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ABSTRACT BOOKLET

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OC1

Real world exclusive enteral nutrition practices over time in > 500 patients: persisting use as induction for paediatric Crohn's disease with emerging combination strategy with biologics

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This study aims to examine the changing practices of exclusive enteral nutrition (EEN) in paediatric Crohn's disease (CD) including the influence of the 2021 ECCO-ESPGHAN guidelines and the COVID-19 pandemic.

We analysed a prospectively identified cohort of newly diagnosed CD patients in two paediatric regional centres between 01/01/15 and 30/06/22. Data were retrospectively collected from electronic medical records. CD patients who received EEN were divided into biannual epochs for analysis. Continuous outcome measures were analysed using Mann-Whitney U or Chi-squared tests, and linear regression modelling for longitudinal comparison.

Of 503 patients (62.2% male; median age 13.0 years, IQR: 10.9 – 14.8), primary EEN was used in 383 (76.1%) with a median course length of 8 weeks (IQR: 7.2 – 8.3). An increasing incidence of CD diagnosis and total EEN courses were observed (p=.01, Figure 1). Remission/response rates, nasogastric tube (NG) usage and completion rates were examined; there were no changes in these parameters over time (p=.153, p=.913, p=.601, p=.337 respectively). Weight z-scores increased (pre-EEN -0.11 vs post-EEN 0.33, p=<.001). An increased rate of EEN as induction therapy was observed (first 12-months 66.7% vs last 87.7% - p=.004), with dual induction (EEN combined with biologics) an emerging strategy over time (first 12-months 2.6% vs last 18.7% - p=.018). (Figure 2)

During the COVID-19 pandemic, primary EEN was less frequently used (63/96, 65.6% vs 320/407,78.6% - p=.007), completion rates were lower (41/68, 60.3% vs 236/315, 74.9% - p=.015) but remission rates were comparable (37/67, 53.7% vs 181/315, 57.3% - p=.59).

Repeat courses of EEN occurred in 47/503 (9.3%) with no difference in remission rates (2nd course 23/47, 46.7% vs 1st course 217/383, 56.7% - p=.463).

This large real-world cohort demonstrates EEN usage has increased together with CD incidence despite an increase in biologic use. The use of dual induction therapy with biologics is an emerging trend; further research is required to ascertain the clinical benefit above dose-optimised biologic induction and its cost-effectiveness.

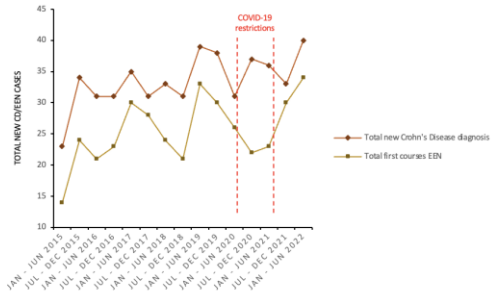

Figure 1: Line graph showing the total number of new CD diagnosis and total new first EEN courses

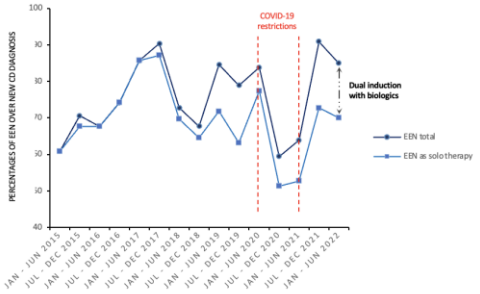

Figure 2: Line graph showing percentage of all newly diagnosed CD patients receiving EEN at induction and those receiving EEN as their sole induction therapy. The difference between the two representing those receiving dual induction with biologics.

OC2

Cancer and mortality in paediatric IBD: 20 year population based data from Scotland

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Paediatric inflammatory bowel disease (PIBD; which encompasses Crohn's disease (CD), ulcerative colitis (UC) and IBD-unclassified (IBD-U)) has an increased overall risk of mortality and malignancy compared to the general population however this is not fully characterized in children. Scotland has a population of 5.5 million people and one of the highest PIBD prevalences worldwide; we aim to present long-term nationwide PIBD cancer and mortality data.

We identified all PIBD patients within paediatric care who developed cancer or died between 01.01.2003 - 30.11.22. Accurate case accrual was ensured through review of the yearly national PIBD in Scotland audit (PISA) data and a follow-up questionnaire to each regional centre. Electronic medical records were reviewed retrospectively.

10 patients were included in the analysis. 6 patients (100% male, all CD) were diagnosed with malignancy with a median age at cancer diagnosis of 16.7 yrs (IQR 13.2 – 17.5) and a median disease duration prior to diagnosis of 3.0 yrs (IQR: 1.5 – 3.1). These included 2 cases of acute myeloid leukaemia, 3 Hodgkin's Lymphoma and 1 Non-Hodgkin's Lymphoma. No patients had a family history of cancer or primary sclerosing cholangitis. 5/6 (83%) received an immunomodulator or anti-TNF within the past 3 months and all patients had been exposed during their disease course. 4/6 (67%) are currently in remission, 1/6 (17%) is receiving treatment and 1/6 (17%) died.

5 patients died, including 1 cancer patient, (80% female, 2 UC, 2 CD, 1 IBDU) with a median age at death of 11.9 years (12.2 years (IQR: 11.7 – 18.0). 2/6 (33%) likely died as a direct consequence of their disease (1 late surgical complication and 1 thiopurine associated malignancy).

Death and malignancy are a rare but significant consequence of PIBD. While causation cannot be proved, six cases of malignancies associated with thiopurine/anti-TNF use were observed over a 20 year period.

OC3

Rising incidence of paediatric inflammatory bowel disease in South Wales, United Kingdom

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Rates of paediatric inflammatory bowel disease (pIBD) have increased globally over the last 25 years. In South Wales, incidence was last reported for 1998-2003 (5.4/100,000) (1), and subsequent defined national and global cohorts have described increases, particularly in Crohn's disease. This work aimed to characterise and update the contemporary incidence of pIBD in South Wales, highlighting implications for resource planning in paediatric and adult gastroenterology services across the region.

Data from the paediatric gastroenterology tertiary referral centre retrospective/prospective pIBD database were retrieved for 2011-2021. Data were classified by age at diagnosis, gender and type of IBD (IBDU, CD, UC). At risk population (<18 years) was determined using 2011 and 2021 UK census data (2,3). Population numbers were extrapolated between censuses. Potential differences in age at diagnosis were analysed though ANOVA. Pearson's correlation coefficients were calculated for analysis of total IBD, UC and Crohn's incidence by year.

365 patients were included (mean age at diagnosis 12.7, 37.3% female). There were no differences in age at diagnosis over the ten-year period as assessed by ANOVA ($F=1.629$, $p=0.097$). Total incidence rose from 2011 (4.28/100,000) to 2018 (6.93/100,000) and again to 2021 (11.24/100,000), $\beta=0.9$, $p=0.000157$. Male pIBD increased from 5.57/100,000 in 2011 to 14.01/100,000 in 2021 and female pIBD increased from 2.29/100,000 to 8.32/100,000.

The largest increase was seen in Crohn's disease from 2011 (2.85/100,000) to 2021 (8.29/100,000), $\beta=0.8$, $p=0.03$. Ulcerative Colitis incidence did not significantly increase, at 1.43/100,000 in 2011 and 2.76/100,000 in 2021, $\beta=0.46$, $p=0.154$.

The incidence of pIBD continues to rise across South Wales, with increases in affected males and those with Crohn's disease. This is in keeping with contemporary trends. Increases are likely driven by multifactorial environmental triggers and highlight continued need to map incidence. These ongoing increases represent significant implications for services.

Table 1 – Mean age at diagnosis and incidence rates per 100,000 by year for 2011 to 2021

Year	Mean age at diagnosis (Decimal Years)	Incidence per 100,000 - Total IBD	Incidence per 100,000 - CD	Incidence per 100,000 - UC
2011	13.2	4.28	2.85	1.43
2012	11.4	3.93	1.43	2.50
2013	13.3	3.59	1.61	1.97
2014	13.3	4.32	2.52	1.80
2015	12.8	4.15	2.71	1.08
2016	13.0	6.16	2.90	2.54
2017	13.3	5.82	2.73	2.00
2018	11.4	6.93	2.55	3.65
2019	12.3	8.42	4.76	3.48
2020	13.3	7.53	5.69	1.65
2021	12.7	11.24	8.29	2.76

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OC4

Double Trouble: Urinary and Faecal Incontinence in Children with Functional Constipation

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Introduction: Bladder dysfunction has been associated with functional chronic constipation and faecal incontinence (CCFI) in children (1-2). The pathophysiology of the coexistence of CCFI and urinary incontinence (UI) remains rudimentary. The interaction between the bladder and the anorectum in children, using diagnostic investigations such as high resolution anorectal manometry (HRAM) is unknown. This study aims to investigate the symptomatologic and physiological outcomes in children with CCFI, with UI compared to children without UI.

Methods: Patients with functional CCFI who presented to our specialised service from September 2016 to September 2022 were included. Measures included: demographics, bowel scores: St Marks Incontinence Scores (SMIC) and Cleveland Constipation Scores (CCS), HRAM parameters and colonic transit x-rays.

Results: Out of 341 patients with functional constipation, 40% (135/341) had UI. There were no significant demographic differences between those with or without UI. Patients with UI and CCFI, had significantly higher SMIC compared to patients who did not have UI ($p<0.001$) (Figure 1). Patients with UI, significantly had lower mean threshold on maximum squeeze increments ($p < 0.05$) and endurance squeeze ($p <0.05$).

Conclusion: This is the first study, to demonstrate the outcomes in children with CCFI and UI using HRAM. We have shown:

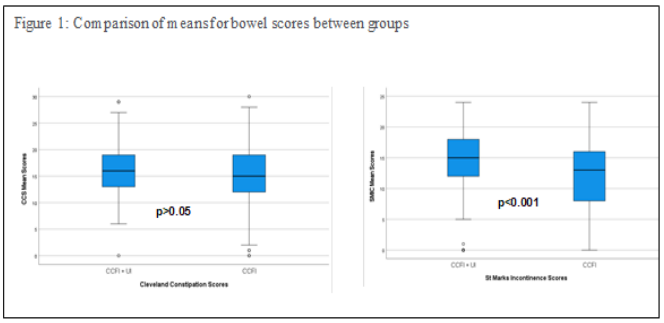
- Despite normal sphincteric function, children with both CCFI and UI, have abnormal external anal sphincter endurance.
- UI and FI are often managed in isolation with unsatisfactory outcomes.
- It is hoped that simultaneous colorectal and urological specialist input, will improve our understanding and outcomes in children with double incontinence.
- We look forward to reporting on the routine use of urological questionnaires, assessment, investigation, review and follow up.

Table 1: Patient Outcomes

MEASURES	CCFI n =200 M, ± *(range)*	CCFI with UI n = 135 M, ± *(range)*	Significance (p value)
Sex	119 males; 81 females	66 males; 69 females	NS
Age	M: 10 ± 3.96 *(1-17)*	M: 9 ± 3.44 *(1-17)*	NS
Cognitive Function	54/200 (77%)	109/135 (81%)	NS
Perception of Pain			
HRAM	M:3 ± 2.30 (0-10)	M: 3 ± 2.29 (0-10)	NS
Venipuncture	M:6 ± 3.41 (0-10)	M: 6 ± 3.74 (0-10)	NS
Perception of Severity	M:8 ± 2.09 (1-10)	M: 8 ± 1.85 (2-10)	NS
HRAM			
Anal Canal (cm)	M: 2.17 ± 0.37	M: 2.18 ± 0.35	NS
Resting pressure	M: 57.11 ± 15.62	M: 58.19 ± 12.97	NS
Squeeze increment	M: 114.40 ± 38.96	M: 107.00 ± 30.66	NS
Maximum squeeze increment	M: 148.82 ± 58.92	M: 136.87 ± 44.09	**p<0.05**
Enhanced squeeze	M: 120.61 ± 66.76	M: 105.51 ± 32.55	**p<0.05**
Rectal Sensation			
First sensation	M: 44.47 ± 39.15	M: 42.16 ± 43.88	NS
Urge to defaecate	M: 80.86 ± 54.46	M: 77.14 ± 51.86	NS
Maximum tolerable volume	M: 155.67 ± 66.95	M: 156.31 ± 63.08	NS
RAIR	97% * present	98% * present	NS
Dyssynergia	75 (54%)	53 (53%)	NS
CTS	44% Normal 39% RED 18% STC	50% Normal 33% RED 16% STC	NS NS NS
Bowel Scores			
SMIC	M: 11 ± 6.32 (0-22)	M: 13 ± 6.18 (0-24)	**p<0.001**
CCS	M: 16 ± 5.27 (1-30)	M: 16 ± 5.05 (0-29)	NS
Abdominal pain severity	M: 7 ± 2.53 (0-28)	M: 7 ± 2.37 (0-10)	NS

RAIR * present – the remaining patients we were unable to elicit RAIR due to low sphincter pressures. ± (Standard deviation); $p< 0.05$ level of significance

Figure 1: Comparison of means for bowel scores between groups



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OC5

Developing and running an online workshop for children and young people with rumination syndrome to help manage a long waiting list for psychological support

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Introduction: The Paediatric Gastroenterology Psychology Service at the Evelina London Children's Hospital, similar to most psychological services, has a long waiting list. There have been an increasing number of patients who were referred for psychological support with rumination syndrome. In order to offer a timelier intervention to these patients, and to reduce the waiting list, the Paediatric Gastroenterology Psychology Service ran four virtual workshops for young people with rumination syndrome, and their parents. Three of these workshops were for teenagers and their parents, and one for parents of young children or of young people with additional needs.

Participants and Methods: The workshops were run online for 2.5 hours. All attendees were provided with a workbook to use alongside the session which contained additional information and written exercises to consolidate their learning.

This workshop covered the following topics

- What is rumination syndrome?
- What causes rumination syndrome?
- What is the impact of rumination syndrome?
- Treating rumination syndrome
    - Diaphragmatic breathing (and strategies on how to implement this with their child- parent workshop)
    - Additional strategies and support
- Parent self-care (parent workshop)

All attendees were invited to complete brief pre and post questionnaires. Using 5-point Likert scale responses (1 = no confidence/knowledge, 5 = extremely confident/Knowledgeable), they were asked four questions exploring their understanding of rumination syndrome, knowledge of strategies to manage rumination syndrome, confidence in managing rumination syndrome and belief that rumination syndrome would improve.

Results: Overall, we had 17 attendees for the four workshops. Another 9 were invited but did not attend. Unfortunately, we only had data for two of the groups. The quantitative data was analysed using descriptive statistics (M,SD), and free text qualitative date was collated and reviewed. Results are shown below.

Table 1. Pre and post questions mean scores

		Understanding of rumination syndrome	Knowledge of management strategies	Confidence in managing rumination syndrome	Belief that rumination syndrome will improve
Young Person	Pre (n=4)	M=3.8 (SD=1.0)	M=2.8 (SD=1.0)	M=2.8 (SD=1.0)	M=3 (SD=0.8)
	Post (n=3)	M=4.7 (SD=0.6)	M=4.7 (SD=0.6)	M=3 (SD=1.8)	M=3 (SD=1)
	Change	**+0.9**	**+1.9**	**+0.2**	**No change**
Parent	Pre (n=6)	M=3.5 (SD=1.2)	M=3.3 (SD=1.9)	M=3 (SD=1.5)	M=2.7 (SD=0.8)
	Post (n=5)	M=4.6 (SD=0.6)	M=4.4 (SD=0.6)	M=4 (SD=1.2)	M=3 (SD=0.6)
	Change	**+1.1**	**+1.1**	**+1**	**+0.3**

Summary and Conclusion: Running the workshops in this way has meant that 17 patients have had psychological support and have been taken off the waiting list.

After attending the workshop, attendees report improvement in their understanding and confidence in managing rumination syndrome. There was less change in their belief that rumination syndrome would improve

The service plans to continue to run these workshops, incorporating attendees' feedback, and monitor the clinical impact of the workshops.

OC6

A 10-year single tertiary centre therapeutic Endoscopy experience for the management of Gastrointestinal strictures

by A Mallikarjuna and D Basude. *Department of Paediatric Gastroenterology, Bristol Royal Hospital for Children, Bristol, BS2 8BJ, UK.*

The joint European Society of Gastrointestinal Endoscopy and European Society of Paediatric Gastroenterology Hepatology and Nutrition guidelines on therapeutic indications of endoscopy recommend the use of esophagogastroduodenoscopy in the dilatation of oesophagus and upper gastrointestinal strictures and the use of ileocolonoscopy for dilatation of ileocolonic and colonic stenosis[1].

We conducted a retrospective observational study of gastrointestinal dilatations performed by Paediatric Gastroenterologists over a 10yr period (1 January 2012 to 31 December 2022) in our centre. Prospective records of procedures were used to identify the patients and the electronic health records were reviewed.

163 endoscopic dilatations were performed in 52 patients during this period. The procedures were either performed or supervised by Gastroenterologist with an interest in therapeutic endoscopy. The age group of the patients ranged between 3 months to 17yrs with a mean of 12.3 years. Anatomical sites dilated were as follows: 92 oesophageal, 7 pyloric/antral, 16 duodenal, 9 ileal, 2 colonic, 1 ileostomy, 3 rectal stumps and 32 rectal.

Hagar dilators with or without balloon dilatation were used in 15 of 32 rectal strictures. Through the scope balloon dilatation was used for all other strictures 130 of 163. In 40 procedures there were additional interventions performed alongside dilatation which included 2 endoknife cut, 1 argon laser and hemostatic clip application, 26 injection of Triamcinolone acetonide, 9 Mitomycin c spray and 4 injection of botulinum toxin.

Underlying aetiologies were recorded as follows: 34 congenital stricture/web, 46 Crohn's disease, 11 anastomotic stricture, 1 radiotherapy-induced stricture, 41 strictures secondary to gastroesophageal reflux disease, and unknown aetiology in 7. Inflamed mucosa at the stricture site was noted in 63 procedures. 158 of these procedures were performed by the Gastroenterologist independently and 5 procedures were done jointly with a Paediatric Surgeon.

Children were discharged the same day for 124 procedures. The rest were done during an inpatient stay. Four procedures were associated with serious complications. Two had perforation requiring surgery and intensive care admission; both children were 2 yrs or younger. One patient became unwell with the intraabdominal collection but no perforation. One patient required 24hrs of observations for persistent rectal bleeding but did not require further intervention. One patient was readmitted within 8 days of discharge.

In conclusion, endoscopic dilatation when performed by a gastroenterologist with therapeutic interest appears to be a safe alternative to surgical management of strictures. The risk of complications is small and the majority can be performed as a day case. Procedures in children under 2yrs of age carry the most risk of bowel perforation.

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OC7

Maralixibat-treated patients with Alagille syndrome demonstrate event-free survival in a natural history comparison with patients with from the GALA database: Application of real-world evidence analytics

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Real-world evidence (RWE) analytics continue to advance natural history comparisons in rare diseases. The Global Alagille Alliance (GALA) is the largest global clinical research database for Alagille syndrome (ALGS). Maralixibat (MRX) is an ileal bile acid transporter inhibitor approved by the FDA for the treatment of cholestatic pruritus in patients with ALGS ≥1 year of age. A pre-specified analysis plan applied novel analytical techniques to compare RWE from GALA with a MRX cohort with the aim to compare event-free survival (EFS) in patients with ALGS.

GALA contains retrospective data for clinical parameters, biochemistries and outcomes. The MRX database comprises of 84 ALGS patients with up to 6 years of data. EFS was defined as the time to first event of hepatic decompensation (variceal bleeding, ascites requiring therapy), surgical biliary diversion, liver transplantation (LT), or death. GALA was filtered to align key MRX eligibility criteria. The index time was determined via maximum likelihood estimation. Balance among baseline (BL) variables was assessed. Selection of patients and index time was blinded to clinical outcomes. Sensitivity and subgroup analyses, and adjustments for covariates, were applied. Missing outcomes data were censored at last contact.

Of 1,438 patients in GALA, 469 were eligible. Age, total bilirubin (TB), gamma glutamyltransferase (GGT) and alanine aminotransferase (ALT) were well balanced between groups and no statistical differences were observed for age, mutation, region, TB, GGT and ALT. Median BL serum bile acids (sBA) was significantly higher in the MRX cohort (p=0.003); 85% of sBA data was not available in GALA. EFS rates in the MRX cohort were significantly better than those reported in the GALA control, (crude 6-year EFS: 73% and 50%, respectively, and adjusted for age, sex, TB, ALT: HR=0.305; 95% CI: 0.189–0.491; p<0.0001). Varied index times, weighted inverse probability of treatment weights, average treatment effect in the treated, LT and death only, regions, sBA sub-group, pruning events to 12 months were consistent with the primary result. Limitations include no standardised measure of pruritus and limited sBA data in GALA, and inherent bias for patients who enter a clinical trial.

This 6-year analysis suggests the potential for improved EFS with MRX in patients with ALGS. This RWE analysis provides a potential method to evaluate outcomes in long-term intervention studies where placebo comparisons are not feasible. Limitations will always be present given lack of prospective conduct and inherent biases, though sensitivity analyses can help mitigate and aid interpretation.

OC8

Long-term efficacy and safety of odevixibat in patients with progressive familial intrahepatic cholestasis: results with ≥96 weeks of treatment

by R.J. Thompson[1], Ö. Durmaz[2], T. Grammatikopoulos[1,3], A. Di Giorgio[4], Q. Ni[5], P. Stein[5], C. Clemson[5], and E. Sturm[6], *[1]Institute of Liver Studies, King's College London, London, UK SE5 9RS; [2]Istanbul University, Istanbul Faculty of Medicine, Istanbul, Turkey; [3]Paediatric Liver, GI, and Nutrition Centre and MowatLabs, King's College Hospital NHS Trust, London, UK SE5 9RS; [4]Paediatric Hepatology, Gastroenterology, and Transplantation, ASST Papa Giovanni XXIII, Bergamo, Italy; [5]Albireo Pharma, Inc., Boston, MA, USA; [6]Paediatric Gastroenterology and Hepatology, University Children's Hospital Tübingen, Tübingen, Germany.*

Patients with progressive familial intrahepatic cholestasis (PFIC) may present with elevated serum bile acids (sBAs), growth deficits, and impaired hepatic function. The phase 3 PEDFIC 1 and PEDFIC 2 studies evaluated odevixibat, an ileal bile acid transporter inhibitor, in patients with PFIC. Using pooled data from these studies, we describe key outcomes in patients treated with odevixibat for ≥96 weeks.

PEDFIC 1 (NCT03566238) was a 24-week, randomised, placebo-controlled study in children with PFIC1 and PFIC2. PEDFIC 2 (NCT03659916) is an ongoing, 72-week open-label extension study in patients with any PFIC type; an optional extension period follows PEDFIC 2. This pooled analysis spans from patients' first dose of odevixibat to 31 January 2022. Outcomes included sBAs, hepatic parameters, growth, and safety.

Of the 111 patients in the pooled population (69 ongoing at data cut-off), 36 had ≥96 weeks' odevixibat exposure and an sBA measurement at week 96 (36% PFIC1, 61% PFIC2, 3% MYO5B deficiency). At baseline, patients had elevated mean sBA, transaminase, and total bilirubin levels, and impaired growth (**Table**). At week 96, mean sBA levels were significantly reduced (P<0.001); no changes were observed in bilirubin levels (**Table**). All 36 patients (100%) had treatment-emergent adverse events (TEAEs); most were mild or moderate in severity. No drug-related serious TEAEs were reported.

Odevixibat treatment for ≥96 weeks was associated with improvements in sBAs, transaminase levels, and growth in patients with PFIC. Odevixibat was generally well tolerated.

Table. Outcomes in Patients With PFIC Treated With Odevixibat for ≥96 Weeks

	Baseline		Week 96[a]	
	n	**Mean (SE)**	**n**	**Mean (SE)**
sBAs, µmol/L	36	266 (22)	36	118 (20)[b]
ALT, U/L	36	102 (23)	32	38 (7)
AST, U/L	36	86 (10)	32	49 (5)
Total bilirubin, µmol/L	36	56 (13)	32	50 (17)
Height Z score	36	–1.5 (0.3)	36	–0.9 (0.2)
Weight Z score	36	–0.9 (0.3)	36	–0.2 (0.2)

[a]Visit window spans weeks 94–104; [b]Mean change in sBAs from baseline to week 96, P<0.001.
Inferential statistics were not calculated for hepatic parameters or growth.
ALT, alanine aminotransferase; AST, aspartate aminotransferase; PFIC, progressive familial intrahepatic cholestasis; sBA, serum bile acid.

