diagnosis and CD8+ T-cells isolated from a peripheral blood sample using magnetic bead sorting. Following, RNA was extracted and genome wide expression profiling performed on Affymetrix Human Gene ST 2.0 Arrays. Detailed clinical phenotype and disease outcome data was recorded for all patients (minimum follow-up: 1.5 years). Bioinformatic analysis were performed using "R" and various Bioconductor packages and included consensus clustering, differential gene expression analysis (DGEA), survival analysis (Kaplan Meier), and weighted gene co-expression network analysis (WGCNA).

Results: Preliminary results are currently available from a discovery cohort of 42 children (22 Crohn's (CD), 20 ulcerative colitis (UC). Unsupervised consensus clustering identified two main groups indicating distinct differences in global CD8 expression signatures amongst IBD patients. DGEA between these groups revealed 1324 annotatable genes, 20% of which were found to overlap with the prognostic expression signatures previously identified in an adult IBD cohort. WGCNA performed separately in children with CD (n=22) and UC (n=20) identified a number of highly significant modules (groups of genes), some of which correlated strongly (Pearson correlation index >0.6, $p<0.05$) with individual outcomes including number of treatment escalations, use of biologics and surgical interventions. Strikingly, performing unsupervised hierarchical clustering using genes derived from these modules followed by Kaplan Mayer survival analyses revealed highly significant differences in disease outcome between patient groups (Figure).

Conclusion: Our preliminary results derived from a subset of available patients supports CD8+ T-cell gene expression signatures as a promising prognostic biomarker for children newly diagnosed with IBD. Interestingly, a limited overlap with a previously identified adult biomarker suggests distinct differences between adult and paediatric onset IBD.

Figure.

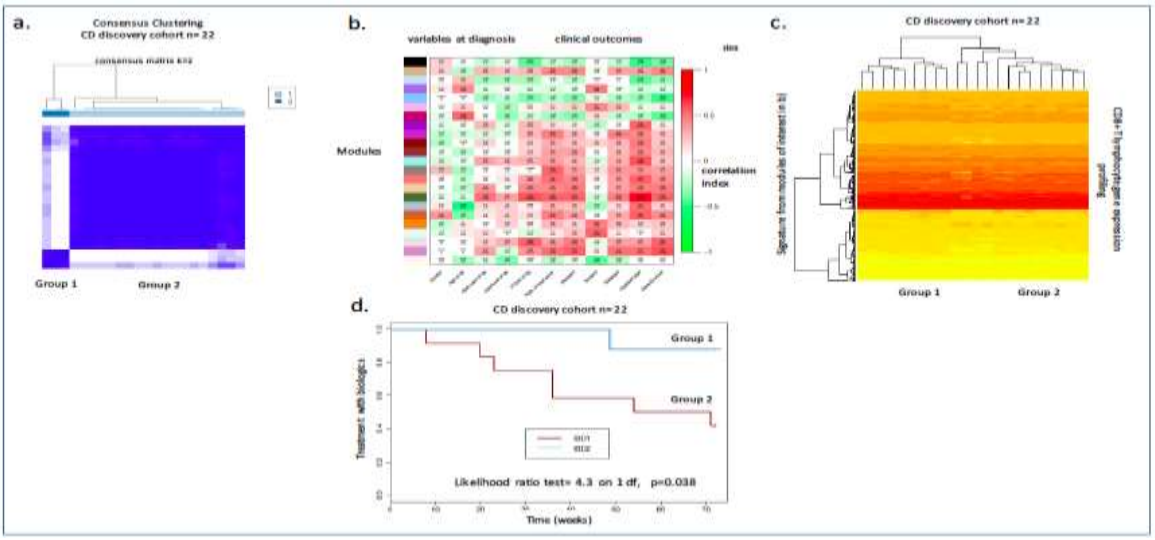

Figure Legend: Summary of key results derived from genome wide CD8+ T-cell gene expression profiles (n=22, children newly diagnosed with CD). a) Unsupervised consensus clustering highlights distinct differences in CD8+ T-cell gene expression profiles. b) Data derived from applying WGCNA to gene expression profiles. Summary of modules (groups of genes) and their correlation to key clinical outcomes. c) Hierarchical clustering based on expression profiles of genes derived from most significant modules (i.e. b) identifies two distinct patient groups. D) Survival analysis (Kaplan Meier) of patient groups identified in "c)" demonstrating significant differences in their disease outcome (here escalation to biologics p=0.038).

Review of Specialist Paediatric Hepatology Service for Children with Autoimmune Liver Disease. Are we meeting the standards?

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Introduction: NHS England Specialist Liver Disease Service Standard specifies that care for children and families includes carefully monitored shared care arrangements with referring clinicians, and delivery as close to home as possible by a close working relationship with local children's services. The recent RCPCH and BSPGHAN standards suggest that (i)services are delivered within clinical networks, (ii) access is equitable and designed across geographical, political and NHS/health board boundaries through the network, (iii) gastro-enterology network is linked to a lead specialist centre for hepatology and includes agreed patient pathways, specialist hepatology outreach clinics and shared care arrangements.

Aims: To determine whether outpatient pathway for children with autoimmune liver disease (AILD) is delivered in accordance with standards. **Objective**: To assess pathways of care for all children with AILD referred to a single specialist centre.

Subjects and Methods: All children seen by a Specialist Liver Service between 2011 and 2015 were identified from activity database. Retrospective data collection was performed using electronic patient records to determine follow up arrangements until Dec 2015 or discharge. Children local to centre, those with IBD or SLE or those assessed for transplant were excluded.

Results: 71 children were identified; of who 20 were from own region and 51 were extra-regional referrals (from 6 regions). Regular outreach clinics were in place at all 6 extra-regional tertiary centres. Of 20 referred from secondary care within own region, 18 had follow up (FU) exclusively at specialist centre, and two had shared FU with referring hospital. 28/51 extra-regional referrals were from tertiary centre, and 23/51 from secondary care. 26/28 (93%) referred from tertiary centre had follow up only at outreach clinic +/- secondary care, two had FU at both outreach clinic and specialist centre (one for transition education and one for complex disease). All 23 referred from secondary care were from 3/6 regions, and represented 40%, 67% and 81% of their referrals. 12/23 (52%) had ongoing specialist centre FU: 8 with shared care at tertiary centre and/or secondary care hospital. 11/23 (48%) had FU only at tertiary centre or outreach clinic and/or secondary care hospital. There were no formal shared care documents except for an outreach outpatient clinic standard from the Specialist Centre.

Summary: Through clinical networks, all extra-regional patients had care shared with local or tertiary services, but 14/51 still had appointments at specialist centre. Ongoing specialist centre FU was more common following extra-regional secondary care rather than tertiary centre referral (52% v 7%). For referrals from within local region, only 2/18 had shared care with secondary centre. Pathways were varied, being tailored according to tertiary and secondary service expertise, geography and patient preference.

Conclusion: Current standards are partially met when applied retrospectively to specialist service provision. The need for shared care documentation and improved network arrangements in some areas is highlighted.

Looking the truth in the eye - benchmarking care and outcomes against the world's largest paediatric IBD registry

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Introduction: Medical management of children with inflammatory bowel disease (IBD) is challenging, complex and informed by numerous guidelines. Despite the well-described benefits of benchmarking clinical practice, the vast majority of paediatric IBD centres in the UK do not measure clinical outcomes. ImproveCareNow (ICN) is a US health-learning network that includes data on over 25,000 children. It includes monthly webinars to share learning and innovation, offers access to an extensive online 'exchange' of ideas, and supports improvement initiatives to improve individual care and population outcomes. Membership also provides units with monthly data on their patient population, benchmarked against over 100 other paediatric IBD centres.

Aims and Objectives: To assess outcomes of care and identify areas for improvement in children with IBD managed at Addenbrookes Hospital, Cambridge.

Subjects and Methods: We report IBD related outcome data of 164 children currently active within the ICN2 Registry (> 75% of all children eligible for enrolment in Cambridge). 54% have Crohn's disease (CD), 31% ulcerative colitis (UC) and 15% with IBD-unclassified (IBD-U). Data is presented as % of eligible patients, compared to the average of all participating centres with ≥75% of patients enrolled, versus the network's defined targets (control numbers given in brackets).

Results:

	Cambridge	>75% enrolled	ICN Target
Clinical remission	70	81	83
Not taking prednisolone	94	96	95
CD in remission	74	82	N/A
UC in remission	69	78	N/A
Satisfactory nutrition	85	83	90
Satisfactory growth	98	93	90
TPMT measured	100	83	90
Correct dose of thiopurine	97	64	80

Summary: Children with IBD managed in Cambridge appear to fall below US peer group and network targets for clinical remission. Having identified such gaps, work is now underway to understand which aspects of care drive this difference and need change to achieve best possible outcomes.

Conclusion: Without accurate, population-based measures, improvements in clinical outcome cannot be documented. Embedding quality improvement and benchmarking of clinical outcomes in children with IBD allows i) identification of areas for improvement, and ii) impact of any change in management. Such large health-learning networks provide substantial advantages over standard audit / registry data, with responsive, clinically-relevant metrics allowing clinical teams to reliably progress toward the best possible outcomes for children with IBD.

Plenary Abstracts

Friday

26th January 2018

Assessing the risk of transplantation in Alagille patients.

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Introduction: Liver transplantation (OLT) is required in up to 40% of Alagille syndrome (ALGS) patients. In the setting of cardiac involvement, with increased right sided heart pressures, the mortality has been historically high. In order to manage this risk, we have used a Dobutamine stress test (DST) to mimic the increased cardiac output required during reperfusion. We have previously recommended threshold criteria of increase of cardiac index of at least 40% with 20 ug/kg/min Dobutamine and/or right ventricular pressure < 50% of systemic pressure during DST.

Aims and Objectives: The aim of the study was to review our management of cardiac lesions before OLT and the effect of these lesions on the short term outcome according to our evaluation and management.

Subjects and Methods: Retrospective data collection of 194 patients with ALGS from over 30 years period focussing on OLT and associated mortality. DST assessment was previously performed in the catheter laboratory; the recent protocol utilizes interventional Hybrid Cardiac MRI Catheterization (XMR) for accurate simultaneous flow and cardiac output assessment.
Patients with pre-transplant cardiac assessment (catheter DST or XMR DST) were recorded and comparison of the outcome post-transplant between no assessment, catheter DST and XMR DST eras was made.

Results: 89% patients (173/194) had a cardiac abnormality and 16% (31/194) had a heart intervention/correction. 44 patients (23%) were transplanted at a median age of 6.62 years (SD 6.85). 3 of 15 (20%) died early post-OLT in the pre-evaluation era for cardiac reasons. Pre-transplant cardiac assessment with catheter DST or XMR DST was performed in 33 cases. 15 had catheter DST before OLT, 9 were transplanted and all are alive. 19 patients had catheter XMR before OLT: 14 of them underwent OLT and all are alive; 3 were not listed for liver transplantation because of the severity of the cardiac pathology - 2 died subsequently from end stage liver disease and one is alive. Two had acceptable risk at XMR but were not listed due to stable liver disease.

	Results	Pre DOB	DOB 10 µg	DOB 20 µg
Catheter DST summary	HR (/min)	100 (SD 23)	110 (SD 26)	128 (SD 25)
	CI (l/min/m^2)	4.4 (SD 1)	5.6 (SD 1.4)	6.4 (SD 1.5)
XMAR DST summary	HR	86.7 (SD 19.5)	108.3 (SD 22.8)	132.1 (SD 25.5)
	CI (l/min/m^2)	3.5 (SD 1)	5.0 (SD 1.0)	5.5 (SD 0.9)

Summary: 20% of the patients transplanted in the pre-evaluation era died post-transplant for cardiac reasons. 17% of the patients had a Dobutamine stress test evaluation (catheter DST or XMAR DST) in our series. 67% of them underwent a liver transplant with only one death, not related with cardiac involvement.

Conclusion: Patients with ALGS have high risk for complications in the transplant period. Assessing the cardiac status using DST has improved transplant outcomes, preventing early post-transplant deaths**.**

"Causes of Infantile Acute Liver Failure in the West of Scotland"

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Introduction: Fulminant acute liver failure is a rare event in infants. It is defined as a hepatic based coagulopathy, with evidence of liver injury, in the absence of established liver disease. It does not require the presence of encephalopathy. The data defining the aetiology and incidence of liver failure originates from regional liver units. This may introduce selection bias and not provide a true reflection of the aetiology or extent.

Aims and Objectives: The aim of this study is to examine all the coagulation screens performed in our centre and identify those that fit our current definition of acute liver failure. By reducing the impact of bias we are able to more accurately define the aetiology and characteristics of liver failure in infants.

Subjects and Methods: All of the coagulation screens performed at the Royal Hospital for Children, Glasgow between June 2015 – June 2016 were examined. The inclusion criteria were infants aged less than 1 year with a prothrombin time greater than 18 seconds associated with evidence of liver injury. The notes of these patients were then retrospectively reviewed.

Results: Over the year, 9989 coagulation screens were performed. 669 tests were from 155 individuals less than one year old with a PT greater than 18 seconds. Results showed 24 out of 155 (14%) had a hepatic based coagulopathy. The aetiologies of these patients included hypoxic ischaemic encephalopathy (33%), ischaemic insult (54%), neonatal haemachromatosis (4%), metabolic disease (4%) and paracetamol toxicity (4%). 75% of patients survived while 25% patients died without liver transplant. None underwent liver transplantation or transfer to a regional liver unit; this is due to either spontaneous recovery or death prior to transfer. 120 patients were identified with a non-hepatic-based coagulopathy. Of these patients 63 (53%) were post-operative results, mainly from cardiac by-pass surgery, 14 (11%) were ECMO patients, 19 (15%) severe sepsis, with a further 23 (19%) other aetiologies. 10 premature patients were identified and excluded due to a lack of normative reference ranges.

Summary: The aetiology of infant liver failure is currently defined by data from regional liver units. This study identified several infants in fulminant hepatic failure who would not be included in this data due to death prior to transfer. Expanding this format to include a greater age range and time scale will help more fully define the cause and characteristics of patients with acute liver failure in the west of Scotland.

Designing the first pan-European paediatric IBD database to allow the study of incidence and prevalence of UC, CD and their rare and severe complications

Introduction: The incidence and prevalence of paediatric-onset inflammatory bowel disease (PIBD) and related complications are currently unknown or unclear in several European and non-European countries.

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Aims and Objectives: We designed an electronic survey system, which allows us to collect PIBD denominator data, annually. With this study we aim to determine the incidence and prevalence of PIBD and its rare and severe disease or treatment related complications and reveal important differences between regions, populations and other variables as well as trends over time.

Subjects and Methods: An electronic survey system based on the Redcap database was designed to capture the referral population and patient population of PIBD experts in 23 participating countries. This is part of the study examining rare complications of PIBD within the H2020 funded PIBD-SET Quality. PIBD experts are invited to complete an annual survey that collects data regarding the location and type of clinical service, and number of new and current cases of PIBD. It also identifies from where their patients are mainly referred (regional coverage). A key element is the use of the Nomenclature of Territorial Units for Statistics (NUTS3) as defined by Eurostat. This structure allows us to combine the collected regional PIBD data with the 4600 validated datasets that are available in the Eurostat databases. In order to validate the quality of collected data, the source of information for entered data in each survey is also captured.

Results: Within one year we have gathered responses from 62 paediatric gastroenterologists based in different regions of 16 European countries and Israel. So far, we have reviewed reports from 5 countries with the population covered reaching 16.8 million children and estimated incidence and prevalence through this system. For the United Kingdom (UK), the Netherlands (NL), Sweden (SE), Italy (IT) and Israel (IS), the coverage of the paediatric population reaches 49%, 77%, 22%, 35% and 57% respectively. Preliminary results are presented below:

Country	Incidence per 10^5 (Poisson 95% CI)	Prevalence per 10^5 (Poisson 95% CI)
UK	**6.5** (5.9-7.4)	**28.9** (27.6-30.1)
NL	**5.6** (4.8-6.5)	**28.3** (26.5-30.2)
IS	**6.9** (5.7-8.2)	**27.3** (25 - 30)
IT	**1.9** (1.5-2.3)	**16.7 (**15.5 - 18)
SE	**7.6** (5.4-10.5)	**48.8** (42.8-55.3)

Summary: By collecting PIBD denominator data through an annual survey from paediatric gastroenterologists in multiple countries, we have been able to calculate incidence and prevalence of PIBD in five different countries. As data collection is still ongoing, the numbers of other countries will be collected as well.

Conclusion: This has shown our ability to obtain incidence and prevalence data of PIBD, compatible with previous data, through studying a very large Pan European referral population allowing direct comparison between different countries and regions. These data will allow us to feedback precise figures to our responders in regard to their local populations and further examine reasons for reported differences in disease incidence and prevalence between countries and regions.

Rare and severe complications in children with paediatric-onset IBD; the international PIBD-SETQ Safety Registry by PIBD-NET

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Introduction: Paediatric-onset inflammatory bowel disease (PIBD) reflects a more severe disease compared to adult forms, which leads to more intensive therapy strategies earlier in the disease course. This exposes children to a risk of serious disease- and treatment related adverse events.

Aims and Objectives: Although severe adverse events following PIBD or its treatment seem rare, it is essential to characterize patients at risk in order to prevent those outcomes. With this study, we aim to identify risk factors for severe outcomes in PIBD.

Subjects and Methods: An electronic safety registry was designed to prospectively identify PIBD patients <19 years of age that develop a severe IBD- or IBD treatment related complication. Paediatric gastroenterologists in 23 countries are requested to register every month whether they observed one of the 10 listed complications. This list, created from the PIBD SETQuality consortium includes; cancer, death, severe neurological disease, renal failure, venous thromboembolism, liver failure, sepsis, opportunistic infections, bone marrow failure and hemophagocytic lymphohistiocytosis (HLH). In addition, other rare and severe complications outside the listed categories are also requested and checked for eligibility. For every registered complication detailed anonymized information on patient- and disease characteristics, therapy and the specific complication is obtained. Denominator data for several regions in participating countries are being collected.

Results: Since October 2016 we have received 790 responses from 20 European and 3 other countries. In total, 124 doctors are actively participating in this safety registry. Since the start of this project, the monthly response rate has increased from 42% in the first month to 61% in the last month. In total 65 complications were reported of which 28 were followed up after checking eligibility and removal of duplicates. Opportunistic infections were most frequently reported (n=4), followed by bone marrow failure (n=3), cancer (n=3), renal failure (n=3), venous thromboembolism and sepsis (n=3). The remaining reported cases were neurological disease (n=2), death (n=1) and liver failure (n=1). No cases of HLH were registered. Five complications were reported as other, including leucocytoclastic vasculitis, amyloidosis, orbital abscess, severe leucopenia and an air embolism.

Summary: During a one-year period since the start of this international safety registry, 28 rare and severe complications in PIBD patients were prospectively identified. The number of participating PIBD experts is increasing while additional information on every reported complication and denominator data per region are being collected.

Conclusion: This prospective safety registry in combination with the collection of denominator data for reporting countries will enable us to calculate incidence numbers on rare and severe complications in PIBD. Follow up forms with additional information on every reported complication are being collected to allow further analysis and understanding of the possible causes, management and outcomes.

Audit of yield of 'paediatric endoscopy and/ or colonoscopy (PE/C)' procedures in a tertiary gastroenterology centre

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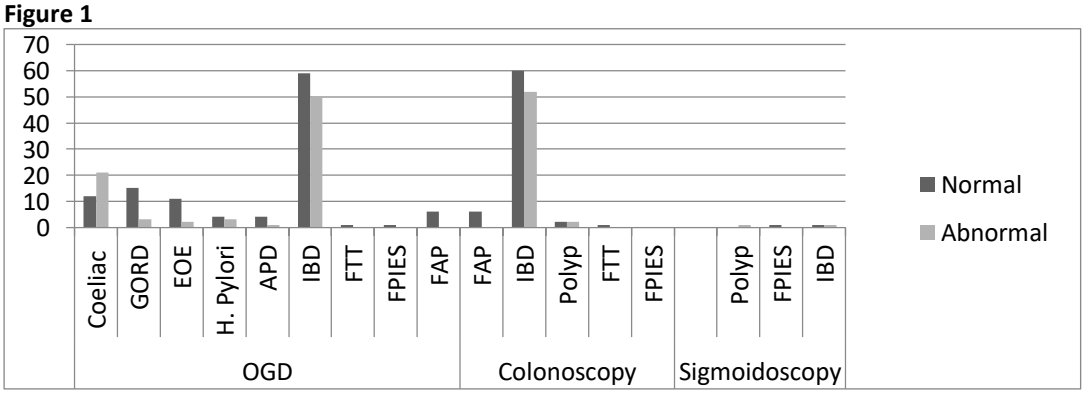
Introduction: Access to PE/C service varies across paediatric gastroenterology centres in England based on local resources and pathways (1). The indications for these high demand procedures are not standardised and yield from these procedures varies and is difficult to measure.

Aims and Objectives: To review the indications and results of PE/C procedures at Tertiary gastroenterology centre at Leicester and to gain an understanding of the common indications. We also aimed to compare rates of these procedures to available national data (1).

Subjects and Methods: This was a retrospective review of the patient records to identify indications (pre-agreed with the team) and the results for endoscopy (OGD), colonoscopy, OGD and colonoscopy and sigmoidoscopy for a 10 month period (Jan-Oct 17). Inclusion criteria were children aged 2-17 who attended for diagnostic and reassessment PE/C during study period. Indications were broadly classified into solitary polyps, inflammatory bowel disease (IBD), familial adenomatous polyposis (FAP), gastroesophageal reflux disease (GORD), H. Pylori, Acid Peptic Disease (APD), Eosinophilia oesophagitis (EOE) and coeliac disease.

Results: 310 procedures were performed in 10 months (equates to 372 procedures/year) of which 242 were diagnostic and 68 were reassessment of the disease. Below are the results of the histology (abnormal or normal) based on indication for each procedure.

Figure 1



193 OGDs, 123 Colonoscopies and 4 sigmoidoscopies were performed with rates of abnormal results being 41.45%, 43.9% and 50% respectively.

Conclusion: Tertiary gastroenterology centre for East Midlands South region is achieving 372 procedures per year which comparable with previously published national data (1).

Results suggest that pre agreed indications helped to measure yield from these procedures. Standardisation of indications for PE/C procedures may help to reduce variation in accessing these services within different regions.

This will also help to compare yield from these procedures in different centres across the country to rationalise threshold to investigate children with these procedures.

Ref: 1. NHS Atlas of variation in diagnostic services Nov 2013.

Portal hypertension and management of oesophageal varices in paediatric patients: Experience of one paediatric gastroenterology unit and comparison to new proposed national guidelines.

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Introduction: Oesophageal varices are a serious complication of portal hypertension due to their liability to rupture once they reach a critical point, which can result in life threatening bleeding. The current lack of national guidance for the assessment and banding of paediatric oesophageal varices has been highlighted following a coroner's report, and new BSPGHAN guidelines have been proposed. Of relevance, these recommend surveillance endoscopy in patients with portal hypertension, including evidence of splenomegaly and a platelet count of $<120{,}000 \times 10^9$/L. The guidelines also recommend what follow-up and time frame a patient should undergo repeat endoscopy dependant on findings and treatment.

Aims and Objectives: To review all paediatric patients with portal hypertension over a five-year period managed at the Royal Hospital for Children, Glasgow and compare their investigation, treatment and follow-up against the proposed paediatric varices guideline.

Subjects and Methods: All clinically active patients in the paediatric service were identified from the local database. All ultrasound scans from the last 5 yrs were reviewed. Any patient with any evidence of splenomegaly was included in the portal hypertension group. Once these patients were identified recent full blood counts were reviewed to look for evidence of hypersplenism (platelet count of <120,000x109/L) in the last 12 months. For each of the patients with evidence of splenomegaly, endoscopy dates were collected, along with data on the type of procedure (elective or emergency), presence and grading of varices, any intervention, and plan for review. Data was also collected on unexpected bleeding episodes requiring admission, and the treatment required for these acute bleeding episodes.

Results: From 273 active liver patients, 49 had ultrasound evidence of splenomegaly and 24 (8.8% of total cohort) was identified as meeting the criteria from the proposed guidelines warranting surveillance endoscopy. The most common underlying aetiologies were: biliary atresia (25%), autoimmune hepatitis (18%), genetic syndromes (12%), cystic fibrosis (10%), and portal vein cavernoma (8%). 50% (N = 12) had been scoped in the last year, 17% (N = 4) had been scoped in the last 1-3 years, and 33% (N = 8) had never been scoped. Of the 12 patients who received endoscopy within the last year, 33% (N = 8) had varices, of whom 2 required intervention. When taking into consideration the presence and grading of varices on last endoscopy, 58% of patients had received endoscopy within the recommended time frame according to proposed guidelines. 3/24 (13%) patients meeting the surveillance criteria had previous admissions within 5 years with emergency bleeding requiring endoscopy, despite 2 of these having undergone surveillance that year.

Conclusion: 8.8% of chronic paediatric liver patients followed up at the Glasgow Royal Hospital for Children met the criteria for endoscopic screening under the new varices guidelines. Historic practice suggests at least one third of patients were not receiving appropriate endoscopic surveillance against this new standard, hence there may be resource implications for implementing this nationwide

Posters

P1 – P57

***P1* Should measurement of Ascorbic acid become routine in the management of patients on Home PN?**

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Introduction: Children on home PN programmes are routinely monitored for selected micronutrient deficiency. Stability of micronutrients in PN bags is assumed but rarely confirmed.

Aims and Objectives: To present a case of vitamin C deficiency that manifested in a child with pseudo-obstruction who is fully dependent on parenteral nutrition and raises the question as to whether current estimates of Ascorbic acid requirements should be reviewed.

Subjects and Methods: A 5-year-old girl with pseudo-obstruction due to long segment Hirschsprung's disease developed hypercalcaemia on a stable PN prescription. There was a preceding one-month history of gradual reduction in mobility. Hypercalcaemia remained persistent on serial measurements. PTH was appropriately suppressed and intermittent hypophosphataemia was observed. Vitamin D was normal,

The child ceased to fully weight-bear despite use of a buprenorphine patch for pain. She localised pain to the back and the right leg. X-ray of the right leg showed mild irregularity of the distal right femoral metaphysis but no clear bony abnormality that explained the symptoms. MRI showed no abnormality of the lumbar spine or of the pelvis. MRI of the right knee showed mild non-specific oedema and a metaphyseal abnormality similar to that seen on X-ray. Complicating the presentation were episodes of fever associated with a CRP rise, which was treated as presumed line sepsis.

Results: MRI changes were not in keeping with a diagnosis of partially treated osteomyelitis. A neuropathic cause was excluded and dermatomyositis was ruled out by documenting normal CK, LDH and AST. Ongoing unexplained hypercalcaemia, reduced mobility and radiological bony lucencies led to considering Multiple Endocrine Neoplasia (MEN) that has an association with Hirschsprung disease. Serum total HCG, Ca125, calcitonin and alpha-fetoprotein were normal.

An X-ray of the contralateral knee confirmed that the metaphyseal irregularities were in fact symmetrical, in keeping with a metabolic or systemic cause. A skeletal survey excluded skeletal dysplasia. Eventually going back to the textbook proved valuable; the bilateral metaphyseal irregularities were consistent with radiological appearances of vitamin C deficiency. Whilst vitamin C levels could not be quantified, replacement of vitamin C at thrice the baseline dose led to a dramatic improvement in symptoms.

Summary: This perplexing case highlights thoughtful insight from multiple specialities in tertiary paediatrics, concluding with a textbook diagnosis of vitamin C deficiency made possible by familiarity with the radiological changes in scurvy.

Conclusion: Vitamins in PN are at risk of degradation with significant risk of degradation of vitamin C by oxidation.[1] This case highlights the need for vigilance around micronutrients that are rarely measured in children on parenteral nutrition.

References

1. Ferguson T et al (2014). A review of stability issues associated with vitamins in parenteral nutrition. e-SPEN Journal 9 (2) , e49-e53.

***P2* Outcomes of a Parental Survey with Nurse led gastrostomy service**

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Aims and Objectives: Our previous studies have aimed to improve gastrostomy service provision at the Evelina London Children's Hospital. Introduction of a nurse specialist had significantly increased parental satisfaction rates and improved documentation. Our study explored the impact of this intervention upon development of buried bumper, a serious complication of gastrostomy. Furthermore, we investigated parental satisfaction with different feeding systems; comparing percutaneous endoscopic gastrostomy (PEG) systems to a balloon device.

Subjects and Methods: Data was collected prospectively on PEG and PEG with jejunal extension (PEG J) via telephone survey, from June to August 2017. All were contacted two years following the initial procedure. Evidence of antibiotic use and clinic follow-up was collected using the electronic patient records (EPR) system.

Results: 42/53 patients responded (36 PEG and 6 PEG-J; 79% response rate). 44.4% of PEG patients developed complications versus 83.3% of PEG-J patients. Only one patient had a positive culture for wound infection. None of the PEG patients developed buried bumper whilst one PEG-J patient developed this complication. Average satisfaction score (on a scale of 1 to 10) of care received at Evelina was 9/10 for both groups. 66.7% of PEG patients had at least 1 follow-up at Evelina versus 83.3% of PEG-J patients. The average contact with the nurse specialist was 1.4 appointments for PEG patients versus 1.33 for PEG-J patients. 100% of PEG patients were satisfied with support from the community team whilst 83.3% of PEG-J patients had satisfactory support. 52.8% of PEG patients versus 50% of PEG-J patients had a balloon device inserted as replacement. Average time of change from PEG to button was 18 months and 15 months for PEG-J. The majority of PEG systems (63.1%) were changed electively to a balloon device (37% changed due to complications in PEG) whereas all the of PEG-J systems replacements to a balloon device were due to complications. Majority of the patients who underwent a change reported that they would have preferred a primary balloon device initially (63.2% of PEG patients and 66.6% of PEG-J patients).

Conclusion: Although most complications are not serious, rate of complications reported by parents are very high. We continue to achieve high satisfaction rates in our Gastrostomy service. There remains a higher chance of developing buried bumper with the PEG-J system and this warrants greater parental education. The majority of both PEG and PEG-J patients would have preferred a balloon device at the outset; parents should be given a choice between the two feeding systems at initial consultation.

Appendix:

Complication	Number of PEG patients with complication (16/36)
Infection	12/16
Leakage	5/16
Bleeding	2/16
Granulation	6/16

Complication	Number of PEG patients with complication (5/6)
Leakage	2/5
Granulation	1/5
Leakage and Granulation	2/5

P3 **Nasogastric Feeding Tube Bedside Care Plan Audit at the Leeds Children's Hospital**

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Introduction: Nasogastric feeding tubes (NGT) are used to feed infants and children who cannot for various reasons eat or drink by mouth. A misplaced NGT is a Never Event, "a serious, largely preventable patient safety incident that should not occur if existing national guidance or safety recommendations have been implemented by healthcare providers"[1]. Use of misplaced NGT's was first recognised as a patient safety issue by the National Patient Safety Agency (NPSA) in 2005 and three further alerts were issued by the NPSA and NHS England between 2011 and 2013[2-4]. The NPSA alert in July 2016 highlighted that staff responsible for checking the tube placement, required competency based training and that every patient at Leeds Children's Hospital should have a NGT bedside care plan. A pH test (pH 1-5) is gold standard for position confirmation and must be documented along with external length upon insertion and every time tube is accessed to ensure the tube has not moved. Patients are also given NGT information booklets upon insertion of the tube.

Aims and Objectives: The aim of this audit is to reduce harm from misplaced NG tubes by monitoring standards across the Children's Hospital. In particular the audit will aim to improve completion of the bedside care plan, identify cases where pH is not used first line to check position and monitor compliance with standards for on-going checks of tube position.

Subjects and Methods: There were 20 subjects. The audit was conducted in June 2017. An audit tool used in 2016 was modified to reflect feedback from previous data collection. Some paediatric ward areas were combined for data collection purposes. Up to 5 patients per clinical speciality were audited. The standard data collection form was used in each case.

Results: A total of 19 care plans were audited. In 11 cases, pH was documented to confirm position of the NGT after insertion. Where pH was used, a pH <5 was successfully confirmed in all cases. External length was recorded on insertion for 8 out of the 19 patients. Following insertion, 6 were given information leaflets. Confirmation of NGT position is mandatory prior to each feed or administration of medication via the tube; 5 patients had a complete record of this having taken place every time the tube was accessed.

Summary: All but one patient had a care plan in their bedside documentation and the majority of care plans were completed. There were gaps within the documentation with four common inaccuracies identified. The external length was not always documented on insertion of the NGT or when transferred from another hospital or ward. First line method to confirm position was not always documented; pH and external length were not documented every time the tube was accessed. Information booklets were not given to each patient with an NGT placed, or if they were it had not been documented.

Conclusion: Ward staff should continue to routinely document pH and external length upon insertion and each time the NGT is accessed. They should ensure the patient is given the relevant NGT information booklet following placement. A further audit is planned for summer 2018. On-going competency based training for all staff responsible for NGT includes 3 yearly theory and practice assessment of insertion and on-going care of NGT. For newly qualified nurses, practical experience is gained on an infant or child. For experienced nurses, competency assessment can be on a manikin practice (peer assessment). From April 2017 to March 2018, face to face theory training on NGT insertion and on-going care is being delivered as part of the mandatory training programme for all registered nurses and health care assistants working in the Leeds Children's Hospital. There is also a learning package for staff not able to access face to face training.

***P4* A review of Copper Deficiency in Paediatric Patients on Long-Term Jejunal Nutrition**

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Introduction: Copper is a trace element essential for a number of vital processes within the body. It is mainly absorbed in the duodenum. Copper deficiency is a rare but potentially serious condition, manifesting as haematological derangement, sensory impairment and neuropathies. It is most commonly seen in patients who have had bariatric surgery. Patients who are jejunally-fed long-term will logically be at risk of copper deficiency due to where their nutrition enters the GI Tract, as feed is delivered beyond the site of copper absorption.

Aims and Objectives: First, to review the detection and management of copper deficiency in jejunally fed patients managed at a single tertiary care centre. Second, to investigate feeding tube position as a potential contributor to copper malabsorption. Finally, to close the loop on a previous audit performed in our department, which found that 40% of jejunally fed patients were copper deficient.

Subjects and Methods: Case notes of all patients on long-term jejunal nutrition were reviewed. Type of Jejunal feeding was noted. Results from trace element screens were recorded and imaging was checked by specialist radiologists to assess tube position. These results were then correlated with serum copper levels, to look for any links to low copper and an inadequately positioned tube.

Results: 22 patients were included in this study, (14 Males, 8 Females, mean age 8 years (range 1-21 years)). 7 of these patients had a Roux-en-Y Jejunostomy, 12 had a PEGJ, 2 had a GJ Button, and 1 had an NJ tube. Nearly half of the patients were suffering from Cerebral Palsy. 2 patients had never had copper levels checked. There was no relationship between type of feed, its copper content, and serum copper levels. 3 of these patients (13.6%) had been diagnosed with a low serum copper level, down from 40% in our previous audit. 1 patient had a low serum copper and was receiving copper sulphate solution as treatment. 1 patient had previously low copper levels with normalisation on recent testing, but remained on treatment. The third patient was on a lower than standard maintenance dose for previous low copper that had subsequently normalised. As an additional point, 4 patients also had low zinc picked up on their micronutrient screen, one of whom was documented as being prescribed supplements. Tube position was generally found to be good; we did notice that a poorly positioned tube of any type, or a tube requiring regular adjustment/change resulted in poorer micronutrient absorption. Assessment of neurological complications from copper deficiency was difficult given the co-morbidities of this patient cohort. No obvious haematological derangement was observed.

Summary: Following a previous audit in our department, we reviewed the detection and management of copper deficiency in jejunally fed patients, and also investigated feeding tube position as a potential contributor to copper malabsorption. Overall, our screening rates have improved, and 13.6% of patients were found to have had a low serum copper, down from 40%. Once picked up, treatment was successful. Type of tube was found to be unrelated, however a tube requiring regular adjustment or maintenance correlated with micronutrient absorption issues.

Conclusion: The number of enterally-fed patients is increasing in the UK, both in the adult and paediatric populations. Micronutrient management is becoming an increasingly recognised part of care. Copper deficiency is under-recognised in children on jejunal nutrition. When detected, treatment was established and successful. Work must be done in implementing guidance on the screening of micronutrient deficiencies as a whole, however there are obvious practicality issues with achieving this in such a population.

***P5* Use of a standardised enteral feed regimen post gastrostomy insertion – how does it impact on dietetic time?**

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Introduction/Background: Previous dietetic practice at Birmingham Children's Hospital for children admitted for insertion of a gastrostomy involved a face to face ward-based assessment for a bespoke feed regimen. The process was felt to be time consuming and not always achievable if parents were not present or the child was in theatre. Following discussions with the surgical team (and as part of a wider project) it was agreed to introduce a standardised feed regimen for patients, initially trialling with 2 surgeons' patients. The regimen involved initiating feeds 2 hours post-op with a water bolus, followed by 4 hourly bolus feeds of increasing volumes (50%, 75% and 100% of usual bolus/hourly feed volume). This enabled target feed volumes to be reached 12 hours post gastrostomy insertion as well as multiple teaching opportunities for parents to use the gastrostomy.

Aims and Objectives: To compare the number of dietetic contacts and dietetic time spent with a patient following gastrostomy insertion prior to and following the implementation of a standardised enteral feed regimen.

Subjects and Methods: From July to October 2017, 39 patients were admitted for a gastrostomy insertion. 16 patients were excluded from the data collection (8 fundoplication, 7 had no dietetic input and 1 was a long-term inpatient). Number of dietetic contacts and time spent was gathered retrospectively on 23 patients via an internal electronic system. 7 of these patients (30%) were put on the standard feed regimen with agreement from their surgeon (highlighted via a red triangle within the figures).

Results:

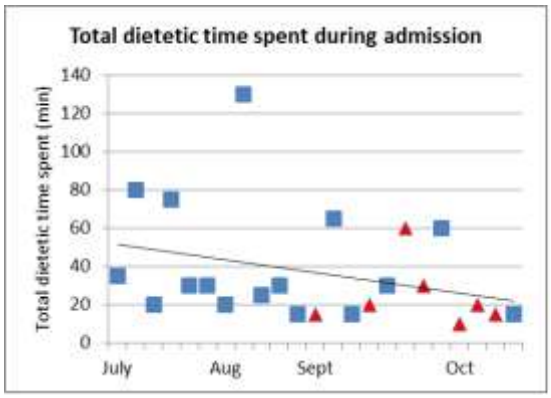

Fig 1

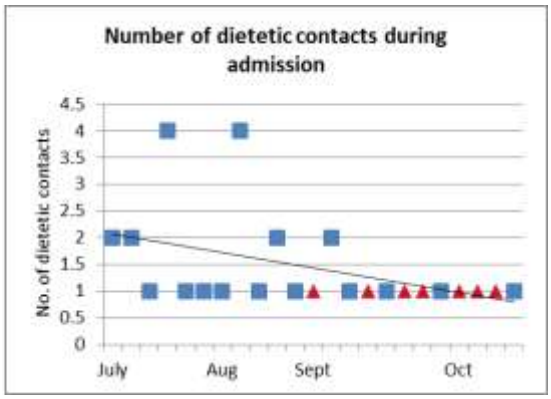

Fig 2

Over the course of 4 months, a downtrend in the total dietetic time spent (fig. 1) and the number of dietetic contacts (fig. 2) was noted. Comparing those on the standardised feed regimen to previous there was a 39% reduction in number of dietetics contacts (dropping from a mean of 1.63 to 1 contact) and a 43% reduction in dietetic contact time during the admission (from a mean of 42 to 24 minutes).

Summary and conclusion: The use of a standardised feed regimen following gastrostomy insertion has reduced the number of dietetic contacts and total time spent with patients during their admission.

P6 **Prevalence of Refeeding syndrome and its associated factors among children admitted with Severe Acute Malnutrition in Kenyatta National Hospital.**

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Introduction: Refeeding Syndrome is a complication of SAM occurring in the initial feeding period. It results in grave morbidity and mortality. Change of feed from special milk to F75 has resulted in a marginal drop in mortality. We seek to find out if these deaths have a relationship with the prevalence of refeeding syndrome.

Aims and Objectives: To establish the prevalence of refeeding syndrome in children with SAM. To determine the associated factors such as oedema, dehydration, type of starter feed, presence of HIV and provision of top up feeds. To observe the outcomes of Refeeding syndrome such as death, recovery and persistence.

Subjects and Methods: In an observational short longitudinal study of children aged 6-59 months who were diagnosed with Severe Acute Malnutrition (WHO weight for height Z score of -3SD) and admitted for treatment were recruited. Levels of potassium, phosphorus and magnesium at admission and after 48 hours of initiating feeds were measured. We also investigated the anthropometric measurements, presence of oedema, dehydration, HIV status, type of feed and followed the children up for 7 days to check outcomes.

Results: 160 children with SAM with a median age of 12.5 months were recruited into the study. The prevalence of Refeeding Syndrome was 21% (95% CI 15.2 to 28.4). Refeeding Syndrome was significantly associated with HIV infection (p=0.032), the odds of Refeeding Syndrome increased six-fold with HIV infection (OR=5.99, 95% CI 1.23 to 29.1) after adjusting for the effect of age and sex. Out of the children who developed RS, 65% recovered when appropriate treatment was administered while one patient died (3%). Out of 34 children who developed Refeeding Syndrome 20% were lost to follow up while 12% had persistent hypokalemia and hypophosphatemia.

Summary: A significant proportion of severely malnourished children developed refeeding syndrome as a complication. This was propagated by certain factors notably the presence of HIV infection. However with treatment many of these recovered.

Conclusion: One in every five children admitted at Kenyatta National Hospital with SAM developed Refeeding Syndrome. However, this is a drop from the previous 93% observed in 2006. HIV was significantly associated with developing Refeeding Syndrome. Majority of the participants who developed Refeeding Syndrome recovered with appropriate treatment.

***P7* Cost, availability and nutritional composition comparison between gluten free and gluten containing food staples provided by food outlets and internet food delivery services.**

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Introduction / Aims: The study aim is to compare the cost, availability and nutritional composition of traditionally wheat based manufactured gluten-free (GF) products with their gluten-containing (GC) counterparts, provided by food retailers (physical stores and online).

Methods: A cross-sectional survey of 26 food categories was conducted in May 2017; data was collected on the cost, availability and nutritional composition of in excess of 1,000 GF and GC foods. Food categories included traditionally wheat based and everyday foods usually containing gluten. Fifty physical stores were surveyed, inclusive of convenience stores, budget supermarkets, regular supermarkets, quality supermarkets and health food shops (10 in each category). In addition, online retailers of manufactured GF products which offered a delivery service to the areas under study were also included.