Disclosures:

R. J. Thompson: Albireo and Mirum – Consultant; Generation Bio – Consultant and stock options; Rectify Therapeutics – Consultant and stockholder
Ö. Durmaz: Nothing to disclose
T. Grammatikopoulos: Albireo – Consultant
A. Di Giorgio: Albireo – Travel support and speaker's fee
Q. Ni, P. Stein, and **C. Clemson**: Albireo – Employment
E. Sturm: Albireo and Mirum – Consultant and research support; Univar – Consultant; Orphalan – Speaker's fee

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OC9

30 year experience of children with intestinal failure associated liver disease - A tale of changing 'color' and persisting stiffness

by J. Valamparampil[1], R. M. Brown[2], J. Rajanayagam[3], J. Hartley[1] and G. L. Gupte[1]. *[1]Liver Unit (including small bowel transplantation), Birmingham Women's and Children's NHS Foundation Trust, Birmingham, B4 6NH, UK.*
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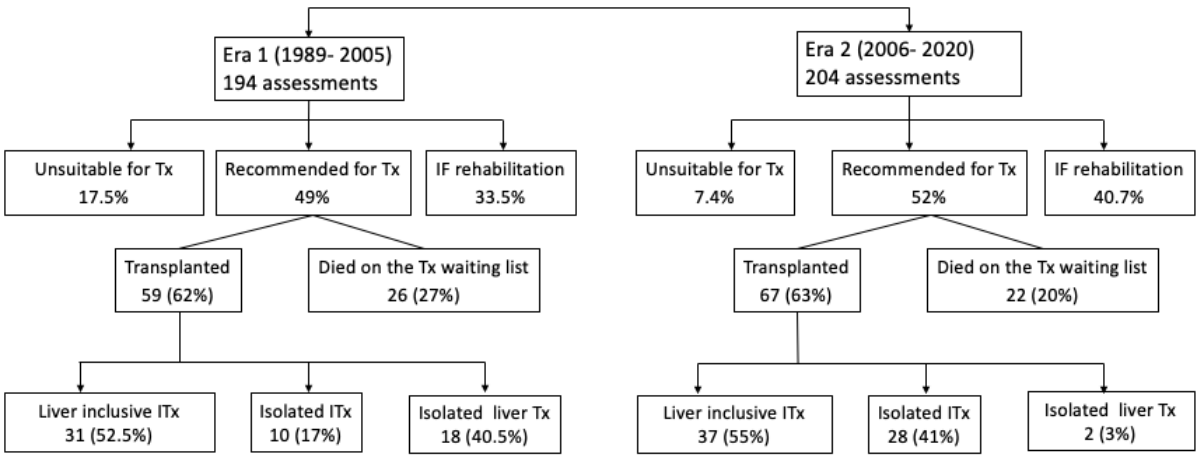
Objectives and Study: Children developing intestinal-failure-associated liver disease (IFALD) presented with jaundice historically, but with recent advances in IF rehabilitation, children are no longer jaundiced, yet can have advanced IFALD as per literature reports. Hence, assessment of severity, management of IFALD with a timely referral for intestinal transplantation (ITx) can be challenging. Aim was to describe 30-year experience on referral patterns, assessment & overall outcomes of IFALD.

Methods: Retrospective analysis of IFALD assessments at a tertiary IF-center between 1989-2020 and grouped into 2 eras- Era-1 (1989-2005) & Era-2 (2006-2020). Demographics, severity of IFALD assessment with/without portal hypertension (PHT), outcomes after disease assessment (Transplantation (Tx)/Unsuitable for Tx/Intestinal rehabilitation) were recorded.

Results and Discussion: 398 (194-Era-1, 204 Era-2) assessments were performed over 32years. The outcomes of assessment are detailed in Fig-1. In era-2, children were older (29mths vs. 9mths) had better nutritional status (weight z-score, -0.75 vs. -1.6), showed a significant decline in median bilirubin (18µmol/L vs 290µmol/L) and had less thrombocytopenia (17% vs 43%). But, moderate-severe fibrosis was still present in 57% of children with IFALD. The presence of moderate-severe fibrosis in non-jaundiced children with no thrombocytopenia led us to introduce hepatic venous wedge pressure gradient (HVWPG) measurement to assess presence/absence of PHT. 43 HVWPG measurements were performed in 33 patients (median age- 42 months, IQR- 16- 93). Bilirubin level, platelet count, splenomegaly and presence of oesophageal varices did not correlate with the severity of HVWPG. Only 11 children had elevated HVWPG. Applying HVWPG measurements, 16 children were continued with intestinal rehabilitation who would otherwise have been recommended for ITx. In era 2, children precluding ITx declined significantly (7.4% vs. 17.5%) suggesting early referral, children not needing transplantation increased (40.7% vs 33.5%) while similar proportion of children with IFALD were recommended for Tx (52% vs 49% - $p<0.05$).

Conclusions: In the modern era, severe IFALD may exist in children without significant jaundice or conventional markers of PHT. HVWPG measurements can guide in the decision making process in children with IFALD for ITx. Appropriate timing of referral /dialogue with a transplant center to assess severity of IFALD is crucial to ensure correct treatment option for improving long-term survival.

Figure 1. Outcomes of assessment and transplantation in children with IFALD



OC10

Identifying paediatric non-alcoholic fatty liver disease (NAFLD) in weight management services

by A. Whiting, S. Moynihan and J. Chan. *St David's Children's Centre, St David's Hospital, Cowbridge Road, Cardiff CF11 9XB*

12.6% of children in Wales were classified as obese in 2018/19 (1). Guidelines advocate for the routine assessment of comorbidity in children with a body mass index (BMI)≥98th centile (2). Currently available guidelines for comorbidity assessment and subsequent investigation and diagnosis of NAFLD for a population at scale have implications for clinical diagnostics, paediatric phlebotomy and ultrasonography resource.

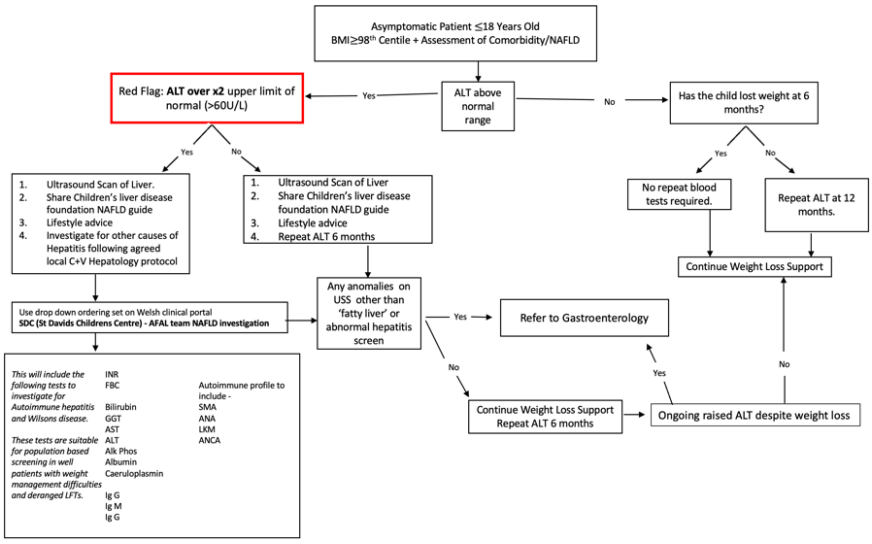
We aim to propose a pathway for implementation within our weight management service, with the intention of initiating future work to devise an all-Wales paediatric NAFLD investigation pathway for children >95th centile BMI.

61 asymptomatic children aged between 3-17 years with a BMI≥98th centile presenting to the tier 3 weight management service between June 2021 and March 2022 were included. The service routinely offers an assessment of comorbidity at referral (3). Retrospective data was obtained from the electronic clinical records of the participants. All children and their families are supported by the service to implement lifestyle modifications.

46% of patients were found to have an elevated (>28U/L) alanine transaminase (ALT), which is suggestive of NAFLD. Of those entering the pathway, 10/61 (17%) had raised ALT>60U/L suggesting screening for other causes of liver disease. Of 11 children with raised ALT who went on to receive a liver ultrasound scan, hepatic steatosis was confirmed in 100% of the radiology reports. At 6 months, complete follow-up data was available for 8/29 patients with raised ALT, of which 100% had achieved a reduction in their BMI. Upon retesting, ALT normalised in 50% (4/8) of these patients and no patient exhibited a rise in their ALT.

The scale of population of asymptomatic children requiring assessment for NAFLD has emerged from this data. There appears a need for urgent revision of guidelines to produce a safe and sustainable pathway. We propose the following pathway (figure 1) which aims to detect those in whom alternative underlying diagnoses might exist.

Figure 1



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OC11

Systemic inflammation in paediatric chronic liver disease

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Systemic inflammation (SI) in chronic liver disease (CLD) has a role in the development of sarcopenia(1, 2) and neuroinflammation(2, 3). The objective of this study is to characterise 10 cytokines in a cohort of 100 children with CLD and 25 disease controls and to evaluate these against basic anthropometry and laboratory results. The aim is to identify the clinical characteristics of the children with SI to inform recruitment in future studies. Here we present the interim data of TNFα and IL17, cytokines that synergistically mediate SI, from children recruited so far.

Single site prospective analysis of biobank samples from paediatric patients with and without CLD. Cytokines measured by ELISA. IBM SPSS Statistics 28 was used for independent t-test and Pearson's correlation(r).

40 children currently recruited (23M:17F). Age 4 months to 15.6 years (median 8.2 years). 27 had CLD and 13 were disease controls.

TNFα and IL17 were detected at 15.6pg/ml and above for 30 and 26 children respectively. 13 children with TNFα >50pg/ml had a higher γGT (160 vs 46 IU/L)*, ALP (470 vs 309 IU/L)*, bilirubin (96 vs 21umol/L)* and IL17 (65 vs 19pg/ml)**.

3 children with IL17 >150pg/ml had higher bilirubin (221 vs 49 IU/L)*, INR (1.6 vs 1.0)* and lower albumin (38 vs 46g/L)**. They also had TNFα >150pg/ml. IL17 levels had a negative correlation with haemoglobin (r -0.38)* and albumin (r -0.52)* and positive with INR (r 0.39)*, bilirubin (r 0.41)* and TNFα (r 0.5)**.

Children with bilirubin >300umol/L, INR >1.4, albumin <35g/L and weight z-score < -1.5 had higher IL17 levels (148 vs 38pg/ml*, 108 vs 34pg/ml*, 176 vs 36pg/ml*, 88 vs 34pg/ml* respectively).

Overall, raised TNFα associated with higher biliary enzymes and raised IL17 associated with impaired synthetic function. Children with more advanced liver disease had higher IL17 levels.

*p<0.05 **p<0.01

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OC12

Transcriptomic analysis of biliary atresia finds ongoing hepatic hematopoiesis with elevated IGF2

by J. P. Mann[1], P. N. Newsome[2], Y. H. Oo[2] and G.L. Gupte[1]. *[1]Liver Unit (including small bowel transplantation), Birmingham Women's and Children's NHS Foundation Trust, Birmingham, United Kingdom, B4 6NH, [2]Centre for Liver Research, University of Birmingham, Birmingham, United Kingdom, B15 2TT.*

Recent single-cell transcriptomic data has implicated foetal immune cells in the pathogenesis of biliary atresia (BA) but why there are immature B-cells present in the postnatal liver is unknown. To understand the mechanisms behind this, we performed a meta-analysis of RNA sequencing data from infants with BA at the time of Kasai and age-matched controls. Previous analyses had been limited by small numbers of controls; however, here, we have increased power to detect associations.

We obtained transcriptomic data from n=177 children with BA and n=78 controls from Gene Expression Omnibus. After filtering and quality control, we performed differential gene expression analysis comparing BA versus control (using DESeq2), with p-value adjustment for multiple testing.

We identified 1,815 significantly differentially expressed genes (**Fig. 1A**). Many were involved in extracellular matrix remodelling/fibrosis (e.g. *CAPG*, *TGFBI*) and phagocytic activity (e.g. *CHIT1*, *CYBA*). We observed increased expression of *TREM2* (**Fig. 1B**), which is implicated in profibrogenic scar-associated macrophages in adults and has not previously been described in children.

Patients with BA had increased expression of multiple markers of haematopoiesis, including erythroid lineage (e.g. *GYPA*, *HBA1*) and early B-cell (e.g. *RAG2*) (**Fig. 1C**). We identified up-regulation of insulin-like growth factor 2 (*IGF2*, log2 fold change 2.5, pFDR = 2.8×10^{-40}, **Fig. 1D**). *IGF2* is a growth factor for haematopoietic stem cells in the foetal liver. IGF2 is secreted by hepatocyte progenitors and we observed upregulation of markers of these progenitors (e.g. *DLK1*, **Fig. 1E**).

Postnatal liver haematopoiesis is active in children with BA but not controls. Ongoing haematopoiesis may be driven by *IGF2* secreted by hepatocyte progenitors. In contrast to other causes of neonatal hepatitis, specific interactions between haematopoiesis and other immune cells may play a role in exacerbating intra-hepatic injury in BA.

OC13

The epidemiology and long-term outcomes in Gastrointestinal Dystonia (GID): longitudinal single centre data from a tertiary gastroenterology unit over 8 years utilising the BSPGHAN criteria for GID

by R. Poole[1], M. Brooks[1], D. Flynn[1], N. McConnell[1], A. Sabharwal[2], J. Woods[1], A. Campbell[2], T. Bradnock[2], G. Walker[2], K. Fraser[1], S. Fraser[1], J. Andrews[2] and A.R. Barclay[1]. [1]*Paediatric Gastroenterology, Royal Hospital for Children, Glasgow, G51 4TF.* [2]*Paediatric Surgery, Royal Hospital for Children, Glasgow, G51 4TF, UK.*

The complex and progressive debilitating symptoms associated with severe neurodisability and the gastrointestinal tract have long been recognised by specialist nutrition support teams, however have only recently been characterised as GID (1). Whether progressive disease represents specific GI dysfunction or heralds global decline in patient health is poorly defined. Insights into longitudinal treatment success, disease progression, morbidity and mortality would help inform complex decision making for professionals and carers looking after patients with life limiting conditions. We describe outcomes for patients meeting agreed criteria of GID in a tertiary specialist complex feeding (CEN) service.

Patients prospectively gathered on departmental database were identified from referrals 03/2013-12/2020 (entry criteria being ≥2yrs follow up) and were assessed for: baseline demographics; medications used; surgical interventions; blenderised diet (BD); jejunal feeding; parenteral nutrition (PN) episodes; mortality. Patient notes were independently reviewed by 2 authors to agree if they fulfilled criteria for GID, with a third author adjudicating if discordant.

163 patients were referred to the CEN team over the timeline with 40 fulfilling GID. Median age 5.3 years. Diagnosis included: 80% cerebral palsy; 12% genetic syndrome associated with neurodisability; 8% epileptic encephalopathy. 53% had a fundoplication, 58% jejunal feeding and 73% BD over timeline. 85% had ≥3 medication at any time for GID symptoms (fig 1). Patients received 1-3 tone medications, 55% also had botox muscle injections and 18% intrathecal baclofen pump therapy. The majority also received multiple sedating and analgesic medications. There were 3 episodes of PN in 3 patients, with one patient being trained for home PN. One patient received PN in what became evidently a palliative phase. Overall mortality was 6 (15%) over 3 years from entry into CEN service. 3 deaths were from GID and 3 from other causes related to primary condition. Associated therapies included: 25% respiratory home ventilatory support; 35% spinal team assessment; 25% salivary gland botox or surgery.

We present the first case series using defined criteria for GID. We describe the complexity, co-morbidity and disease burden of this condition. Polypharmacy is substantial and requires multi-speciality co-ordination of prescribing and monitoring to inform overall strategy. We would advocate symptom management plans led by palliative and supporting care colleagues. We highlight the resource implications and need for a co-ordinating case holder for these patients with complex multisystem needs. It is of note that the minority of patients benefited from PN for symptom control and went onto HPN. Mortality is significant from GID or co-morbidities in this population.

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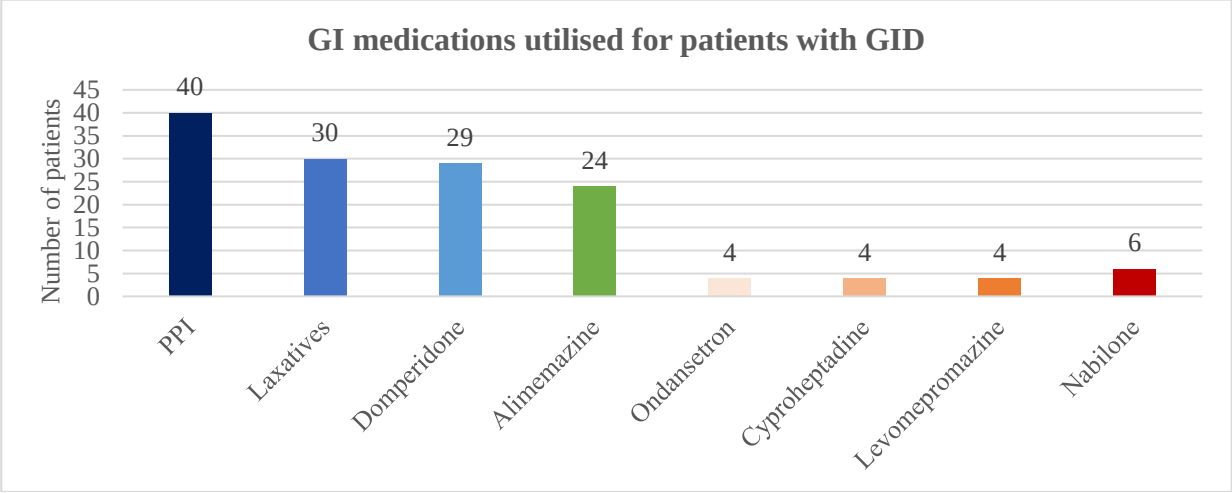

Figure 1

OC14

Outcome of extreme short bowel syndrome, no gut syndrome in children – a multicentre study

By E. Cernat[1], A. Batra[2], J. Koeglmeier[3], R. Wood[4], S. Buxton[5], A. Urs[6], S. Rajani[6], C. Salvestrini[7], N. Onyeador[8], D. Rawat[1] and S. Hill[3]. *[1]Department of Paediatric Gastroenterology, Leeds Children's Hospital NHS Trust, Leeds, LS1 3EX, UK; [2]Department of Paediatric Gastroenterology, Southampton Children's Hospital, Southampton, SO16 6YD, UK; [3]Deparment of Paediatric Gastroenterology, Great Ormond Street Hospital NHS Foundation Trust, London, WC1N 3JH, UK; [4]Department of Paediatric Gastroenterology, Royal Manchester Children's Hospital, Manchester, M13 9WL, UK; [5]Department of Paediatric Gastroenterology, Great North Children's Hospital, Newcastle, NE1 4LP, UK; [6]Department of Paediatric Gastroenterology, Sheffield Children's Hospital NHS Foundation Trust, Sheffield, S10 2TH, UK; [7]Department of Paediatric Gastroenterology, Addenbrooke's Hospital, Cambridge, CB2 0QQ, UK; [8]Department of Paediatric Gastroenterology, St George's University Hospitals NHS Foundation Trust, London, ST16 3SR, UK.*

Children are now surviving with extreme short bowel syndrome (SBS) i.e. resection of the entire jejunum and ileum which has been termed "No gut syndrome" (NGS) in adults. We aimed to describe the population characteristics and outcome in children with NGS.

A questionnaire was sent to the paediatric intestinal rehabilitation centres in the UK via the national intestinal failure (IF) network to identify patients discharged home with long-term IF (>27 days) and NGS. Our inclusion criteria were all infants/children < 18 years with a maximum jejunal remnant < 5 cm from the ligament of Treitz after major excision of jejunum, ileum (and in some cases duodenum) +/- colon. Information obtained included patient demographics, outcome and school attendance. We also requested total number of cases of ultra-short bowel syndrome (U-SBS) < 10cm from duodenojejunal flexure. A multicentre Health Research Authority approval was obtained via the integrated research application system (IRAS).

Nine of 20 (45%) IF centres managing a total of 204 patients on home parenteral nutrition (HPN) replied - 11/204 (5.3%) had U-SBS and 12/204 (5.8%) had NGS. Median age [IQR] was 7 years 6 months [5y 5m–10y 10m]. Two patients were excluded since were recently transitioned to adult services.

The aetiology of NGS was mid-gut volvulus in 7/12 (58%) cases, ischemia in 2/12 (17%), and one of each of necrotising enterocolitis (NEC) 1/12 (8%), post-resection of failed intestinal transplant 1/12 (8%) and intestinal atresia 1/12 (8%).

Eight/12 (67%) patients had duodenum alone and 4/12 (33%) < 5cm jejunum. Eleven/12 had a colonic remnant (2/11 full colon, 6/11 from transverse and 3/11 sigmoid-rectum) which was in continuity in 7/11 cases (2 full colon, 4 transverse, 1 sigmoid-rectum).

There were no patient deaths. One/12 (8%) gained enteral autonomy and one/12 (8%) underwent intestinal transplant (for irreversible IF). The child who gained enteral autonomy was a term infant with neonatal volvulus and colon in continuity from 6 months, weaned from PN aged 4 years 1 month.

The other 10/12 (84%) patients remained stable on HPN, with median [IQR] 7 nights/week [6.75-7], over 12 hours/night [12-13], 2 lipid-free nights/week [0-4]. Seven/11 tolerated enteral nutrition - 1/7 blended diet and 6/7 oral food (3 only trophic). Four cases had mild IF associated liver disease (IFALD), 2 nephrocalcinosis and 1 hypoglycaemic episodes. Information about school attendance was obtained in 11/12 with 9 attending school (8/9 full time) and two nursery.

Advances in PN and health care have enabled long-term survival of patients with no gut syndrome. Children appear to have a reasonable quality of life - they attend school/nursery and PN is only infused overnight. It is also possible for a no gut child to gain enteral autonomy.

OC15

Use of oral spray to maintain normal B12 levels in children with short bowel syndrome and history of vitamin B12 deficiency.

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Children with short bowel syndrome (SBS) are prone to long-term health consequences associated with micronutrient deficiencies.[1] The shortened small intestinal length predisposes to deficiencies with ileal resection resulting in high risk of vitamin B12 deficiency leading to hematologic (megaloblastic anaemia) and neurological (myelopathy, demyelinating diseases)[2] abnormalities. The standard treatment is regular intramuscularly (IM) Vitamin B12 injections if vitamin B12 blood levels are low and the child is no longer on parenteral nutrition/PN.

The aim of our study was to see if vitamin B12 could be maintained in the normal range with oral vitamin B12 spray rather than regular IM injections in patients with SBS and vitamin B12 deficiency.

We retrospectively reviewed children with SBS who were selected to trial oral B12 spray because they were expected to comply with treatment and in a position to purchase it. They were all weaned off PN, were regularly followed up in our clinic with regular vitamin B12 monitoring and had a history of vitamin B12 deficiency. The children were offered *Boost* (*contains B12, green tea and chromium*) *oral* spray as a substitute for IM injections. Children aged >12 years were prescribed *1200 µg=4 sprays/day and those aged<12 years 600ug=2 sprays/day* as explained to them by the dietitian.

Five children, 3 male, 2 female, aged 10 years 4 months-16 years 3 months (mean 12 yrs 10 months) when reviewed were treated with oral B12 spray. The remaining small intestinal length after neonatal intestinal resection was 7-77cm, median 50cm with ileo-caecal valve present in 3 children. All 5 children initially received B12 IM and were transferred to B12 spray. One child had developed a rash with B12 injections so was shifted to spray, while the other 4 preferred spray to injections. The spray had been used for 10-78(mean 31.4) months when reviewed. Serum B12 levels were 179-366 (mean 246.6)ng/L before initiation of spray. When reviewed on spray treatment vitamin B12 levels were 238-727 (mean 453.6)ng/L. None of the patients required B12 injections while they were on regular spray therapy and none of them wanted to change to IM injections. One child had to stop treatment and restart IM B12 due to oral aphthous ulceration associated with spray.

In summary, our study showed that use of B12 spray maintained normal vitamin B12 levels in patients with SBS and vitamin B12 malabsorption. In conclusion, Boost vitamin B12 oral spray can be used as a substitute for IM injections in families willing to comply with therapy.[3]

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OC16

Induction and maintenance therapy using topical steroid preparations in eosinophilic oesophagitis in children: a practical guideline

by *J. Chan[1], *D.M. Flynn[2], M. Gordon[3], M. Gasparetto[4] and M.K.H. Auth[5] on behalf of the BSPGHAN Eosinophilic Oesophagitis (EoE) working group. *[1]Noah's Ark Children's Hospital for Wales, Cardiff, CF14 4XW, [2]Royal Hospital for Children, Glasgow, G51 4TF, [3]University of Central Lancashire (UCLAN) PR1 2HE, [4]Norfolk and Norwich University Hospital, University of East Anglia (UEA), NR4 7UY, [5]Alder Hey Children's Hospital, Liverpool, L12 2AP. *joint first authors. EoE working group: MKH Auth, J Chan, DM Flynn, A Cordell, J Epstein, M Furman, E Gaynor, L Goh, L Jackman, H Kannappan, K Moolenschot, R Parmar, J Thompson.*

Topical steroid use in Eosinophilic Oesophagitis (EoE) has become common practise in recent years for induction and maintenance treatment. Several different steroids have been proposed and newer delivery methods are becoming more readily available for children, with several guidelines published over the past 10 years. As a result of this and exponential increases in wider research into EoE, UK practise is not standardised and varies widely. The EoE working group and research leads of BSPGHAN together carried out a systematic literature review to determine evidence based guidance for preparation, dosing and duration of use of swallowed topical steroid (STS) formulations in EoE in children.

A systematic literature review (no date limits) was carried out using Cochrane guidance and this technical review has informed an evidenced based, best practice guideline, with easy access flow chart for use by healthcare professionals involved in the management of paediatric EoE. It is hoped that this BSPGHAN guideline will be adopted and shared with other professional societies.

2638 citations were identified and 18 Randomised controlled trials pertaining to steroid use were included. Evidence exists for the use of STS for induction and for maintenance therapy in EoE, especially with regards to histological improvement. Using AGREE criteria, dosing of steroids by age (0.5mg BD <10 years and 1mg BD ≥ 10 years) for induction of at least 3 months was suggested based on evidence and practical consideration. Once histological remission has been accomplished, use of maintenance dosing of steroids appears to reduce the frequency and severity of relapse. A maintenance weaning regime was proposed with half dosage (evaluated by repeat endoscopy) and an option to further reduce to a quarter of the dosage. We provide an overview of the practical considerations including; once or twice daily dosing and treatment concordance. Oral viscous budesonide (OVB) and, if agreed by local regulatory committees as current license ≥18 years old, oro-dispersible budesonide (Budesonide 1mg tablets) were selected as the easiest to use preparations with most improvement in histology. A practical 'how to prepare and use' appendix is included. Side effects were identified including candidiasis and adrenal gland suppression, mainly restricted to those receiving other topical steroids for atopic conditions e.g. asthma. The use of oral systemic steroids in strictures may have a role and this was discussed briefly. Dual therapies and when to change therapy were not included as part of this protocol.

In summary, a practical, evidence-based guideline and flow chart with consensus from the EoE Working Group, education and research representatives of BSPGHAN was developed with detailed practical considerations for use in the UK. We plan to demonstrate the algorithm at the annual conference for wider society member review using a QR coded survey system for feedback to ensure stakeholder input before wider dissemination.

OC17

Oral Viscous Budesonide (OVB) technique: a patient adherence improvement project

by L. Jackman, K. Kite, K. Aris, L. Goh and E. Gaynor. *Great Ormond Street Hospital, Great Ormond Street, London, WC1N 3JH, UK.*

Eosinophilic oesophagitis (EoE) is a rare allergic condition of the oesophagus. Untreated inflammation can lead to oesophageal strictures / dysphagia; impacting quality of life (QOL).

There are three recommended treatments' for EoE; proton pump inhibitors (PPI's), dietary exclusions or swallowed topical steroids (budesonide and fluticasone). Until recently there has been no commercial budesonide preparation available for use in EoE. Oro-dispersible tablet, Jorveza®, is licenced only in > 18 years of age.

In paediatrics, families are provided with viscous budesonide vial and instructed to make into a slurry swallow. OVB should have a standardised consistency to ensure adequate contact time in the oesophagus. Incorrect preparation technique can result in inadequate management, unnecessary treatment escalation / additional endoscopic investigations; potentially reducing QOL.

Our multidisciplinary (MDT) clinic established that incorrectly preparing OVB was a frequent problem; suspected to affect compliance and treatment outcomes.

The current recommended ratio for budesonide to sucralose sweetener to make OVB is 5g per 0.5mg/2ml budesonide vial. The use of other vehicles e.g., apple puree and linctus are less well studied.

Our aim was to improve patient adherence and QOL by simplifying OVB mixing technique with digitally-delivered patient education.

We confirmed that 1 x Nutricia® 50ml blue scoop (BS) was equivalent to 5g of sucralose sweetener (Splenda®, supermarket own brands from Asda® & Aldi®). We calculated that one 0.5mg/2ml vial of budesonide should be mixed with one BS to achieve the recommended consistency of OVB. Using this novel technique for mixing OVB we recommended

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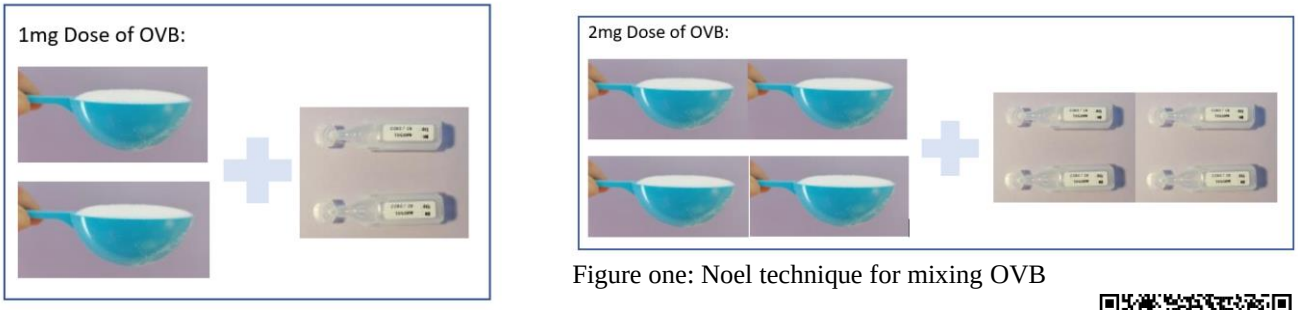

Figure one: Noel technique for mixing OVB

To further support the families, we developed a patient education leaflet and demonstration video

Figure two: QR code for accessing patient education video

Following these measures, we contacted thirty-two patients (age range 4 years 9 months – 17 years 10 months) (7/21 female; 14/21 male) with EoE treated with OVB under the care of a quartnary GI-allergy service. They were asked to review their experience using a standardised telephone survey. developed by the MDT.

66% (21/32) of patients responded. 86% (18/21) were using the BS technique. Of the 14% (3/21) who were not using the BS; 1 patient preferred manually weighing, 1 patient was non-compliant, and 1 patient changed treatment modality. 67% (14/21) of patients reported improved ease to make up OVB following the introduction of the BS. 29% (6/21) had only ever used the BS; therefore, were unable to comment of ease of use. 86% (18/21) of patients watched the education video; 90% (19/21) were provided with written information for mixing OVB. 100% of those who watched the video found this supportive.

In conclusion, our cohort reported the introduction of the BS, educational video and written patient information improved their technique to mix OVB correctly.

Further analysis is required to understand if the BS and enhanced patient education modalities improved clinical outcomes, histological remission and QOL of children with EoE treated with OVB.

OC18

Is it safe to diagnose coeliac disease using the no-biopsy pathway for those with TGA-IgA ≥5x upper limit of normal: a regional study from Southwest England?

by V. Gangalam[1], D. Basude[2], H. Khattak[3], C. Knight[3], L. Tashtoush[4], S. Broad[4], N. Zurick[5] and S. Paul[1,2]. *[1]Yeovil District Hospital. [2]Bristol Royal Hospital for Children. [3]Musgrove Park Hospital, (Taunton). [4]Derriford Hospital, (Plymouth). [5]Royal United Hospital, (Bath), UK.*

Introduction: The ESPGHAN 2020 guidelines have streamlined the diagnostic pathways for paediatric coeliac disease [CD] and have recommended that all children with TGA-IgA ≥10xULN can be safely diagnosed via the no-biopsy pathway [NBP][1]. During the COVID-19 pandemic there has been greater emphasis on the use of the NBP for lower TGA-IgA titres than that recommended by the ESPGHAN for diagnosing CD across the world including the UK[2]. This practice has been supported by papers published from tertiary gastroenterology centres in Italy, India and England where a cut-off value of TGA ≥5xULN have been deemed to be safe for NBP[3-5]. An English study published in 2017 have highlighted the variation in the TGA-IgA titres used across the centres in the UK including southwest England where the titre varied from 4 IU/ml to 30 IU/ml[6].

Aims: To establish whether the lower TGA-IgA ≥5xULN can be safely used for diagnosing CD via NBP across various TGA-IgA titres used in the non-specialist regional centres in the Southwest England [SWE].

Methods: Anonymous data between 2013 and 2020 were obtained from 5 regional centres across the SWE for the patients with TGA-IgA between ≥5xULN and <10xULN at the time of referral and as to whether they were finally diagnosed with CD following upper GI endoscopy and histology. Data was also collected on the age of the patients at first TGA-IgA titres, sex, symptoms and other high risk factors e.g. Type-1 diabetes mellitus, first degree relatives.

Results: Five centres participated in the study. A total of 8 TGA-IgA titres have been used with values varying between 4 IU/ml to 30 CU/ml. 67 patients were identified and 48 underwent UGIE and biopsies, 45/48 (93.75%) got confirmation for CD on histology. The 3 who did not have confirmatory histology had repeat TGA-IgA <5xULN at the time of endoscopy. Out of rest 19, 13 were diagnosed via NBP and rest 6 did not undergo a formal diagnostic process to confirm or rule out CD. There were 44 females and 23 males; mean age was 10.1 years [range 2 – 17 years]. Out of 67 children, 5 had family history, 4 had type 1 diabetes, 1 each had auto-immune condition and Down's syndrome, rest 8 had no documented indication for undergoing a screening for CD. Figure 1 shows the breakdown for the 5 centres and their conversion for diagnosed CD.

Conclusions: This 5 centre study shows 93.75% correlation of TGA-IgA 5-10xULN at referral to final diagnosis of CD despite 8 variations in assay methods. This appears to improve to 100% when repeated at time of endoscopy. 19.4% of this group were given a diagnosis of CD via NBP based on clinician's decision. There is need for similar collaborative studies through the BSPGHAN PeGHAN network to firmly establish similar findings for various TGA-IgA titres used across the UK.

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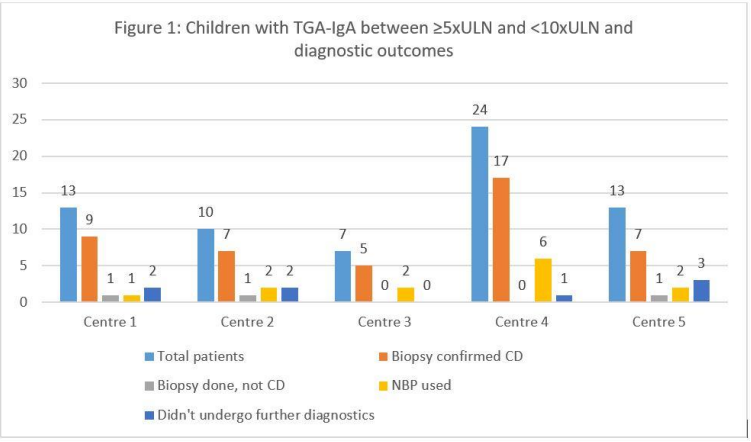

OC19

An unusual genotypic case of very early onset inflammatory bowel disease

by E.J. Imoisili and A. Mukhopadhyay. *Great North Children's Hospital, Newcastle-upon-Tyne, NE1 4LP, UK.*

Very Early Onset Inflammatory bowel disease (VEOIBD) is a growing concern in the paediatric population. This category of patients has certain peculiarities, among which are the single gene causation found in a good number of these patients and unusual course of the disease.[1]

The patient was a 3-Year-Old girl who was initially brought for a General Paediatric assessment following concerns of new onset constipation. She was treated for 6 months with Movicol and sodium picosulfate with partial resolution. On further assessment however, she was noted to have perianal mucosal protrusions thought to be internal haemorrhoids and weight loss. She was referred for specialist assessment where she was found to have pancolitis on endoscopy. She also had an unusual finding of splenomegaly. She was started on infliximab infusions, azathioprine, and topical tacrolimus (for rectal lesions).

In view of her age and clinical findings, she had additional investigations as required for children with VEOIBD. She was found to have IL-27 serum auto-antibodies (IgG). She however showed normal response to IL-10 panel testing. She had additional immunological, genetic testing and glycogen storage disease screening done which were normal. She is currently on 4 weekly vedolizumab treatment, with steroids, following poor response to high dose infliximab and is being considered for colostomy and colectomy if current treatment fails.

Many literatures highlight the difficulty of diagnosing VEOIBD due to overlapping clinical features.[2,3]The patient presented in this case was on long-term treatment for constipation and anal protrusions were initially thought to be mere complications of this, however incidental findings of pancolitis on endoscopy made the diagnosis apparent in this child. Such initial misdiagnosis is not uncommon.

The relationship between IL-27 and IL-10 have been described by some authors, but whereas there is documented cases of IL-10 signalling defects resulting in VEOIBD, data is lacking on IL-27 related disease.[3] It would therefore be interesting to see if our patient would have any disease peculiarity as is the case with IL-10 related cases.

As highlighted above, our patient is currently on vedolizumab therapy, some authors suggest 'considering anakinra, an IL-1 receptor antagonist, in patients with IL-10 signalling defects who are too ill to undergo transplant or while searching for a suitable donor, as this has led to marked clinical, endoscopic, and histologic improvement in some patients'.(Schwartz et al, 2022)[4] It remains to be seen if our patient would respond to a particular type of treatment, in preference over another. Currently, the patient has failed repeated treatment with high dose infliximab. Response to further treatment will be closely monitored.

In conclusion, this case highlights a rare direct IL-27 association with VEOIBD. We hope that this case stimulates a search for similar genetic cases, and that observation of the course of the disease and response to current therapies provides useful insight into this rare entity.

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OC20

Exclusive enteral nutrition to induce biochemical remission in Crohn's disease: characteristics of treatment failure

By R. Stanley, A. Elliott Horn and D.I. Campbell, *Great North Children's Hospital, RVI, Newcastle-upon-Tyne, NE1 4LP, UK*

Mucosal healing is considered the goal of Crohn's disease (CD) treatment (1) with Faecal Calprotectin (FCP) <250ug/g confirming good response. Treatment with exclusive enteral nutrition (EEN) has previously shown a rising FCP in 40% of patients despite apparent treatment compliance (2)

This study measures clinical disease activity scores and FCP across 8 weeks treatment with EEN comparing disease distribution between biochemical responders (FCP <250ug/g) and non-responders.

Patients treated with EEN, were identified retrospectively via clinical records between 2018-2021. Inclusion criteria: <18 years, newly diagnosed CD, completed 8 weeks. FCP and weighted Paediatric Crohn's Disease Activity Index (wPCDAI) were measured baseline, week 4 and 8. Montreal classification for disease site at enrolment, age and gender documented.

A total of 100 patients met inclusion criteria, with 30 having complete data. Table 1 shows demographic of patients with complete data.

Table 1: Demographic information

Montreal classification	L1 (isolated small bowel)	3
	L2 (isolated colonic)	8
	L3 (ileocolonic)	19
Age (mean SD)	12.0 (2.3y)	
Male:Female	M=23 F=7	
wPCDAI <12 (remission)	L1+ L3 = 18/22 (82%)	L2 3/8 (38%)

21/30 patients achieved clinical remission (wPCDAI <12 points), 3 patients wPCDAI did not fall all had increased FCP.

9 achieved FCP <250ug/g. Their average baseline FCP were lower compared to non-responders [723 (SD 263) v 2138.3 (SD 1906), p>0.05].

8 children (26%) FCP increased at week 8, with no significant difference in disease distribution from responders (chi L1, L2, L3 p>0.2). These 8 children tended to have a higher baseline wPCDAI compared to responders (20.6 (SD 21.8) v 8.9 (SD 11.3), p=0.06). Week 8 50% had wPCDAI <12.

This series reports data on real clinical practice. Treatment for CD with 8 weeks EEN, results in 70% having a clinical improvement, yet a 70% chance of failing to respond biochemically (FCP<250mmol/L) i.e. failed mucosal healing. Disease distribution does not predict failure.

High dropout rates, poor biochemical response alongside baseline clinical symptoms, should be considered when offering EEN therapy.

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OC21

Post-induction therapeutic drug monitoring of Infliximab enables early optimisation of biological treatment in the maintenance phase

by C. Bakewell[1], J.J. Ashton[1,2,3], C. Davies[4], C. Barnes[1], T. Coelho[1], N.A. Afzal[1] and R.M. Beattie[1.]
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Routine measurement of infliximab levels and anti-drug antibodies before the 4th infliximab dose is recommended by ESPGHAN in paediatric IBD. This proactive approach is intended to prompt early optimisation of dosing during the maintenance phase. In our Children's Hospital we routinely measure infliximab trough levels and anti-drug antibodies prior to the 4th dose. We aimed to assess if this practice leads to changes in treatment post-induction.

Patients aged 0–17 years commencing infliximab between January 2021 and May 2022 who had trough infliximab levels and anti-drug antibodies measured at their 4th dose were included. Infliximab level ≥5mg/L was considered therapeutic and the threshold for antibody positivity was >10AU/L. Outcome data were collected from our hospital database including demographics, biochemistry, and clinical course to assess for a change of infliximab dose/frequency, switch of biologic or introduction/increase of thiopurine.
Of the 44 patients that met inclusion criteria, infliximab levels were sub-therapeutic in 27 (61%) and antibodies were elevated in 17 (39%). Treatment was changed for 21 patients (48%) following therapeutic drug monitoring (TDM) and both low infliximab levels and elevated antibodies independently predicted subsequent treatment escalation (p=0.012 and p=0.023 respectively). The most common change was to increase infliximab dose from 5mg/kg to 10mg/kg (14/21). Of the 23 patients whose treatment was not changed, 14 (61%) had both therapeutic infliximab levels and negative antibodies. Serum albumin correlated positively with infliximab level (p=0.035) and negatively with CRP (p=0.0001) but neither CRP, albumin, age, underlying diagnosis or thiopurine use predicted subsequent treatment change.

Routine measurement of infliximab levels and anti-drug antibodies before the 4th dose was associated with treatment change in a substantial proportion of patients, the majority of whom increased infliximab dose. These data reinforce the importance of optimising infliximab dose based on TDM to achieve 'adequate' levels, although longer follow-up is needed to understand how these early adjustments affect clinical outcome.

OC22

App for paediatric inflammatory bowel disease (PIBD) patients – a success?

by E.L.Wilkinson, *Cambridge University Hospitals NHS Foundation Trust, Hills Road, Cambridge, CB2 0QQ, UK.*

To determine the impact of the introduction of the app IBDMate to the paediatric inflammatory bowel disease (PIBD) service on patient satisfaction, medication adherence and understanding.

The PIBD department and IBDrelief have developed an app - IBDMate for children with inflammatory bowel disease (IBD) and their guardians. The app aims to improve the health literacy and quality of life for patients. With IBDMate, the PIBD team can 'prescribe' educational courses from hundreds of videos, articles and quizzes featuring the PIBD team. Topics include living with IBD, medication, meet your team, tests and research. The project was ethically approved by the patient engagement team. A baseline measure of patient satisfaction with the PIBD service and understanding of medications was established prior to the app introduction using a paper-based questionnaire, distributed to patients after their hospital visit from 01/10/2020 - 31/12/2020. Following the introduction of the app, a second online questionnaire was distributed from 01/05/2021 - 30/06/2021. The PIBD helpline was also reviewed for medication related enquiries during the study period. Data from patients/guardians who had not accessed the app before completing the second questionnaire were excluded. Patients were given a unique identifier and no personal information was collected. Descriptive and comparative statistical analysis was undertaken to assess impact.

The first and second questionnaires were completed by 33 and 31 patients respectively. Patient satisfaction with the quality and way information is received improved from 88% to 100%. Understanding of how medication works and side effects of medicines improved by almost 20%. After using IBDmate patients were able to remember 10% more information about their medicine and unintentional medication omission reduced from 10% to 0%. Responses to open questions revealed patients felt that the app helped them understand their medicines better and they found it useful to get to know the clinical team and hear other patients' stories. Participants felt the app was a trusted, reliable and relevant source of information. Suggested improvements were having a section for younger children to engage with, and retention of login details. 55% used the app to look at information about their medication. The number of calls to the PIBD helpline that were related to medication dropped from 25% to 15% following introduction of the app.

The introduction of IBDMate has had numerous positive impacts on patients through increased knowledge, accessibility, medication adherence and trust in information. This project demonstrates the benefits and further potential of the app although several areas would benefit from additional work:

- Monitor service satisfaction to ensure high standards are maintained as the app is developed
- Encourage broader use of the app
- Further research on the impact of the app on freeing up clinical resources by reduced helpline call volume and clinic visits
- Further development of the app to include resources for younger children and retention of login details

OC23

National survey of exclusive enteral nutrition protocol used in paediatric tertiary inflammatory bowel disease centres in the UK

by L. Jackman, L. Arpe and G.O'Conner. *Great Ormond Street Hospital, Great Ormond Street, London, WC1N 3JH, UK.*

Exclusive Enteral Nutrition (EEN) is the first line treatment for Crohn's Disease (CD) in the UK and has been proven to be an excellent steroid sparing option with remission rates reported between 60 – 80%[1]. Although this dietary treatment is well established, there continue to be significant variation in practice; including type of formula used, length of EEN, protocols for food reintroduction and use of naso-gastric tube (NGT) feeding versus oral diet. This survey aimed to detail the current practices of implementing EEN across tertiary inflammatory bowel disease (IBD) centres in the United Kingdom (UK).

We performed a UK based survey to ascertain current practice with regards to EEN in the treatment of CD across 20 tertiary IBD centres. Dietitians were contacted at these centres and asked to complete an electronic questionnaire regarding the following topics: 1. Feed choice 2. Duration, route and exclusivity 3. Dietetic support 4. Food reintroduction approach 5. Alternative dietary management options.

17/20 (85%) of invited centres responded. 100% of centres used polymeric feeds as their first choice. 70% of centres recommended EEN for 6 weeks. 58% of centres did not audit the use of NGT but of the 6 centres that did found that 0-25% required an NGT as some stage of EEN. Foods and drinks allowed whilst on EEN varied greatly. Dietetic review frequencies also varied significantly between IBD centres. 43% introduces solid foods over 5-7 days, 19% introduced food over 7-14 days and 37.5% introduced food over a minimum of 14 days. 18% of the centres were offering the CDED as a treatment for IBD.

This survey shows that there is a varied practice when it comes to implementing EEN for CD patients across the UK. These findings mirror those which were reported globally in in 2018[2]. There has been little progress made in standardising practice despite multiple clinical guidelines supporting this treatment methodology[3 4 5 6].

Further evidence is required to understand if variations in practice have an impact on remission rates in CD.

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OC24

Evaluating remission rates of standard polymeric feed use for Exclusive Enteral Nutrition in a Paediatric Crohn's Disease

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Exclusive Enteral Nutrition (EEN) is the first line treatment for the induction of remission in Paediatric Crohn's Disease (CD) (1,2). There are no studies which evaluate the effectiveness of one polymeric formula over another in terms of rates of remission (1,2). Our centre, prescribes feeds based on age, weight, taste preference, convenience, and allergic history.The aim of this retrospective chart review is to evaluate our local remission rates for paediatric Crohn's disease patients having EEN and assess if standard polymeric feeds result in remission rates that are concurrent with published data. We retrospectively reviewed EEN patients between the 1st of November 2019 to 31st October 2022. The wPCDAI was calculated at baseline and at the end of treatment (6 weeks). Remission was defined as wPCDAI of below 12.5 (3); clinical response was defined as a reduction of more than 17.5 in wPCDAI (3). Fifty patients (30 Male and 20 Female, age range: 2y to 17y) were treated with EEN. 15/50 patients were excluded from the final cohort: 4/50 did not complete the prescribed course of EEN; 5/50 had incomplete clinical data (wPCDAI could not be calculated), 6/50 patients had EEN in combination with biologic treatment. 25/35 (71%) were in clinical remission following completion of EEN at 6 weeks. Furthermore, 29/35 (83%) had a clinical response to the treatment. Of the 25 patients who entered remission, 18 received standard polymeric sip feeds (Ensure plus/Paediasure Plus/ Paediasure). 6/35 patients did not achieve clinical remission, however demonstrated a clinical response (minimum reduction of 17.5 in wPCDAI), with an average reduction of 36 in 6 weeks. Our remission rates (71%) were concurrent with published data. Standard polymeric remission rates were 70% at 6 weeks, which is comparable to oral Modulen IBD® (76%) at 8 weeks (4). This shows that standard feeds are effective for inducing remission in CD. It's likely that increased feed choice may improve adherence to EEN. Furthermore, many publications evaluate the remission rates of EEN after 8 weeks (4). In future it may be beneficial for our centre to aim for a minimum of 8 weeks as some of the wPCDAI scores were just above the range that demonstrated clinical remission. Furthermore, including faecal calprotectin results at baseline and end of treatment may provide further insight regarding clinical remission.

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OC25

Aiming for growth targets and optimised bone health in children with Crohn's disease

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Growth failure occurs in up to 85% of children newly diagnosed with Crohn's Disease (CD) and persists in up to 40% throughout their disease course.(1) Undernutrition and chronic inflammation appear to be the main driving factors behind growth failure and poor bone health.(2, 3) Adequate disease control and good nutritional status contribute to achieving growth target potential and minimising the long-term implications related to bone health.

To explore correlations between CD characteristics, bone health and growth parameters at diagnosis and during follow-up (Fig. 1A) and assess ways to optimise these parameters.

Retrospective data were collected from children (age at diagnosis < 18 years) newly diagnosed with CD between January 2018 and December 2019, from the Electronic Patient Records at our Institution. Baseline information surrounding CD (including age of diagnosis, disease location and severity score), bone health (including bone age, densitometry scan (DXA) Z-scores for bone mineral density (BMD) and bone mineral concentration (BMC), Vitamin D level) and growth (including height and weight centiles, BMI and pubertal status) was retrieved. Growth parameters were reassessed at follow-up to understand the changes in anthropometric measurements over time. Growth delay (Paris classification G1) was defined as involuntary stable weight or weight loss of 10% or more over the previous 6 months, and/or a decrease in height by >= 1 centile or a decrease in height velocity by >= 1 standard deviations (SD). Inferential statistical analyses performed using IBM SPSS Statistics v28.0 and *R* Bioconductor, included Pearson's Chi Squared test, Bonferroni correction for multiple testing, Fishers Exact Test, Spearman's Rank Correlation Coefficient, linear and multiple regression.

Seventy-six children were included (53 males (69.7%); age at diagnosis: mean 12.8 years, median 13.7, range 4.05-17.2; disease location (Paris classification): 28 L1, 12 L2, 36 L3; follow-up duration: mean 3.2 years, median 3.4, range 1-4.3). Thirty-one (81%) children had a DXA scan at diagnosis while only 2 children had a repeat scan during follow-up. Children with poorer DXA Z-scores had a younger age at diagnosis (P=0.021), more severe disease (P=0.074 for BMD, P=0.017 for BMC) and a lower BMI (P=0.002) at diagnosis (Fig. 1B and 1C). Seventeen (22.4%) children had growth delay at diagnosis, and at follow-up 44% of these children continued to have growth delay. Five children had bone fractures during the follow-up (2 had growth delay at diagnosis and 4 received one or more courses of steroids). Multivariate regression analysis demonstrated an older age of diagnosis to be a significant predictor of a lower height velocity at follow-up.

Disease severity and age of diagnosis are important CD-related factors that influence bone health and growth (Fig. 1D and 1E). These factors are closely related and impact on each other, therefore monitoring and optimising each aspect is pivotal to support children towards achievement of their growth and bone health potential. Monitoring should include repeat DXA scans during the disease course and optimisation of calcium and vitamin D intake throughout follow-up.