Results: A third of all stores surveyed did not stock any traditionally wheat based manufactured GF items; this was comprised of budget supermarkets and convenience stores. The online GF food suppliers superseded all of the physical stores in the number of manufactured GF items available. However, over half of the GF items were more expensive in online stores than in regular supermarkets. In fact, 74% of GF foods surveyed were more expensive than their GC counterparts, including food staples such as GF bread and bread rolls were 294-449% more expensive than the GC counterparts ($p<0.001$), plain flour 94% ($p=0.006$) and flaked cereals 93% more expensive ($p<0.001$). Nutritional composition comparison revealed higher energy, fat and saturated fat and less protein content for surveyed manufactured GF products.

Summary / Conclusion: Availability of manufactured GF products remains poor, especially in convenience stores and budget supermarkets, serving those from poor socio-economic cohorts, the elderly and physically disabled. The stores where availability has improved from previous published findings are associated with the greatest additional cost. The inferior comparative nutritional quality of manufactured GF products emphasises need for those on a medically indicated GFD to be advised and monitored by adequately trained health professionals and also provides evidence towards dispelling false claims that removing gluten from the diet has health benefits.

Acknowledgments: Funding from Coeliac UK

***P8* Incidence of paediatric stricturing duodenal Crohn's Disease in South-East Scotland: a 12 year population-based cohort study.**

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Introduction: Stricturing duodenal Crohn's disease (CD) is a rare but serious presentation of CD causing significant morbidity. There are no data on its incidence rate in paediatric CD, and only estimates for adult CD.

Aims and Objectives: Our aim is to provide the first incidence data and case studies outlining this severe presentation in children.

Subjects and Methods: A regional cohort of prospectively acquired incident cases of PIBD diagnosed less than 16 years of age in paediatric services within a strict geographical area of South-East Scotland (based on postcode) was captured over a 12-year period (10.2005 – 09.2017). Incidence rates for all CD and for duodenal stricturing CD were calculated and standardised for age and sex with the Scottish Census of 2011. A retrospective review was conducted on the medical records of patients presenting with stricturing duodenal CD together with a detailed review of the available literature and consensus guidelines.

Results: In total 157 new cases of paediatric CD (<16yrs) were diagnosed within the study period. Median (IQR) age at diagnosis 13.1 (10.7-14.4) years; 64% male predominance. Overall CD incidence rate 5.80/100,000/year (95% CI 4.91-6.76) in South-East Scottish children, with a specific duodenal B2 phenotype disease incidence rate of 0.07/100,000/year (95% CI 0.01-0.27); representing 1.3% of incident cases at diagnosis. The 2 incident cases of stricturing duodenal CD (male aged 13.4yrs and female aged 15.5yrs) presented with typical systemic symptoms of weight loss, abdominal pain, anorexia and lethargy, together with recurrent vomiting suggestive of obstruction. Both cases partially responded to intensive and rapidly escalated medical therapy but eventually required surgery (laparoscopic gastroduodenostomy without resection). A detailed literature search confirmed there are no paediatric guidelines or case-reports relating to duodenal stricture as either a presentation or complication of CD.

Summary & Conclusion: Duodenal stricture is a rare but serious presentation of CD causing significant morbidity and not currently covered in the paediatric literature or consensus guidelines. Best practice medical and surgical management remain uncertain and require further research.

P9 **The role of thiopurine metabolite monitoring in the management of paediatric inflammatory bowel disease. A retrospective study done in Royal Aberdeen Children Hospital, Scotland.**

Png, YD, Foundation Year 1 Doctor; Kishore S, Consultant Paediatric Gastroenterologist; Bisset M, Consultant Paediatric Gastroenterologist; Mahdi G, Consultant Paediatric Gastroenterologist; Cameron C, Paediatric Gastroenterology Nurse Specialist; Smart B, Paediatric Gastroenterology Nurse Specialist; Department of Paediatric Gastroenterology of Royal Aberdeen Children Hospital, Aberdeen, Scotland.

Introduction: Thiopurines has been proven to be effective in the treatment of inflammatory bowel disease (IBD). However, the use of thiopurines was often limited by its narrow toxicity profile and the difficulty with predicting individual patient response. Measuring the concentration of 6-thioguanine nucleotides and 6-methylmercaptopurine nucleotides produced by thiopurines had been proposed as a strategy to identify patient variations in drug response and toxicity. Thiopurine metabolites monitoring was introduced into the Royal Aberdeen Children Hospital (RACH) in 2014 about three years ago. The service of providing metabolite monitoring test in RACH was reviewed. The indications and outcomes of thiopurine monitoring were evaluated.

Aims and Objectives: This study aim to investigate the indications for performing thiopurine metabolite testing, the clinical actions taken in accordance to the metabolite concentration and in which situation metabolite measurements could lead to a change in management.

Subjects and Methods: Patients with inflammatory bowel disease who were on thiopurine medication for at least eight weeks and had metabolite bloods taken between January 2014 and February 2017 were recruited. Hospital records were reviewed.

Results: A total of 37 patients were included. 70 metabolite samples were performed on these patients. All the metabolite samples were grouped into 51 episodes based on the reason for checking metabolite levels. The most common reason for performing metabolite test in RACH was routine measurement (43%), active disease (41.0%), adverse effects (7.9%), drug adherence assessment (5.9%) and a combined indication of flare + adverse effect (2.0%). The clinical actions taken in accordance to metabolite concentration include dose optimization (48.7%), further investigation (18.4%), education about compliance (9.2%), switch drug class (7.9%), add biologic (6.6%), cease drug (5.3%) and surgery (3.9%). Overall, metabolite testing lead to a change in management plan in 38 out of the 51 episodes (74.5%). Greatest rate of change was found in adverse effect and compliance groups (100% versus 100%), followed by active disease group (90.5%) and routine group (50.0%).

Conclusion: Metabolite testing led to a change in management in most cases especially in patients with toxicity and active disease. The most common change was dose change. The use of metabolite test to guide dose changes was useful particularly for patients with routine assessment and active disease. Prospective studies are needed to determine whether the use of metabolite testing to guide dose is of benefit. The clinical action taken in accordance to metabolite concentrations and the interval at repeating routine/repeat metabolite test in Royal Aberdeen Children Hospital should be standardized.

P10 **Faecal Microbiota Transplantation as a Treatment for Inflammatory Bowel Disease: a National Survey of Adult and Paediatric Gastroenterologists in the UK**

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Introduction: Interest in the use of faecal microbiota transplantation (FMT) as a therapeutic treatment in inflammatory bowel disease (IBD) has increased following promising outcomes in patients with Clostridioides difficile infection (CDI). To date, four double blind randomised control trials have been conducted investigating the efficacy of FMT in patients with ulcerative colitis (UC)[(1)(2)(3)(4)]. Whilst research exploring clinician awareness and attitude towards the use of FMT in CDI has been carried out[(5)(6)], data for IBD is currently lacking, despite growing patient and clinician interest in the treatment.

Aims and Objectives: The primary aim of this survey was to assess the perceptions of gastroenterologists and current practice relating to FMT as a treatment for IBD in the UK.

Subjects and Methods: The survey was developed using Snap Survey software, following literature review and author consensus. It was distributed amongst UK-based gastroenterology trainees and consultants through the British Society of Gastroenterology and British Society of Paediatric Gastroenterology, Hepatology and Nutrition e-newsletters, and at the BSG Conference in June 2017.

Results: In total, 61 respondents completed the survey including pre-subspecialty trainees, gastroenterology specialists, associate specialists and consultants. 82% (n=50) of respondents were adult gastroenterologists and 18% (n=11) were paediatric gastroenterologists. Almost all (95%; n=58) respondents stated that they had heard of FMT being used as a treatment for IBD prior to participating in the survey. Based on current evidence, 34% (n=21) of respondents would consider using FMT in patients with IBD, 26% (n=16) would not and 39% (n=24) were undecided. The majority of respondents (95%) would consider entering a patient with IBD into a clinical trial. A total of 43% (n=26) respondents said that a patient had expressed interest in FMT and a small proportion (10%; n=6) said that they were aware of a patient that has undertaken FMT on their own without medical supervision. When asked to rank routes of delivery in terms of preference, nasogastric tube was the least preferred route (39%; n=24) and oral capsule was the most preferred route (34%; n=21).

Summary and Conclusion: A clear majority of UK gastroenterologists recognise FMT as a potential treatment for IBD, however uptake is limited. A significant proportion of clinicians would consider FMT in IBD based on currently available evidence and the majority would consider entering patients into clinical trials. Orally-delivered encapsulated FMT is the preferred route of administration and future work should explore the utility and efficacy of this route.

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P11 **Management of Anaemia in Children and Young People with Inflammatory Bowel Disease – an audit of DGH practice**

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Introduction: Anaemia is commonly seen in children and young people with Inflammatory Bowel Disease (IBD); this can be due to a number of different causes including iron deficiency and anaemia of chronic disease amongst others. Chronic anaemia carries potential short and long-term detrimental effects including poor growth, lack of energy and poor concentration with subsequent consequences for academic achievement and effects on immune function. Therefore anaemia in IBD should be identified early and treated effectively. In 2015, a BSPGHAN Working Group created a guideline for the Diagnosis and Management of Anaemia in IBD.

Aims and Objectives: The author was seeking to benchmark practice and answer the following questions.

- Do I identify significant anaemia and do I initiate treatment early enough?
- Do I use Intravenous (IV) iron at an appropriate threshold?
- Do I monitor haemoglobin after treatment?
- What form of oral iron do I use and do I change if poor tolerance?

Subjects and Methods: A retrospective notes review of 21 cases was carried out to explore management of anaemia. A standard pro-forma was used to record data and allow analysis with identification of patterns and themes.

Results: All 21 patients had haematinics checked regularly as per the guideline. 16 had confirmed anaemia. Two of these were not recognised as such and therefore did not receive treatment, but both had haemoglobins >100. Of the 14 patients who were treated, all received oral iron as first line. 7 had Hb<100 and 8 had Hb>100; the BSPGHAN guideline recommends those with Hb<100 should be treated with IV iron particularly if they have moderately active disease. 2 of the 7 patients did receive IV iron at a later date. Duration of treatment was clear in 71% of patient notes. 4 patients did not tolerate the oral iron preparation and were switched to an alternative but the time taken for this varied from 2 weeks to several months. 13 of 14 patients underwent repeat testing within 4-8 weeks as per guideline but in only 8 patients had the Hb returned to normal levels in this timeframe. Others were sometimes treated for a prolonged period on oral iron.
In total 4 patients received IV iron. Two had Hb<100 and two had Hb>100 but either poor response to oral iron or severe disease. The decision was often made after several months on oral iron. Two patients were transfused. One had severe UC and subsequently underwent subtotal colectomy. In the second child there was a delay in diagnosis of IBD due to parents refusing endoscopies.

Summary: Haematinics were checked regularly in this cohort of IBD patients. Treatment was not consistently initiated for those with Hb >100. There is a need to treat milder anaemia more aggressively as it has been shown this can improve quality of life. Duration of iron treatment is not always clear and there is a need to enquire more thoroughly about tolerance and ease of administration. Some patients could potentially have benefitted from intravenous iron earlier in their course of disease, sometimes at the time of diagnosis. They were much more likely to receive IV iron if Hb was<100. Historically there has been a reluctance, largely due to risk of side effects and limited resources.

Conclusion: Overall whilst patients undergo regular blood tests to look for anaemia, treatment is not always initiated in those with milder anaemia. There is a reluctance to move to intravenous iron treatment early. The author recognises the need to treat anaemia more aggressively and has already changed practice following this audit in Summer 2017.

P12 **Evaluation and management of adrenal suppression in paediatric inflammatory bowel disease (IBD) patients on long term corticosteroids**

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Introduction: Corticosteroid therapy is often used to induce remission or control flares in children with inflammatory bowel disease. However, prolonged courses of steroids predispose patients to adrenal suppression. Patients with adrenal suppression may have nonspecific symptoms including fatigue, headache, loss of appetite, weight loss and nausea which may be missed or mistakenly attributed to inflammatory bowel disease. There remains a lack of data in the evaluation and management of adrenal suppression in paediatric IBD patients on long term corticosteroids.

Aims and Objectives: To review our single centre's experience in the evaluation and management of adrenal suppression in paediatric IBD patients on long term corticosteroids.

Subjects and Methods: Data of all paediatric IBD patients who had cortisol levels and subsequently synacthen tests from February 2008 to October 2017 in a tertiary paediatric gastroenterology unit were extrapolated from Infoflex database, Cerner Millennium, endocrine database and biochemistry log. Patient demographics, diagnosis, duration of corticosteroid treatment, early morning cortisol levels, synacthen test results and management were analysed.

Results: Data expressed as median (range) Over an 8.5-year period, there were only 5 children annually who had morning cortisol levels but in total only 11 children required a synacthen test.

3/11 cortisol <20 nmol/l
8/11 cortisol 73.5 (29 to 190 nmol/l)
Disease distribution: Ulcerative colitis n=6 (5 pancolitis, 1 distal colitis)
Crohn's disease n=5 (4 ileocolonic, 1 left-sided colitis)
M to F ratio 7:4.
Age at diagnosis 10.5 years (2.5 - 15.8 years)
Age of first synacthen test 16 years (range 7 - 19 years)
Duration of steroid 13 months (3 - 38.5 months)

The standard in-house protocol following prolonged steroids and low morning cortisol level was to switch oral prednisolone to hydrocortisone and then undergo a synacthen test after 2-3 months (n=9).

7/11 children had normal synacthen test and their oral hydrocortisone dose or oral prednisolone dose were easily weaned.

4 children with abnormal results did not wean with repeat synacthen tests at median of 7.5 months (range 6 - 9 months). The repeat synacthen tests were normal with ultimate successful weaning of oral hydrocortisone.
One exception was a child with a history of prolonged steroid use who had an ileal resection for stricturing disease. She had been steroid free for 5 months. She developed an Addisonian crisis with cortisol level 29 nmol/l postoperatively responding to intravenous hydrocortisone and required maintenance oral hydrocortisone 5mg TDS for 3 months before a synacthen test.

Summary and Conclusion: We recommend that all children with prolonged steroid course should have a morning cortisol level measured when the dose of oral prednisolone is tapered down to 5mg daily. If cortisol level is low we recommend conversion to oral hydrocortisone followed by formal synacthen test at 2-3 months.

***P13* Inflammatory Bowel Disease Teams Working in Partnership with Out of Hospital Healthcare Providers Improve Patient Experience**

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Introduction: Many patients with chronic conditions like inflammatory Bowel Disease (IBD) prefer to receive high quality healthcare close to where they live. However many specialist services including paediatric IBD care is provided only in very few centres in the UK. Healthcare at Home Ltd, a provider of healthcare out of hospital settings, has started enhanced nursing care services in April 2017 for patients with Crohn's disease (CD) managed in our hospital receiving treatment with Humira (Adalimumab). This service is funded by Abbvie, manufacturer of Humira. Patients who opt-in to receive this service receive nurse visits at home 2-3 weeks prior to their hospital appointments. During these visits, nurse will review Humira administration, collect blood and stool samples as requested by the hospital team, measure height and weight and complete history aspect of disease activity scores. Blood and stool sample are transported to the hospital within few hours and the clinical information is passed on to the hospital team securely within one day.

Aims and Objectives: To assess the patient and parent satisfaction of enhanced nursing care service offered to patients with Crohn's disease on treatment with Humira.

Subjects and Methods: We have sent patient/parent satisfaction survey questionnaires via post in August 2017 to the 29 patients registered for enhanced nursing care service. The completed questionnaires were returned to Healthcare at Home team. Parents and patients were asked to complete the survey questions independently.

Results: 16/29 (55%) families have returned the completed survey questionnaires. Majority of patients and parents reported the enhanced nursing care service very helpful. Nurse review of Humira administration, the options to have bloods taken at home and the results available prior to the clinic visit have been rated highly by both patients and parents. They have found it easier to make decisions about treatment changes because of the availability of results prior to clinic visit. Details in Table 1

	Parents in agreement n (%)	**Patients in agreement n (%)**
Satisfaction with the enhanced nursing care service	16/16 (100%)	14/14 (100%)
Nurse visit and Humira administration review added value to my care	13/16 (87%)	10/14 (71%)
It is beneficial to have blood tests at home prior to outpatient clinic visit	11/15 (73%)	11/14 (79%)
Decisions about treatment changes were easier because of the timely availability of blood and stool test results	14/16 (88%)	9/14 (65%)

Summary and Conclusion: Enhanced nursing care service offered by out of hospital healthcare provider in collaboration with hospital IBD team was found to be very beneficial by both patients and parents.

***P14* Role for Local Small Scale IBD Family Education Day**

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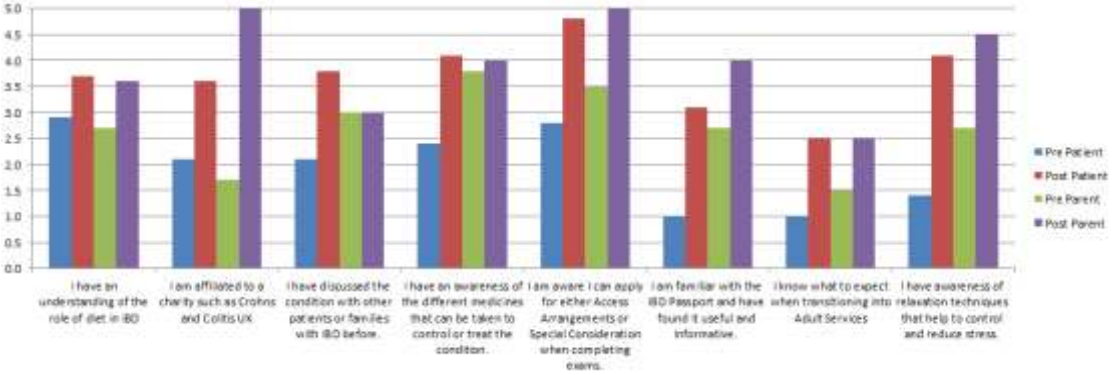
Introduction: Patient and family knowledge is crucial in optimising management of IBD and facilitating the partnership between family and the medical team. This need is arguably greatest as the young person passes through adolescence, transition and public examinations. The IBD charities provide a fantastic resource for information and large scale education days. We questioned if there might be an additional useful role for small scale local family days run by local IBD teams.

Aims and Objectives: **1.** To provide information to better equip families of young people with IBD approaching examinations on their exam entitlements, tools to help manage their IBD during this time, general IBD education and the opportunity to meet their Multidisciplinary team and ask questions.
2. The chance to meet other local families with IBD, form links and share experiences.

Subjects and Methods: During the August summer holiday, ten families attended an IBD Education and Information Day, delivered by a multidisciplinary team with time for discussion. Speakers included a Consultant Paediatric Gastroenterologist, IBD Clinical Nurse Specialists from both Paediatrics and Adult/transition services, a Representative from an IBD charity, Pharmacist, Dietician, Clinical Psychologist and Hospital school teacher. The day included lunch funded by the Royal Free Hospital School and a chance for an anonymous Q&A session with the expert panel. Open written feedback was collected as well as a questionnaire evaluating knowledge/awareness before and after the day.

Results: Likert scale from questionnaire converted to numerical value (5=strongly agree)



Pre and post questionnaires showed an increase in knowledge (pooled data).

All families gave written feedback, which was overall extremely positive about the day.

- 'Was unaware of the home invigilation or 5% added onto results''
- 'That I'm not alone in my condition'.
- 'This is so useful, particularly for newly diagnosed. I wish I had done this 3 years ago.''
- The multidisciplinary team also fed back that they enjoyed the day and felt that they had learned from hearing the families' experiences

Summary: Families with IBD found a small scale education and social day with their team very helpful and expressed interest in further similar days.

Conclusion: There is a role for small scale family IBD education days locally in addition the larger ones organised by charities.

P15 **Colectomy-Free Survival and Factors Associated with it in Children with Ulcerative Colitis Managed in a Tertiary IBD centre in the UK**

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Introduction: Colectomy-free survival is an important outcome for children with ulcerative colitis. There is only limited data available about long term outcome of children with ulcerative colitis.

Aims and Objectives: To review the outcome of colectomy-free survival and associated factors in patients with ulcerative colitis managed in our centre.

Subjects and Methods: We have performed a retrospective analysis of all patients diagnosed with ulcerative colitis in our hospital from January 2010 to December 2015. The patients were identified from the medical database of the paediatric gastroenterology unit and paediatric surgical unit. The clinical, laboratory, endoscopy data and medical and surgical treatment were analysed.

Results: 147 patients with ulcerative colitis were identified in the study period, 85 (58%) were male patients. The median age at diagnosis was 12.9 years (2.2 to 17 years) and median duration of follow up was 34 months (12 to 96 months). 105 (72%) recorded pancolitis (E4) at diagnosis while 15 (10%) and 26 (18%) had extensive (E3) and left sided (E2) lesions respectively. Severity of disease at diagnosis, documented as physician global assessment, was mild in 50 (34%), moderate 70 (48%) and severe in 26 (18%) patients. 55 (37%) patients had no relapse in first year after diagnosis. 90 (61%) patients were in clinical remission at both 3 months and 12 months after diagnosis. 86 (59%), 54 (37%) and 46 (31%) patients received steroid treatment at diagnosis, 3 months and 12 months after diagnosis respectively. 145 (99%) patients received treatment with Mesalazine during the follow-up period. 93 (63%) patients were treated with Azathioprine and 66% of these patients were commenced on Azathioprine treatment within 6 months of diagnosis. 31 (21%) patients received treatment with Infliximab and median time to start Infliximab was 1.4 years (3months- 7 years). 12 (8%) patients had surgery (sub-total colectomy) and chronic active severe UC was the indication for surgery in all patients.