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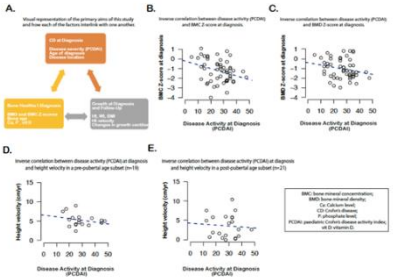
Figure 1.



OC26

An investigation into the correlation between faecal calprotectin and disease activity in paediatric inflammatory bowel disease

by A. De. *Kings College London Guy's Campus, Great Maze Pond, London, SE1 1UL, UK.*

Inflammatory bowel disease (IBD) is a chronic gastrointestinal inflammatory condition commonly presenting in childhood; there are two main subtypes Crohn's disease (CD) and ulcerative colitis (UC). Faecal Calprotectin (FCP) has been utilised as a marker of intestinal inflammation and has become routine in its use for management of IBD; both in monitoring disease activity and predicting relapse and remission in fluctuating pathology and symptomology of patients. This study aimed to assess the relationship between FCP, IBD activity indices and the commonly used blood markers in paediatric IBD.

In a retrospective data study at the Paediatric Gastroenterology Department at Hospital A, we analysed faecal calprotectin and disease activity using the scores Paediatric CD Activity Index (PCDAI) and Paediatric UC Activity Index (PUCAI) for Crohn's Disease subtype and Ulcerative Colitis subtype respectively in 208 IBD patients from the years 2015 to 2021. Paris classification was used for phenotype identification. Spearman's Rho correlation coefficient analysis was performed to draw and quantify correlation between faecal calprotectin and the two clinical scores.

208 patients were included in this study, with 115 CD (18% < 10 years and 82% 10-17 years) and 93 UC (32% < 10years and 68% 10- 17 years). There was a positive correlation between FCP and PCDAI (r_s = 0.546, $p < 0.001$) and between FCP and PUCAI (r_s = 0.485, $p < 0.001$). FCP and activity indices were correlated positively with inflammatory markers/ platelets and negatively with albumin and haemoglobin. FCP positively correlates with PCDAI in all CD phenotypes including isolated ileal disease.

In paediatric IBD, FCP appear to positively correlate with clinical picture and blood markers in all disease phenotypes and can provide an accurate non-invasive measure of disease activity.

OC27

Pharmacist-led post induction review (PIR) for adult IBD patients initiated on biologics

ABSTRACT WITHDREW

OC28

Influence of ethnicity in the clinical profile of paediatric inflammatory bowel disease patients

by S. Gulam Khader and H. Kannappan. *University Hospitals Coventry and Warwickshire NHS Trust, Clifford Bridge Road, Coventry, CV2 2DX, UK.*

The inflammatory bowel diseases (IBD) comprising Crohn's disease (CD) and ulcerative colitis (UC) are idiopathic diseases of the gastrointestinal tract characterized by a relapsing and remitting course. (1) Despite the increasing incidence of IBD, much uncertainty still exists about the relationship between ethnicity and differences in the epidemiology and features of IBD. This study seeks to explore the effects of ethnicity on the diagnosis, management, and outcomes of patients with IBD.

Sixty-two paediatric IBD patients in a university gastroenterology practice in the UK, between 2018 and 2022 were retrospectively analysed to study the variation of clinical presentation by ethnicity. Patient self-reported ethnicity was obtained from the hospital administrative records and three subgroups were identified: Whites, Asians and Blacks. The study cohort included 35 CD patients, 22 UC patients and 5 IBD unclassified patients.

We noted that out of the 35 CD patients, 60% were Whites, whereas 28.6 % belonged to the Asian group and 11.4 % were Blacks. Similarly in the UC subgroup, Whites constituted the majority (54.5%) whereas 40.9% were Asians and less than 5% were Blacks.

Among the Asian population, the proportions of CD and UC were almost equal. In contrast, 58.3 % of Whites presented with CD compared to 33.3% with ulcerative colitis. The difference was more pronounced in Blacks with 80% presenting with CD and 20% with UC, although these results were not statistically significant.

Further analysis showed that among those who first presented with symptoms of IBD at less than 5 years of age, a significantly higher proportion were Asians at 50% and 37.5% were Blacks compared to 12.5% Whites. However, nearly two-thirds of those who first presented at the age of 6-10 years and 11-15 years were Whites ($p = 0.010$).

When comparing disease extent in CD, 80% of the Asians presented with ileocolonic disease whereas half the Black population had localised ileal involvement. However, the incidence of upper gastrointestinal involvement was higher in the White ethnicity. In UC, nearly three-quarters of those presenting with extensive colitis belonged to the Asian ethnicity as opposed to Whites who were more likely to have proctitis or left-sided colitis ($p = 0.013$). Four out of six patients that developed extraintestinal manifestations of IBD were Whites.

Among children requiring less than two hospital admissions for flare-up of IBD symptoms, approximately two-thirds were Whites. Only two patients required more than two hospital admissions and they belonged to the Asian subgroup. With regard to treatment, our results demonstrated that 60% of the Blacks required biological therapy, whereas equal proportions of the Asian and White subgroups were treated with immunomodulators like azathioprine or aminosalicylates and biologicals. Lastly, allergic reactions to azathioprine and biologicals were more commonly noted in the White population.

Despite the relatively limited sample size, this study certainly provides valuable insights into the differences in phenotypes and presentations of IBD across paediatric patients of various ethnicities. There is a defined need for further research to assess the role of ethnicity in the pathogenesis and progression of paediatric IBD patients.

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OC29

An unusual presentation of severe lower back pain 3 months after diagnosis with Crohn's disease and following treatment with systemic steroids

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Patients with Crohn's disease frequently have low bone mineral density (BMD) at diagnosis and through follow-up.[1] Multiple factors contribute to this risk, including poor nutrition, inflammatory cytokines, glucocorticoid treatment, vitamin D deficiency, decreased activity and reduced muscle mass.[2] We present an interesting case of severe lower back pain due to vertebral compression fractures, in a newly diagnosed patient with Crohn's disease, treated with systemic steroids.

A 11-year-old male, diagnosed with Crohn's disease three months prior, presented to outpatient clinic with a two-week history of severe lower back pain. He had been treated initially with two weeks of oral steroid (40mg) followed by a seven-week weaning regime. The pain started in the final week of this course and was described as constant and sharp with intermittent spasms. He had associated morning stiffness in the lower back and struggled getting out of bed. There was no significant past medical history. At time of Crohn's diagnosis, his symptoms included abdominal pain, mouth ulcers, and weight loss. While he demonstrated clinical response to steroid, he was due to escalate to infliximab in view of extensive small bowel involvement on MRI.

On examination, there was a reduction of spinal mobility with mild paraspinal tenderness, especially in the thoracic region. An urgent referral was made to rheumatology who arranged an MRI spine to investigate for IBD-related arthropathy/sacroiliitis. This demonstrated multiple new compression fractures of the lumbar spine but no evidence of sacroiliitis. A subsequent x-ray showed wedge compression fractures of all the vertebrae from T11 to L5. This was associated with a 4cm loss of vertical height. An urgent referral was made to the spinal surgeons although interestingly, the lower back pain had already improved at the time of the referral, having reported significant symptomatic improvement during Infliximab induction. At present, this patient remains under close monitoring, with a particular focus on his bone health and is establishing infliximab treatment. While a vitamin D level was not sent initially in this patient, subsequent blood tests showed an adequate vitamin D level (60nmol/L).

All clinicians managing children with IBD must remain vigilant for low BMD-associated complications. These can often mimic arthropathy. While glucocorticoid treatment often results in prompt induction of clinical remission of IBD, it can have a negative impact on bone health, further compounding the damage inflicted by chronic inflammation. Exclusive enteral nutrition or treatment with anti-TNF agents may be preferable in a selected group of patients, particularly those who have low-BMD at diagnosis.

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OC30

Impact of induction therapies on anthropometric indices in Luminal Crohn's disease

by S. Rajani, R. Pybus, R. Rodley, K. Green, V. Forbes, N. Delnero, A. Urs, D. Schluckebier, R. Rabone, S. Sharma, N. Nedelkopoulou, P. Rao, P. Narula, M. Thomson, Z. Londt and A. Kapoor. *Sheffield Children's Hospital, Clarkson Street, Sheffield S10 2TH, UK.*

We conducted a retrospective study to assess whether there is any additional benefit of Exclusive enteral nutrition (EEN)EEN as primary induction therapy on anthropometric parameters in children with luminal Crohn's disease in secure remission at one year post diagnosis. We evaluated the impact of different induction therapies on anthropometric indices at one year.

A retrospective review of records for all Crohn's disease (CD) patients diagnosed between January 2019 and December 2021 was done. We used clinic notes, letters, investigations to collect our data.80 children were diagnosed with inflammatory CD of which 40 were males and 20 were females. The average age at presentation was 13.9 years. 38(47.5%) patients received exclusive enteral nutrition (EEN) at presentation, and 42(52.5%) received other treatments(steroids/biologics). 16(20%) patients needed steroids at presentation, 26(32.5 %) needed biologics in view of severe disease. All patients were in secure clinical (patient reported outcomes and clinical assessment) and biochemical remission (CRP, ESR and faecal calprotectin) at the end of one year.Maintenance therapy for patients included immunomodulators(azathioprine/methotrexate) and/or biologics.

Paired t test revealed significant improvement in weight z-score($p<0.000$) and height ($p=0.021$) for all patients irrespective of initial induction agent at one year. There was no significant difference between improvement in weight or height z-scores when EEN was compared with other induction therapies ($p= 0.832$ for weight and 0.090 for height)

EEN is established therapy for luminal Crohn's disease with growth failure. Other induction agents were equally efficacious as EEN in improving weight and height z scores at one year. Mitigating systemic inflammation in IBD seems to ameliorate the negative impact on growth parameters. Longitudinal follow up may be needed to establish impact on eventual height.

OC31

Case report and literature review of chronic recurrent multifocal osteomyelitis (CRMO) associated with paediatric Crohn's disease

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Chronic recurrent multifocal osteomyelitis (CRMO) is a rare autoinflammatory condition presenting with gradual onset bony pain, localised tenderness and swelling. Bone marrow inflammation and lytic bone lesions are seen, with elevated inflammatory markers and sterile cultures. The exact aetiology is unclear; however, it is thought to be caused by a disruption in innate immunity regulation because of cytokine imbalances[1].

We present a case of a 7-year-old female who presented with an 8-month history of chronic diarrhoea, nocturnal stooling, occasional bloody and mucous stool and recurrent mouth ulcers. No abdominal pain or weight loss was reported. Initial investigations showed a raised ESR of 16mm/hr, raised faecal calprotectin of 1200mcg/g and a raised platelet count of 449×10^9/L. Upper GI endoscopy, ileo-colonoscopy and histology were in keeping with a diagnosis of Crohn's disease (CD). The MRI small bowel showed significantly thickened small bowel loop, lying to the left of the midline with no retroperitoneal lymphadenopathy or pelvic free fluid. Initial treatment involved exclusive enteral nutrition and azathioprine was commenced later. Prednisolone was started due to lack of improvement, but investigations demonstrated high disease activity. Infliximab at 5mg/kg was started due to failure to induce remission. She responded to infliximab and over the next 3 years, the patient's GI symptoms were well controlled.

She then developed a non-tender right-sided clavicular head swelling and a cervical lymph node. CRP and LDH were normal, ESR was slightly raised at 22mm/h but normalised. Ultrasound of the neck showed features of a reactive lymph node. MRI of right clavicle confirmed CRMO affecting the medial end of right clavicle. She had no GI symptoms, with a reassuring repeat endoscopy. Infliximab infusions were stopped, and the patient was started on methotrexate following rheumatology advice. At the time of writing, the patient has clinically improved with well controlled CD.

CRMO treatment can include NSAIDs, corticosteroids, methotrexate, bisphosphonates, and tumour necrosis factor (TNF)-α inhibitors[2]. However, our patient was in CD remission at the time of CRMO symptom onset and improved after infliximab was stopped and replaced by methotrexate. A case series by Cordesse et al. (3) supports this approach, demonstrating a paradoxical CRMO reaction occurring in 3 patients in remission of CD through anti-TNFα efficiency. The exact physiopathology remains unclear, and management can be complex. As the GI disease was under control in our patient, stopping TNF-α inhibitors was possible and this may be useful in other patients. Detailed multi-professional management will be required for these cohort of patients.

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OC32

Real-world Scottish experience of anti-TNF therapy in paediatric Crohn's disease 2016-2020 against the ECCO-ESPGHAN recommendations

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ECCO-ESPGHAN updated the guideline on the management of paediatric Crohn's disease (CD) in 2021. This promoted a move to early risk stratification and a 'top-down' (anti-TNF within 4 weeks of diagnosis) approach for patients who were deemed high-risk. High-risk was defined by extensive panenteric or severe disease, perianal, stricturing and/or penetrating behaviour. We sought to compare the use of anti-TNF therapy against the new guidance by conducting a Scottish nationwide retrospective multi-centre study (all three regional PGHAN units).

We analysed a prospectively identified nationwide cohort of all new paediatric CD patients (<18 years) diagnosed in Scotland between 01/01/16 and 31/12/20. We retrospectively collected data from electronic medical records on the use of anti-TNF therapy (infliximab or adalimumab) within 18 months of diagnosis. Paris location and behaviour at diagnosis were determined, allowing us to split the patient group into high-risk or low/medium-risk as per latest ECCO-ESPGHAN guidance. Data regarding surgical intervention was collected and defined as: Perianal (Seton suture or drainage of abscess) and/or bowel resection +/- stoma formation.

419 patients were included (259/419 male; median age at diagnosis 13.2 yrs). Disease phenotype is shown in table 1. 225/419 (54%) were classified as high-risk and 194/419 (46%) as low/medium-risk. 171/225 (76%) high-risk and 78/194 (40%) low/medium-risk patients received anti-TNF within 18 months of diagnosis, with median start at 5 months (IQR: 1- 8 m) and 6.5 months (IQR: 3 – 13 m) respectively. 49/171 (40%) high-risk and 12/78 (15.3%) low-risk received anti-TNF within 4 weeks of diagnosis. High-risk patients were more likely to receive anti-TNF (76.0% vs 40.2%, p<0.0001). 37 (8.8%) patients underwent surgical intervention; 31/225 (13.7%) high-risk and 6/194 (3.1%) low/medium-risk (13.7% vs 3.1%, p= 0.0002).

The ECCO-ESPGHAN guidance seeks to shift practice towards "top-down" anti-TNF therapy in high-risk patients. Our real-world data supports this view by showing that high-risk patients are more likely to require biologics within 18 months and more likely to require surgery than low/medium-risk patients. The ECCO-ESPGHAN approach would however have led to unnecessary anti-TNF in 24% of our high-risk cohort within 18 months of diagnosis. The financial cost and risk:benefit profile of this group should be considered carefully both in clinical practice and future guidance.

	Total no. (%)	+ No upper GI disease	+ Upper GI disease	L4a	L4b	L4a+b
L1	62 (15)	38 (61)	24 (39)	11 (18)	7 (11)	6 (10)
L2	114 (27)	77 (68)	37 (32)	30 (26)	5 (4)	2 (2)
L3	198 (47)	77 (39)	121 (61)	80 (40)	20 (10)	21 (11)
Isolated L4	45 (10)					

Table 1: Paris Classification Disease Location

OC33

Adalimumab Dose Optimisation

by C. Ogilvie, J. Joachim, A. Sood, A. Hibbert, B. Kukoyi and B. Hope. *Kings College Hospital, Denmark Hill, SE5 9RS, UK.*

Patients starting adalimumab (ADA) receive 'standard' or 'accelerated' dosing. Guidelines on selecting patients for the accelerated regimen are lacking[1,2].

Our departmental database was used to identify 32 IBD patients treated with ADA between 2013 and 2021. The mean age at ADA commencement was 12.8 years. 22 patients had a diagnosis of ulcerative colitis (UC), and 10 had Crohn's disease (CD). The mean interval between diagnosis and starting ADA was 3.1 years (range = 4months - 8years).

29/32 received a second immunomodulator drug in addition to ADA, usually azathioprine (26/32). 29/32 had previously been treated with infliximab (IFX). Reasons for switching included primary lack of response (12/32); adverse reaction (7/32) and loss of response due to anti IFX antibodies (7/32). No significant adverse reactions to ADA occurred in this cohort.

Faecal calprotectin (FCP) levels were available for all patients both at baseline (before starting ADA) and after induction, and paediatric ulcerative colitis activity index (PUCAI) and paediatric Crohn disease activity index (PCDAI) scores were available for 31/32.

Patients were grouped according to whether they remained on their starting dose (Group A), or increased their dose because of disappointing clinical response plus low or intermediate serum levels (<8µg/mL)[3], (Group B).

28/32 received standard induction doses of adalimumab. Serum levels were checked 12 weeks after induction. The mean post induction serum ADA level of the accelerated regimen group was 5.98µg/mL. Of the 4 accelerated induction patients, only two continued on ADA at the time of data collection, one on an increased dose.

Baseline FCP was similar in both groups, although Group B had higher mean DAI scores. FCP and DAI scores fell after induction for Group A (p=0.027 and 0.5), and after dose increase for Group B (p=0.17 and 0.002).

Table 1 – Adalimumab dose and level

Group	Weight	No. of patients	Starting dose (mg)	Mean post induction ADA level (µg/mL)	Mean ADA level after dose increase (µg/mL)	No. of patients remaining on ADA	
A (17)	<40kg	3	40/20	6.36	n/a	1	10
		3	80/40			1	
	>40kg	11	80/40			8	
B (15)	<40kg	5	40/20	4.33	7.15	3	7
		1	80/40			1	
	>40kg	9	80/40			3	

Table 2 – Mean FCP and DAI score

Patient group	Baseline mean FCP (µg/mg)	Mean FCP post induction	Mean FCP after dose increase	Baseline mean DAI	Mean DAI post induction	DAI after dose increase
A	1832.82	620.06	n/a	17.19	15.31	n/a
B	1695.67	1516.20	960.00	21.00	17.00	13.33

In conclusion, a high proportion (47%) of IBD patients required dose escalation. Patients with higher DAI scores at induction could benefit from higher starting doses.

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OC34

Drug induced eosinophilic pneumonia in a child treated with ustekinumab for ulcerative colitis

by K. Sandhu, P. To, I.M. Balfour-Lynn, J.M.E. Fell, D. Thangarajah. *Chelsea and Westminster Hospital, London, SW10 9NH, UK.*

Background: Ustekinumab is known to cause respiratory side effects, with eosinophilic pneumonia (EP) a rare adverse event only reported in the literature in a few adult cases. We report on an 11-year-old female with ulcerative colitis (UC) with drug induced EP secondary to ustekinumab.

Methods/Case Presentation: An 11-year-old girl with UC who at 9 months from diagnosis was started on ustekinumab due to steroid dependency and anti-TNF failure. After a single loading dose of ustekinumab (6mg/kg) and concurrent prednisolone 0.5mg/kg, she presented within one week with dry cough, fever, night sweats and dyspneoa, raised inflammatory markers (CRP 167, ESR >120) and eosinophil count of 1.6 (range 0.9-1.3) and negative TB screen. CXR (figure 1) showed bilateral lung changes with tenting of diaphragm, antibiotics were started with no improvement after 1 week. After respiratory consult, CT (figure 2) was diagnostic of EP with reduced lung function, FEV1 (forced expiratory volume in 1 second) 52% and FVC (forced vital capacity) 48%. She was treated with 1.5mg/kg of prednisolone for 5 days. 2 weeks later there was resolution of chest symptoms and normal X-ray with improvement in lung function (FEV1 70%, FVC 72%). Immunology consult excluded immunodeficiency and secondary autoimmune conditions. Further steroids given for her UC led to improvement to FEV1 91% and FVC 88% which was maintained for a further year.

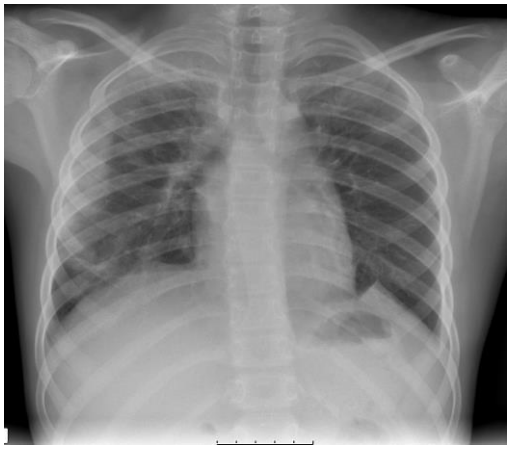

Figure 1: CXR showing dense air space opacification in right lung and left upper zone with tenting of left hemidiaphragm

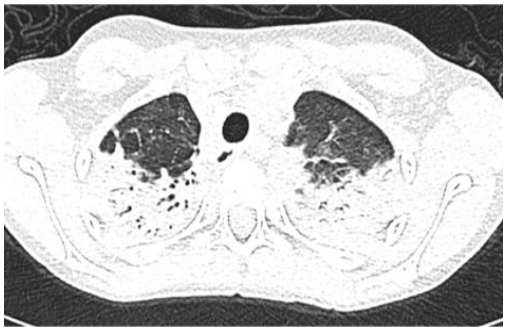

Figure 2: Bilateral upper lobe consolidation

Results/Discussion: We report the first paediatric case of ustekinumab induced EP. In the adult series similar presentation has been observed and with one case of patient had acute respiratory distress syndrome. Most cases occurred after prolonged use of ustekinumab, and fewer occurring after a single dose. It has been postulated the pathogenesis of EP is hypersensitivity reaction or upregulation of TH2 pathway due to ustekinumab blockade of IL-12 and IL-23.

Conclusion: EP is a rare and serious side effect of ustekinumab. We recommend avoiding further doses of ustekinumab, screening for opportunistic infections in the context of an immunosuppressed patient. In 2018, the food and drug administration approved a warning for ustekinumab highlighting the risk of non-infectious pneumonia. Our case highlights that children can present similarly to adults and resolved with a course of steroids.

OC35

Children with new onset IBD accessing paediatric endoscopy services in the post pandemic recovery phase are more transfusion dependent

by K Allan, N Francis, S Loganathan, S Kirkham and D Devadason, *Nottingham Children's Hospital, Derby Road, NG7 2UH, UK.*

Paediatric onset IBD often present with anaemia. The cause of anaemia is multifactorial and include blood loss, iron deficiency, chronic inflammation, malabsorption and vitamin deficiencies. Occasionally blood transfusion is required to ensure a patient is stable for any diagnostic endoscopic procedure under a general anaesthetic. The COVID-19 pandemic disrupted access for children requiring diagnostic endoscopy and has contributed to children presenting with increased levels of morbidity. There is paucity of data on the impact of the pandemic on paediatric IBD services.

To assess whether there has been a difference in the requirement of blood transfusions for children accessing diagnostic endoscopy services post-pandemic, compared with children accessing the service pre-pandemic.

This was a retrospective study looking at the records of new onset PIBD in two distinct 12-month periods; pre-pandemic (01/01/2019- 31/12/2019) and post-pandemic (01/09/2021-31/08/2022). Data collected included type of disease, age at diagnosis, pre-endoscopy haemoglobin, requirement of blood transfusions peri-endoscopy (defined as a week prior to up to 4 weeks following an endoscopy). Decision for transfusion was decided based on haemoglobin level and clinical stability. Results were analysed using an excel spreadsheet.

In the pre-pandemic period 44 children were diagnosed with new onset IBD. The mean age of diagnosis was 12y 4months (range 3 years 5 months to 17 year 11 months. A total of 4 patients required a transfusion pre-endoscopy. Each patient received 1 unit (mean units received 1.0; 3 UC, 1 CD, 0 IBDU). No patients required a blood transfusion within a month post endoscopy.

In the post-pandemic period 58 children were diagnosed with new onset IBD. The mean age at presentation was 12y 6months (range from 3 years 10 months to 17 years 4 months). A total of 11 patients required transfusion peri-endoscopy, requiring 17 units in total between them (mean units received 1.54; 7 UC, 2 CD, 2 IBDU). 9% of children required a transfusion in the pre-pandemic period vs 18.9% of children in the post pandemic period. Compared to the pre-pandemic period, there has been a threefold increase in the use of red cell products for children with new onset IBD undergoing endoscopy.

There appears to be a greater need for red cell transfusion in children undergoing endoscopy than was the case pre-pandemic. This may be the result of children waiting longer for diagnostic procedures and delayed presentations through usual pathways. Ongoing disruption in access to diagnostic endoscopy in children may also impact blood transfusion services and needs to be planned for in the recovery of paediatric endoscopy services.

OC36

A retrospective audit on the outcomes of nutritional therapy in a paediatric Crohn's disease cohort

by F. Mohamedhaji. A. Ocholi. *St George's University of London; St George's Hospital, London, UK.*

Introduction: Crohn's disease (CD) is a chronic, inflammatory bowel disease characterised by transmural inflammation that can affect any part along the gastrointestinal (GI) tract. Approximately 25% of patients diagnosed with CD have onset of disease in childhood[1].Diet has been found to strongly impact gut microbiota, with almost all children with CD presenting with nutritional deficiencies at diagnosis[2].Treatment of CD is largely directed at the induction and maintenance of remission. Paediatric patients with an established diagnosis of CD can receive nutritional therapy which is considered a first-line primary induction treatment with efficacy in achieving mucosal healing[3]. This can be assessed with highlighting any significant decrease of acute phase reactants, such as erythrocyte sedimentation rate (ESR), C reactive protein (CRP) and faecal calprotectin (FC).

Aims: 1. To review the practice of liquid nutrient therapy for Crohn's disease in a paediatric population. 2. To review the effectiveness of nutritional therapy in Crohn's disease

Methodology: A retrospective review was carried out using the data available on electronic hospital records. Cohort contained all paediatric patients with a confirmed imaging and histological diagnosis of CD from 2015 until 2022. The data included: patient demographics, dates of diagnosis, dates for start, mid-point and end review of nutritional therapy treatment course, site of diagnosis, weight percentiles and pathology results. Dates for commencing treatment were confirmed using Dieticians review notes. Patients' past meds and care plans were used to look for any prescribed medications post treatment and endoscopic and radiology scans were used to identify accurate GI sites affected. Data was then collected, filtered and compiled on to an excel spreadsheet.

Results: The overall cohort consisted of 125 paediatric patients with mean age of patients being 12 years old. Every patient was offered a nutritional therapy course a few days after diagnosis of CD. 57.6% of the cohort on nutritional therapy showed complete positive response to treatment while 42.4% did not show a positive response. 16.8% less patients reached normal levels of CRP compared to those who showed a reduction in CRP results whereas the difference in ESR results was 8%. Albumin has the greatest percentage difference (18.4%) between number of patients who normalised compared to those who showed a positive response. 71.2% of total patients achieved an increase in their weight (in kg). However in only 27.2% of total patients the increase in weight was significant enough to allow for an increase in patients weight percentile range. Nutritional therapy showed a more effective response in patients who had a site of disease of upper GI only (100%) and Ileal only (85.71%). It is unclear why 'Colon +Upper GI' and 'Ileal+ Upper GI' sites had a lower efficacy rate. However, this difference in effectiveness result could be limited by the small sample size of initial patients with those sites affected.

Conclusion: Patients diagnosed with Crohn's disease from 2015 to 2022 in receipt of nutritional therapy showed to have a majority effective response to the treatment. This highlights that the use of nutritional therapy for paediatric CD patients does work and continued use is beneficial. A statistical analysis of the data with a follow up period after end of treatment would be helpful in analysing long term effectiveness of nutritional therapy.

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OC37

BSPGHAN survey on the access and prescription of vedolizumab and ustekinumab in paediatric inflammatory bowel disease patients

by M.K.H. Auth *[1], J.J. Ashton*[2], K.D.J. Jones[33], K.Y. Lee[4], E.W. Swann[5], V. Garrick[6], A. Rodrigues[7], F. Torrente[8], P. Deb[9], H. Doble[10], R. Muhammed[11], T. Paul[12], T. Coelho[2], D. Thangarajah [13], V. Zamvar[14], P. Narula[15], A. Fagbemi[16], D. Devadason[17], H.S. Bhavsar[18], G. Lee[19], M. Ayaz[20], H.M. Lee[21] and J. Kammermeier[20] * Joint first authors. *From Paediatric Gastroenterology in, [1]Liverpool (&Univ.); [2]Southampton / Genomics Med.; [3]Great Ormond Street, [4]Bristol; [5]Edinburgh; [6]Glasgow; [7]Oxford; Cambridge; [9]Royal London [10]Pharmacy (Alder Hey); [11]Birmingham; [12]St. Georges; [13]Chelsea & Westminster; [14]Leeds; [15]Sheffield; [16]Manchester; [17]Nottingham; [18]Leicester; [19]Evelina; [20]Epsom & St Helier; [21]King's College.*

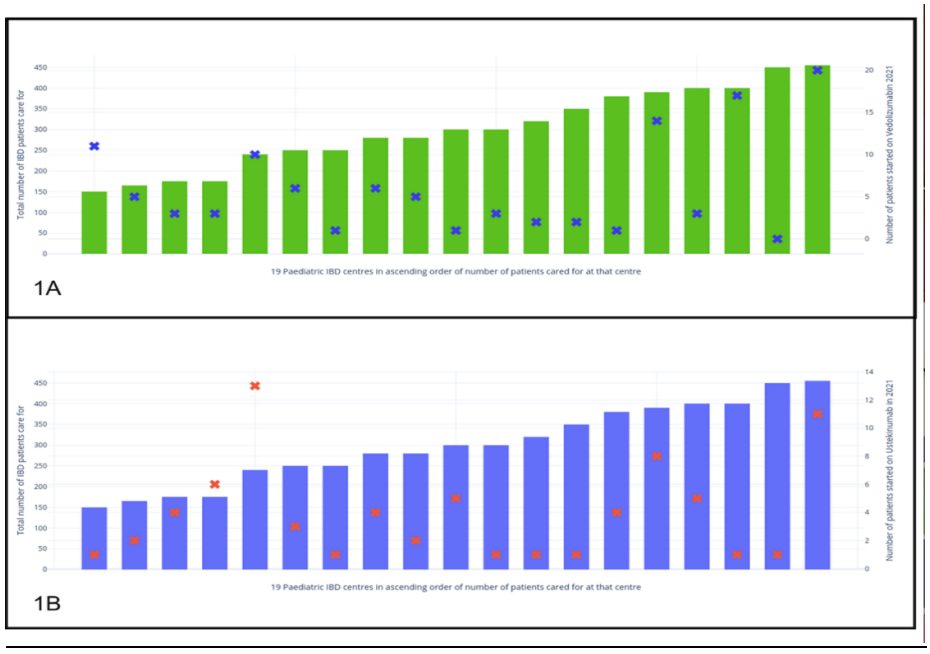
While the incidence of IBD is rising, therapeutic options for paediatric IBD are limited, especially for younger children. Unlike in adults, vedolizumab and ustekinumab are not generally licenced for paediatric use in the UK. We aimed to obtain data on the real-world access to, and use of, these therapies in the paediatric population.

We surveyed all 21 designated UK IBD centres to assess the incident use of vedolizumab and ustekinumab from 1/1/2021-31/12/2021. We collected information on funding, dose escalations and therapeutic drug monitoring (TDM) in an online questionnaire.

Covering an estimated 5710 patients, 19/21 centres responded,. One-hundred-and-thirteen were started on vedolizumab (Fig 1A) , prescription incidence 2%, median prescriptions per centre was 3 (range 0-20). Considering ustekinumab (Fig 1B) , 74 patients were commenced, prescription incidence 1.3%. Median prescriptions per centre was 3 (range 1-13). Prescription rates at each centre were not predicted by patient number cared for at that centre (p=0.34). Dose escalation was common in vedolizumab (66.7% centres) and ustekinumab (52.6%).

Funding strategies varied substantially, and multiple funding sources were used; 13/19 centres (68.5%) reported funding through routine NHS England/Scottish arrangements. There was local NHS trust funding in 8/19 centres (42.1%). Individual funding requests (IFRs) were used in 5/19 (26.3%), although IFRs are reserved for patients with unique additional characteristics. Four centres unable to achieve funding in pre-pubescent children.

Our survey shows that there is widespread use of vedolizumab and ustekinumab across the UK, although practice is highly variable. Access to therapy appeared to differ substantially. There is a growing disparity between real-world practice/feasibility and international guidelines/benchmarking. Establishing effective and early therapy in all eligible patients remains a priority.



OC38

Posterior reversible encephalopathy syndrome following immunosuppressive treatment for ulcerative colitis.

by S.P.F. Smith, A. Abdulla, Z. Zaidi, P. Deb, N. Croft, S. Naik, A. Kadir. *Paediatric gastroenterology, Royal London Hospital, Whitechapel Rd, London E1 1FR, UK.*

Posterior reversible encephalopathy syndrome (PRES) is a rare complication of immunosuppressive treatment in inflammatory bowel disease. PRES is a rare condition with neurological sequelae and characteristic findings on imaging.

To Discuss a case of PRES following a diagnosis of ulcerative colitis that was refractory to treatment.

An 11 year old boy was referred to a tertiary paediatric gastroenterology unit with a 6 month history of abdominal pain, bloody diarrhoea, 6kg of weight loss. Results showed a faecal calprotectin of 6831.
Endoscopy revealed severe pan-colitis with a normal terminal ileum. The patient was started on IV hydrocortisone 2.5mg/kg QDS and a 5-ASA. Unfortunately there was a suboptimal response to IV steroids. On day 5 the PUCAI remained high at 55. A decision was made to start infliximab. The patient remained on IV Steroids and high dose infliximab with refractory disease. Sigmoidoscopy was performed on day 21 which showed severe colitis and a granulated appearance. following this on day 22 a subtotal colectomy and end ileostomy was performed. Steroids were weaned following colectomy. Blood pressure was noted to be at the 95th centile, with some readings up to 160 systolic, with an associated headache and bradycardia. On day 2 post op, the patient presented in status epilepticus. This was managed initially on the ward but due to ongoing seizure activity the patient required transfer to paediatric intensive care for ongoing management. He was intubated and ventilated. MRI Brain scan revealed features in keeping with PRES.

Management of the PRES Syndrome included close control of hypertension, neuroprotection and anti-epileptic medications, and removal of possible risk factors for PRES - in this case the steroids and the infliximab. Follow up MRIs revealed improving PRES, and 2 small infarcts. The patient had some ongoing focal seizure activity so levetiracetam was continued.
The patient required a prolonged period of both neurological and nutritional rehabilitation, including physiotherapy, play therapy, parenteral nutrition and NG feeding. During this time his neurological status showed slow continuous recovery. The patient was discharged home 29 days after the diagnosis of PRES.
The patient was followed up in outpatient clinic and had made a good recovery and was neurologically back to baseline with the exception of some short term memory loss.

PRES syndrome is a severe life threatening neurological disorder, characterised by variable symptoms, which include visual disturbances, headache, vomiting, seizures and altered consciousness.
The Causation of PRES is not fully understood. Possible mechanisms of PRES include hypertension and endothelial injury. There is no direct treatment for PRES, management is with removal of the causative agent, control of hypertension, and stabilisation of the patient. Whilst most cases fortunately are reversible, timely and efficient management is vital to achieve this.
A review of the literature shows steroids and hypertension as a frequent causative mechanism of PRES. The association between PRES syndrome and infliximab is not well defined, however there are a few single case reports published of PRES syndrome in patients treated with either infliximab or Ustekinumab.

OC39

Inflammatory Bowel Disease (IBD) and Eating Disorders: Association or Causation, and Changing Practice after Covid 19

by A. Ibraheem, J. Virtue, P. Sukhtankar. *Gloucestershire Hospitals NHS Foundation Trust, UK.*

We present two young people presenting with IBD and eating disorders and discuss how our management may evolve to prevent eating disorders and support patients with both diagnoses adequately

Case 1: A 16-year-old girl attended clinic with 3-months weight loss, abdominal pain, loose and bloody stools ten times a day. We admitted her for intravenous fluids and 'gut rest'. She has severe anxiety, and her mother reports that she has not been eating well for over a year. Meals are restricted to once a day. 48h after admission a nasogastric (NG) tube was inserted and she remains NG fed for anorexia 4 months later. She has an endoscopic diagnosis of ulcerative colitis treated with prednisolone then azathioprine. Blood inflammatory markers are normal but calprotectin remains elevated.

Case 2: A 13-year-old boy with autism was diagnosed with Crohn's disease in 2021. Initial presentation was with nausea and spasmodic pain. At diagnosis he was noted to be tall for his age (190cm) and slim with BMI 9th centile. He was treated with prednisolone and gained weight rapidly. He remained withdrawn with refusal to get out of bed. 6 months after steroids were weaned his parents noted that he was not eating and was walking compulsively. They had thought his reduced intake was due to pain; he lost 10kg in 3 weeks. His calprotectin and blood tests were normal. On direct questioning he disclosed he was walking 20000 steps a day and restricting intake to lose weight. He is cared for at home by the community eating disorders team and currently does not attend school.

Discussion: These cases show that an eating disorder can make diagnosis and response to treatment in IBD challenging. Additionally, it is well recognized that IBD symptoms and recovery can contribute to developing an eating disorder[1] (crohnsandcolitisdietitians.com/disordered-eating-and-ibd). Case 1 would not have presented with anorexia had she not developed IBD, and it is now unclear whether her abdominal pain and loose stools are related to ongoing inflammation or to enteral feeds after starvation. Case 2 developed anorexia after regaining weight following treatment for Crohn's disease.
During the COVID 19 lockdowns eating disorders and new diagnoses of inflammatory bowel disease both increased for complex reasons including changes in intestinal permeability with Covid infection and the decline in mental health of young people due to pandemic responses. [2,3] It is clear from our cases (and others!) that eating disorders are more severe and complex than pre-Covid. It is widely recognized that IBD is a risk factor for disordered eating due to rapid weight loss, reconstitution of weight and often excessive weight gain with steroid treatment, as well as possible restricted foods in many cases may well predispose young people with IBD to developing eating disorders.

In the time following the Covid pandemic vigilance for eating disorders is needed to recognize these early and provide adequate support and resources for patients. We propose standard questions regarding eating and attitude to weight for all IBD patients attending outpatient clinic.

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European Journal of Pediatrics https://doi.org/10.1007/s00431-022-04773-5

OC40

Tofacitinib Rescue Therapy For New-onset Acute Severe Colitis: A Case Report

by M. Stoian, T. Bildstein, S. Chadokufa, V. Suri, F. Kiparissi and K. Jones. *Great Ormond Street Hospital for Children, Department of Paediatric Gastroenterology, Department of Gastroenterology/Mucosal Immunology, London, WC1N 3JH, UK.*

Tofacitinib, a **s**elective small molecule Janus Kinase (JAK) inhibitor, is well known in treatment of rheumatoid diseases and in recent years became increasingly important in managing adults with Inflammatory Bowel disease (IBD). In the paediatric population, data is scarce, however has proven to show rapid response of symptoms in moderate to severe Ulcerative Colitis (UC) after biologic treatment failure and/or prior to undergoing surgery (1,2,3). Here we describe a potent treatment of a 13-year-old with newly diagnosed unclassified IBD (IBD-U) and severe colitis with Tofacitinib.

The previously healthy male adolescent was diagnosed with IBD-U with a high Paediatric Ulcerative Colitis Activity Index (PUCAI) of 85 and severe colitis macroscopically and histopathologically. Initial OGD and colonoscopy showed macroscopical and histological chronic duodenitis, severe left sided chronic active colitis. Acute inflammation in the transverse, descending and sigmoid colon in the ultrasound was confirmed, the MR enterography showed additional 2cm focal narrowing in the Terminal Ileum. On day 7 of high dose intravenous steroids and on azathioprine, he presented with a persistent PUCAI above 60, weight loss of 2kg within two weeks of admission and ongoing rectal bleeding, therefore first line biologic treatment with intensified Infliximab regime, together with IV antibiotics were added and parenteral nutrition (PN) for gut rest was started. On endoscopical re-assessment on day 20 of steroid treatment ongoing chronic active colitis was seen. Because PUCAI stayed above 45 despite medical escalation and gut rest, potential need for colectomy was discussed. Following a multidisciplinary team discussion, trial of Tofacitinib for colonic salvage was initiated.

There was a rapid and dramatic response to oral Tofacitinib 10mg twice daily. PN was able to be stopped after 10 days since on Tofacitinib and within 28 days he gained 4.1kg of weight, PUCAI dropped from 45 to 0, ESR and CRP normalised (maximum 22 to 6mm/hr and maximum 9 to below 5mg/L, respectively), albumin normalised (lowest 31g/L, latest result 48 g/L) and significant improvement of faecal calprotectin (>10.000ug/g to 934 ug/g). After weaning off steroid completely, currently on Prednisolone 5mg once daily and Tofacitinib 5 mg twice daily, long-term plan is to bridge him to Vedolizumab.

In this case Tofacitinib supports previous data of prompt and effective results. JAK inhibitors are beneficial as rescue therapy for steroid non-responder and primary anti-TNFα non-responder and therefore can avoid major surgical intervention.

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OC41

Cohort study of paradoxical psoriasis in an adolescent population receiving anti-TNFα for inflammatory bowel disease (IBD) in a large teaching hospital with specialist dermatology and paediatric gastroenterology services

by O. Ogunmoye, N. Onyeador, N. Shareef, N. Reps, A. Abdul-Wahab. *Department of Paediatric Gastroenterology St George's University Hospital Trust, London, UK. Department of Dermatology, St George's University Hospital Trust, London, UK.*

Paradoxical psoriasis is well recognised in adults receiving anti-TNFα therapy for IBD [1]. Biologics are increasingly being initiated earlier in the treatment ladder for IBD in the paediatric population and they are also vulnerable to paradoxical psoriasis [2]. This observational study aimed to describe the clinical features and management in this group, as there is limited data on how paradoxical psoriasis differs in children.

8 patients with paradoxical psoriasis were referred between 2019-2022, comprising 5 females and 3 males and age range of 12-19 years (mean 15.4 years). They all had Crohn's disease and were receiving infliximab (7/8 patients) or adalimumab (1/8). The onset of symptoms following anti-TNFα initiation was 3 months - 3 years (average 12.75 months) similar to adults. Involvement included scalp (7), flexural (3), trunk (4) limb (3) and face (1). In adults, palmoplantar surfaces and a pustular morphology are most commonly reported. In our cohort, some presented with eczematous flares requiring anti-bacterial agents which is rarely seen in adults. Management of the paradoxical psoriasis included switching from infliximab to usetkinumab in 3 patients; addition of methotrexate (2). Topical treatments alone were sufficient in 3 patients.

There were florid clinical presentations in our cohort and the scalp predominantly affected (7/8) which tended to be severe. 5 of 8 patients required a switch of their anti-TNFα therapy or an additional systemic treatment. It is crucial for physicians to be aware of this phenomenon and work collaboratively to recognise this early to optimise patients' symptoms and quality of life.

References:

(1) Bucalo et al, 'PP induced by anti-TNFα treatment: evaluation of specific clinical and genetic markers' [2020]
(2) Courbette et al, 'Infliximab PP in Children with IBD' [2019]

OC42

Common, non-pathogenic variants in genes from monogenic disorders in children confer additional risk of liver injury later in life

by J. P. Mann[1], P. N. Newsome[2], Y. H. Oo[2] and G.L. Gupte[1]. *[1]Liver Unit (including small bowel transplantation), Birmingham Women's and Children's NHS Foundation Trust, Birmingham, B4 6NH, UK. [2]Centre for Liver Research, University of Birmingham, Birmingham, B15 2TT, UK.*

Monogenic liver disease is caused by rare, pathogenic mutations. Exome and genome sequencing frequently identifies variants of unknown significance in these genes. It is not clear whether such non-pathogenic variants confer increased risk of liver injury beyond childhood. Here, we found that these variants increase the severity of liver damage and may act as a 'second hit'.

We identified 77 monogenic paediatric liver diseases. For each gene, we searched for evidence of a liver phenotype in individuals not known to have genetic disease using population-based datasets. We identified genome-wide significant associations ($p<5x10^{-8}$) between variants (e.g. single nucleotide polymorphisms) and liver biochemistry (ALT, bilirubin, GGT, ALP) in n=1,654,950 participants from the Common Metabolic Disease Portal and n=394,841 from UK BioBank using GeneBass.

We found 89 genome-wide associations for biomarkers of liver injury in otherwise apparently healthy individuals across genes from 44/77 (57%) monogenic disorders (**Fig 1**). For example, common variants in *ABCB11* (the cause of PFIC type 2) were associated with GGT ($p=2.0x10^{-33}$) and ALT ($p=8.4x10^{-39}$). Similarly, common polymorphisms in *JAG1* (that do not cause Alagille's syndrome) were associated with GGT ($p=2.3x10^{-9}$) and ALT ($p=5.9x10^{-10}$).

Significant associations were found most frequently for 5/7 (71%) of cholestatic disorders and 5/7 (71%) bile acid metabolism disorders, compared to 3/8 (38%) of congenital fibrotic disorders.

In addition to affecting liver enzymes, serum lipid profile (e.g. total cholesterol) was affected by genes from 23/44 (52%).

Common variants in genes that cause rare monogenic liver disease also confer a risk of liver injury later in life. Understanding the mechanisms of these genes provides an opportunity for recognition and treatment of common liver diseases.

OC43

Transaminitis in paediatric practice

by T. Dunne, M. Roberts, M. Al-Jubouri and A Elbadri. *St. Helens and Knowsley Teaching Hospitals NHS Trust, Warrington Road, Rainhill, Prescott, L35 5DR, UK.*

Transaminitis (elevated liver enzymes) is a common, often incidental, finding in paediatric practice, frequently found when investigating non-hepatic complaints. Alanine aminotransferase (ALT) is one such enzyme. It predominantly resides in hepatocytes and plays a major role in amino acid metabolism and gluconeogenesis. Hepatocellular injury leads to release of ALT, increasing its serum concentration, making it a marker of liver damage. Investigations of liver problems can be complex.

A quality improvement audit project was devised to study the assessment, management, and outcomes of transaminitis (raised ALT) in a busy paediatric unit in a large district general hospital to inform the production of a standardised and streamlined transaminitis guideline.

We retrospectively audited children (1-16 years) with elevated serum ALT identified between June 2019 and June 2021. Exclusion criteria were transaminitis on neonatal prolonged jaundice screening, children who passed away and those with unavailable records. Data were collected using a special audit tool and analysed using Microsoft Excel and SPSS.

Total 99 children met the inclusion criteria; most were ≥11y (53.1%) followed by 6-10y (20.4%). Children were categorised into one of 10 presentation groups with nearly 2/5 being 'acute non-febrile illness with no clinical evidence of liver disease' followed by 1/4 being 'acute febrile illness with no clinical evidence of liver disease'. Only 3 children had an acute presentation with clinical evidence of liver disease. Mean initial ALT was 255 (mode = 57, range = 50-5896) and mean peak ALT was 397 (mode = 57, range = 50-11900). Around 80% of initial and 70% of peak ALTs were <200. Only 2/5 of children had any aetiological investigations on initial presentation. In 64% of presentations an aetiological diagnosis was not reached; the most common diagnoses were Epstein-Barr Viral infection, fatty liver disease and secondary to medication use, respectively. About 60% of patients with elevated ALT had normal levels on repeat testing within 1 month, with only 13% persisting beyond 6 months.

In conclusion, most transaminitis was mild (<200), with greater ALTs corresponding with clinical evidence of liver disease being present. Approach to investigation and management was variable, with differing aetiological investigations performed at different points following initial recognition. It was common to not reach a diagnosis for elevated ALT and to not follow-up until complete resolution. It was noted that 7 children were diagnosed as fatty liver disease on BMI and USS findings without the full panel of aetiological investigations recommended by the British Society of Paediatric Gastroenterology, Hepatology and Nutrition.[1] This work has highlighted a variable approach to this common presenting issue. We would recommend working towards a standardised approach or guideline both to ensure important underlying clinical conditions are not missed but also children are not exposed to unnecessary investigation.

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OC44

Prevalence of secondary antibody deficiency following paediatric intestinal transplant and intravenous immunoglobulin use

by K. Xenofontos[1], J. Crook[2], W. Lau[1], C. Bambridge[3], J. Hind[3] and D. Hazdic[3]. *[1] University College London, Gower Street, London, WC1E 6BT, UK. [2] Pharmacy Department, King's College Hospital NHS Foundation Trust, Denmark Hill, SE5 9RS, UK. [3]Paediatric Liver Unit, King's College Hospital NHS Foundation Trust, Denmark Hill, SE5 9RS, UK.*

Secondary antibody deficiency (SAD) is a severe complication following intestinal transplant (ITx)[1]. The aim of this study is to describe the prevalence of SAD in children following ITx, describe their clinical presentation, including those who received intravenous immunoglobulin (IVIG).

A single-centre retrospective observational case series of paediatric patients (age <18 years) who had ITx between August 2009 and October 2021 at specialist paediatric unit in England. Deceased patients were excluded. Descriptive statistics were performed. SAD was defined as immunoglobulin G (IgG) levels below the normal reference range for age. The audit was registered locally, and ethical approval was not required.

17/28 ITx patients were included in the analysis (age 10months – 13years). All patients received similar levels of immunosuppression post-transplant. 10/17 patients (58.8%) developed SAD within four months following ITx, 1 patient had pre-existing SAD prior to transplantation. 6/10 (60%) SAD patients and 6/7 (86%) without SAD contracted at least one infection post-transplant. Eight patients received IVIG during the study period, 3 patients received IVIG for SAD, 2 within one month of transplant and 1 pre and post-transplant.

The median fluid loss in all patients, SAD group and IVIG group was 24ml/kg (inter quartile range (IQR) 17-60), 21.2ml/kg (IQR 13-33) and 29.4 ml/kg (IQR 22-36) retrospectively. The average length of stay for all patients, SAD group and IVIG group was 13 (3-71), 10 (3-27) and 37 (13-71) intensive care (ICU) bed days and 62 (28-190), 65 (28-190) and 127 (63-190) total hospital bed days retrospectively.

The prevalence of SAD in this cohort was higher in comparison to those previously reported following SOT[2,3,4]. High fluid losses and increased length of ICU stay may predispose patients to lower IgG levels post ITx. Routine IgG monitoring post ITx should be considered alongside future studies of B cells and functional antibody assays.

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OC45

Maralixibat improves growth in patients with Alagille syndrome: A 4-year analysis

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Maralixibat (MRX) is an ileal bile acid transporter inhibitor (IBATi) recently approved by the Food and Drug Administration for the treatment of cholestatic pruritus in patients with Alagille syndrome (ALGS) 1 year of age and older. Recent data suggest that MRX is associated with improved event-free survival in this population suggesting that the drug may potentially improve liver disease outcomes beyond pruritus control in ALGS. Patients with ALGS clearly have impaired growth and therefore we evaluated the impact of long-term MRX treatment on the nutritional status of these patients.

Height and weight Z-scores were evaluated in patients who participated in 3 clinical trials of MRX and their long-term, open-label extensions for treatment of cholestatic pruritus in ALGS; only patients for whom we had height and weight data at baseline and week 204 follow-up were included. T-tests and Pearson correlation coefficients were used to evaluate the association between height with other parameters known to correlate with growth.