Factors associated with colectomy were steroid treatment at 3 months after diagnosis(75% v 34% p value 0.05), steroid treatment at 12 months after diagnosis (92% v 26% p value 0.01) and longer time interval from diagnosis to initiation of Infliximab treatment (10.4 months v 19.8 months p value 0.01).

Age, extent and severity at diagnosis, the laboratory parameters at diagnosis including Hb, ESR, CRP, albumin, platelets, number of relapses in first year after diagnosis, number of episodes of hospitalisation for intravenous steroids and need for treatment with Azathioprine or Infliximab were not associated with colectomy.

Summary and conclusion:
Only a small proportion (8%) children needed colectomy in our cohort of patients with UC and the need for steroid use at 3 months and 12 months after diagnosis and longer interval to start treatment with Infliximab were associated with colectomy.

***P16* Use of Omegaven in Reducing Conjugated Bilirubin During Parenteral Nutrition**

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Introduction: Intestinal failure-associated liver disease (IFALD) is a major cause of morbidity and mortality in children requiring long-term parenteral nutrition (PN). Some reports suggest that using fish oil emulsion (omega 3 fatty acids) has several advantages, including a high concentration of a-tocopherol and the absence of phytosterols. It has also been reported that ameliorates IFALD by improving bile flow, inhibiting steatosis and through an immunomodulatory effect.

Aims and Objectives: To report experience of using Omegaven in 4 paediatric patients with IFALD.

Subjects and Methods: Review of patients with IFALD (conjugated bilirubin > 80 µmol/l) who failed to respond to treatments including using ursodeoxycholic acid (10mg/kg TDS) and reduction in parenteral lipid (SMOFlipid®) intake to 0.5g/kg/day. Omegaven® (10% fish oil emulsion) was given at a starting dose of 0.5g/kg/day and increased up to 1g/kg/day. Liver function tests (LFTs) were monitored and effectiveness of Omegaven® on reducing conjugated bilirubin assessed.

Results: 4 patients who received PN for more than 40 days developed severe IFALD (conjugated bilirubin range from 81-239µmol/L) and were treated with Omegaven®. All the patients were on some amount of enteral feeding at the time that Omegaven® was started (ranging from 11-90mL/kg/day). Patient 1 was weaned to enteral feeding before a fall in bilirubin was seen. The 3 remaining patients all showed a marked fall in conjugated bilirubin. Patient 2 conjugated bilirubin was 202µmol/L when Omegaven® was started and decreased to 55µmol/L before PN was stopped; patient 3 decreased from 239µmol/L to a total bilirubin of 27µmol/L (no conjugated bilirubin was measured at the time) and patient 4 conjugated bilirubin decreased from 163µmol/L to 10 µmol/L. All patients were weaned to enteral feeds and there were no adverse side effects noted with the use of Omegaven®. Patients 1-4 were on Omegaven® for a period of 24, 33, 52 and 68 days respectively.

Conclusion: Our small experience with these 4 patients shows that Omegaven® might have a role as a rescue therapy for patients with IFALD due to long-term parenteral nutrition and may be effective in decreasing plasma bilirubin concentration. Further studies will be necessary to evaluate fully the role of this nutritional intervention in the management of IFALD.

***P17* A dietary related case of megaloblasic anaemia in an adolescent with eating disorder.**

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Introduction: Megaloblastic anaemia in children is mainly related to dietary deficiencies of vitamin B12 and (or) folate, is predominately seen in developing countries and less commonly seen in paediatric populations within western society[1-2]. However, in recent years there has been a rise in the prevalence of eating disorders in western society and thus a consequential rise in those presenting with anaemia, of which, megaloblastic anaemia is usually rare[3-4].

Aims and Objectives: We therefore present a case of megaloblastic anaemia due to severe folate and vitamin B12 deficiency in a patient with eating disorder.

Subjects and Methods: A 15-year-old Caucasian girl presented with a four-year history of dizziness, pallor, background of eating disorder, dysmenorrhoea, anxiety and asthma. She was noted to have a poor diet mainly comprising of small portions of plain pasta/rice, yoghurt, chicken, fish, cheese and potatoes. A review of the history, case notes, blood samples and follow-up appointments was undertaken over a one-year period.

Results: She initially presented with tachycardia (130bpm), shortness of breath and dizziness on standing. She appeared very pale, with weak pulses and faecal loading within the abdomen. The remainder of her general examination was unremarkable. Initial bloods showed Hb 44 g/l, Hct 0.13 l/l, MCV 112 fl, Plts 143 x10^9/l, WCC 3.3 x10^9/l, Neuts 1.7 x10^9/l, Retics 14.7 x10^9/l. Blood film showed a macrocytic anaemia with basophilic stripping, tear drops and polychromasia. Neurtophils were reported left shifted to myelocytes with hypersegmented nuclei. Haematinics showed vitamin B12 90 pmol/L, folate 0.1 µg/L and ferritin of 263.5 ng/mL along with a vitamin D deficiency. The remainder of bloods were normal. During her admission she was jointly reviewed by the haematological, dietary and gastroenterology teams. The patient and her parents were advised that her blood count would gradually improve with B12, folate and iron supplementation with out the need for transfusion and the risks surrounding this in her case. However, despite this advice the patients' parents wished for her to receive a blood transfusion and she went on to receive 1 unit of red packed cells. During admission she was also reviewed by psychiatry who arranged further out-patient follow-up. Prior to discharge her bloods showed Hb 64 g/l. She was discharged to her GP with a plan for two further B12 loading doses of intramuscular B12 followed by three monthly intramuscular injections up to 1 year, folate supplementation to be taken for 3 months then stopped and ferrous fumerate (iron) supplements to continue with plan for review of these by her GP.

Summary: A case of megaloblastic anaemia in a young adolescent secondary to dietary deficiencies due to an eating disorder which was treated with folate, B12 and iron replacement.

Conclusion: This case highlights the importance of addressing nutritional deficiencies in individuals with eating disorders. It also presents a unique example of how megaloblastic anaemia secondary to dietary deficieincies which are not commonly seen within western paediatric populations can present in individuals with eating disorders, how as clinicians we need to be aware of this potentially growing subgroup, can seek to recognise and hopefully actively treat these cases early.

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P18 **Nasal Retaining Loops (nasal bridle) causing nasal trauma in Children: a case series**

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Introduction: The Nasal Retaining loop uses catheter mounted magnets to place umbilical tape around the septum and attaches the tape to the naso-enteral feeding tube with a clip. This is suitable for use in infants and children on long term naso enteral feeding with frequent tube dislodgement or self-removal; in patients with skin conditions which precludes normal fixation with tape and is generally safe. We report 3 cases in children in whom long term use of the nasal bridle resulted in trauma to the nasal septum, in one case resulting in a permanent facial deformity. To our knowledge this has not been reported before. Following these case reviews the current practice was reassessed and new pathways developed to prevent such injuries.

Case 1: Patient with complex medical problems, congenital nephrotic syndrome, diarrhoea on long term parenteral nutrition (PN). Initially had naso-gastric tube (NGT) for medications and enteral feed which was changed to naso-jejunal tube (NJT) due to vomiting. Due to frequent self-removal of the NGT it was agreed that a nasal bridle should be used to secure the NJT. The nasal bridle was inserted and a plan was made to regularly change it; although manufacturer did not recommend regular change. The child had the nasal bridle changed every 2-3 months. The first 3 were traumatic; the next 4 changes were uneventful. The 8th change was delayed due to the patient been unwell and before it was changed the nasal bridle eroded through the septum resulting in a permanent facial deformity.

Case 2: Patient with skin problem and faltering growth required a NGT tube. A nasal bridle was inserted to secure the tube. The nasal bridle was changed regularly. It was then found that the tape from the nasal bridle had cut into the septum. ENT initially felt the damage to the septum was due to an adverse response to the tape caused by the child's skin condition. The NGT was then removed and the skin healed with no long-term damage to the septum. The child went onto have a gastrostomy inserted.

Case 3: Patient with skin problem and feeding difficulties had a NGT tube and required a nasal bridle to secure the tube. The nasal bridle was inserted but not changed for 3 months. During this time the tape cut into the nasal septum. Following removal of the nasal bridle the skin healed and no long-term damage was caused.

Conclusion: Nasal retaining loops (nasal bridles) are useful in securing the naso-enteral tubes but it must be acknowledged that harm can be caused with long term use and it should not be used as an adjunct to gastrostomy or jejunostomy. We therefore recommend that a nasal bridle should not be used for more than 6 months and should be changed every 30 days (following new licensing guidance). With adequate monitoring and changes in practice we hope that we would be able to prevent similar harm.

P19 **Hypoallergenic and anti-inflammatory feeds in Malawian children with complicated severe acute malnutrition (SAM): an open randomized controlled trial**

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Introduction: Globally, undernutrition accounts for nearly half of under five deaths. About 20% of children with severe acute malnutrition (SAM) require admission because of medical complications, inability to take feeds or failure to improve under community management. Despite following a well-established WHO protocol, case fatality in "complicated" SAM remains up to 35% with additional mortality and morbidity after discharge. Children with SAM have an enteropathy that 1) has similarities to intestinal inflammation in non-IgE mediated gastrointestinal food allergy and Crohn's disease, 2) fails to improve with existing management and 3) likely contributes to poor outcomes.

Aims and Objectives: We evaluated whether therapeutic feeds that are effective in treating intestinal inflammation in non-IgE mediated gastrointestinal food allergy and Crohn's disease also benefit children with complicated SAM.

Subjects and Methods: In a randomized, open study in Queen Elizabeth Central Hospital, Blantyre, we recruited 95 children aged 6-23 months with SAM (weight-for-height z score <-3 and/or mid-upper arm circumference <11.5 cms and/or nutritional edema). After initial clinical stabilization, children were allocated randomly to 2 weeks of exclusive enteral nutrition either with an elemental feed, a polymeric feed or standard feeds (F-100 and/or ready-to-use therapeutic food). The primary outcome was change in intestinal inflammation evaluated by faecal calprotectin.

Results: Mean (SD) fecal calprotectin concentration (normal <50 ug/mg stool) was 547 (744) at baseline and remained highly abnormal at the end of the intervention (697 (735)) with no significant differences between the study arms. Biomarkers of mucosal integrity (fecal α_1-antitrypsin, plasma intestinal fatty acid binding protein and IgG anti-endotoxin antibodies) and systemic inflammation (platelets, C-reactive protein, α_1-acid glycoprotein) were also highly abnormal at baseline and generally persisted in all three arms.
Weight gain and changes in insulin-like growth factor-1 and binding protein 3 were similar in each arm. The alternative feeds were tolerated less well than standard feeds. Case fatality was 7/95 (7.4%) with a similar frequency of serious adverse events in each arm.

Summary: The enteropathy in complicated SAM did not respond to standard feeds or the alternative therapeutic feeds administered for up to 14 days in this study.

Conclusion: Improving survival and long-term outcomes in children with severe malnutrition is an urgent priority. A better understanding of the gut pathology in complicated SAM is needed to inform the development of novel therapeutic interventions.

***P20* The effect of inflammation on body composition in children admitted to a tertiary hospital.**

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Introduction: Malnutrition is prevalent in children and adults with chronic illness. The development of disease related malnutrition is multi-factorial. The presence of chronic inflammation has been associated with altered body composition (BC) in certain inflammatory diseases.

Aims and Objectives: We tested the hypothesis that chronic inflammation in children, resulting from any pathological process, is associated with malnutrition, lower lean mass (LM) and higher fat mass (FM), compared to children with other diseases requiring specialist in-patient paediatric care.

Subjects and Methods: A prospective study at a tertiary paediatric hospital recruited 154 patients at admission (4.6-17.6 years, 50% female). Baseline data was collected on diagnoses, steroid prescription, nutritional support and activity level. Anthropometric and BC measurements were performed using a range of techniques including Dual-energy X-ray Absorptiometry (DXA).

A subgroup of 44 subjects with primary inflammatory disease and/or systemic inflammation (defined by elevated CRP and/or platelets) were analysed compared to (1) healthy reference children and (2) subjects within the cohort without inflammation.

Results: Subjects with inflammation had lower height (mean -0.72 SDS, p=0.007), lower LM (mean -1.04 SDS, p=0.002), lower Lean Mass Index (LMI, LMI=LM(kg)/height^2) (mean 0.62 SDS, p=0.036) and higher Fat Mass Index (FMI) (mean 0.54, p=0.035), compared to healthy reference children. There was, however, no significant difference when compared with patients without inflammation.

Linear regression modelling of the whole cohort suggested useful predictors for low LMI were artificial nutrition and low activity levels (enterally fed β -0.62, p=0.048, parenteral nutrition β -0.9, p=0.004, wheelchair user not involved in sport β -3.36, p=0.002). For FMI high-dose steroids were found to be a useful predictor (β 0.81, p=0.023). Presence of inflammation was not a useful predictor for LMI or FMI.

Summary: Malnutrition was common in children with inflammation, when compared to healthy children, although no significant difference was identified between inpatients with and without inflammation.

Conclusion: The indicators used were insufficient to demonstrate that inflammation had a significant adverse effect on anthropometric and BC measurements when compared to non-inflammatory chronic diseases affecting an inpatient paediatric population.

P21 Positive safety applied to Total Parenteral Nutrition

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Introduction: Never events are defined as those that have the potential to cause serious patient harm or death and are wholly preventable[i]. NHS England have stated that the primary purpose of reporting and investigating "never events" is to learn from them and to improve the provision of care, a sentiment echoed by the Care Quality Commission[ii]. Undoubtedly this has value however; the relative infrequency of such events provides rare learning opportunities to improve the provision of care. In 2016/17 NHS England reported only 424 "never events"[iii]. In a similar period the NHS dealt with over 1 million contacts every 36 hours[iv]. This equates to a failure rate of just 1 in approximately every 573,000 contacts. Positive safety is an approach to safety improvement that emphasises learning from what goes right every day. To apply positive safety in the NHS it is necessary to establish metrics of the performance standards of safety measures (known as barriers) such that the emergence of weaknesses in the function of barriers would be detected. This would then enable the organisation to take action to restore the function and improve overall resilience in patient safety.

Aims and Objectives: Bristol Royal Hospital for Children conducted a pilot study to examine the benefits of applying a positive safety approach to the management of a common and routine clinical procedure, the administration of parenteral nutrition (PN) in paediatric care. The Hospital worked with a safety specialist (ERM) to develop a set of Bowtie models of hazards associated with PN. The hazard scenarios considered were: incorrect infusion, infection of the patient's central venous line (used to give the PN), and PN demand exceeding the hospital's capacity to manufacture it.

Subjects and Methods: The Bowtie models were developed over a four day workshop attended by relevant stakeholders from all aspects of the PN process within the hospital.

Results: The work identified 97 independent risk controls for these scenarios: 81% were preventative in nature and 19% were mitigative (intended to minimise the consequences of the hazard rather than prevent it). It also revealed that 93% of the control measures were either procedural (65%) or behavioural (28% i.e. that depend on a person's knowledge or memory). The control measures were rated for their inherent strength (i.e. ability to control the risk) and their reliability of function (i.e. how well people comply with a procedure). Based on these ratings a number of controls were identified as having high strength but relatively poor reliability.

Summary: The process enabled better communication between team members with overlapping responsibilities and provided each individual with a valuable overview. It highlighted the role and significance of existing barriers and empowered staff to continue to support the application of individual barriers and to challenge deviations from optimal practice. Opportunities to improve barrier performance for better risk management were identified, leading to recommendations to optimise control measures across the entire chain of events from the prescription and manufacture of PN to its administration and monitoring.

Conclusion: The study has demonstrated the utility of the positive safety method for application in healthcare to improve patient safety. The outputs of the process have proved useful in a number of ways that are potentially transferrable to other areas in healthcare.

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***P22* Chronic granulomatous disease mimicking Crohn's disease**

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Introduction: Chronic granulomatous disease (CGD) is a rare primary immunodeficiency disease caused by mutation in any of the genes encoding subunits in the nicotinamide adenine dinucleotide phosphate (NADPH) oxidase enzyme complex present in many cells including phagocytes. The hallmark of CGD is recurrent bacterial and fungal infections and granuloma formation. CGD tends to present before the age of five and is more common in boys as two third of cases are inherited in an X- linked recessive mode. The gastrointestinal manifestations are relatively common in patients with CGD. However, gastrointestinal manifestation as initial presentation of CGD is very rare and has been reported in only 5% of cases. The gastrointestinal symptoms seen are weight loss, failure to thrive, loose stools, bowel obstruction, perianal ulcerations, fistulas, rectal bleeding, anaemia, hypoalbuminaemia and colitis. These can mimic Crohn's disease.

Aims and Objectives: We present siblings who were initially diagnosed and treated with Crohn's disease and a few years later were diagnosed with CGD. The diagnosis of CGD was particularly challenging, as the presentation was atypical.

Subjects and Methods: Retrospective notes reviews of two patients using the Infoflex database and Cerner Millennium.

Results: Both female patients presented age five and half and there was no history of recurrent infections. Both siblings had minimal macroscopic findings on upper and lower endoscopy but extensive microscopic finding of granulomatous inflammation. The younger sibling subsequently developed hepato-splenomegaly and had complete work up including normal nitroblue tetrazolium test. Her liver biopsy revealed granulomatous inflammation. As part of the COLORS study an abnormality was detected and further genetic analysis confirmed the diagnosis of CGD.

Summary: The two siblings with CGD had a mild phenotype mimicking Crohn's disease initially. The gene abnormality identified in both cases was $p22^{phox}$ mutation that is inherited in autosomal recessive mode. Of interest parents are consanguineous.

Conclusion: The mild phenotypic disease with profuse granulomatous inflammation should raise the suspicion of alternative diagnosis including CGD even in the absence of recurrent infections. These cases highlight the need for clinical suspicion of alternative diagnosis especially in cases where the disease presents unusually or are refractive to mainstay treatment. We suggest early genetic testing for CGD.

***P23* Superior Mesenteric Artery (SMA) syndrome mimicking Gastroenteritis with an unusual complication of oesophageal perforation - case report**

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Aims: To report a case of Superior Mesenteric Artery (SMA) syndrome complicated by an oesophageal perforation with an initial diagnosis of gastroenteritis.

Case report: A 15 year old boy who is normally fit and well with no significant past medical or surgical history was admitted under the surgeons with a very short history of severe crampy abdominal pain following a takeaway a few hours prior. He denied any fever, no recent travel, no diarrhoea or vomiting and no recent weight loss, bowels were opened that day. Initial abdominal examination revealed generalised tenderness with some voluntary guarding but was not distended. Observations, bloods and gas were unremarkable. Initially managed as Gastroenteritis due to suspected food poisoning with Intravenous (IV) rehydration. The paediatric team were asked to review the patient the following day. At this point he was vomiting with worsening abdominal pain. On examination, he was pale, dehydrated with tenderness and guarding over the whole abdomen, bowel sounds were absent. Gas revealed metabolic acidosis, Nasogastric tube drainage produced around 2 Litres of coffee ground fluid, the patient became tachycardic and received a 20ml/kg bolus of 0.9% NaCl. He was commenced on IV Cefotaxime, Metronidazole and IV omeprazole. An AXR was performed which was – featureless, an erect CXR was normal. CT showed an oesophageal perforation with an extremely distended stomach and proximal duodenum and cut off of gas beyond that which raised a strong possibility of a diagnosis of SMA syndrome. The patient was stabilised and immediately transferred to the tertiary paediatric surgical team. He was managed conservatively with analgesia, IV hydration and nutritional support. The symptoms resolved and he made a good recovery.

Discussion: Superior mesenteric artery (SMA) syndrome is an extremely rare condition caused by the D3 portion of the duodenum being compressed between the aorta and SMA leading to intestinal obstruction. SMA syndrome can present acutely with signs and symptoms of proximal small bowel obstruction and or can present with chronic symptoms. Diagnosis can be made by an upper gastrointestinal series or CT scan. Treatment is usually always conservative with nasogastric decompression and IV fluids with total parenteral nutrition and surgery if medical therapy fails.

Conclusion: This case highlights the importance of considering SMA syndrome as a differential in patients presenting with acute abdominal pain or gastroenteritis. As seen in our case here SMA syndrome might well present acutely and might be misdiagnosed as gastroenteritis. Superior Mesenteric Artery syndrome is a rare but potentially life threatening condition – recognition and awareness is paramount.

***P24* 'Is this going to hurt?' – Understanding acute post-endoscopy pain in a tertiary paediatric gastroenterology centre.**

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Introduction: Information for paediatric gastroenterologists about the nature and quantity of pain immediately post gastrointestinal (GI) endoscopy under general anaesthetic (GA) has been anecdotal. ESGE/ESPGHAN[1] guidance suggests best practice for paediatric GI endoscopy is to be under GA and that there is a lack of evidence to recommend for or against use of CO2 insufflation in ileocolonoscopy. A randomised controlled trial showed a reduction in pain post-endoscopy with CO2 insufflation in the paediatric population under sedation[2].

Aims and Objectives: To understand the quantity and severity of acute pain experienced by children following GI endoscopy with air insufflation under GA. The hypothesis was that procedures requiring upper GI endoscopy (OGD) only would report a lower incidence and severity of pain, compared to colonoscopy (+/- OGD).

Subjects and Methods: Nursing records of patient/carer-reported post-procedure pain and physiological observations were analysed from 100 cases undergoing endoscopy at a regional tertiary centre for paediatric gastroenterology. Cases were collected prospectively between November 2016 and February 2017.

Results: The median age was 12 years (range 11 months-17 years); 56 patients were male. For 75 patients it was their first experience of an endoscopy. 70 of the patients had no comorbidities. The procedure type was: OGD+colonoscopy (n=40), OGD (n=38), colonoscopy (n=12), OGD+sigimoidoscopy (n=9), sigmoidoscopy (n=1). The diagnoses of the patients were normal endoscopy or functional GI disorder (n=48), inflammatory bowel disease (n=17), oesophagitis/gastritis (n=16), coeliac disease (n=13), polyp or polyposis (n=3) and Other (n=4). The following analgesia was given during the procedure: none (n=22), paracetamol (n=65), opiate (n=9), paracetamol and opiate (n=4). The mean length of stay post-procedure was 3 hours (range 1-5 hours). The median Paediatric Early Warning Scores (PEWs) across stay = 0 (range 0-4) and the median pain score = 0. Where a PEWs score of 0 is entirely within range and up to 4 requires nurse-in-charge input. A pain score of 0 indicates no pain. Two cases out of 100 had a transient pain score >0 lasting one hour, with no intervention recorded. One patient at hour 1 (pain score 1/10, PEWS 2, had received paracetamol), and the second patient at hour 3, (pain score 2/10, PEWS 4, after no analgesia). Both cases had only had an OGD undertaken. No cases had persistent pain recorded. Pain score and PEWs score were not documented in the first hour in 33 and 13 cases respectively.

Summary: Pain after endoscopy under GA is infrequent, transient and mild, being documented in only 2 of 100 patients. There was incomplete recording of pain and PEWs score post-procedure, especially during the first hour after the procedure.

Conclusion: These findings would suggest that in a setting where general anaesthesia is available for paediatric GI endoscopy the use of CO2 insufflation is not required as acute pain is already minimal and there is little physiological disturbance. It may also be that a more nuanced tool is required to capture paediatric experiences of pain post GI endoscopy and this might require future work on a pain score for paediatric endoscopy.

References:

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***P25* Validation of Direct Observation of Procedural Skills (DOPS) for Paediatric Colonoscopy**

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Introduction: Direct observation of procedural skills (DOPS) are tools designed by the Joint Advisory Group (JAG) to assess competence in endoscopy. These were expanded in July 2016 (new DOPS) to include those specific to paediatric colonoscopy. However, paediatric colonoscopy DOPS assessments have not been validated.

Aims and Objectives: To correlate overall trainee competence with components of the paediatric colonoscopy DOPS.

Subjects and Methods: We performed a prospective UK-wide analysis of formative paediatric colonoscopy DOPS submitted to the JETS e-Portfolio over one-year (August 2016-2017). Scores were averaged across procedural domains (pre-procedural, procedural, post-procedural and endoscopic non-technical skills – ENTS). Each DOPS item, except for ENTS, were grouped into cognitive and technical skillsets by two independent investigators, and correlated with the overall performance score. Correlation analyses were performed using Spearman's test (rho >0.70 indicating high positive correlation).

Results: 61 DOPS assessments were completed by 13 unique trainers for 14 trainees. Overall performance score comprised: 1: Maximal supervision (1.6%), 2: Significant supervision (13.1%), 3: Minimal supervision (47.5%) and 4: Competent (37.7%). By domain, overall competence correlated most with scores for the 'Procedural' domain (rho: 0.849, p<0.001), ENTS (0.666, p<0.001), 'Post-procedural' (rho 0.635, p<0.001) and pre-procedural (rho 0.471, p<0.001). By domain, overall score correlated more with performance in predominantly 'Cognitive' (rho 0.834, p<0.001) and 'Technical' (rho 0.815, p<0.001) domains compared to ENTS. In terms of DOPS items, overall competence score correlated most with 'Proactive Problem Solving' (rho 0.836, p<0.001) and 'Patient Comfort' (rho 0.826, p<0.001), and weakest with 'Confirms Consent' (rho 0.228, p=0.115) and 'Equipment Check' (rho 0.302, p=0.020).

Summary: In colonoscopy, performance in the 'Procedural' domain, Proactive Problem Solving' items, and 'Cognitive' skillsets had greatest correlation with overall procedural competence.

Conclusion: Competencies in paediatric colonoscopy, as assessed within DOPS, vary in their correlation with overall competence. As assessors are completing the new DOPS in a consistent manner, this provides novel validity evidence for the new paediatric colonoscopy DOPS.

***P26* Uptake of new Paediatric Direct Observation of Procedural Skills (DOPS) for Colonoscopies**

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Introduction: The Joint Advisory Group on GI Endoscopy (JAG) was set up in 1994 and JAG Endoscopy Training System (JETS) went live in UK in 2009. Since 2012 Paediatric endoscopy has also been logged via the JETS system. A new Paediatric formative DOPS form was introduced in July 2016. There are currently 24 GRID trainees in PGHAN. Trainees are encouraged to fill out DOPS throughout their Endoscopy training. Adult data suggests that the new style DOPS indicate better construct validity with the new rating scale (Ref1).

Aims and Objectives: To survey the uptake of Paediatric DOPS forms in the 12 months following release.

Subjects and Methods: All Paediatric DOPS Paediatric Colonoscopy form data recorded via JETS from July 2016 – July 2017 were reviewed.

Results: 61 DOPS forms were completed by 14 trainees (range 1-20, mean= 4, median =3). Number of DOPS varied between regions

Table 1: Variation in Regions

Region	**Mean DOPS per trainee (range)**
Mersey	2 (2)
W Scotland	1 (1)
W Midlands	1 (1-2)
Yorkshire	5 (1-8)
London	7 (1-20)

Table 2: A range of individuals in training and non-training roles, across all levels, completed DOPS

Level of Trainee	Number
Consultant	2
ST8	4
ST7	3
ST6	1
SPR Y4	1
Staff Grade 1	1
Clinical Research fellow	2

Table 3. Number of DOPS varied between individuals

Level of Trainee (Colon DOPS)	Mean number of DOPS per individual (range)
Consultant	13 (6-20)
ST8	3 (1-6)
ST7	1 (1-2)
ST6	4 (4)
SPR Y4	8 (8)
Staff Grade 1	4 (4)
Clinical Research fellow	1 (1)

Summary: Completion of DOPS forms for Paediatric OGDs varied widely between individuals and regions.

Conclusion: A more uniform uptake of Paediatric DOPS should be encouraged, perhaps by targeting the barriers to DOPS being performed that create inequalities between individuals and centres – for example some centres are not registered on JETS.

Ref: Siau, K. Changes in scoring of direct observation of procedural skills (dops) forms in endoscopy training and their impact on competence assessment 2017 Gut; Volume 66, Issue Suppl 2

***P27* Uptake of new Paediatric Direct Observation of Procedural Skills (DOPS) for OGDs**

Levi, R, Locum Consultant Paediatric Gastroenterology [1]; Siau, K[2] JAG Research Fellow; Howarth, L[3], Consultant Paediatric Gastroenterologist ; Broughton, R[2], Senior Programme Manager JAG/RCP; Tzivinikos, C[4]Consultant Paediatric Gastroenterologist; Feeney, M[5],Consultant Gastroenterologist; Narula, P[6,] Consultant Paediatric Gastroenterologist

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Introduction: The Joint Advisory Group on GI Endoscopy (JAG) was set up in 1994 and JAG Endoscopy Training System (JETS) went live in UK in 2009. Since 2012 Paediatric endoscopy has also been logged via the JETS system. A new Paediatric formative DOPS form was introduced in July 2016. There are currently 24 GRID trainees in PGHAN and 24 SPIN trainees in the UK. Trainees are encouraged to fill out DOPS throughout their Endoscopy training. Adult data suggests that the new style DOPS indicate better construct validity with the new rating scale (Ref1).

Aims and Objectives: To survey the uptake of Paediatric DOPS forms in the 12 months following release**.**

Subjects and Methods: All Paediatric DOPS form data recorded via JETS from July 2016 – July 2017 were reviewed.

Results: 158 Paediatric OGD DOPS were recorded by 17 individuals. The number of DOPS an individual filled in varied from 1 - 24. (Mean 9.2, Mode 10).

Level of Trainee (OGD DOPS)	Mean no procedures per trainee (range)
ST8	13 (10-18)
ST7	7 (3-13)
ST6	16 (6-24)
ST4	4 (4)
SPR Y4	10 (10)
LAT	4(4)
Staff Grade	11 (11)
Clinical Research fellow	1 (1)

Mean number of DOPS per trainee varied between regions

Region	Mean DOPS per trainee (range)
W Midlands	9 (1-24)
London	12 (10-15)
Yorkshire	6 (1-10)
Mersey	3 (3)
W Scotland	10 (10)

Summary: Completion of DOPS forms for Paediatric OGDs varied widely between individuals and regions**.**

Conclusion: A more uniform uptake of Paediatric DOPS should be encouraged, perhaps by targeting the barriers to DOPS being performed that create inequalities between individuals and centres **–** for example some centres are not registered on JETS.

Ref: Siau, K. Changes in scoring of direct observation of procedural skills (dops) forms in endoscopy training and their impact on competence assessment 2017 Gut; Volume 66, Issue Suppl 2

***P28* Shifting Practice in the Diagnosis of Paediatric Coeliac Disease in English District General Hospitals**

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Background: The diagnosis of paediatric coeliac disease (CD) within the UK has long mirrored global practice, with endoscopic biopsy for all children key for diagnosis. National Institute for Health and Care Excellence[1] (NICE) guidelines on CD developed in 2009 and further updated in 2015 recommends endoscopic biopsy, reserving the use of Human Leukocyte Antigen (HLA) testing only in certain circumstances. On the other hand, The British Society of Paediatric Gastroenterology, Hepatology and Nutrition[2] (BSPGHAN) guidelines on CD released in 2013 embraces an approach endorsed by the European Society of Paediatric Gastroenterology, Hepatology and Nutrition[3] (ESPGHAN). This involves diagnosing CD by combining serological testing with HLA detection in symptomatic patients. The extent to which this shift has led to changes in practice is unknown.

Aim: We set out to investigate the uptake of these different national guidelines within English district general hospitals (DGH).

Subjects and Methods: This is a cross-sectional study of paediatric units in English DGHs completed between March- May 2017. A questionnaire was sent by e-mail with preference to paediatric consultants with a special interest in gastroenterology followed by general paediatricians.

Results: We contacted a total of 117 units with a response rate of 60% (n=70). The questionnaires were completed by 38 paediatric consultants with special interest in gastroenterology, 2 paediatric gastroenterologists, 29 general paediatric consultants and 1 paediatric dietitian. Of these, 96% (n=67) reported to having some form of guidance for their practice and 81% (n=57) reported of recent changes in their practice. This was reflected in the source of guidance used, with 58 units now using the new BSPGHAN guidelines either in combination with other guidelines or as the primary source and only 5 units reported to the sole use of NICE guidelines. For those using BSPGHAN guidelines, 96% (n=56) would make local diagnosis without endoscopic biopsy if meeting the criteria, only referring to tertiary centre if a biopsy was indicated following this policy

Summary and conclusion: This almost universal uptake of BSPGHAN guidelines across England is clearly out of step with UK NICE guidance. There may be a case for these national guidelines to be standardised, thus avoiding confusion and providing clarity on the pathway of diagnosis. More noteworthy is that the North American Society of Paediatric Gastroenterology, Hepatology and Nutrition[4] (NASPGHAN) guidelines still recommend endoscopic biopsies for all diagnosis of paediatric CD. The shifts in practice in England identified in this study clearly demonstrate that global practice for the diagnosis of CD is diverging between Europe and North America. Future work is clearly needed to ascertain the sensitivity and specificity of diagnostic pathways employing HLA testing and to rationalise best practice globally.

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***P29* Fecal calprotectin in pediatric juvenile polyps versus inflammatory bowel disease.**

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Introduction: Faecal calprotectin (FC), a stool biomarker of bowel inflammation is increasingly used as part of the non-invasive investigations of gastrointestinal symptoms in children.
Elevated FC levels are often seen in inflammatory bowel disease (IBD), but some studies have also reported elevated FC amongst children with juvenile polyps (JP).

Aims and Objectives: The aim of the study was to investigate the use of FC in the diagnosis of JP and to compare the average calprotectin levels amongst children with JP and those with IBD**.**

Subjects and Methods: We collected data retrospectively of patients aged ≤18 years diagnosed with JP and IBD with available FC levels in our centre over a 5 years' period (2011-2016).

FC levels were obtained from electronic biochemistry records and hospital clinic letters.

The comparison of FC levels between children with JP vs. IBD was limited to those with a calprotectin result within 3 months (90 days) of the diagnostic biopsy. Possible demographic confounders (age) were investigated.

Results: 26 cases of JP were identified between 2011-2016 in our centre. 14/26 (54%) had a FC result available, of which 13/14 were within 3 months of the date of the diagnostic biopsy for JP.
In the comparison group (IBD) we identified 33 children with biopsy diagnosis of IBD and a calprotectin result within 3 months of the biopsy.

All children (JP and IBD) had calprotectin levels above 'normal' (60 μg/g).
Average calprotectin levels seemed to be higher amongst children with IBD than those with juvenile polyps, although this did not reach statistical significance (p=0.07, Wilcoxon rank sum test). Interestingly, the children diagnosed with IBD were on average significantly older (mean 12.0 years, SD 3.5) than those with juvenile polyps (mean 5.5 years, SD 4.1) (T-test: p<0.001).

Calprotectin (μg/g)	IBD (n=33)	Juvenile polyp (n=13)	Significance test
Mean (SD)	1175 (1214)	625 (690)	p=0.06 (T-test)
Median (IQR)	872 (408-1352)	330 (230-662)	p=0.07 (Wilcoxon)

Summary: Calprotectin levels were available for about half of cases of JP, and were on average elevated, although perhaps to a lesser extent than in children with IBD.

There is also potential confounding by age, as children with JP were on average significantly younger than those with IBD. However, as other studies suggest that younger children have higher average calprotectin, it may be that the true difference in calprotectin levels between the groups is actually under-estimated in this study.

Conclusion: The differentiation between IBD and JP, as well as other inflammatory conditions of the colon and/or small intestine, cannot be based solely on FC. The high FC found in these non-IBD conditions may be one of the reasons for the low specificity of FC as compared to its sensitivity.

P30 **Qualitative evaluation of Psychological Input with children and Families within a tertiary paediatric Gastroenterology Service**

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Introduction: An evaluation was commissioned after completion of the first year of the Psychology service to examine the experiences of patients and their families who received psychology sessions and been discharged up until March 2017.

Aims and Objectives: The aims were to provide an insight into the experiences of patients and their families who received Psychology to understand what, if any, benefits they gained and secondly, to inform best practice in this area.

Subjects and Methods: Ethics approval gained from the Questionnaires Committee. 20 families were randomly selected from discharged patient. 14 parents and 7 young people responded and took part in semi-structured telephone interviews. The interviews were transcribed and subjected to thematic analysis.

Results: A thematic analysis identified three themes shown below:

I'm a patient - get me out of here	**The importance of being 'normal'**	**Don't judge me**
Timeliness is key	Not alone in this	Judged as a parent
Riding the roundabout	Working hard to provide my child with normality	My child being judged
I'd have walked over hot coals	Progress allowed me to feel normal again	Just listen to me

I'm a patient-get me out of here: Encompasses the feelings of desperation parents and patients were feeling before psychology offered. They felt they had gone around in circles, sometimes for years, and were keen to try anything which might work. They wished they had been offered the service earlier. Identified sub-themes are 'timeliness is key', 'riding the roundabout' and 'I'd have walked over hot coals'.

The importance of being normal: Patients and parents seek to be seen as "normal"; both find it of great comfort to know that they are not alone in their situation. Parents sought to provide a semblance of normality for their children. Young people believed that one of the benefits of psychology was being able to feel "normal" again. Sub-themes identified are 'not alone in this', 'working hard to provide my child with normality' and 'progress allowed me to feel normal again'.

Don't judge me: The central concept around this theme is that parents and patients/young people fear being judged; by health care professionals, their own children, peers or others such as school staff. Interventions work best when the patient/parent is heard, understood and helped with no judgement involved. Identified sub-themes are 'Judged as a parent', 'My child being judged' and 'Just listen to me'.

Summary: The study discusses the effects of service delivery on families and the effect of delayed or non-delivery. It concludes that not only is this service overwhelmingly of benefit to its recipients, but that an integrated psychology service should be part of standard practice for caring for young people with a chronic illness

Conclusion: The present study found that patients and families who used the service believed that they would have benefitted by accessing the service earlier. In addition, those interviewed believed that a key benefit of the service was in moving them toward feeling "normal". They also valued the non-judgmental approach offered by the psychologist and this was not something they had always felt in previous exchanges with health professionals.

***P31* Coeliac disease in the no biopsy era: Scope to do less scopes?**

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Introduction: The incidence of coeliac disease (CD) is increasing in many countries. ESPGHAN guidelines published in 2012 simplify the diagnostic process and reduce the need for biopsy to secure the diagnosis in symptomatic patients. This represented a significant shift in practice, requiring modifications throughout the multidisciplinary team.

Aims and Objectives: To assess compliance with ESPGHAN 2012 guidance and review individual cases in which new guidance was not followed.

Subjects and Methods: We performed a retrospective cohort study of patients diagnosed with CD in Southeast Scotland between 01.01.10 and 31.12.16. All raised anti-tissue transglutaminase (anti-tTG) levels are reported to the local lead for CD. We included 2 years of data collection prior to ESPGHAN guideline introduction, allowing analysis of a greater number of individual cases who had biopsy-confirmed diagnosis, acting as a form of control group. Results were cross-referenced with endoscopy lists and case-notes. Data were collected on sex, age at diagnosis, presentation (classical/non-classical/identified by screening, as per Oslo criteria), initial anti-tTG and biopsy result. For those not biopsied, repeat anti-tTG and HLA-DQ type were recorded. Data were recorded using a specific proforma.

Results: 382 (261 female) patients with CD were identified over this 7 year period. Median age at diagnosis was 8.3years (IQR 5.1-12.3). Increasing incidence was noted year on year. Since 2012, 177 patients met the criteria for no biopsy consideration. All had CD confirmed on further blood testing or biopsy. In total, since 2012, 44 patients have been diagnosed with no need for biopsy and in 2016, 23 of 43 (53.4%) eligible patients were diagnosed using the no biopsy strategy with the consultant doing a virtual consultation by telephone and sending informatics using e-communication. Diagnosis was confirmed by repeat anti-tTG and HLA-DQ2/8, performed on the day of the dietetic appointment, all within a month of the initial blood test.

Summary and conclusion: Our modified ESPGHAN 2012 strategy is a cost-effective option for the NHS and allows quicker diagnosis for the patient. Confidence in this approach is increasing. This study confirms that all eligible patients in our region could have had a secure diagnosis made without a biopsy.

***P32* The rising incidence of childhood coeliac disease – a 7-year regional cohort study**

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Introduction: The incidence of coeliac disease (CD) during childhood is rising in the United Kingdom, however many cases remain undetected as current prevalence is certainly not approaching 1%. CD may present with classic symptoms, detected following screening of at-risk populations or during investigation of non-specific symptoms if awareness levels are high. We have previously reported data from our region and have shown regional differences due to increased clinical awareness.

Aims and Objectives: To determine the incidence of childhood CD in Southeast Scotland over a 7-year period.

Subjects and Methods: We performed a retrospective cohort study of patients diagnosed with CD <16 years of age during 2010-2016, identified by direct laboratory notification of raised anti-tissue transglutaminase (anti-tTG) levels, endoscopy records and case-notes. Annual incidence was calculated using population figures published by Scottish government and further classified by mode of presentation (classical/non-classical symptoms as per Oslo criteria, or targeted screening) and method of diagnosis (biopsy/no biopsy). Poisson regression was used to calculate changes in incidence over time.

Results: 382 children were diagnosed during the 7 year period (31 diagnoses in 2010 and 85 in 2016 respectively) with a significant increase in incidence from 13.8 to 36.7 per 100,000 population. Female to male ratio was consistent (2.2:1 throughout period). Median age at diagnosis was 8.3 years (IQR 5.1-12.3). Only 8% were identified through targeted screening, with the remainder presenting classically/non-classically in equal proportion. Diagnosing CD without biopsy (as per European Society of Paediatric Gastroenterology Hepatology and Nutrition guidelines) began in our region in 2012, with 27% of total cases diagnosed with this strategy by 2016. 20% of diagnoses occurred in the least deprived 10% of the population, as determined by national multiple deprivation scores by postcode.

Summary and conclusion: The incidence of coeliac disease in children and young people <16 years of age increased 2.5-fold during the 7 year period from 2010-2016. A no biopsy strategy is becoming more common over time. To our knowledge the incidence of 36.7 per 100,000 in 2016, is the highest reported in the UK, whether in childhood or adult studies, and shows no signs of diminishing.

***P33* Paediatric Gastric Xanthoma: a case report**

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Introduction: Gastric xanthomas are single or multiple non-malignant lesions and are most frequently found in the gastric antrum. Even though multiple cases are reported in adults, gastric xanthomas are a rare finding in the paediatric population. We report a case of a thirteen year old female with an isolated gastric xanthoma, and review of the literature.

Subjects: A thirteen year old female with a diagnosis of Chronic Fatigue Syndrome was referred to our department with a five year history of intermittent chest pain. She was previously investigated with ECG, echocardiogram, barium swallow and MRI thorax, all of which were normal. She also had an empiric trial of proton pump inhibitors for possible gastro-oesophageal reflux disease, without good response. She subsequently underwent an oesophago-gastro-duodenoscopy (OGD) to explore anatomical or inflammatory causes of her ongoing symptoms.