Data were available for 34 patients with baseline mean (SD) age of 6.7 (3.8) years, height Z-score of -1.66 (1.17) and weight Z-score of -1.46 (0.95). Overall, mean height Z-score increased to -1.29 (1.03) at week 204 (change from baseline: 0.37; p=0.0004). The greatest catch-up height gain was observed among those within the lowest baseline quartile height Z-scores, increasing from -3.1 (0.71) at baseline to -2.38 (0.82) at week 204 (change from baseline: 0.72; p=0.018), and there was a significant correlation between change in height and baseline height (r=-0.48; p=0.004). Similarly, greater catch-up weight gain was observed with lower baseline weight Z-scores, with a significant correlation between change in weight and baseline weight (r=-0.39; p=0.02). The change in height Z-scores correlated with the change in weight Z-scores such that greater catch-up linear growth was observed in patients with greater catch-up weight gain (r=0.73; p<0.0001). There was no clear change in vitamin D levels or albumin throughout treatment. Among patients with sBA <200 µmol/L at week 48, height Z-score increased from -1.58 (1.23) at baseline to -1.16 (1.00) at week 204 (change from baseline: 0.42; p=0.001), whereas there was no significant change in height Z-score among patients with sBA ≥200 µmol/L.

Catch-up height and weight are observed in patients with ALGS treated with MRX, and importantly patients with the greatest height disadvantage at baseline had the greatest catch-up in height. Increased catch-up height was also seen in patients that achieved lower sBA with MRX, suggesting an improvement in bile acid homeostasis may be a factor. Further analyses are needed comparing growth trajectories in MRX-treated patients to a natural history cohort of patients with ALGS to fully understand the attributability of MRX.

OC46

Native liver survival in odevixibat serum bile acid responders: Data from PEDFIC studies in patients with progressive familial intrahepatic cholestasis

by R.J. Thompson[1], C. Clemson[2], V. Valcheva[2], E. Sturm[3], Q. Ni[2], Q. Yu[2], J.P. Mattsson[2], and H.J. Verkade[4], *[1]Institute of Liver Studies, King's College London, London, UK WC2R 2LS; [2]Albireo Pharma, Inc., Boston, MA, USA; [3]Paediatric Gastroenterology and Hepatology, University Children's Hospital Tübingen, Tübingen, Germany; [4]Department of Paediatrics, University of Groningen, Beatrix Children's Hospital/University Medical Centre Groningen, Groningen, the Netherlands.*

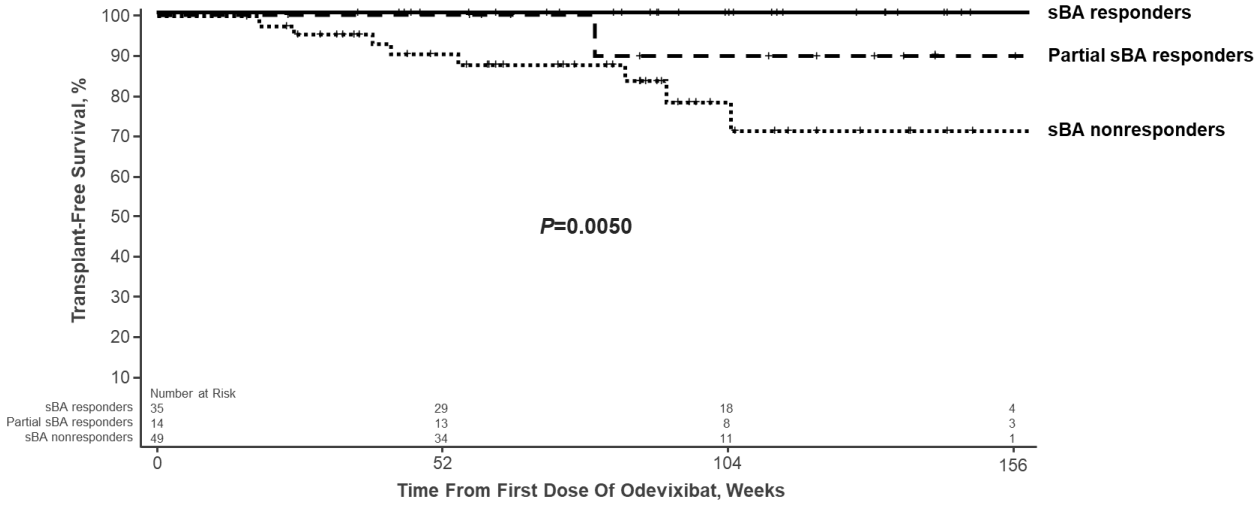
Patients with progressive familial intrahepatic cholestasis (PFIC) may have continued hepatic damage leading to liver transplantation (LT). Efficacy and safety of odevixibat, an ileal bile acid transporter inhibitor, were assessed in patients with PFIC in the phase 3 PEDFIC 1 and PEDFIC 2 studies. In a pooled analysis of data from these studies, we analysed native liver survival (NLS) in odevixibat-treated patients who met serum bile acid (sBA) treatment response criteria (sBAs reduced ≥70% or levels ≤70 µmol/L at 6 months). NLS was also analysed in partial sBA responders (patients with sBA reductions ≥30% to <70% at 6 months) and nonresponders (patients with sBA reductions <30% at 6 months or who underwent LT or discontinued treatment before 6 months).

PEDFIC 1 was a 24-week, randomised, placebo-controlled study in children with PFIC1 or PFIC2. PEDFIC 2 is an ongoing 72-week extension study in patients of any age with any PFIC type. This pooled analysis spans from patients' first dose of odevixibat to a cut-off date of 31 January 2022.

Of 98 patients analysed (mean treatment duration, 88 weeks), 35 (36%) were sBA responders, 14 (14%) were partial sBA responders, and 49 (50%) were nonresponders. Mean sBA reductions at 6 months were 87% in responders and 44% in partial responders; there was a mean increase of 27% in nonresponders. All 35 sBA responders and 13 of the 14 partial sBA responders remained transplant free; 8 of the 49 nonresponders underwent LT (**Figure**). sBA responders had mean improvements at week 24 of treatment vs baseline in alanine aminotransferase and total bilirubin levels.

sBA decreases at 6 months were strongly associated with NLS for up to 3 years in odevixibat-treated patients with PFIC.

Figure. Native Liver Survival in Serum Bile Acid Responders, Partial Responders, and Nonresponders to Odevixibat in the PEDFIC Studies



P value is based on log-rank test (serum bile acid responders vs nonresponders). +, censored; sBA, serum bile acid.

Disclosures:

R.J. Thompson: Albireo and Mirum – Consultant; Generation Bio – Consultant and stock options; Rectify Therapeutics – Consultant and stockholder
C. Clemson, V. Valcheva, Q. Ni, Q. Yu, and **J.P. Mattsson:** Albireo – Employment
E. Sturm: Albireo and Mirum – Consultant and research support; Univar – Consultant; Orphalan – Speaker's fee
H.J. Verkade: Ausnutria BV, Albireo, Danone/Nutricia Research, Intercept, Mirum, Orphalan, and Vivet – Consultant

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OC47

Odevixibat treatment in a patient with undefined cholestasis and no unified genetic diagnosis: A case report

by T. Grammatikopoulos[1,2], B. Tucker[2], L.S. Casale[3], P. Nagraj[4], V. Valcheva[4]. *[1]Institute of Liver Studies, King's College London, London, UK. [2]Paediatric Liver, GI, and Nutrition Centre and MowatLabs, King's College Hospital NHS Trust, London, UK. [3]Gibraltar Health Authority, Gibraltar. [4]Albireo Pharma, Inc., Boston, MA, USA.*

Cholestasis, a condition where bile flow is disrupted, has clinical features such as jaundice, severe pruritus, and elevated levels of serum bile acids (sBAs) and other hepatic parameters. Progressive familial intrahepatic cholestasis (PFIC) is a group of cholestatic liver diseases with known genetic aetiologies. However, some patients with cholestasis have symptoms consistent with PFIC but ambiguous genetics. Odevixibat, an ileal bile acid transporter inhibitor, diverts intestinal bile acids away from the liver.

We describe a case of a patient with undefined cholestasis and no unifying genetic diagnosis who was treated with odevixibat.

Within months of birth, a male patient with prolonged jaundice and raised liver parameters (**Table**) was found to have mild expansion of interlobular portal tracts and giant cell hepatitis with canalicular cholestasis on liver biopsy. Genetic sequencing identified a heterozygous mutation in *AKR1D1* and common benign homozygous mutation of *ABCB11*. By age 5, the patient had progressive liver fibrosis and continued abnormal liver function tests (**Table**), including elevated sBAs with severe pruritus and sleep disturbance that were refractory to conventional treatments (ie, rifampicin, ursodeoxycholic acid). At age 6, odevixibat (800 µg/day) was started for cholestasis and pruritus. Within 6 months of treatment, there was complete resolution of pruritus and dramatic improvements in his sleep. sBAs decreased from 413 µmol/L to 45 µmol/L at latest follow-up.

In a patient with undefined genetic cholestasis, odevixibat effectively resolved pruritus, improved sleep disturbance, and reduced sBAs.

Table. Hepatic Parameters Over Time in a Patient With Undefined Cholestasis and No Unifying Genetic Diagnosis

Time Frame	ALT, IU/L	AST, IU/L	GGT, IU/L	Total Bilirubin, µmol/L
Within months of birth	307	274	158	120
Age 5 (before treatment)	219	154	230	70

ALT, alanine aminotransferase; AST, aspartate aminotransferase; GGT, gamma-glutamyl transferase.

Disclosures:

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B. Tucker: Nothing to disclose
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OC48

Odevixibat therapy in patients with FIC1-deficient progressive familial intrahepatic cholestasis and severe diarrhoea following liver transplantation: A retrospective case series

by G.F. Vogel[1,2], E. Lainka[3], S. Kathemann[3], D. Aldrian[1], P. Rauschkolb[4], C. Maucksch[4], V. Valcheva[4], and C. Clemson[4]. *[1]Department of Pediatrics, Medical University of Innsbruck, Innsbruck, Austria. [2]Institute of Cell Biology, Medical University of Innsbruck, Innsbruck, Austria. [3]Department of Pediatric Gastroenterology, Hepatology and Liver Transplantation, University Children's Hospital, Essen, Germany. [4]Albireo Pharma, Inc., Boston, MA, USA.*

Features of progressive familial intrahepatic cholestasis due to deficiency in FIC1 (encoded by *ATP8B1*; PFIC1) may include disrupted bile acid handling, pruritus, poor growth, other extrahepatic manifestations, and progressive liver disease that can necessitate liver transplantation (LT). While certain symptoms of PFIC1 may resolve after LT, patients frequently have persistent malabsorption, diarrhoea, and failure to thrive that can impact quality of life. In this case series, we present clinical details of 3 patients with PFIC1 and post-LT diarrhoea that impacted daily activities who received odevixibat, an ileal bile acid transporter inhibitor, in clinical practice.

Retrospective patient data were collected by treating physicians using standardised forms and included demographic, clinical, and treatment information.

Data from 3 male patients with post-LT diarrhoea were collected. Prior to LT, patient symptoms included cholestasis, elevated serum bile acids, pruritus, dystrophy, and/or vitamin deficiencies. Patients experienced symptoms ranging from 1.8 to 4.8 years before LT and had unsatisfactory responses to conventional medical therapies. All patients underwent split LT, and common indications for LT included cholestasis, dystrophy, and intractable pruritus. After LT and prior to odevixibat initiation, patients had steatosis (patients 2 and 3), inflammation (patient 3), and diarrhoea (all patients). Patient 3 underwent surgical biliary diversion at 4 years post-LT, which resolved the steatosis and inflammation. Post-LT diarrhoea impacted the daily life and/or school functioning of all patients. Attempts to treat the diarrhoea with cholestyramine (patients 2 and 3) and Oralpädon (patients 1 and 2) were unsuccessful, with unresolved diarrhoea as the reason physicians cited for initiating odevixibat. After odevixibat initiation, patients had less-frequent and/or firmer stools at last available assessment, as well as improvements in aspects of daily life.

In patients with FIC1 deficiency, chologenic diarrhoea after LT, which may be due to physiologic bile acid levels in the FIC1-deficient bowel, can be a frequent and severe symptom. The real-world data presented here indicate that odevixibat can improve diarrhoea and quality of life in patients with PFIC1 and severe post-LT diarrhoea.

Disclosures:

G. F. Vogel: Takeda and Albireo – Consultant; Albireo – Scientific grant
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OC49

The role of therapies in early intervention in young children with liver disease

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Children with liver disease, and those who undergo liver transplantation, are at significant risk of developmental deficits across all areas; motor, cognitive, language and behaviour. Early childhood adversity may have an impact on the developing brain with lasting consequences throughout childhood and into adulthood. With improved survival rates following liver transplantation in the paediatric population the emphasis is on children 'not only surviving but thriving', with focus on quality of life, functional abilities and social/emotional wellbeing for these children. The wider multi-disciplinary team, such as Physiotherapy, Occupational Therapy and Speech and Language Therapy have an important role in optimising functional abilities. Here we present 2 children with biliary atresia, a progressive fibro-obliterative cholangiopathy of infancy that frequently leads to end-stage chronic liver disease and liver transplantation. They were assessed by physiotherapy before and after liver transplantation.

Child A was assessed at 5.5 months (PELD score 33.1). Weight and length z-score was -2.68 and -2.35 respectively. He had gross motor developmental delay; had not developed head control, or intermittent sitting balance and had difficulty with transitioning. Progressing well in social and fine motor aspects of development. The parents were given advice in regards to adapting tummy time to help with strengthening and help to develop head control and in how to promote transition between positions. He was transplanted at the age of 8 months after a period of steady deterioration. Post-transplant he stayed in hospital for 46 days, 16 of these he was intubated. Post-transplant he had a portal vein thrombosis and recently post-transplant lymphoproliferative disease requiring chemotherapy. He is now 13 months old, is sitting unsupported but not pulling to stand or walking.

Child B was assessed at 11 months (PELD 13.5, weight and length z-score -0.42 and 0.41respectively) and found to have mild gross motor delay with no concerns about fine motor, cognition and language. He was unable to sit in a high chair due to poor sitting balance. A referral was made to local community therapy. At 16 months (PELD 13, weight and length z-score 0.7 and 0.74 respectively) developmental progress in all areas of development was noted, but still had global developmental delay, predominantly gross motor; not yet pulling to stand or furniture walking. He had good transitioning to crawling, and crawling ability. Following liver transplant at 17 months of age, he stayed in hospital for 23 days, only 2 days in critical care. After transplant he had 2 episodes of mild rejection.He received therapy input in the acute recovery phase and on discharge at 17.5 months he had regained his baseline developmental skills.

These two cases illustrate how advanced liver disease impacts development and outcomes after liver transplant. Adult data is supporting the role of pre-habilitation whilst waiting for transplant. Assessing these children early and systematically and using a multi-therapy approach may have a significant role in supporting them and their families both pre and post-transplant. A period of bespoke pre-habilitation whilst awaiting transplant can help optimise developmental and functional outcomes.

OC50

Severe acute liver failure with a *de novo* missense variant in *EIF2B1*

by C. Ashton, K. Jayaprakash, M. Mtegha, S. Karthikeyan and S. Rajwal. *Paediatric Hepatology, Leeds Children's Hospital, Great George Street, Leeds, LS1 3EX, UK.*

The EIF2B1 gene is a protein coding gene which has been implicated in pathways for insulin translation and the cellular stress response. Previously described patients with novel EIF2B1 heterozygous variants presented with neonatal onset diabetes and transient liver dysfunction. Rarely it can result into fatal acute liver failure. We describe a rare case of a 2 year old male with a known heterozygous missense variant in *EIF2B1* (NM_001414.3: c.131G>A; p.(Gly44Asp)). Location: GRCh37 (hg19) – Chr12:g.124115065) who presented with recurrent fulminant liver failure which unfortunately was fatal.

This patient was born at term (39 weeks) with no dysmorphic features to non-consanguineous parents. At 2 months old he presented neonatal diabetes and liver dysfunction (ALT 1907) which recovered.

At 2.5 years old, he presented with fever and vomiting and was discharged home. He presented two days later with hypoglycaemia, acute liver failure and renal impairment. He was transferred to intensive care with INR 5.6, ALT 956 and lactate 3.5. His illness progressed to multiorgan failure making him unsuitable for liver transplantation.

Over three months he recovered from acute liver failure with supportive treatment (ALT 52, INR 1.2). CT head confirmed a severe hypoxic cerebral insult needing intensive neuro rehabilitation. In hospital he developed a further episode of acute liver failure after three months (ALT 700, INR 3.5) with a rapid deterioration in intensive care which proved fatal.

This case highlights the spectrum of presentation of EIF2B1 variants. Parents need to be counselled about risk of recurrent acute liver failure with subsequent fatality.

OC51

To determine pneumococcal antibody levels, dispel myths and explore UK vaccination policy in children with coeliac disease

by D.G. Cairney[1], A. Richards[2], K. Hogg[1], E. Furrie[3] and P. Gillett[1]. *[1]Paediatric Gastroenterology, Hepatology and Nutrition, Royal Hospital for Children and Young People, Edinburgh, EH16 4TJ, UK. [2]Medical School, University of Exeter, Exeter, EX1 2HZ, UK. [3]Blood Sciences Department, Ninewells Hospital and Medical School, Dundee, DD1 9SY, UK.*

Pneumococcal infection is notifiable in the UK and is rare in paediatric coeliac disease (CD) patients[1]. Vaccination in the UK began in 2006 with the conjugate vaccines PCV-7 in 2006 and PCV-13 (Prevenar) from 2010, given at 2, 4 and 12 months and now a two-dose schedule (from 2020). The COVID-19 crisis escalated parental and healthcare concern about viral infections, subsequent risk of secondary infection, underlying risk of hyposplenism and pneumococcal immunisation and its role in reducing infection. We looked at vaccine response in a group of paediatric coeliac disease patients to help alleviate those concerns.

Consecutive patients attending for CD follow-up at a regional tertiary service in Scotland had pneumococcal antibody levels measured as part of their routine blood monitoring and standard care. Electronic records of these patients were retrospectively reviewed to identify immunisation dates, potential episodes of pneumococcal infection, positive microbiology or evidence of hyposplenism.

46 (73%) were female. Median age at CD diagnosis was 9 years. Median time from immunisation to CD diagnosis was 6.5 years (range -3 to 14 years). Six patients (born before 2006) had never received immunisation. No patients had antibody levels less than 20 units/ml.

Pneumococcal antibodies were measured in 63 patients from July 2020 to February 2021. 46 (73%) were female and median age at CD diagnosis was 9 years. Median time from immunisation to CD diagnosis was 6.5 years. The lowest antibody level was 21 units/ml and there was no significant difference in antibody level between children who had received 0, 1, 2 or 3 doses of immunisation. No patients had hospital presentations or positive microbiology samples suggestive of pneumococcal infection and none had evidence of hyposplenism.

All patients had levels above 20 units/ml (this represents a ‘protective level’ in unimmunised and PPV-23 immunised patients). The protective level post PCV-13 immunisation is unknown. We assume (as with the general population) that lifelong immunity develops and boosters should not be required. Immune memory theoretically should be better with conjugate vaccines, but longer-term antibody response in CD has not been definitively investigated. It is currently recommended that unimmunised CD patients receive PPV-23 (Pneumovax) then lifelong 5-yearly boosters[2] despite ‘hyposplenism’ affecting 30% of adult CD patients at most. Uptake and understanding of the optimal schedule is poor in the CD community. Further detailed evaluation of vaccination response for CD patients is required for clarity of patient protection and avoidance of potentially unnecessary and expensive immunisations.

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OC52

Trientine is a safe and effective treatment for Wilson's disease

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Wilson's disease (WD) is an autosomal recessive disease caused by a defect in the ATP7B gene, which encodes a metal P-type ATPase responsible for trans-membrane copper transport within hepatocytes (1). Standard treatment of Wilson's disease includes copper chelators such as penicillamine and Zinc. Penicillamine is used first line, however 30% of patients discontinue therapy due to adverse effects (2). Trientine is indicated in WD patients who are intolerant to penicillamine.

We undertook a retrospective review of our centre's WD patients aiming to describe use of trientine. 25 WD patients were identified. 16, 2 and 1 patients were on pencillamine , zinc and zinc + penicillamine treatment respectively. 3/25 were on trientine. 3/25 who presented with acute liver failure underwent liver transplantation.

Trientine group:

All three children presented with deranged liver function tests (LFTs). All were started on Penicillamine initially. Reasons for switching were drug-induced hepatitis, inadequate response to penicillamine and proteinuria. No patients reported side effects from trientine.

	Age at Diagnosis (years)	Duration of penicillamine treatment (years)	Duration of Trientine Treatment (years)	Median (Range) ALT	
				Pre-Trientine	Post-Trientine
Patient 1	13	1.2	3.3	220 (182 - 260)	110 (95 - 244)
Patient 2	11	2.9	2.2	36 (18 - 120)	153 (128 - 164)
Patient 3	4	1.1	2.3	234 (94 - 381)	306 (91 - 652)

Table 1: Characteristics of patients treated with trientine

Patient 1 was also noted to be non-compliant with gluten free diet for coeliac disease which may have contributed to their raised ALT.

In conclusion, trientine is effective as Penicillamine in treating Wilson's disease with good tolerability.

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OC53

The role of MAIT cells in children with autoimmune liver disease

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Mucosal-associated invariant T (MAIT) cells, defined as CD3+ Valpha7.2+ CD161++ T lymphocytes, are central in the immunosurveillance, tissue maintenance and repair at mucosal sites[1,2]. They are considered integral in protecting the biliary mucosa from microbiota due to their predominant location around the portal tracts[2,3]. In autoimmune liver disease (AILD), MAIT cell cytokine production of interferon gamma (IFNγ), tumour necrosis factor alpha (TNF-α) and granzymes are significantly reduced compared to healthy subjects[1,2,3]. Furthermore, chronic MAIT cell activation predisposes the liver to the profibrotic state which was shown to be interleukin-17A (IL-17A) dependent in AILD[3]. The biological characteristics and functional activity of MAIT cells in children with AILD has not been investigated. We performed mass cytometry by CyTOF (Cytometry by Time of Flight) on peripheral blood mononuclear cells (PBMC) from children with AILD (AIH type 1, N=8, median age 14yrs (range 9-14), and PSC, N=8, 14yrs (range 12-15) and in healthy children (HC) (N=8, 12yrs (range 5-15)). PBMCs treated with a cell stimulation cocktail of phorbol 12-myristate 13-acetate (PMA) and ionomycin were incubated for 4hours prior to downstream CyTOF investigation. Untreated paired PBMCs from the same patients were used as controls. MAIT cell and conventional CD3+ cytokine production, measured as the median metal intensity (MMI), were recorded. Our results show stimulated (STIM) blood MAIT cells from children with PSC had higher TNF-α production than AIH patients and healthy children (Figure 1). Reduced MAIT cell Granzyme B and IFNy expression were observed from the AIH and PSC cohorts compared to HC. IL-17A production was induced in the 4hour incubation period, an observation not previously reported in adult MAIT cells which requires longer activation. The patients' CD3+ T lymphocyte cytokine expression levels were also assessed and are presented for comparison. Of note, although the trend of CD3+ TNF-α production is similar, it is 20-fold less than observed in MAIT cells. No CD3+ IL-17A production is observed in all three cohorts. In conclusion, reduced IFNγ and Granzyme production in our paediatric AILD cohort is consistent with MAIT cell dysfunction described in adult AILD. However, higher MAIT cell TNF-α and IL-17A production in the PSC cohort following acute activation suggests TNF superfamily pathway preservation and a greater potential for early release of the chronic inflammatory cytokine IL-17A, both of which may contribute to autoimmune liver disease progression.

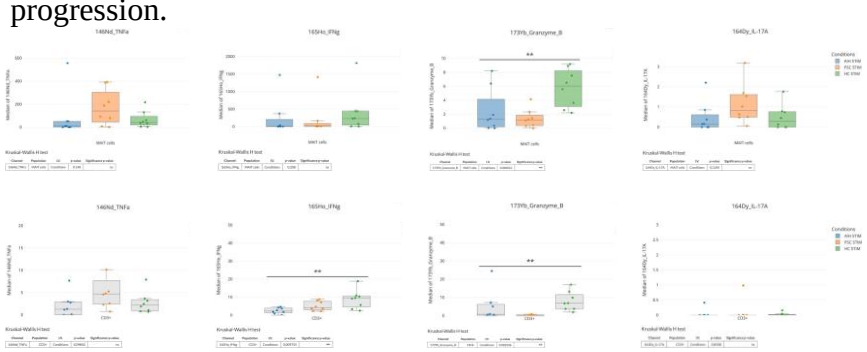

Figure 1. Box – and- whisker plots showing the MAIT cell post-stimulation (STIM) response in paediatric patients with AIH N=8, PSC N=8 and in healthy children N=8. MAIT cell expression levels of the cytokines TNF-α, IFNy, Granzyme B and IL-17A are displayed in the top row. Their paired CD3+ post-STIM responses are shown in the bottom row.

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OC54

Children with coeliac disease have a low risk of reduced bone mineral density

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The objectives of this study are to determine the incidence of reduced Bone Mineral Apparent Density (BMAD) in children diagnosed with coeliac disease, to identify risk factors for reduced BMAD and to quantify seroconversion rate to negative IgA anti-tissue transglutaminase antibodies (TTG) on gluten-free diet (GFD).

A list of coeliac patients is kept by the gastroenterology department. Relevant information was extracted from patient records. All children diagnosed between 2004-2021, with > 1-year follow-up, were included. GraphPad Prism (San Diego, CA) was used for univariable logistic regression analysis.