Results: OGD revealed a single white well circumscribed lesion approximately 3x3 mm in the gastric antrum. Histology revealed foamy cells which were confirmed to be macrophages. Routine staining and immunostaining for Helicobacter pylori were negative and there was no evidence of malignancy. A second gastric antrum biopsy showed normal architecture. Oesophageal and duodenal biopsies were normal.

Summary: This is a rare case of an isolated gastric xanthoma which was found incidentally in an adolescent girl investigated for chronic episodic chest pain. Paediatric gastric xanthomas are rare and there are only four other case reports: one was an isolated gastric xanthoma, whereas multiple lesions were found in the rest of the patients (2, 4 and >10). Gastrointestinal xanthomas are an unusual finding in the adult population. Most adult xanthomas are found in the gastric mucosa, but they have been reported in the oesophagus, duodenum and large bowel. They are non-malignant lesions that can either be single or multiple and ranging in diameter from 0.5-10mm. They are characterised by the accumulation of foam cells in the lamina propria. In adults, gastric xanthomas have been associated with hyperlipidaemias, chronic gastritis, Helicobacter infection and intestinal metaplasia. Only in one of the paediatric cases was there an association with Helicobacter Pylori.

Conclusion: We present a thirteen year old female with an isolated gastric xanthoma as an incidental finding. In the adult population, they have been associated with hyperlipidaemias, chronic gastritis, Helicobacter Pylori and intestinal metaplasia. Gastric xanthomas are a rare finding in the paediatric population, with only four other cases reported and no known associations with other conditions.

***P34* The worrying headache – A case of cerebral sinus thrombosis in a patient with Ulcerative Colitis.**

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Introduction: Inflammatory bowel disease (IBD) is associated with an increased risk of arterial and venous thromboembolism due to its procoagulatory state[1, 2]. Cerebral venous sinus thrombosis (CVST) is a very rare complication of IBD in children[3].

Aims and Objectives: To report a rare case of CVST in a child with newly diagnosed ulcerative colitis and to discuss the importance of early recognition of CVST.

Subjects and Methods: Details were collected from a review of the case notes.

We present a 7 year old boy who was referred to Watford general hospital with a 3 month history of loose stools, per rectal bleeding and weight loss. Blood tests showed thrombophilia (721), Haemoglobin of 77 g/L and his faecal calprotectin was high (5268 mg/Kg). He had a gastroscopy and colonoscopy which showed pancolitis macroscopically with multiple colonic ulcers. Histology of the biopsy specimens showed chronic gastritis, colonic crypt distortion, occasional crypt abscesses and diffuse moderate chronic laminal propria inflammation with no granulomas consistent with ulcerative colitis. He was treated with three days of IV methylprednisolone and was discharged on a weaning course of oral prednisolone and mesalazine. He presented to our accident and emergency department 9 days later with a few days history of headaches, which were rapidly progressing in severity. He had a generalised tonic clonic seizure which was unresponsive to benzodiazepines and he was intubated and ventilated. He was transferred to a nearby Paediatric intensive care unit. A CT Head done at presentation showed no abnormalities but a subsequent cerebral venogram revealed an extensive cerebral venous sinus thrombosis. He developed intracranial hypertension, severe papilloedema and encephalopathy as a result. A heparin infusion was ineffective, so he had an endovascular thrombectomy and thrombolysis by interventional radiology. He had significant motor and speech deficits requiring intensive neurorehabilitation and also required psychological support for his mood.

Results: He has now regained normal motor and speech function. He was commenced on Azathioprine a month after the CVST to maintain remission and he remains on regular antiepileptic medication. It was challenging to achieve adequate anticoagulation with Warfarin which might be due to interaction with Azathioprine metabolites[4] and he required high doses of Warfarin to achieve therapeutic levels of INR with bridging low Molecular weight heparin.

Summary: This presentation discusses the case of a 7 year old boy with Ulcerative Colitis who presented with headache and rapidly deteriorating neurological symptoms secondary to a cerebral venous sinus thrombosis. This patient had a CT Head which showed no abnormalities, however a cerebral venogram revealed a cerebral venous sinus thrombosis. He developed significant neurological sequelae as a result and anticoagulation was challenging.

Conclusion: When a patient with IBD presents with headache or other neurological symptoms, clinicians should have a high index of suspicion for CVST as it could be potentially fatal. A cerebral venogram would be more useful than a plain CT in the diagnosis of CVST. Anticoagulation might be difficult in patients who are on Azathioprine due to drug interactions.

***P35* A rare cause of iron deficiency anaemia in childhood**

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Case Presentation: An 8-year-old boy presented to his local hospital with a few weeks history of lethargy, palpitations and headaches. His haemoglobin was found to be low at 49 g/L with a ferritin of 8 ug/L. There was no history of bleeding, nosebleeds, haematemesis, rectal bleeding and no family history of bleeding disorders. There were no other symptoms of note. He was given packed red cell transfusion and commenced on iron supplements. He represented 10 months later with similar symptoms of lethargy and palpitations. Haemoglobin had dropped to 57 g/L. He was referred to the paediatric haematologist at Royal Manchester Children's Hospital for further investigations.

Investigations: Blood tests revealed normal Vitamin B12, folate, renal function, liver function and CRP. ESR was raised at 40 mm/1st hour and orosomucoid 1687 mg/L. Bone marrow aspirate was normal with reactive features. Chest X-ray was normal. Ultrasound abdomen showed 4 rounded hypoechoic lesions in the epigastrium, superficial to the stomach at the tip of the left lobe of liver. They were in an atypical position for nodes and the largest measured 15.5mm. MR scan abdomen did not demonstrate any abnormality but the scanning was inadequate due to significant artefact movement. An oesophagogastroduodenoscopy performed revealed a large sessile gastric polyp at the middle of the greater curvature of the stomach measuring 8-10 centimetres in diameter. A rapid urease test was negative but a course of treatment for H.pylori was given based on the endoscopy findings.

Treatment: He proceeded with surgery and had a wide local excision of the lesion with adequate clear margins. Intraoperatively, he needed a partial gastrectomy due to the size of the lesion, leaving approximately 40% of the stomach.

Result: The histology of the lesion showed an ill-defined spindle cell lesion that involves the mucosa, submucosa, muscularis propria, serosa and focally some adherent fibrofatty/mesenteric tissue. The histomorphology and immunohistochemical profile confirmed an inflammatory myofibroblastic tumour. The tumour was completely resected with no evidence of spread outside the stomach or elsewhere. Lymph nodes sampled as part of the surgery were negative. No radiation therapy or chemotherapy was required.

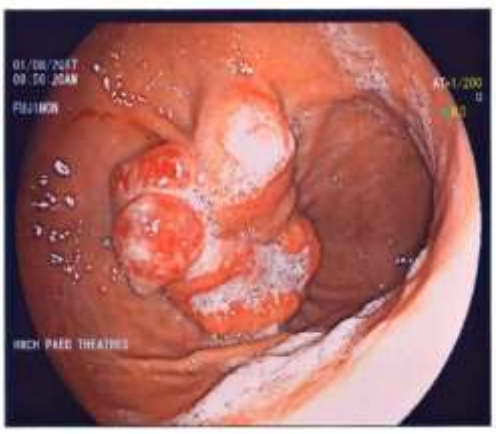

Figure1. Endoscopic image of large sessile gastric polyp at the greater curvature of the stomach measuring 8-10cm in diameter with surface ulceration and bleeding

Outcome and follow-up: Post-operatively he made a good recovery and was tolerating a full diet. He was discharged with regular follow-up with the oncologist, surgeons and gastroenterologist.

Discussion: Inflammatory myofibroblastic tumour (IMT) is a rare spindle cell neoplasm comprising of fibroblasts and myofibroblasts with a non-specific inflammatory infiltrate. IMT is rare and most often affect young adults and children. It has been reported in practically all organ systems with the liver and biliary tract the commonest affected region followed by respiratory tract and gastrointestinal tract[1]. Gastric IMT in children is very rare and the available literature comprises of a small number of case reports. Common presentation

includes malaise, pallor, weight loss, fever and abdominal pain[2]. Primary excision of the tumour remains the most widely reported treatment and is generally successful in effecting long-term cure. Recurrence has been reported and some resulting in death[2-5]. Recurrence is mainly treated with further surgical resection and chemotherapy[2-4]. Gastric IMT has an unpredictable outcome and all patients require careful clinical, biological and radiographic follow-up after resection to monitor for recurrence of tumour.

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***P36* A Single Centre 2-Year Experience of Paediatric Wireless Capsule Endoscopy: Benefits, Risks and Considerations**

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Introduction: Wireless capsule endoscopy (WCE) for the investigation of small bowel pathology has been established in our tertiary paediatric gastroenterology service since July 2015. In this report, we describe our two-year experience with WCE.

Methods: All paediatric patients aged ≤18 years who underwent WCE from 1 August 2015 to 1 August 2017 were included in our study. The wireless capsule system used was the MiroCam, manufactured by Intromedic. We audited compliance to standards from NICE and NHS England. These were as follows: correct indications for WCE, procedure to be performed within six weeks of request, successful end-to-end visualisation of the small bowel and no significant complications of WCE. We aimed for 100% compliance to these targets. Data was collected electronically.

Results: Over a two-year period, 26 patients underwent WCE. 14 (54%) were male and 12 (46%) were female. Median age was 11.5 years (range 2-18). 13 patients (50%) were able to swallow the wireless capsule, with the youngest patient being 8 years old. The rest were placed endoscopically. Of the indications for WCE, 11 (42%) were for investigation and surveillance of polyposis, 10 (38%) for suspected Crohn's disease, 3 (12%) for occult gastrointestinal bleeding, and 2 for other gastrointestinal symptoms.

4 (15%) of procedures were unsuccessful or provided only limited views. Of these 1 was due to a battery failure while 3 were secondary to food debris. One had previous bowel surgery (swallowed WCE) and 2 were endoscopically placed but were allowed to eat after 2 hours of placement.

Of the 26 procedures, 15/26 (58%) revealed pathology. These included polyps (8, 30%), small bowel ulcers (5, 19%), small bowel bleeding (1, 4%) and lymphangiectasia (1, 4%)

21/26 (81%) procedures were completed within 6 weeks of request. We had no significant complications.

Discussion: In our experience, WCE is a safe and useful method for assessing small bowel pathology. Through careful patient selection, we experienced no complications and more than half of patients had clinically significant findings. To avoid failures in procedure, we recommend that bowel preparation procedures and the timing prior to food re-introduction be altered in cases where the wireless capsule is endoscopically placed, or if there are risk factors of slow motility.

P37 **Medical and surgical therapies for the treatment of gastrointestinal dystonia in children with severe neurodisability; A systematic review.**

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Introduction: In children with severe neurodisabling conditions the clinical constellation of, pain behaviour, retching, bloating, abdominal distension and constipation can be referred to as gastrointestinal dystonia (GID). These problems tend to present in enterally fed patients who may have already has antireflux surgery. Suggested therapies have included extensive pharmacopenia and jejunal feeding, but the evidence base for such therapies remain obscure.

Aims and Objectives: To inform a joint BSPGHAN/RCPCH guideline development, for GID, we aimed to appraise the published evidence on the medical and surgical treatments for GID via systematic review of the literature and to suggest where deficit in the literature could be addressed with guidance.

Subjects and Methods: Systematic retrieval of data for patients <18yrs with severe neurodisability and gastrointestinal symptoms. Outcomes sought; weight gain, longitudinal growth, pain, quality of life and mortality. We excluded papers where outcomes were the treatment of gastro-oesophageal reflux alone. Electronic searches of Cochrane library, Pubmed (to Nov 2017) and Medline (1946-Nov 2017) were made using the following keyword and MeSH terms; cerebral palsy, neurodisability, child, nutrition, foregut dysmotility, neurogastroenterology, prokinetic, domperidone, baclofen, aprepitant, fosprepitant, levopeamazine, nabilone, jejunostomy and retching. Hand searches of meetings of relevance and personal collections were also perfomed. Two authors independently assessed the level of evidence (EL) was assessed using SIGN (Scottish Intercollegiate Guidelines Network) methodology (http. //www.sign.ac.uk) a third author arbitrated.

Results: Search strategy yielded 2811405 hits. Combination searches reduced this to 6064 titles and abstracts. 32 medical and 27 surgical studies were identified as potential studies and reviewed in full, including 1 guideline and 3 Cochrane reviews. 30/32 medical and 18/27 surgical studies were excluded as they were not relevant to our focused clinical question or where specific patient group could not be separated from general data. 2 medical studies related to parenteral nutrition (PN) were included and 9 surgical studies relate to jejunal feeding were included (**Table**)

Therapy	**No of studies (no of patients)**	**EL**	**Outcomes and comment**
PN	2 (13)	3	Weight gain, cessation of pain, improved feed tolerance post Rx.
Gastro Jejunal (transpyloric) tube	4 (401)	3	Some weight improvement, frequent tube replacements, 1 study 15% mortality
Witzell jejunal tube	1 (33)	3	Some growth improvement, volvulus, no mortality data.
Primary jejunal button	1 (16)	3	Improvements in weight, volvulus, 0 mortality
Roux and Omega jejunostomies	3 (27)	3	Long term tolerance and growth report, 1 mortality reported.

Summary: Medical data are limited to PN for treatment of GID. Surgical data, although low EL, supports jejunal feeding for growth and symptoms, permanent jejunal feeding is reported with Roux, omega ostmies and primary jejunal buttons.

Conclusion. The data for treatment of GID are limited and make developing evidence based guidelines inappropriate. Invested health professional should focus on sharing of experience of specialist multidisciplinary teams, increasing experience and data on novel pharmacopenia. The authors are now developing consensus based guidelines via Delphi process that will aid the standardisation of care, audit of practice and development of research of medical therapies.

***P38* Pseudothrombocytopenia in a patient with Crohn's disease**

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Introduction: We report a case of a patient with Crohn's disease who had low platelet counts on multiple occasions and was eventually found to have EDTA-dependent thrombocytopenia.

Case: A 14-year old boy of mixed English and Spanish ethnicity was seen for follow-up of Crohn's disease. He had previously responded well to exclusive enteral nutrition, but disease activity had worsened with severe changes in his terminal ileum. As part of his monitoring, a full blood count (FBC) was sent. This reported platelet clumping, and an accurate platelet count was unable to be provided.

At his next appointment three months later, a repeat FBC was sent, which returned a platelet count of 33x 10^9/L. He showed no signs of thrombocytopenia (easy bruising, bleeding), and a repeat FBC showed platelet counts of 141 x10^9/L. Sequential FBCs after that showed thrombocytopenia ranging between 58 to 108 x10^9/L on more than 10 occasions. This did not improve despite adequate treatment of his Crohn's disease with escalation to infliximab. We sought a haematology opinion, who advised for a repeat sample to be sent in a citrated tube. Previous samples were sent in ethylenediaminetetraacetic acid (EDTA) tubes. The citrated sample showed a normal platelet count of 185 x 10^9/L, confirming suspicions of EDTA-dependent thrombocytopenia. No further investigation was therefore indicated.

Discussion: Pseudothrombocytopenia (PTCP) is an in-vitro phenomenon of platelet aggregation, resulting in spurious reporting of a low platelet count by automatic cell counters, which are typically EDTA- dependent. EDTA, a calcium chelator, is generally considered to be a reliable anticoagulant for FBC testing because of its stability in blood cell counting and sizing. Platelet clumping in the presence of EDTA is caused by an autoantibody against glycoprotein IIb/IIIa located on the cell membrane of platelets, which has no pathological significance[1]. Although possible, other anticoagulants such as heparin and sodium citrate rarely induce such phenomena.

There is a suggestion that malignancy, chronic liver disease, infection, pregnancy, autoimmune disease, and cardiovascular disease may increase the risk of EDTA-dependent PTCP[2]. To our knowledge, PTCP in inflammatory bowel disease has not previously been described in the medical literature. There have been reported cases of infliximab-induced thrombocytopenia in adult patients with ulcerative colitis, and it was therefore important that this was excluded in our patient.

A misdiagnosis of PTCP can lead to unnecessary investigations and treatments, causing anxiety and potential harm to the patient. Patients with PTCP have previously been subjected to platelet transfusions, long-term steroid therapy, and even splenectomy[1]. Therefore, it is important that when low platelet counts are noted that PTCP should also be considered as a potential diagnosis.

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***P39* Can food allergy present as protein-losing enteropathy?**

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Introduction: Protein-losing enteropathy is a presenting feature of several gastrointestinal conditions and is characterised by an excessive loss of serum proteins into the gastrointestinal tract leading to hypoalbuminaemia, generalised oedema and potentially pleural and pericardial effusions. The increase in the intestinal leakage of plasma proteins occurs via one of two mechanisms: the first is mucosal injury with or without erosions or ulcerations; the second is an increased lymphatic pressure in the gut that causes the dilated lymph vessels to leak protein via the surface epithelium into the gut. Evidence that food allergy can present as protein-losing enteropathy is sparse in the literature and yet clinical experience suggests otherwise.

Aims and Objectives: We looked to the clinical presentation, physical findings, laboratory values and histology results of seven patients with protein-losing enteropathy secondary to food allergy. The aim was to evaluate the clinical response to different therapies. Also to evaluate their histology results for those ones who underwent endoscopy to distinguish any features could explain their clinical presentation.

Subjects and Methods and results: We describe seven children with median age of 15 months (range 5 -35 months) presenting to a single tertiary referral centre over a two years period with significant protein-losing enteropathy secondary to food allergy. Clinical presentation was with hypoalbuminaemia, increased protein loss in the gastrointestinal tract and oedema ranging in severity from peri-orbital to generalised. All patients had peripheral eosinophilia. Five patients did, and two did not, undergo endoscopy. Isolated eosinophilic esophagitis was seen in three patients and two had a marked raise in eosinophil count in more widespread gastrointestinal locations. Clinical and serological markers of allergy including eczema, total immunoglobulin (Ig) E levels and specific IgE to foods yielded mixed results. All patients made a complete recovery with amino acid based feeds.

Conclusion: We conclude that although under-recognised in the literature, food allergy presenting as protein-losing enteropathy is not uncommon. Response to elemental diet is generally excellent and it may be reasonable to trial this therapy even without endoscopic confirmation of eosinophilic mucosal inflammation.

P40 **Functional Gastrointestinal Disorders (FGIDs) and sleep disturbance in children (5-15 years) - a frequent association: an observational study from a District General Hospital.**

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Introduction: FGIDs are one of the commonest presentations in gastroenterology & general paediatric clinics. It is also well known that sleep disturbance is commonly associated with FGIDs, but the opportunity to intervene could be missed during clinical contact unless history of sleep disturbance is specifically asked and identified.

Aims and Objectives: Aim of this observational study was to explore the extent of sleep disturbance & FGIDs from clinical history to highlight the scope for a better future management of sleep disturbance at the primary and secondary care.

Subjects and Methods: A prospective data was collected from patients with FGIDs over 18 months (2016-17) using structured clinical history (based on ROME IV criteria) & separate questionnaires to assess sleep disturbance. Patients with autism, Attention Deficit & Hyperactive Disorder, complex neuropsychiatric diagnosis & chronic neuro-developmental disorders were excluded. A total of 65 cases (20 males, 45 females) between 5 to 15 years with FGIDs were identified and diagnosed using Paediatric Rome IV criteria.11 males & 26 females were between 5-10 years whereas 9 males & 19 females were between 11-15 years.

Results: Commonest presentation was functional abdominal pain disorders, H-2 (38/65 - 58%); Functional defecation disorder, H-3 (24/65 – 37%); functional nausea & vomiting disorder, H-1(15/65 – 23%). A degree of overlap within the three main types was observed in the patient-parent reported symptoms. Sleep disturbance was assessed by using structured clinical history questionnaires. 40/65 (61%) children have one or more types of sleep disturbances, as shown below:

1- Problem of sleep initiation: 7 males & 18 females (n=25)
2- Excessive daytime sleepiness & tiredness: 4 males & 14 females (n=18)
3- Frequent awakening during night: 11 males & 25 females (n=36)
4- Short duration of sleep: 16 males & 24 females (n=40)
5- Perceived degree of severity: Significant (n=12), moderate (n=15) mild (n=13) severity

Summary: It is evident from the above data that abdominal pain is the commonest presentation & nausea/vomiting is the least common presentation in this cohort. In addition, variabilities of sleep disturbance in parental & patient's history is an important finding. However, only a few children have significant sleep disturbance affecting school performance and daytime sleepiness. The history of sleep disturbance is essential in children with FGIDs in a clinical setting. More awareness and effective early intervention using standard sleep hygiene advice should be included in clinical management as a routine to improve the quality of life in these children. In our clinical setting, we tend to use 'NHS-Choices' web resources & printed parental information for sleep hygiene. Keeping the concept of 'Gut-Brain Axis' in mind, the symptomatic improvement of FGIDs may be linked to sleep hygiene. If standard measures fail, the severe spectrum of cases may benefit from structured sleep assessment in a sleep laboratory.

Conclusion: Use of ROME IV criteria for the diagnosis of FGIDs is helpful for diagnostic labelling and should be used while taking focussed history. The abnormal sleep is a frequent finding in FGIDs & opportunity of early intervention through patient/parent education should be encouraged. This practice may help to improve the quality of long-term care in children with FGIDs. The advice on sleep hygiene should be offered in primary & secondary care using educational leaflet and NHS web resources.

P41 **Diagnostic yield for scheduled endoscopy at a tertiary paediatric gastroenterology centre in London**

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Introduction: There is significant variation in the paediatric admission rate for gastrointestinal endoscopy across England. It is unclear if this reflects variation in prevalence of gastrointestinal disease, or in access to services. The latter might be reflected as variation in diagnostic yield between regions/centres. Here we provide the diagnostic yield for a single tertiary gastroenterology unit in London.

Methods: Retrospective case-notes analysis covering a seven-month period in 2016. All children undergoing scheduled upper and/or lower gastrointestinal endoscopy on a diagnostic list in our centre were identified from an electronic diary system. Endoscopies were categorised as 'unremarkable' (no pathological features identified macroscopically or microscopically), or 'pathological'. The studies was registered locally as a departmental audit and formal ethical approval was not required.

Results: 169 children underwent gastrointestinal endoscopy (upper (OGD-only): lower (colon-only): both (OGD-colon) = 82:17:70). 32 were having surveillance procedures for known gastrointestinal disease (mainly inflammatory bowel disease or eosinophilic oesophagitis). 50% (41) of OGD-only, 59% (41) of OGD-colons, and 69% (11) of colon-only procedures were pathological. There were 26 new diagnoses of inflammatory bowel disease.

Conclusion: In our centre most children undergoing gastrointestinal endoscopy had detectable pathology. Comparison of diagnostic yield between centres may help in the interpretation of variation in endoscopic investigation rate.

***P42* Upper GI Endoscopy is an Expensive Accessory Investigation in the Diagnosis of Crohn's Disease in Children**

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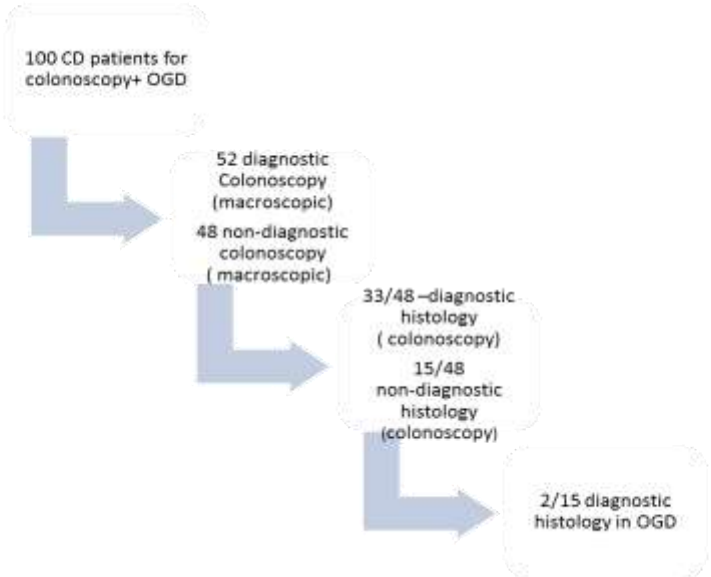
Introduction: ESPGHAN Porto revised criteria recommends upper gastrointestinal endoscopy (OGD) and ileo-colonoscopy with small bowel imaging for all suspected patients with Crohn's disease (CD).OGD is recommended with an aim to improve the diagnostic yield in patients suspected to have Crohn's disease (CD).

Aims and Objectives: To analyse the additional diagnostic yield gained from OGD in patients who had diagnostic colonoscopy for suspected IBD.

Subjects and Methods: We have done a retrospective analysis of the data of 100 consecutive CD patients diagnosed in the time period of 2013-2017. All these patients had OGD and colonoscopy. . Endoscopy reports and histological reports were reviewed. We have analysed the cost of OGD in these patients.

Results: Out of 100 CD patients (L1 33%, L2 36%, L3 31%) who underwent both OGD and colonoscopy, 52 colonoscopies and 34 OGD were diagnostic macroscopically (Ulcers typical of Crohn's disease were seen). Ileal intubation rate was 80%. Histology was diagnostic of CD in 76% colonoscopies and 41 % of OGDs. Of the 48 patients in whom colonoscopies were non diagnostic macroscopically, only 15 patients (31%) had macroscopic abnormalities of Crohn's disease in OGD. All patients who had diagnostic colonoscopy features of Crohn's disease, histology was confirmative of CD. Of the 48 patients who had non-diagnostic colonoscopy appearance of CD, 33 had diagnostic histology of CD. Only 2 patients had diagnostic histology from OGD out of the 15 patients who had non-diagnostic histology from colonoscopy. In summary OGD provided additional diagnostic yield only in 2% patients (picture 1). Approximately 66,000 GBP could have been saved by avoiding OGD in 85% of these patients (average cost of diagnostic OGD is £775/-).

Conclusion: 85% of patients with Crohn's disease can be diagnosed by colonoscopy and histology. OGD provided additional diagnostic yield in only 2% patients with Crohn's disease.



***P43* Adalimumab Monotherapy is as effective as Combination Therapy in Paediatric Crohn's Disease**

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Introduction: Adalimumab is an effective treatment for induction of remission and maintenance of remission in children with Crohn's disease (CD). The value of combination therapy with Adalimumab and immunosuppressive agents including Thiopurines and Methotrexate is unclear.

Aims and Objectives: To compare the outcome of CD patients receiving Adalimumab mono and combination therapy.

Subjects and Methods: We have done a retrospective review of the medical records of CD patients receiving Adalimumab treatment. The data was collected for a period ranging from January 2013 to August 2017. We collected data on patient demography, disease characteristics, co-immunosuppression, laboratory parameters, clinical remission (defined by Physician Global Assessment), mucosal healing (assessed by Simple Endoscopic Score-CD), serious side effects including malignancy and serious infections, Adalimumab drug levels and the need for dose/frequency adjustment of Adalimumab.

Results: 55 CD patients have received Adalimumab treatment. 17 patients were on monotherapy and 38 patients on combination therapy. Patient characteristics and disease characteristics were similar in both the groups. Thiopurines and Methotrexate were the co-immunosuppression in 33 (87%) and 5 (13%) patients respectively. There was no statistically significant difference between rate of clinical remission, mucosal healing, CRP normalisation, dose/frequency adjustment of Adalimumab. No serious infections or malignancies were recorded in patients in both the groups. Details in table 1

	Adalimumab monotherapy N=17	**Adalimumab Combination therapy N=38**	**P value**
Median age at diagnosis (range)	13 years (2.5-17)	12 years (6-15.5)	n.s
Male	14/17 (82%)	24/38(58%)	n.s
Disease distribution	L1 8 (47%) L2 2(12%) L3 7(41%)	L1 6(16%) L2 12 (32%) L3 20 (52%)	n.s
Indication for Adalimumab treatment Luminal CD	16 (94%)	30 (79%)	n.s
Adalimumab as primary biologic agent	10 (59%)	30 (79%)	n.s
Median follow-up (months)	24(8-40)	22(1-47)	n.s
Clinical remission at last follow-up (PGA) Remission Mild to moderate	 12 (71%) 5 (29%)	 28 (74%) 10 (26%)	 n.s
Complete mucosal healing (SES-CD score 0)	6/10 (60%)	19/24 (79%)	n.s
% of patients with normal CRP (<10)	13 (76%)	34 (89%)	n.s
% of patients needing dose/frequency adjustment	10 (59%)	16 (42%)	n.s
Adalimumab levels <5 5-10 >10	 5/17 (25%) 10/17 (59%) 2/17 (12%)	 1/31 (3%) 19/31 (61%) 11/31 (35%)	 n.s n.s n.s

Summary and Conclusion: Monotherapy with Adalimumab is as effective and safe as combination therapy in children with Crohn's disease.

***P44* Remsima® is cost effective and safe in managing Paediatric Inflammatory Bowel Disease: A Prospective Study**

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Background and Aims: Remsima®/CT-P13 the biosimilar of Remicade® has recently entered the European market. There is limited data available on its use in Inflammatory Bowel Disease (IBD) in children. In this study we aimed to prospectively investigate the safety and cost implications of 1) switching from Remicade® to Remsima® in children with IBD 2) Initiating treatment with Remsima in children with IBD.

Methods: All children who were treated with Remicade® for IBD in the Department of Paediatric Gastroenterology at The Royal London Hospital were switched to Remsima®. All children with IBD whose treatment required escalation to a biological were started on the biosimilar, Remsima® instead of Remicade®.

Primary endpoints include a change in inflammatory markers, change in disease activity score, and the development of any adverse effects.

The total number of infusions and their cost were recorded from January 2017 to November 2017. The C-reactive protein (CRP) and Erythrocyte Sedimentary Rate (ESR) was measured prior to infusion. Adverse effects were recorded in all patients' immediately after infusion and reassessed at both virtual and face to face clinics post infusion.

Results: Our cohort consisted of 63 IBD children (34 male and 29 female) with a median age of 14 (range 7-16). 48 children with Crohns and 15 with UC. 45 (35 Crohns, 10UC) children were switched from Remicade® to Remsima®. 18 (13 Crohns, 5UC) children started treatment with Remsima® having never previously been on Remicade®. Median follow up was over 8 months (range of 1-9 months).

280 infusions have been performed since January 2017. 235 of these were Remsima® and 45 were Remicade®. The cost of a single vial of Remsima® 100mg vial is £162.00, 53.6% cheaper than Remicade® 100mg. The average infusion dose in this study was 350mg which equates to a cost saving of approximately £150,000 over 9 months.

84% of patients tolerated the medication without any adverse effects. 84% of patients had improved disease activity scores, which correlated to improved or normalised inflammatory markers. 9.5% of patients developed an adverse reaction to Remsima®. 4.8% of patients developed anaphylaxis to Remsima®, and less than 5% developed antibodies.

Conclusion: We demonstrated that switching from Remicade® to Remsima® was cost effective, safe and feasible. Switching was not associated with significant side effects and did not impact the short term clinical outcomes in children with IBD.

P45 **Medical treatment and surgical outcome of children and adolescents with ulcerative colitis improved in the 4th IBD audit round**

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*All Consultant Paediatric Gastroenterologists.

Introduction: Children with ulcerative colitis are affected by steroid dependency, anaemia, and complications of surgery. Steroid sparing strategies and surgical outcomes had not been assessed on a representative national level.

Aims and Objectives: We examined morbidity and escalation treatment for children with active UC, steroid sparing strategies, proportion of second line treatment and surgical outcome, highlighting geographical differences, areas of improving practice and areas for future development.

Subjects and Methods: The 4th round of our national prospective audit was conducted for the inpatient period of all children with ulcerative colitis for medical or surgical treatment in the UK from 1.1. -31.12.2013. 32/34 invited centres participated and recruited 224 children in 298 admissions. We compared results with two previous paediatric audit rounds.

Results: Over six years, recording of PUCAI score (median 65)(23% to 55%, p<0.001), guidelines for acute severe colitis (43% to 77%, p 0.04), and ileal pouch surgery registration (4% to 56%, p<0.001) have increased.
Corticosteroids were given in 183/298 episodes (61%) with 61/183 (33%) not responding and requiring second line therapy or surgery. Of those treated with anti-TNFalpha (16/61, 26%), 3/16 (18.8%) failed to respond and required colectomy. Prescription of rescue therapy (26% to 49%, p=0.04) and proportion of anti-TNFalpha (20% to 53%, p=0.03) had increased, colectomy rate (23.7% to 15%) was not significantly reduced (p=0.5).
Subtotal colectomy was the most common surgery performed (n=40), and surgical complications from all procedures occurred in 33%. In 215/224 (96%) iron deficiency anaemia was detected and in 51% treated, orally (50.2%) or intravenously (49.8%).

Summary: Our national audit programme has proven effective to reduce steroid side-effects and iron deficiency anaemia in children with UC. Although over 6 years in the era of biologics there was a trend of decreasing colectomy rates, nearly half of children requiring colectomy had to be operated non-electively, indicating the importance of early recognition, optimising treatment, and collaborative gastro-surgical assessment.

Conclusion: Oral and intravenous iron therapy was efficient and safe. More than half children with rescue therapy received anti-TNFalpha, and nearly 20% of those failed to respond and required colectomy. Subtotal colectomy was required in 13.7% of patients admitted, and complications occurred in one third of surgical patients with UC.

***P46* Role of faecal calprotectin in the diagnosis of paediatric inflammatory bowel disease (PIBD)**

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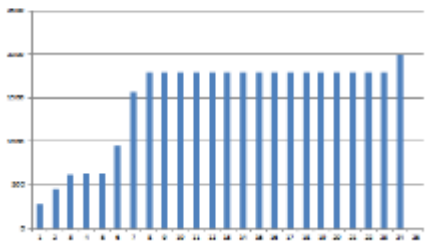
Introduction: Faecal calprotectin is increasingly used in primary and secondary care to screen for PIBD and this often leads to increased anxiety for healthcare professional and families in addition to increase in referrals to tertiary gastroenterology services.

Aim: To review usefulness of faecal calprotectin in prediction of PIBD diagnosis

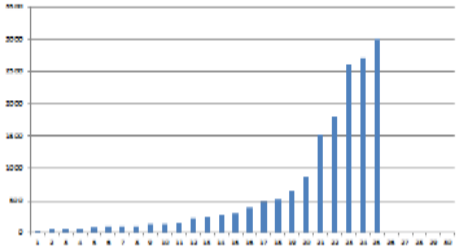
Method: Retrospective review of faecal Calprotectin results in patients' undergone diagnostic colonoscopy (with or without endoscopy) was done during 8 month period (Jan – Aug'17). Colonoscopies done for reassessment and polyposis screening were excluded.

Results: 72 patients underwent diagnostic colonoscopy over study period. 42 patients had normal colonoscopy findings and 30 had abnormal findings. Abnormal colonoscopy findings included PIBD (20), solitary polyps (5), coeliac disease (1), infection (1) and H. Pylori (2). Faecal calprotectin was done in 30/42 and 25/30 in patients with normal and abnormal colonoscopy results respectively.

Calprotectin in patients with abnormal colonoscopy results (n=25); mean=1497



Calprotectin in patients with normal colonoscopy result (n=30); mean=528



Faecal calprotectin in patients with normal colonoscopic findings (n=30) was requested by GP (13), paediatrician (8), tertiary gastroenterology team (7) and paediatric surgeon (2).

Conclusion: Faecal calprotectin is an important marker in deciding definitive diagnostic investigations for PIBD. It is a very sensitive test for PIBD however it is not very specific and can be raised in other inflammatory and infective conditions affecting bowel. Faecal calprotectin can be raised in absence of specific pathology due to transient bowel upsets (normal colonoscopy findings) and normal range in children can be above that in adults leading to low threshold for referral and investigations.

Understanding of differential diagnosis of raised faecal calprotectin and age appropriate faecal calprotectin levels in primary and secondary care can help referral pathways and better utilisation of resources.

***P47* An investigation of azathioprine on autophagy pathway activity**

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Introduction: Autophagy is an intracellular process that degrades damaged proteins and organelles to maintain cellular homeostasis. Defective autophagy has been strongly linked to inflammatory bowel disease (IBD) pathogenesis, with evidence that enhancing autophagy may be therapeutically beneficial by regulating inflammation and clearing intestinal pathogens. Due to the difficulties and high costs associated with the development of new drugs, a more comprehensive characterisation of existing IBD drugs and their mechanism of action in the context of autophagy is required.

Aims and Objectives: To investigate the effect of azathioprine on autophagy pathway activity and to determine the molecular mechanisms involved.

Subjects and Methods: We have characterised the autophagy response to azathioprine by monitoring the autophagy marker LC3 using several complimentary methods including live-cell imaging, flow cytometry and western immunoblotting. Additionally, using the Human Autophagy RT² Profiler, we characterised changes in autophagy signalling genes in response to azathioprine.

Results: Our results identify the immunosuppressant azathioprine as a strong inducer of autophagy. We show that azathioprine induces autophagy in part through inhibition of the mTORC1 pathway. Further, using the Human Autophagy RT² Profiler, we show that the PKR-like ER kinase (PERK) gene, which initiates the unfolded protein response, is up-regulated by azathioprine.

Summary: Azathioprine is a strong inducer of autophagy and the ER-stress/unfolded protein response.

Conclusion: The modulation of autophagy represents an exciting therapeutic option for the treatment of IBD, and evidence is already emerging that drugs currently used for the treatment of IBD can affect the autophagy pathway. The crosstalk between autophagy and ER-stress offers new options for how IBD could be targeted and combination treatments aimed at modulating both the UPR, and autophagy, warrant further investigation.

***P48* Audit of referral to Leicester Children's Hospital for suspected Inflammatory Bowel Disease (standard 5 of new BSPGHAN / RCPCH quality standards[1])**

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Introduction: In January 2017 the new BSPGHAN / RCPCH quality standards were published. The first step to implementation is to audit current practice. We present an audit of Standard 5 "Children with suspected inflammatory bowel disease are seen by a specialist service within four weeks in an age appropriate facility by a multi-disciplinary team"

Aims and Objectives: The aim was to see how we performed during 2016-2017 compared to this standard of care, and to see what changes we may need to make to improve.

Subjects and Methods: We used our IBD database to identify children diagnosed with IBD over 2016 and 2017. We used the HISS system to note the date the original referral was received, the date they were seen either as an inpatient or in outpatients

Information about all suspected cases is not easy to obtain, so a decision was made to start with those cases we confirmed with IBD in 2016 and 2017 (up to November 2017) to see how we performed with these cases in the first instance. We aimed to look at the cases where there was most delay to see how we can reduce this. Cases diagnosed in the private sector, or those with incomplete information, were excluded.

Results: Total patients diagnosed with IBD = 62

Incomplete information 1, diagnosed privately 4, leaving 57 patients

Time seen	Number (Percentage) of patients
As an inpatient	12 (21%)
After 28 days or less	29 (51%)
After 28-42 days	11(19%)
Over 42 days	54d, 55d, 62d, 68d, 96d (9%)

Summary: Out of 57 patients diagnosed with IBD in 2016-2017 in NHS 72% met the new quality standard, with 91% seen within 6 weeks. 5 patients waited longer. None were felt very unwell when first seen.

Conclusions:

1) In our centre we meet the suggested standard in 72% of cases, and are close to this in a further 19%
2) This was achieved by consultants doing many extra adhoc clinics. The appointment of a 4th consultant should help us to get even closer to the standard
4) The children who waited were felt not to be so unwell according to the information on the referral letter, or family cancelled appointment initially, so we will consider education to GPs and general paediatricians regarding information we would find helpful in referral letters
5) The next step would be to audit all suspected cases, but this information is not easy to obtain retrospectively

[1]https://bspghan.org.uk/sites/default/files/bspghan_rcpch_standards_for_pghan_final.pdf

P49 **Single Centre Experience of Incidental Findings in MRI-enterography (MRE) in Paediatric Patients Diagnosed with Inflammatory Bowel disease (IBD)**

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Introduction: In the recent years, MRI-enterography (MRE) has become a main tool in the diagnosis and assessment of inflammatory bowel disease (IBD). Sometimes MRE detects unexpected findings that were previously unidentified. Such incidental findings may lead to further diagnostic workup including imaging and laboratory investigations.

Aims and Objectives: Incidental findings in inflammatory bowel disease patients detected on MRE have been extensively discussed, in the adult population, but only very few papers have discussed such findings in children. The primary aim of this study was to determine the nature and frequency of incidental findings detected in children with confirmed IBD undergoing MRE for diagnostic/staging purposes in our centre. The secondary aim of the study was to evaluate the clinical impact of such incidental findings.

Subjects and Methods: All MRE performed at a single centre over a 3 year period (Jan 2014 to Dec 2016) were retrospectively analysed; only patients with a confirmed diagnosis of IBD were included. A random selection of MRE without incidental findings was reviewed by a consultant radiologist to inform that incidental findings were not missed previously (double reporting). The medical case notes of children with incidental findings were retrospectively reviewed to inform the clinical impact of such incidental findings.

Results: A total of 190 patients underwent MRE over the 3 year study period, but 102 patients with a confirmed diagnosis of IBD were only included. Incidental findings were noted in 16 patients (15.6%); two were intestinal findings (probable small bowel intussusception) and the rest were extra intestinal findings. 50% (8 patients) of the incidental findings related to renal pathology (solitary kidney, cyst, duplex kidney, dilated pelvi-calyceal system), 18.7% (3 patients) to spleen pathology (splenomegaly, cyst), 12.5% (2 patients) to prominent mesenteric lymphadenopathy and 6.2% (1 patient) to gallbladder pathology (gallbladder stones) respectively.

Summary and Conclusion: Incidental and unrelated findings were found in 15.6% of IBD patients undergoing MRE for diagnostic/staging purposes. Although many (43.7%) of these children required further imaging studies, only one patient from the entire cohort (1%) needed further active management for a significant, previously unidentified pathology.

P50 **Budd Chiari syndrome – a single centre experience**

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Introduction: Budd-Chiari Syndrome (BCS) is associated with obstruction of hepatic veins (HV) independent of the level of obstruction from HV to atrio-caval junction. BSC is more commonly seen in Asia and few reports are from western literature. The natural history is poorly defined and delay in the diagnosis can lead to poor outcome.

Aims and Objectives: To review experience in diagnosis, management and the natural history of children with BCS at Birmingham Children's Hospital.

Subjects and Methods: Retrospective review of children aged 0-18 years diagnosed with BCS between 1996 and 2016.The diagnosis of BCS was made on the basis of occlusion of 2 or more HV or occlusion of HV with supra-hepatic inferior vena cava (IVC) occlusion on USS. Data collected: mode of presentation, diagnosis, medical management, therapeutic interventions and follow-up.