179 (118 F) patients were identified. Mean age at diagnosis is 7.6 years. (Range 11 months – 17 years). 133 patients underwent at least one DEXA scan. At first scan, 100 children had normal BMAD (Z score ≥-1), 19 had osteopenia (Z score <-1 and ≥ -2) and 13 had osteoporosis (Z score <-2). At their latest scans, 3 still had osteopenia and only one still met the criteria for osteoporosis. The median interval between starting gluten-free diet and first DEXA was 1.2 years (range 1 week – 10 years). 159 children had TTG levels checked >1 year after starting GFD. Of these, 81 achieved full seroconversion to negative TTG. Low body mass index (BMI) correlated with low BMAD (p=0.02). Using BMI for risk discrimination by ROC curve analysis was valid. A significant relationship between TTG, vitamin D level, ALP or serum calcium, and low BMAD was not demonstrated (P>0.5).

Persistent osteoporosis among children with coeliac disease following a gluten-free diet is very rare. Low BMI is a predictor of osteoporosis. Targeted DEXA screening has a place in the current management of coeliac disease whereas routine screening does not.

DEXA scan number	BMAD Z score ≥-1	BMAD Z score <-1 and ≥ -2	BMAD Z score < -2	Total
1st	100	19	13	132
2nd	39	13	8	60
3rd	15	5	3	23
4th	5	3	1	9
Total	159	40	25	224

Table 1: Classification of DEXA scan results according to BMAD Z scores.

OC55

A pilot study to monitor non-invasive markers of liver fibrosis in paediatric patients on home parental nutrition

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Intestinal failure associated liver disease (IFALD) can cause liver fibrosis without jaundice in the modern era[1]. Liver biopsy may be misleading in assessing the severity owing to patchy hepatic involvement, and does not help in defining the presence or absence of portal hypertension[2].

The aim was to evaluate common non-invasive tests for liver fibrosis among a cohort of patients on home parenteral nutrition (HPN) in a tertiary paediatric gastroenterology unit.

We conducted a prospective study between March 22 and Nov 22. Data on abdominal ultrasound, fibroscan and biochemical markers (Enhanced Liver Fibrosis ELF test, AST-to-platelet ratio index APRI, fibrosis-4 score Fib-4)were collected.

15/31 patients on HPN were included. While 16 patients were excluded due to either age or behavioural issues. Median age was10.8 years (range 3y10m-17y), 73% male. The median duration of HPN was 47 months (range 12-189), 67% of patients had short bowel syndrome. Results for fibroscan, APRI, FIB-4 and ELF test are shown in Table 1. Abdominal ultrasound showed splenomegaly in five patients. No correlation was identified between Fibroscan score, APRI score, FIB4, ELF test or splenomegaly (r = 0.09,0.1,-0.3,0.12 respectively). 13.3% (2/15) children had splenomegaly and F score of > 7.7 suggestive of progressive fibrosis. Of these, one patient is undergoing intensive rehabilitation to stop HPN and the second patient's monitoring has been intensified.

Non-invasive modalities for assessment of liver fibrosis maybe helpful in identifying fibrosis at earlier stages in children with IFALD. This would allow different treatment options to be considered in a timely manner before progression to end stage IFALD needing intestinal transplantation.

Table 1: Results of Fibroscan, APRI, Fib-4 and ELF test

	Range (median)	Patients with splenomegaly N =5 Range (median)	Values in detecting fibrosis in chronic liver disease
Fibroscan (F)	4.2-8.8kPa (5.7kPa)	5.7-8.8 (6.66kPa)	F0 (2 to 7 kPa) normal to mild scarring ,F2 (7.5 to 10 kPa) indicates moderate scarring
APRI score	0.18-1.3 (0.5)	0.46-1.04 (0.71)	APRI score > 1.0 had a sensitivity of 76% and specificity of 72% for predicting cirrhosis
Fib-4	0.11-1.5 (0.35)	0.12-1.5 (0.49)	<1.45, 45% probability that the patient has significant fibrosis
ELF	7.9-10.3 (9.35)	7.9-10.3 (9.3)	<7.7 limited to no fibrosis 7.7—9.8 moderate fibrosis > 10.5 - advanced fibrosis

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OC56

Improving medicines information provided to families of infants recovering from Kasai portoenterostomy procedure

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Biliary atresia (BA) is a rare cholangiodestructive disease affecting infants. Management involves a Kasai portoenterostomy procedure (Kasai) to re-establish bile flow and alleviate jaundice, with liver transplantation required if bile flow is not restored.[1]

Several unlicensed medications are prescribed post procedure, including fat-soluble vitamins, antibiotics, steroids and phenobarbitone. The availability of relevant medicines information (MI) for unlicensed medicines is limited and can affect adherence and increase the risk of medication errors. In addition, poor health literacy and adherence to medications are known to negatively impact on the outcomes of children with chronic conditions.[2] To support families through the procedure MI is provided by the specialist nurse, prior to, during admission and on discharge via shared care. This project aims to evaluate parental/carer experience of MI provided post Kasai and identify areas for improvement.

Families of children who underwent Kasai between June 2020 and 2022 were invited to complete a questionnaire. Ethical approval was not required for the service evaluation, local committee approval and prior parental/carer consent was obtained.

Out of 38 families identified, 23 agreed to take part and were included in the analysis. Families originated from a wide range of geographies across 13 countries, with 48% identifying as British nationals. 52% of families spoke English as a first language and all as a second language. The majority found it either 'very easy' (61%) or 'easy' (30%) to understand written MI in English, those who found it difficult, independently translated the MI. There was a high level of parental satisfaction (91% 'very satisfied'/'satisfied') regarding amount and quality of MI received. Despite this, 91% families contacted the specialist nurse, pharmacist or consultant team via shared care for further MI queries and 52% conducted independent internet searches for further information. Families felt least confident when asked about the risks and benefits of medication use, side effects and interactions with other medicines. One family reported a medication error occurred due to lack of understanding of dose changes post discharge.

The majority of families are satisfied with the current provision of MI post Kasai procedure, with a high use of shared care services for MI post discharge. There is an opportunity to improve the MI we provide, using targeted medicines leaflets, which include risks and benefits of unlicensed treatments, side effects and their management and medication interactions. Improved availability of accessible MI in a variety of languages and translation services for families may improve future satisfaction, adherence and help reduce some health inequalities currently experienced by our families.

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OC57

Diagnosing coeliac disease in adults; the paediatric way

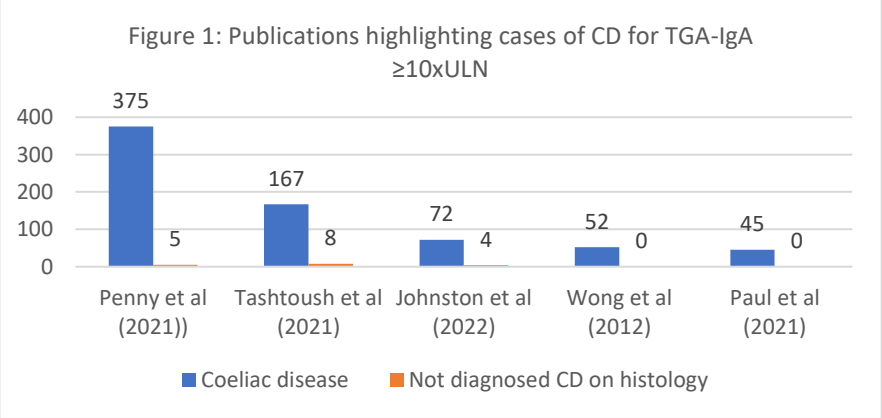
By V. Gangalam, P. Heaton, S.Paul. *Yeovil District Hospital, Yeovil, BA21 4AT, UK.*

Introduction: The no-biopsy pathway [NBP] has been successfully being used for diagnosing coeliac disease [CD] in children since 2012. However, the need for upper GI endoscopy and biopsy has remained an essential step for diagnosing CD in adults and there has been reluctance amongst the adult gastroenterologists to adopt the NBP for diagnosing CD in adults. During the COVID-19 pandemic, the British Society of Gastroenterology (BSG) published interim guidelines for diagnosing CD, including the NBP for those under 55 years and with no other concerning features[1].

Aims: To establish, by analysis of published articles and abstracts, whether the ESPGHAN recommended NBP is a safe approach to the diagnosis of CD in adult practice

Methods: We included UK-based prospective and retrospective studies identified in a search of PubMed, Google Scholar, EmBase, databases with the terms: adults, CD, NBP, ESPGHAN guidelines, BSG.

Results: A total of 5 publications[2-6] were identified from the UK: 4 papers and 1 abstract. A total of 728 patients with anti-tissue transglutaminase antibody (TGA)-IgA values ≥10x Upper Limit of Normal (ULN) were identified from the 5 publications, 711/728 were confirmed to have CD following upper gastrointestinal endoscopy (UGIE) and biopsy with Marsh 2-3 identified on histology. Figure 1 shows the breakdown for the studies included in the analysis. 97.66% of patients with TGA-IgA ≥10xULN received histological confirmation of CD.



Conclusions: Our analysis of UK-based adult CD studies have shown that the NBP can be safely implemented in the UK, and supports the recommendations made in the BSG COVID-19 interim guidelines for CD. The NBP prevents unnecessary delay in the management of patients with CD. Studies from Italy, India, Finland and New Zealand alongside the UK-based data have shown that life-threatening concomitant pathologies are extremely rare in the adult CD population, and that a NBP is accurate and safe.[7-10] We hope the next revision of the NICE CD guidelines will acknowledge and include the NBP for diagnosis of CD which will streamline the diagnostic pathways across both paediatric and adult gastroenterology practice.

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OC58

Single centre experience for children who presented with acute non-A-E hepatitis in March & April 2022: 10 months on

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Aim: Presenting features and current status of children who presented with acute non-A-E hepatitis in a district general hospital

Method: We looked at all cases of children (<16yrs) who presented with acute hepatitis during March & April 2022. Presenting symptoms, clinical findings, level of transaminitis, presence of coagulopathy were noted. Investigations were based on the guidance issued by the RCPCH on acute non-A-E hepatitis during the same time period. Patients were discussed with the tertiary centre. After discharge patients were assessed monthly for the first 6 months and then 2 monthly where possible in the outpatient clinics.

Results: 9 patients presented with transaminitis during a period which coincided with a spike in acute non-A-E hepatitis paediatric cases nationally. 77% of patients were females. The ages of patients ranged 2.5 to 6 years corroborating what has been observed nationally. .All patients had some gastrointestinal symptoms (anorexia, vomiting, loose stools or abdominal pain). 44% abdominal pain, 44% loose stools and 22% vomiting. 11% had lethargy and 55% were clinically jaundiced. 44% had acute liver failure either at presentation or during their admission. Two patients were readmitted after their initial admission episode because of worsening transaminitis trend. Surprisingly none had pruritus. Acute non-A-E hepatitis investigative work up as per RCPCH guidance was negative in all cases except finding the presence of Adenovirus & Enterovirus. 66% (6 patients) had PCR positive for Adenovirus and 11% (1 patient) for Enterovirus. On presentation 66% had hyperbilirubinemia and 33% presented coagulopathy. 66% were treated with Vitamin K but only 55% were managed with Urso-deoxycholic acid and fat-soluble vitamins. Only two patients needed transfer to the local tertiary hospital, rest were effectively managed locally

Conclusion: Nationally there was a spike in children presenting with acute hepatitis. We had similar cases. Although only 66% cases have their transaminitis completely resolved, all children are asymptomatic and clinically well at the last clinical review. We hope our data supports other hospital findings and adds to the national database for future surveillance.

OC59

Dietetic involvement in paediatric parental nutrition; is it essential?

by R. Stanley. *Great North Children's Hospital, Newcastle Upon-Tyne, NE1 4LP, UK.*

Provision of parental nutrition (PN) is complex, enteral and PN requirements frequently change (1,2). It is imperative that patients undergo detailed regular nutritional assessment to avoid growth faltering. This includes identifying individual nutritional needs. Generic guides exist for PN requirements however, individual assessment is multifactorial (1,2) which dietitians are uniquely placed to evaluate and ensure adequate growth (2,3). Despite this, dietitians are not always included in managing paediatric PN. This study assesses if dietitian involvement in PN is essential to ensure adequate growth.

Children receiving PN were identified retrospectively via clinical records between January2022-June 2022. Patients excluded if on critical care ward or >18 years. Dietetic assessment identified through electronic notes. Growth was assessed calculating changes in weight-for-age Z-scores (delta-Z-Score) from start to end of PN.

78 children identified. 73 complete data.

Table 1: Cohort demographics

Speciality commenced PN (n=73)	Neonates	11
	Immunology (including bone marrow transplant)	24
	Gastroenterology	8
	Oncology	12
	Surgery	15
	Other	3
Age commencing PN (mean SD)	5.11y (5.9y)	
Male:Female	M=45 F=28	

50/73 patients had dietetic assessment prior to commencing PN. Further 8 assessed on average 6 days after PN commenced [range 2-15 days]. Dietitian initial PN assessment of these 8 recommended 25% no changes, 50% to increase macronutrients due to not meeting requirements and poor growth, 25% to reduce macronutrients due to requirements being exceeded.

Delta Z-scores changed on average by +0.31 (range -2.21 to + 4.96) for patients assessed by a dietitian and -0.724 (range -1.89 to +0.39) for patients not seen by a dietitian. The interquartile ranges of each group was 1.06 and 1.01 for no dietitian and dietitian involvement respectively.

Average length of time patients received PN when assessed by dietitian 55.5days (SD 56.5d) and 21.06days (SD 16.63d) for those not seen by dietitian.

In total, 79% PN patients were assessed by a dietitian, those without dietetic assessment prior to commencing PN, have 75% risk of inappropriate nutrient provision.

Patients with dietetic assessment are more likely to have positive growth trajectory. As it is essential to meet energy and protein requirements for adequate growth, it can be concluded that PN patients not reviewed by a dietitian are less likely to meet their individual requirements. This study has shown that it is essential for paediatric patients receiving PN in an acute setting to be assessed by a dietitian to ensure adequate growth. It highlights the importance of dietitians being integral members of teams that manage paediatric PN in acute settings.

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OC60

Temporal trends in complex feeding decisions a decade in a specialist clinic in a tertiary paediatric setting

by R. Poole[1], M. Brooks[1], D. Flynn[1], N. McConnell[1], A. Sabharwal[2], J. Woods[1], A. Campbell[2], T. Bradnock[2], G. Walker[2], K. Fraser[1], S. Fraser[1], J. Andrews[2] and A.R. Barclay[1]. [1]*Paediatric Gastroenterology, Royal Hospital for Children, Glasgow, G51 4TF, UK .*[2]*Paediatric Surgery, Royal Hospital for Children, Glasgow, G51 4TF, UK.*

Decision making for patients with complex feeding issues have previously been made in isolation. It is advocated for cohesive decisions to be made in a multidisciplinary setting(1). Since the 2013 appointment of a nurse specialist, complex feeding decisions are made in our tertiary MDT complex enteral feeding (CEN) clinic. We aim to describe clinical outcomes with reference to changes in medical and surgical therapies.

From a prospectively gathered departmental database, all patients reviewed in CEN clinic (03/2013-12/2022) were assessed for: demographics; primary diagnosis; referral mode. Interventions of interest: fundoplication; jejunal feeding; blenderised diet (BD). Patients commenced on jejunal feeding or BD each year were calculated. Comparison of proportionate interventions for when dietetic support for BD was and was not resourced was performed by fisher's exact test (sig $p \leq 0.05$).

Of 201 patients reviewed: 61% had a severe neurodisability; 26% multisystem disease; 7% complex GI surgery; 4% complex cardiology; 2% isolated complex feeding issue. 62% male, median age at referral was 3.3 years. 58 fundoplications were performed, 99 received jejunal feeding (initiated with 37 nasojejunal; 58 gastrojejunal device; 4 surgical jejunostomy) and 57 patients of the 72 recommended started BD. BD has been advocated by team since 2016 as a feeding strategy for gastric intolerance of milk formula. Dietetic support to establish BD was resourced 2018-2020. Out-with this timeframe, availability was limited in certain areas. After rising incident cases of jejunal feeding, we observed a reduction with the introduction of dietetic resource to support BD initiation (2018-2020). This trend was reversed 2021-2022 with significantly higher jejunal initiation being required (p=0.0005) (fig 1). We observe a reduction in fundoplication from 11 to 3 per year, in line with increased in jejunal feeding and BD. A small minority required a period of parenteral nutrition (PN) support including 2 patients who received PN for > 12 months.

We report the longitudinal trends in intervention in a CEN clinic with reduction of fundoplication and rise then reduction in jejunal feeding when dietetic support for BD was available. The reduction in BD initiation 2021-2022 sits chronologically with the withdrawal of dietetic support and resulted in significant rise in jejunal feeding. We highlight the success of fundoplication for patients in our service with careful selection. This has driven service redesign within our institution. Jejunal feeding has become secondary to a trial of BD in our hierarchy due to BD benefits including: less invasive feeding modality; apparent better symptom control; parental wishes and cost saving. Effects of the pandemic and dietetic resource may have restricted access to BD in the last 2 years.

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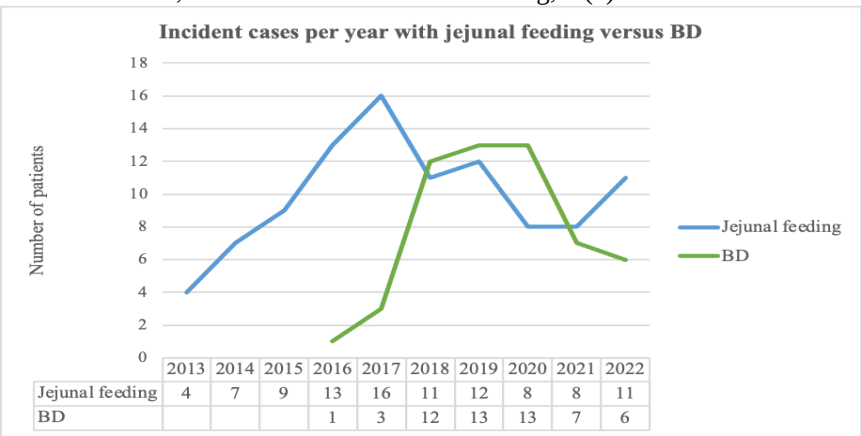

	2013	2014	2015	2016	2017	2018	2019	2020	2021	2022
Jejunal feeding	4	7	9	13	16	11	12	8	8	11
BD				1	3	12	13	13	7	6

Figure 1

OC61

Rare complication with parenteral nutrition

by A. Mallikarjuna, A.E. Wiskin. *Bristol Royal Hospital for Children, Bristol, BS2 8BJ, UK.*

Parenteral nutrition is essential as a short-term measure or for long-term use in children with various gastrointestinal issues[1]. Here we report two cases with an untoward complication following the use of parenteral nutrition.

Case 1: 11yr old girl was transferred from district hospital with suspected fistulating small bowel Crohn's disease evidenced on MRI scan. Due to poor oral intake, recurrent vomiting, even with nasal tube feeds, she was started on parenteral nutrition. A joint decision with the surgical team was to initiate medical treatment with infliximab. Five days into admission to the children's hospital, she had blurred vision; difficulty with speech; wobbly gait; bilateral facial palsy; ptosis; intermittent confusion. She was investigated for possible metabolic, infective or autoimmune aetiology. MRI brain scan showed a high-intensity signal in the medial thalami, periaqueductal white matter, inferior colliculi, midbrain, pontine and medullary tegmentum. Fundoscopy showed chronic papilloedema. On retrospective review, although she had adequate weight for height, her recent nutritional intake was poor; she had been nil by mouth for one week in her local hospital before transfer and had lost 2kgs in weight. She was treated with intravenous thiamine for Wernicke's encephalopathy. Her neurological signs recovered with treatment. She subsequently underwent a right hemicolectomy and recovered well post-surgery.

Case 2: 8m old boy with short gut secondary to volvulus and intestinal failure presented 15 days after first discharge on home PN with recurrent vomiting. He was admitted, had a normal upper gastrointestinal contrast study and less vomiting. He was discharged after a period of observation. Three days later, he re-presented with episodes of leaning to the right side, twitching of the right arm, and reduced movements of the right side with weak vocalisations. Blood gas showed high lactate and normal pH. Parenteral nutrition was stopped and he was started on intravenous fluids. Home parenteral nutrition prescription was reviewed which showed no water-soluble vitamin due to an error in transcribing from inpatient to home prescription. He was started on intravenous thiamine. He was also investigated with CSF analysis and MRI head, which were normal. His neurological signs gradually showed improvement. Wernicke's encephalopathy is an acute life-threatening neurological condition due to thiamine deficiency characterised by a triad of ataxia, ocular dysfunction and altered mental status. While it can be reversible it can also lead to permanent neurological impairment. It can be prevented by thiamine supplementation in children at risk of thiamine deficiency or in circumstances of increased thiamine requirement. Treatment for Wernicke's encephalopathy should be with prompt administration of parenteral thiamine[2]. A thorough understanding of the patient's nutritional status and their recent nutritional intake before initiating nutritional rehabilitation with enteral or parenteral route can help to avoid this hazardous complication.

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OC62

Biochemical markers, radiological findings and clinical characteristics for the early detection of intestinal failure associated liver disease

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Intestinal failure (IF) associated liver disease (IFALD) is the progressive liver injury secondary to IF and long-term use of parenteral nutrition. IFALD can present with cholestasis, steatohepatitis and if untreated, progresses to hepatic fibrosis. Non-invasive tests (biochemical and radiological) can enable its early detection and guide nutritional modifications and early treatment.

Objectives & Methods: Retrospective review of patient records to detect the incidence of early IFALD for children on long term PN over a 3-year period (2020-2022) using a low threshold of ALT>1.5 of upper limit of normal persistent for ≥2 weeks and correlation of their clinical, biochemical and radiological findings.

Results: 22 children (13M, 1.3-18 years) on long term PN (median time of 6.86 years) were identified. 7 (32%) patients had biochemical evidence of early IFALD and 15 (68%) had not (table I). The type of SBS, the presence of ICV, colon length and parenteral or enteral support did not differ between the 2 groups. Biochemical markers differed in these 2 groups: median [IQR]- ALT 63IU/L [61-80.5] vs 26.3 [18-31.2], p<0.001, AST 47.7IU/L [47-65.3] vs 24 [19.7-34.3], p<0.001, total Bilirubin 10.3 umol/L [7.3-14.5] vs 6[5.3-10.2], p=0.034 and AST/PLT ratio 0.24 [0.18-0.47] vs 0.14 [0.11-0.16], p=0.013 respectively. Enlarged fatty liver on abdominal ultrasound was more common on the IFALD group (71.4% vs 20%, p=0.03) (table II). 6 (85.7%) of the children with IFALD had short bowel syndrome (SBS) and 2 (33%) of those had ultrashort bowel. Bowel lengthening procedures were more common in the IFALD group (71.4% vs 6.7%, OR 35, 95%, p=0.004). Children with IFALD had been reliant on PN for longer 10.8 (6.5-15.5) years vs 4.63 (3.1-7.4) years, p=0.017. Ursodeoxycholic acid and fat free PN nights aimed to reverse IFALD progression (Table I). **Conclusion:** Combining biochemical markers and radiological findings could be useful for the early detection of IFALD for high-risk groups such as SBS children with lengthy PN dependence.

Table I: Trajectory of Biochemical and Clinical characteristics of children with early IFALD

Variable	**2020**	**2021**	**2022**
ALT, *Median IU/L [IQR]*	59 (42.5-69)	72 (55-123)	53 (45-67.5)
AST, *Median IU/L [IQR]*	50.5 (40-84.2)	72 (53.5-97)	46 (45.3-56.5)
GGT, *Median U/L [IQR]*	17 (12-32.5)	17 (14.5-27.5)	20 (15-41)
Bilirubin, *Median umol/L [IQR]*	9 (7-11)	10 (7.5-18)	12 (7.5-16)
AST/PLT, *Median [IQR]*	0.21(0.19-0.78)	0.44 (0.29-0.64)	0.26 (0.17-0.35)
PN nights, *Median n [IQR]*	4 (3-7)	5 (3.5-7)	5 (3.5-7)
Fat free nights, *Median n [IQR]*	0 (0-0.5)	1 (0-2.5)	0 (0-3)
PN admin, *median hours* [IQR]	12 (11.5-14.5)	12 (11-13.5)	13 (12-14)
PN Lipid, *median g/kg/night [IQR]*	2 (1.7-2)	1.9 (1.5-2)	1.9 (1.6-2)
No enteral nutrition, *n (%)*	1 (6.7%)	1 (6.7%)	1 (6.7%)
CLABSI, n *(%)*	2 (28.6%)	3 (42.9%)	1 (14.3%)
UDCA treatment, n *(%)*	3 (42.9%)	5 (71.4%)	5 (71.4%)

OC63

The use of Omegaven rescue therapy (ORT) in the treatment of Intestinal Failure associated liver disease (IFALD) in patients already received SMOF lipid: a tertiary single centre experience.

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The role of lipid manipulations in parenteral nutrition (PN) in changing the natural history of IFALD is well established. European centres report utilising a mixed lipid formula 'SMOF' (Soy, Medium chain (coconut), Olive and Fish) for prevention and treatment of IFALD, with North American centres using a pure fish oil 'Omegaven' in established IFALD (1). Whilst there has been a suggestion that the use of Omegaven may have additional benefit for 'rescue therapy' for patients on SMOF lipid who have still developed IFALD (2), either with cholestasis or deterioration of LFTs, to date there are limited published 'real world' cross-over data. We report our use of Omegaven rescue therapy (ORT) in paediatric Intestinal Failure (IF).