Results: 25 children (18M: 7 F) were diagnosed with BCS from 1996 to 2016.The median age at presentation was 10 (range: 1-15yrs). Most prevalent presenting symptoms included were: splenomegaly 22 (88%); abdominal distension 21 (84 %); hepatomegaly 20 (80%); refractory ascites 12 (48%). Supra-hepatic IVC thrombus was seen in 4 (16%) patients in conjunction with one hepatic vein while 21 (84%) had obstruction in two or more of the hepatic veins. 11 patients had involvement of all three hepatic veins and in 10 patients two hepatic veins were obstructed. Liver biopsy had been done in 22 patients revealing histological findings consistent with BCS. The most common diagnosis was a protein C and S deficiency in 12 (48%) patients, while myeloproliferative disorder was present in 6 (25%) patients (Jak-2 mutation identified in 1/6). All patients received anticoagulation initially but anticoagulation alone was successful in 6 patients. Three needed thrombolytic therapy and 16 had radiological intervention; balloon angioplasty and stent placement in 6 (24%) and TIPS in 10 (40%) patients. Median time between start of anticoagulation and radiological intervention was 26 days (range 15-183 days). Three patients had balloon angioplasty initially which was not successful and needed TIPPS later. Two (8%) patients had liver transplantation of which one patient had TIPPS which was not successful. None of the patient required a surgical shunt. One patient died within 4 days of presentation of acute BCS. All patients were maintained on long-term anticoagulation. In one patient anticoagulation was withdrawn and BCS recurred, which was treated successfully with TIPPS and maintained patency on HV on long term follow-up. Twenty four patients are still alive with a median follow-up 13.8 years (range 2.1-23.5) with 22 patients being transitioned to adult care while 2 still remain under FU at our hospital.

Summary: BCS is rare, but does present in the western population. Our patients differ from the reports of the Asian series: BCS may present at any age from infancy to child hood (in Asian studies most children present under 2 years of age) and the main site of obstruction was hepatic veins (in Asian studies, supra hepatic IVC is the common site of obstruction). Anticoagulation as the main stay of treatment works alone only initially but later on it needs to be augmented primarily by radiological intervention.

Conclusion: BCS may present at any age from infancy to childhood and has a good long-term outcome. Anticoagulation treatment and radiological interventions (Balloon angioplasty, stents, TIPS) can avoid the need for surgical shunts and liver transplantation.

P51 **The development of liver disease in childhood acquired hepatitis C infection in the UK**

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Introduction: The long term outcome of childhood HCV is unclear, but most infected children develop chronic HCV with a lifetime risk of significant liver disease.

Aims and Objectives: To describe the development of liver disease and the effect of treatment in patients infected with HCV in childhood in UK.

Subjects and Methods: Retrospective review of patients infected with HCV in childhood. Data was collected from a national UK clinical database (HCV Research UK) covering 51 adult and 7 paediatric centres. Patients registered in the database who acquired HCV infection between 0-18 years were included.

Results: 1049 patients, 756 (72%) males, were included. The most prevalent route of infection was IV drug use: 560 (53%), blood products: 251 (24%); perinatal exposure: 119 (11%) and other routes of infection: 119 (11%). Overall, 334 (32%) had a diagnosis of cirrhosis, 55 (5%) hepatocellular carcinoma (HCC); and liver transplantation (LTx) performed in 46 (5%). Median time to the development of cirrhosis was 33yrs (range 12-53yrs). There was no significant difference in time to the development of cirrhosis between any of the risk groups (IV drug group: 33yrs; blood group: 32yrs; perinatal group 36yrs; other infection route group: 36yrs), (p=0.76). 663 (63%) had been treated (413 interferon based; 250, direct acting antivirals). Disease progression in 370 patients with SVR according to diagnosis of cirrhosis is presented in table 1.

Summary: 33% had cirrhosis independent of infection route; 5% had HCC; 5% required LTx. More patients who were treated after diagnosis of cirrhosis compared to those patients treated pre cirrhosis developed HCC, required LTx, or died.

Conclusion: HCV infection in young people causes significant liver disease which can now be prevented with antiviral therapy. Early treatment, especially before development of cirrhosis in young people (<18 years), significantly decreases morbidity and mortality associated with HCV infection.

Table 1.

Disease progression	Pre cirrhosis	Cirrhosis	
With PEG/INF+RBV	n=194	n=48	p
Cirrhosis	10 (5%)	NA	-
HCC	1 (0.4%)	6 (2%)	<0.001
LTx	0	9 (19%)	<0.001
Death	0	2 (4%)	0.04
With DAA's	n=65	n=63	p
Cirrhosis	12 (18%)	NA	-
HCC	0	11 (17%)	<0.001
LTx	1 (2%)	7 (11%)	0.03
Death	0	3 (5%)	0.01

P52 **Aetiology and outcome of Acute liver failure in children in the United Arab Emirates (UAE)**

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Introduction: Paediatric Acute liver failure(ALF) is characterized by severely impaired liver function, with or without encephalopathy in children without previous liver disease. Geographic location affects aetiology, with Non–A-E hepatitis and drugs accounting for the majority of cases in the west.

Aims and Objectives: We aim to review the aetiology, presentation and outcome of ALF in children in the UAE.

Subjects and Methods: This is a retrospective, single centre study of children presenting with ALF from birth to 16 years over a 7-year period (September 2010-2017).
We used the Paediatric ALF Study Group criteria for defining ALF: 1) absence of a previously known history of chronic liver disease, 2) biochemical evidence of acute liver injury, and 3) hepatic-based coagulopathy defined as PT≥15 s or INR≥1.5 not corrected by vitamin K in the presence of clinical HE or PT≥20 s or INR≥2 regardless of the presence or absence of clinical HE

Results: 81 patients were identified (48 males and 33 females). Median age at presentation was 18 months (range 2 days-16 years). 12% presented in the first 4 weeks of life and 88% between 1-16 years of age.
The aetiology was identified in 86% and included; 49% infection, 16% metabolic (The main cause of metabolic disease was Wolcott Rallison syndrome, seen in 46%), 15% acute circulatory failure, 14% indeterminate, 4% toxic and drugs, 1% infiltrative disease and 1% autoimmune hepatitis.

Jaundice was seen in 42% at presentation (Median bilirubin 43 (range 2.2-600)) and didn't favour any aetiology. Encephalopathy was more significantly seen in the metabolic disease (77%, P=0.013). Renal failure was seen more significantly in acute circulatory failure (83%, P=0.008).

INR was highest in toxic group (Median 6.5 (range 2.7-7.2)) and infiltrative disease (INR >10). Highest AST was in Indeterminate (Median 1059 (range 116-1435)) and Metabolic groups (Median 947 (range 38-9515)). Ammonia levels were highest in urea cycle defect (Median 455 (range 264-646)).

Overall survival was 57% with improved survival in patients presenting after 1 month of age (68%). Only three patients were transplanted and they all survived. Metabolic, toxic and autoimmune disease had most favourable outcome with 60%, 100% and 100% survival respectively.

Summary and Conclusion: ALF in the UAE has unique aetiology; with increased number of infections and reduced incidence of autoimmune hepatitis. The main metabolic disorder contributing to liver failure was Wolcott Rallison syndrome (a syndrome commonly seen in the Arab world and seen in children of consanguineous marriage). The presenting features and biochemical tests alluded to different aetiologies and can help target investigations. The Mortality rate was high in our group, we feel this can be secondary to the different aetiology spectrum in our group in addition to lack of availability of liver transplantation in the UAE, and the need for children with ALF to travel for transplantation.

P53 **Congenital porto-systemic shunts – who is at risk of developing hepatic tumours?**

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Introduction: Hepatic tumour formation is a recognised complication of congenital porto-systemic shunts (CPS).

Aims and Objectives: In patients with CPS we aimed to compare the subgroup of patients with hepatic tumours to those who did not form tumours.

Subjects and Methods: Single-centre retrospective cohort study of all patients with CPS referred from 1990 to 2016. Data are quoted as median (IQR). Categorical data were compared using a two-tailed Chi-squared test, and continuous data using a two tailed Mann-Whitney test. A P value of 0.05 was considered significant.

Results: 46 patients were investigated for CPS at a median age of 8 months (IQR 1 month – 14 years). In the tumour group 11 where type 1 and 10 were type 2, whereas in the non-tumour group there were 1 type 1 and 24 type 2 shunts. The group with tumours presented at a significantly higher age compared to those without (P <0.001). There was a significantly higher proportion of patients with tumours in the group who did not have identifiable intrahepatic portal veins on imaging (P <0.001) Congenital cardiac anomalies were common overall, and found in 17 patients (8 of those with tumours and 11 of those without). A total of 8 patients had associated cutaneous haemangiomata (3 in the tumour and 5 in the non-tumour group). 3 patients developed pulmonary hypertension (1 of which had a tumour), and 4 developed hepatopulmonary syndrome (2 of which had a tumour). There were no significant differences in the ammonia levels between the two groups. Of the 14 intrahepatic shunts, 8 closed spontaneously by 2 year of life.

Summary and conclusion: Lack of intrahepatic portal veins on imaging, and presenting later in life are associated with the development of liver tumours in patients with CPS. Associated respiratory complications are not related to the formation of liver tumours

	Tumour (n=21)	**No Tumour (n=25)**	**P value**
Age at presentation IQR (years)	12 0.7-17	0.1 0-2.3	<0.001
Sex (M:F)	13 : 8	14 : 11	0.77
Perinatal presentation	5 (24%)	16 (64%)	0.009
Absent hepatic portal veins on radiology	11 (53%)	1 (4%)	<0.001
Operative closure Age at operation IQR	14 (67%) 8.4 5.1-14.1	13 (52%) 3.1 2.9	0.38 0.53

P54 **Long term motor developmental outcomes of children following intestinal transplantation**

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Introduction: Medical and surgical advances have contributed to improving long-term survival following intestinal transplantation (ITx). Effects on developmental progress are not well documented in children. In our centre, children assessed for ITx undergo a standardised age appropriate developmental assessment pre-transplant. Children are formally reassessed post-transplant in order to direct appropriate therapy intervention and optimise outcomes.

Aim: To report on the long term motor developmental outcomes of children following ITx.

Methods: A retrospective review of physiotherapy records and Liver unit database was performed to identify children with ITx who had motor developmental assessment. Children who had pre and post-transplant assessment (> 9 months post ITx) were included in the study. Developmental progress pre and post-transplant was measured using the Bayley Scales of Infant Development (BSID II and III) and/or the Movement Assessment Battery for Children (MABC I and II). Children were classified into normal, mild delay and significant delay according to the scores for the respective tests. Excluded from the study were children: in the initial years of the programme who were too unwell to have a developmental assessment; whose physiotherapy records were not traceable; who had pre but no post-transplant assessment; or those who had only one post-transplant assessment.

Results: 31 children were initially in the study. 23/31 children had pre-transplant and post-transplant developmental assessment. 8 children had only post-transplant assessment, of which two children had only one post-transplant assessment and were excluded from the study.

12/23 had significant delay in motor skills, 6/23 had mild delay and 5/23 were normal at pre-transplant developmental assessment. In 6/12 with significant delay, there was improvement in further follow-up transplant assessment. In 11/23 of children with mild delay and normal development, 8 remained stable and showed an improvement, while there was a decline in motor development in 3 children (2 children with mild delay and one child with normal development).

Discussion: In the majority of children, an improvement was seen after 12 months and 60% enjoyed motor development scores comparable to the peers. The 3 children who had delayed motor development had different tests in pre (BSID) and post (M-ABC) transplant assessment. Hence it was difficult to interpret if a low score was an actual decline or a misrepresentation of scores in different tests. Children who had a delayed motor development had additional physiotherapy input to encourage and facilitate developmental progress and achieve optimal physical outcomes.

Conclusion: A majority of children following intestinal transplantation improve in their motor development. On-going physiotherapy input is an important part of the management of post Intestinal transplant children in their long-term follow-up.

***P55* Natural History of Paediatric Intestinal Failure: A Systematic Review and Meta-Analysis**

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Introduction: Intestinal failure (IF) is a major cause of morbidity and mortality in children, and its prevalence is increasing due to improved neonatal care. To date, cohort studies have primarily been from single centres and of small size.

Aims and Objectives: to describe the natural history of clinical outcomes in childhood-onset IF

Subjects and Methods: A MEDLINE search was performed using terms including: 'intestinal failure', 'paediatric', 'parenteral nutrition (PN)', 'liver disease', and 'IFALD'. The inclusion criteria were: IF before 18 years, reporting of clinical outcomes (e.g. liver failure, death, transplant), and >12 months follow-up. IF was defined as PN dependence for at least two months, or according to the authors' own definition. Studies were grouped by patient cohort: IF; IFALD; previous bowel lengthening procedure; and evaluated for intestinal transplant. Risk of bias was assessed using the ROBINS-I tool.

Results: 1584 articles were screened and 184 were included with a total of 9806 patients, sub-divided into 7371 with IF, 852 with IFALD, 681 with IF and bowel lengthening, and 902 being evaluated for intestinal transplant (ITx). Mortality was highest in the IFALD group at 0.23 (95% CI 0.15-0.31) at 28.7 months, and similar for the other 3 groups: 0.14-0.16 (95% CI 0.13-0.20) at 47-59 months. Liver-related mortality was more common in those with IFALD (50% of deaths) and those evaluated for small bowel transplant (56% of deaths), compared to 36% of deaths in the IF group. Sepsis was the second leading cause of mortality in all groups (25.0-34.6% of deaths). Patients evaluated for ITx were the most likely to develop IFALD (54.1%) and liver failure (17.9%) compared to the other non-IFALD groups (32.4-46.1% for IFALD and 7.5-11.3% for liver failure). The IFALD group had the overall highest rate of liver failure (44.2%) compared to 7.5% in IF ($p<0.0001$).

The proportion of patients reaching enteral autonomy was significantly lower in patients evaluated for ITx (21.1%), but was similar between the other 3 groups (54.3%-57.8%, $p<0.0001$ compared to IF group, $p=0.0006$ compared to bowel lengthening group, and $p=0.0042$ compared to IFALD group). Mean septic episodes per patient were higher in the evaluated for ITx group compared to the IF group (4.94 vs 2.23). Patients with IFALD were born at a younger gestational age (32.4 vs. 33.6 weeks) and with lower birth weight (1725g vs 1948g) than those with IF. Small bowel length and presence of ileocaecal valve (ICV) were not associated with IFALD. Patients evaluated for ITx had a shorter small bowel length (34.0cm vs 46.4cm) and decreased presence of ICV (38.3% vs 49.6%) compared to patients with IF.

Summary and Conclusion: The prognosis for patients with paediatric IF is strongly influenced and worsened by the development of IFALD. The outcome for children undergoing bowel lengthening procedures is similar to other children with IF. These data provide a clearer description of the natural history of IF which is essential for parental counselling and the design of treatment protocols.

P56 **Absence of oesophageal varices in children with splenomegaly and advanced intestinal failure associated liver disease**

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Introduction: Children with splenomegaly and portal hypertension due to primary liver disease demonstrate presence of gastro-oesophageal varices (GOV) on oesophago-gastroduodenoscopy (OGD). Intestinal failure-associated liver disease (IFALD) is a secondary liver disease caused by continuous use of parenteral nutrition (PN). IFALD leads to hepatobiliary dysfunction which may progress to biliary cirrhosis, portal hypertension and end stage liver disease. Children on PN for intestinal failure who develop end stage liver disease have a 100% mortality rate after 5 years without a combined liver and small bowel transplantation (SBTx).

Aims and Objectives: To evaluate the current methods for identification of liver disease in children with IFALD referred for SBTx assessment.

Subjects and Methods: Retrospective chart review of children with IFALD, who underwent assessment for SBTx between Sept. 2004 - Dec. 2016. IFALD was defined as: ≥one episode of a total Bilirubin persistent over 100 mmol/L for >4 weeks (not caused by a line infection), combined with features of portal hypertension such as splenomegaly or thrombocytopenia. Children undergoing SBTx assessment have a standard program over 2 weeks including: liver function tests, abdominal ultrasound, OGD, liver biopsy if no GOV or grade 1 varices. Spleen size, liver biopsy and presence of GOV were recorded.

Results: 110 children, 65 (59%) males, were included. Median age at time of SBTx assessment was 1 year (range 0.4-20 years) with a background of: 70 (64%) short bowel syndrome; 32 (29%) dysmotility disorders; 8 (7%) primary mucosal disorder. Abdominal ultrasound was performed in all 110 children: 94/110 (85%) splenomegaly; 51/110 (46%) hepatomegaly; 13/110 (12%) ascites. In 3 patients the spleen was not measurable due to splenectomy or haemorrhagic ascites. OGD was performed in 62 (56%) children revealing varices in 12 (19%): 11/62 (18%) grade I; 1/62 (2%) grade III. Of the 62 patients, 8 (13%) had a normal size spleen and no varices on OGE. There was no difference in prevalence of varices in patients with short bowel syndrome 12 (17%) and full length bowel 1 (3%), p=0.8. Liver biopsy was done in 66 (55%) showing: 5 (8%) no fibrosis; 27 (41%) mild/moderate fibrosis; 34 (52%) moderate/severe fibrosis. OGD and liver biopsy was performed on the same day in 44/66 (67%) patients. There was no difference in prevalence of varices in patients with mild fibrosis on liver biopsy compared to patients showing severe fibrosis (5 [19%] vs 3 [13%]; p=0.7). We found a significant association between degree of fibrosis and bilirubin level (95% CI 0.002-0.009, p=0.003) but no association between platelets (95% CI -0.003-0.003, p=0.9) or prothrombin time (95% CI -0.04-0.17, p=0.2) and degree of fibrosis.

Summary: In children with IFALD, splenomegaly is common and GOV are infrequent. Children with IFALD and a normal size spleen did not have varices at OGD. The degree of liver fibrosis does not correlate with presence of GOV.

Conclusion: In children with IFALD, splenomegaly is common and GOV are infrequent. Children with IFALD and a normal size spleen did not have varices at OGD. The degree of liver fibrosis does not correlate with presence of GOV. Therefore, conventional methods for determining portal hypertension (splenomegaly, gastroesophageal varices) are not useful in assessing the severity of intestinal failure associated liver disease.

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[i] NHS England Patient Safety Domain, Revised Never Events Policy and Framework, 27 March 2015.

[ii] Care Quality Commission, Briefing: Learning from serious incidents in NHS acute hospitals, a review of the quality of investigation reports, June 2016.

[iii] NHS Improvement, Provisional publication of Never Events reported as occurring between 1 April 2016 and 31 March 2017, Published 16 June 2017

[iv] NHS England http://www.nhs.uk/NHSEngland/thenhs/about/Pages/overview.aspx**IP%&**

P57 Obesity is an additional risk factor contributing to hepatotoxicity in children on long-term methotrexate

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Introduction: Methotrexate (MTX) is a commonly prescribed systemic therapy for childhood rheumatological conditions including Juvenile Idiopathic arthritis (JIA) and psoriasis. MTX is associated with serious adverse reactions such as myelosuppression, interstitial pneumonitis and hepatoxicity (abnormal liver function tests)
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Aims and Objectives: To evaluate risk factors & liver biopsy changes in hepatotoxicity secondary to long-term MTX in rheumatological conditions

Subjects and Methods: A retrospective review of Liver Unit database to identify children referred for suspected MTX hepatotoxicity (transaminases 1.5 -2 times above normal limits) between 2002-2013. Data regarding underlying diagnosis, liver function tests, and additional risk factors were documented. The liver biopsies were reviewed by a paediatric liver histopathologist who did not have access to the clinical details .The Roenigk classification of methotrexate-associated liver damage was used to evaluate different histological features (fatty change etc) and a final score was applied. The Kleiner's staging was applied for evaluation of changes secondary to NAFLD.

Results: Fifteen patients with suspected hepatotoxicity (intermittently/persistently abnormal LFT's) secondary to MTX over 10 years (2002- 2013) were enrolled. M: F ratio was 1.1:1 (M-8,F: 7). The underlying rheumatological conditions were JIA (6 children), uveitis in 3, sarcoidosis and psoriasis in 2 children each and sjogren's syndrome and systemic sclerosis in 1 each. Six children had a BMI over the 85th centile (Overweight-1, Obese- 5) at presentation. MTX dose varied between 2.5mg per week to maximum of 7.5 mg per week. Unfortunately we could not obtain the total cumulative dose of MTX for most of our patients. 5/15 children had raised (1.5 to 2 times) transaminases (all of them had a high BMI) and one child had > 5 fold increase in AST and ALT. Synthetic liver function in all 15 children was normal.12/15 underwent liver biopsy (total 13 biopsies; one patient hadt biopsy twice to monitor fibrosis). Liver biopsy was not done in 2 patients as LFT' were entirely normal at presentation and during subsequent monitoring. One child's parents did not give consent for the biopsy (although this child had abnormal LFT's). The Roenigk scoring system showed the following grades of methotrexate induced liver injury -6/13 biopsies – Grade 1, 3/13- Grade 3a, 3/13 - Grade 3b and 1/13 - Grade 4. Five children had an additional risk factor of obesity (BMI > 95th centile) – 2 did not have a biopsy and 3 children had grade 1c,Grade 1c, Grade 2 fibrosis as per the Kleiner staging, none had steatohepatitis. Children who had intermittently abnormal LFT's but normal LFT's at the time of admission had a normal biopsy, whereas children with abnormal LFT's had obesity and changes of methotrexate hepatotoxicity on liver biopsies. Children with methotrexate hepatotoxicity were advised alternative treatment and management of obesity.

Discussion: Liver biopsies can be helpful in the decision making process of continuation of MTX in children with intermittently abnormal liver function tests . Additional risk factors i.e obesity was seen as a significant factor leading to fibrosis in our cohort of patients.

Conclusion: Children on long term MTX with additional risk factors i.e. obesity increases the risk of liver injury and needs a careful evaluation including liver biopsy to assess the fibrosis/inflammation and determine if it is safe to continue therapy.

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