Since Jan 2019, under the advice of the specialist nutrition team, ORT was used selectively in the apriori agreed context of: IF, continued evidence of worsening liver function (total bilirubin (Tbil) >20umol/l or x3 rise in ALT/AST) despite anti-sepsis measures and prior lipid reduction to 1g/kg/day. Omegaven was given at 1-1.5g/kg/day. Data from Jan 2019- Dec 2022 was gathered including: demographics, sepsis, serial change in Tbil, Conjugated bilirubin, AST/ALT, with trend analyses over multiple time points with date of commencement ORT was performed using Dunnett multiple comparisons with control (CI 95% $p<0.05$). Exclusions to analyses included: inadequate rise in LFTs/Tbil, death during Rx and additional liver diagnoses.

15 included patients (8 in NICU) received ORT. ORT was given for 7-33 days. Median Tbil levels were rising significantly prior to ORT and then fell significantly both at T+14 days and T+28 days. (Fig 1). ALT/AST fell in the group but was not statistically significant. Including retreatments in individual patients also reduced treatment effect.

We present provisional data of ORT in patients already established on SMOF and lipid lowering strategies. We see that the most common indication for Omegaven is rising liver function tests both in context of sepsis/suspected sepsis and longstanding IFALD. We observed a rapid significant fall in Tbil with 14 days of therapy, despite a significantly rising Tbil prior to initiation. These data could be improved by additional cases, and defining a contemporary control group that are well matched for IF co-morbidity, age and deteriorating liver function however they differ to the natural history of IFALD. The possibility for a more sustained switch to Omegaven in a very select group of IF patients with longstanding deteriorating liver function merits exploration, as does the use of ORT in IF patients at risk of liver dysfunction during sepsis episodes.

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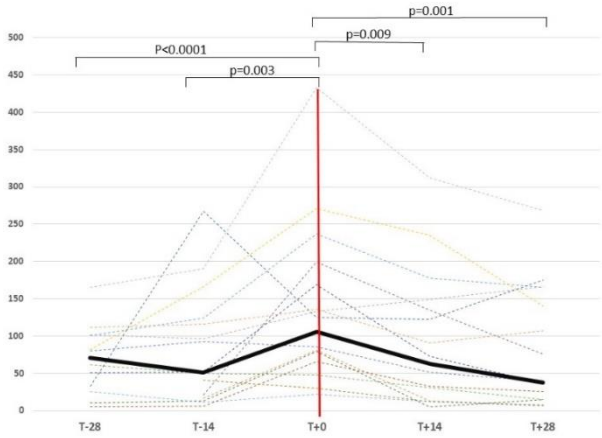

Figure 1: Tbil levels in 15 patients prior to and after initiating ORT (Median in bold)

OC64

Outcomes of children with short bowel syndrome (SBS) on home parenteral nutrition: experience at a tertiary centre hospital

by S, Rajani, K. Green, R. Rodley, H. Shapland, H. Berry, P. Tablot, T. Kaldez, J. Hadfield, J. Kirton, R. Rabone, Z. Londt, S. Marven, R. Lindley, A. Urs**.** *Sheffield Childrens Hospital, Clarkson Street, Sheffield S10 2TH, UK.*

This study describes the outcomes of children with SBS by the multidisciplinary intestinal rehabilitation program (IRP) at the Sheffield Children's Hospital A retrospective review of children with SBS on home parenteral nutrition between 2012 to 2021 using the Hospital electronic medical records.

A total of 23 patients with SBS were discharged on HPN. 14 were male and 9 were female. All except one presented in neonatal period. The aetiology in our cohort was gastroschisis (n 9%) followed by Necrotising enterocolitis (n 6%), volvulus (n 5%), intestinal atresia (n 2%), one was trichobezoar leading to intestinal perforation. The average small bowel length % length was 15.51%. 9 (39.1%) of our patients did not have ileo-caecal valve. 4(17%) patients underwent partial colectomy and 2(8.6) % underwent subtotal colectomy while 17 (73%) had a preserved colon. Intestinal continuity achieved in all except one patient. 5(21.7%) underwent gut lengthening procedure (STEP or Bianchi). 2 (8.6%) patients had IFALD%, d lactic acidosis in 6(26%) of our patients. 5(21.7 %) had vitamin D deficiency and none had any trace element deficiencies. Overall, a full enteral autonomy was achieved in 17 (73.9 %), parenteral nutrition dependent in 6 (26%), transplanted 3 (13%), one underwent isolated small bowel transplant, one combined bowel and liver and one isolated liver transplant.

Survival of children with SBS was 100% and achieved excellent enteral autonomy with reduced morbidity secondary to complications under the guidance of IRP. Children with bowel length >30 cm achieved full enteral autonomy. Absence of ileo-caecal valve did not impact enteral autonomy in our group.

Table showing characteristics of our patient group

	Group A (SBL <10%)	**Group B (SBL 10-25%)**	**Group (SBL 25-50%)**
Number	8	10	5
M:F	8:0	4:6	2:3
Median SBL in %	5.2 %	18.45%	38.4%
ICV intact	2	3	4
Colon	5 intact 2 Partial	8 intact 1 total 2 partial	2 intact 1 total 2 partial
Bowel lengthening procedure	2	3	0
Enteral autonomy	4	8	5
Transplant	2	1	0
Complications	IFALD 1 Dla 1	IFALD 1 Dla 4	IFALD 0 Dla 1
Duration of PN	51.5 months	46.4 months	31.6months

SBL – Small bowel length, IFALD- Intestinal failure associated liver disease, Dla – D lactic acidosis

OC65

Blood phytosterol levels in children with intestinal failure on home parenteral nutrition, and relation to liver disease

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Intestinal failure-associated liver disease(IFALD) in children on parenteral nutrition(PN) probably has a multifactorial aetiology. In particular, soya lipid emulsion has been associated with accumulation of phytosterols and associated cholestatic liver disease.[1] More recently mixed lipid emulsions which include fish, as well as soya, olive and coconut oil, >vitamin E and <phytosterols[2] have largely replaced pure Soybean lipid.

Our aim was to investigate whether patients on long-term treatment with PN at home on mixed lipid emulsions such as SMOF(soya, MCT, olive, fish oil)lipid have raised phytosterol blood levels and IFALD.

Retrospective Data was collected from our hospital's electronic patient records for all patients on home PN in 2022 for >6 months. Data collected included: age, gender, diagnosis (categorised as short bowel syndrome[SBS], motility disorder or intestinal mucosal disorder), years on PN, nights/week PN infused, nights/week with lipid, whether tolerating some oral/enteral nutrition and liver US. Blood results reviewed were phytosterol, albumin, liver enzyme(ALT, ALP, GGT), Vitamin A, E, D, clotting screen, and platelet levels.

There were 29 children,16 female,13 male, aged 2-18years. Eleven, 38%, had SBS, 9, 31% motility disorder and 9, 31% a mucosal disorder. Twenty-three, 79% tolerated some oral/enteral nutrition and 6, 21% did not.

Eighteen patients, 62% were on PN since infancy. Patients had 3-7(mean 6.3) PN infusions/week, administered over 10-24(mean 13.5)hours, with lipid included from 0-5(mean 2.6) nights/week. The lipid used was SMOFlipid.

Three patients, 10.3% had mildly elevated phytosterol levels, three, 10.3% borderline raised levels (within 10% of normal range) and 23, 79.3% had normal levels. One child on anti-fungal medication had a raised fungal related sterol, lanosterol. The elevated phytosterols in the three patients were:

1. mild elevation of whole phytosterol profile
2. raised Sitosterol and a cholesterol precursor level
3. raised Sitosterol

Two patients with high phytosterol levels had low blood albumen, 28g/l. Albumin was normal in the other 27 patients. ALT was normal in 27/29 patients and raised in 2/29(range 9-138U/L, median 28 U/L). Bilirubin was normal/borderline raised in all cases ranging from 2-20(median 8)umol/l. GGT ranged from 10-14(median 22) U/L. ALP ranged from 95-324U/L, median 202U/l. Vitamin A levels were 0.62-2.13(median 1.37) umol/L, Vitamin E was normal 11.5-46.4 umol/L in all 29 cases. Vitamin D was 33-122(median 68)nmol/L. Coagulation was normal in 20 patients and prolonged in 9 on anticoagulant treatment. Platelet levels were 52-444(median 233) $x10^9$/l. One patient had hepatic steatosis on U/S with with normal phytosterols and liver function tests.

In summary, children on long-term home PN on mixed lipid infusions had normal or only borderline raised phytosterol and liver function tests. In conclusion, phytosterol levels are not usually raised in children on PN with mixed lipid formulations and IFALD does not appear to be a problem.

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OC66

Bone health, BMI and Biologics; an audit of a paediatric IBD population

by J. Porter[1], N. Osborne[1], C. O'Connor[1], E. Renji[2], H. Garrett[1] .[1]*Department of Nutrition and Dietetics, Alder Hey Children's NHS Foundation Trust, Liverpool,* L14 5AB, *UK.* [2]*Department of Paediatric Gastroenterology, Hepatology & Nutrition, Alder Hey Children's NHS Foundation Trust, Liverpool, L14 5AB, UK.*

It is widely accepted patients with active inflammatory bowel disease (IBD) need dietetic input due to exclusive enteral nutrition treatment and/or risk of malnutrition [(1).] However, the input required when in remission is less understood. Therefore, the gastroenterology dietetic team at a tertiary paediatric hospital completed a 12-month nutritional audit (October 2022) of IBD patients undergoing biologic treatment at the medical day care unit.

In line with national IBD guidelines, body mass index (BMI), dietary restrictions, calcium intake and plasma vitamin D was reviewed [(1)]. Calcium intake assessed using dietary history and the British Dietetic Association's 'Calcium: food fact sheet' [(2).] Data was analysed using Excel.

From n=66 patients 67% were male, 33% female with an average age 15 (7-19). Biologic therapy use was 89% first line (e.g., infliximab) and 11% second line (e.g., vedolizumab). Diagnosis consisted of Crohns (71%), ulcerative colitis (21%) and IBD unclassified (8%). Overall, irrespective of remission, there was a correlation between higher BMI and higher PCDAI/PUCAI scores in females (F =0.56 p= 0.03), but not in males.

Based on PCDAI/PUACI, 76% (n=52) were classified as being in remission. In accordance with the World Health Organisation classification of BMI Z scores [(3)]13% were obese, 29% overweight, 54% healthy and 4% thinness. Their modifiable risk factors for bone health are outlined below.

Table I: Vitamin D and calcium of those in remission (n=52) Vs general population

	Adequate calcium	**Inadequate calcium**	**General population [(5)]**
Adequate Vitamin D	24%	32%	81%
Inadequate Vitamin D (<50nmol/L)	4%	18%	19%
General population [(5)]	85%	15%	

Dietary advice was given based on review data and clinical judgement, including improving calcium intake (75%), healthy eating (23%) and healthy eating with weight management (15%). Within the cohort, 69% of children had no food restrictions while spicy food was the most common food avoidance (10%).

Current IBD guidelines focus on supporting malnutrition [(1)]. However, BMIs described above are comparable to the general population [(4)]. IBD teams, including dietitians, must accept the focus of nutritional intervention may change. Guidelines recommend an annual growth assessment regardless of disease state [(6)]. Given our comparison of bone health to the general population, calcium and vitamin D should also be assessed annually to help overcome negative effects of steroids, commonly used in patients with IBD [(1)]. Exercise status could also be assessed. Our data would support further research into the link of disease scores and obesity.

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OC67

Hierarchy of nutrition interventions for gastrointestinal dystonia (GID), clear consensus for the use of blenderised diet ahead of post-pyloric feeding and surgical interventions; output of the BSPGHAN/BAPS/BPNA/APPM RAND panel.

by M. Brooks[1], H. Norton[2], S. Meade[3], C. Richards[4], S. Protheroe[5], M. Samaan[3], A.R. Barclay[1]. .[1]*Department of Gastroenterology, Hepatology and Nutrition. Royal Hospital for Children, Glasgow, G51 4TF, UK.* [2]*Department of Dietetics, Birmingham Women`s and Children`s Hospital. Birmingham, B4 6NH, UK.* [3]*Department of Gastroenterology, Hepatology and Nutrition, Guys and St Thomas NHS Foundation Trust, UK.* [4]*British Association of Paediatric Surgeons, 38-43 Lincoln`s Inn Fields, London, WC2A 3PE, UK.* [5]*Department of Gastroenterology, Hepatology and Nutrition, Birmingham Women`s and Children`s Hospital. Birmingham, B4 6NH, UK.*

Children and young people with severe neuro-disabling conditions can experience debilitating distress from enteral feeding. When this occurs in the absence of commonly associated problems (reflux/constipation); the term 'gastrointestinal dystonia' now has an agreed definition (1). We aim to describe the development of an agreed algorithm on nutritional interventions GID from the described (1) RAND panel.

In brief (2) a writers group structured the questions for expert survey, based on the limited written evidence and their added professional experience. A panel of 27 experts in their field were assembled from 5 stakeholder groups including: Gastroenterology, Neurology/Neurodisability, Surgery, Palliative Care and Allied Health Professionals. Geographic representation was from 13 UK specialist centres (including all 4 nations) and 1 centre from Republic of Ireland to a. The panel rated the appropriateness of definition, investigations and management of GID. A scale of 1-9 enabled scoring of 1-3 to indicate inappropriate, 4-6 uncertain, 7-9 appropriate as criteria for recommendation. Panel agreement index was calculated using a continuous likelihood ratio, with <1 indicated 'general agreement' and >1 'no agreement'. Results were discussed at a moderated meeting before a revised post meeting survey was complete.

27/27 panellists answered questions on nutritional interventions (appropriateness scale/disagreement index). Reducing bolus size (8.0/0.37), continuous feeding (8.0/0.16), empiric trial of hydrolysed feed (7.0/0.24) were all considered appropriate early interventions in GID. Panel agreed a clear preference for the use of blenderised diet ahead of post pyloric feeding (8.0/0.16), due to level of symptomatic benefits, tolerance parental empowerment and cost. The use of temporal undernutrition (permissive feeding) to balance quality of life, or to allow time to consider goals and treatment priorities was appropriate (8.0/0.03). Considering minimal oral feeding for comfort/enjoyment despite aspiration risk was also advocated for (7.0/0.22). From the output the following nutritional strategies algorithm was devised (Fig 1).

We present the first coherent nutritional treatment algorithm in GID devised from the best available evidence and large expert panel opinion. Consensus around the increasing role for blenderised diet is welcome to health professionals and parents who have pioneered this treatment. However there are implication for health care resources, institutions such as schools and community care. Resource education and local guidelines will be required in order to further progress this excellent intervention.

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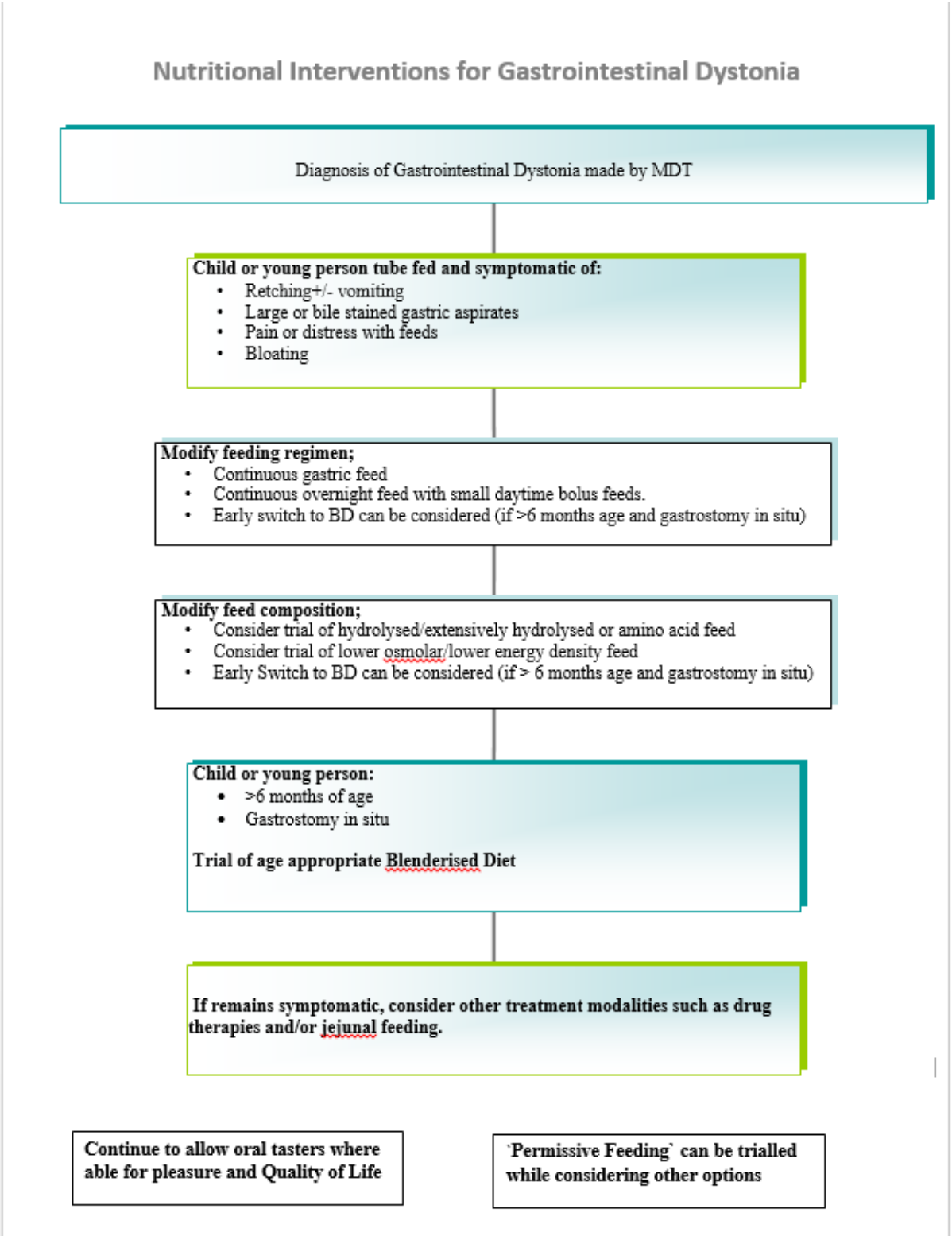
Fig 1.


Nutritional Interventions for Gastrointestinal Dystonia
Diagnosis of Gastrointestinal Dystonia made by MDT
Child or young person tube fed and symptomatic of:
Retching+/- vomiting
Large or bile stained gastric aspirates
Pain or distress with feeds
Bloating
Modify feeding regimen;
Continuous gastric feed
Continuous overnight feed with small daytime bolus feeds.
Early switch to BD can be considered (if >6 months age and gastrostomy in situ)
Modify feed composition;
Consider trial of hydrolysed/extensively hydrolysed or amino acid feed
Consider trial of lower osmolar/lower energy density feed
Early Switch to BD can be considered (if > 6 months age and gastrostomy in situ)
Child or young person:
>6 months of age
Gastrostomy in situ
Trial of age appropriate Blenderised Diet
If remains symptomatic, consider other treatment modalities such as drug therapies and/or jejunal feeding.
Continue to allow oral tasters where able for pleasure and Quality of Life
`Permissive Feeding` can be trialled while considering other options

OC68

A multicentre assessment of adequacy of bowel preparation for paediatric ileocolonoscopy using the Boston Bowel Preparation Scale (BBPS)

by H. Barraclough[1], P. Gillett[2], D. Basude[3], R. Bremner[4], A. Butt[5], I. Chalmers[6], R. Gowda[7], R. Hansen[8], J. Hart[9], A. Kapoor[1], S. Loganathan[10], A. Mallikarjuna[3], R. Mardare[11], A. Pigott[12], E. Renji[13], N. Skoutelis[14], C. Taylor[7], M. Thomson[1], P. Tierney[14], P. Narula[1]. *[1]Sheffield Children's Hospital, S10 2TH, UK, [2]Royal Hospital Children & Young People, EH16 4TJ UK. [3]University Hospitals Bristol & Weston NHS Foundation Trust, BS1 3NU, UK, [4]Birmingham Women's and Children's NHS Foundation Trust, B15 2TG, UK. [5]Royal Alexandra Children's Hospital, BN2 5BE, UK. [6]Royal Aberdeen Children's Hospital AB25 2ZG, UK. [7]Maidstone & Tunbridge Wells Hospital, TN2 4QJ, UK. [8]Royal Hospital for Children, G51 4TF, UK. [9]Royal Devon & Exeter NHS Foundation Trust, EX2 5DW, UK. [10]Nottingham Children's Hospital, NG7 2UH, UK. [11]Barts Health NHS Trust, E1 1BB, UK. [12]University Hospitals of North Midlands, ST4 6QG, UK, [13]Alder Hey Children's Hospital, L14 5AB, UK. [14]Belfast Health & Social Care Trust, BT12 6BA, UK.*

Adequate bowel preparation facilitates ileocolonoscopy completion. Pediatric Endoscopy Quality Improvement Network (PEnQuIN) quality standards recommend unadjusted rates of adequate bowel preparation of ≥ 80% and terminal ileal intubation ≥85%. A Boston Bowel Preparation Scale (BBPS) score of ≥6 is adequate. This study aims to identify whether UK hospitals are attaining PEnQuIN standards, and reviews medications used.

Fifteen UK hospitals participated. Local governance processes were followed. Each hospital provided anonymous BBPS data. Patients were grouped according to bowel preparation drug type. Patients were excluded if procedures were abandoned due to significant bleeding, stricture, previous resections or severe disease and if a subgroup had below 5 patients. A single factor Anova test and two sample T-Test was performed.

Complete data for 209 patients from 12 hospitals was collated. Five of 12 participating hospitals attained the PEnQuIN standard for adequate bowel preparation. Bowel preparation drugs varied greatly among hospitals. Patients were grouped according to drug type for analysis

(see table 1).

Group	Drug/s	N° Patients	Percentage achieving BBPS score of ≥6	Ileal Intubation
A	Picosulphate & Senna	102	65.7%	97.1%
B	Picosulphate	69	79.7%	95.7%
C	Citramag	25	92%	88%

Only Group C achieved the desired adequate bowel preparation rate of ≥ 80%. Not all groups are equal ($p<0.006$) on single factor Anova analysis. Significant differences in BBPS scores between groups A&C and B&C were found on a paired T test. Significant subgroup variability exists regarding drug timing, dosage and frequency resulting in heterogeneity.

This initial study highlighted marked variability in the UK use of bowel preparation medications. Only Group C attained the PEnQuIN standard for adequate bowel preparation. Groups A and B achieved high ileal intubation rates despite not meeting the standard. A large prospective multi-centred trial is required to establish the ideal medication type and protocol for bowel preparation in children.

OC69

Train the paediatric colonoscopy trainer course: an assessment of perceived value to participants in improving their colonoscopy teaching practice.

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Endoscopy teaching practice is variable, significantly affecting training provided. Endoscopy specific train the trainer courses are commonplace in adults, and in the UK are now mandatory[1,2]. There is only one paediatric train the colonoscopy trainer course (PTCTC) in the UK and although informal feedback has been positive, its practical value has never been formally assessed. A small cohort of studies demonstrated a benefit with these courses but highlighted the need for further research, especially in paediatrics. We aim to assess the practical value of the PTCTC and how attendees perceive their teaching practice compared to non-attendees.

A questionnaire based on the PTCTC learning objectives and aims was distributed to two groups of Consultant Paediatric Gastroenterologists who teach colonoscopy in the UK; those who had attended the course and those who had not attended the course (controls).

41 completed responses were received. 25 attended a PTCTC, 3 attended an adult course and 13 had not attended any course. Overall responses indicated participants of the PTCTC rated their confidence and knowledge in teaching practices as higher than controls (4.27 vs 3.56 P = <.001). There was a statistically significant difference in all areas: set (4.21 vs 3.71 P = .011), dialogue (4.29 vs 3.55 P = <.001) and closure (4.37 vs 3.6 P = <.001) with those who attended the PTCTC giving higher ratings. There was evidence of increased understanding of key concepts such as using standardised language, conscious competence, dual task interference and performance enhancing feedback.

Overall, this study demonstrates a higher perceived level of knowledge in fundamental teaching principles and confidence in colonoscopy teaching skills in those who attended a PTCTC. The study was limited by the sample size, but the results support the need for these courses and for ongoing research into their importance.

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2. Anderson JT. Assessments and skills improvement for endoscopists. Best Pract Res Clin Gastroenterol 2016; 30: 453–71.

OC70

Video capsule endoscopy (VCE) capsule retention due to duodenal diverticulain a paediatric patient: A case report and literature review

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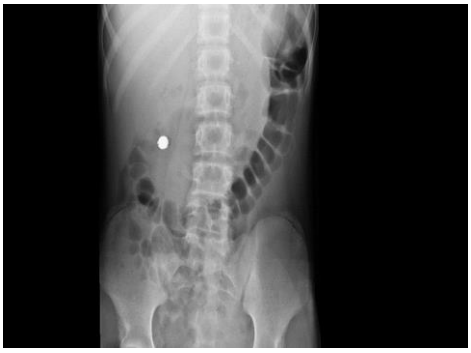
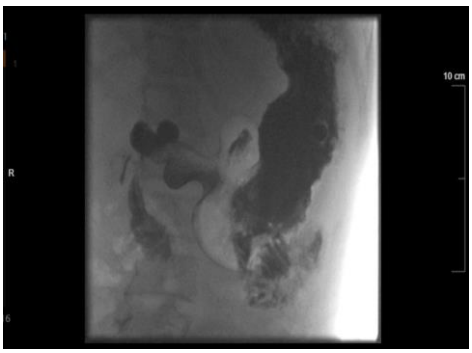
Introduction: Video capsule endoscopy (VCE) has become major diagnostic tool to visualise the gastrointestinal tract, particularly the jejunum and ileum for inflammatory bowel disease andother small bowel pathologies. Capsule retention is the potentially most serious complicationassociated with it.

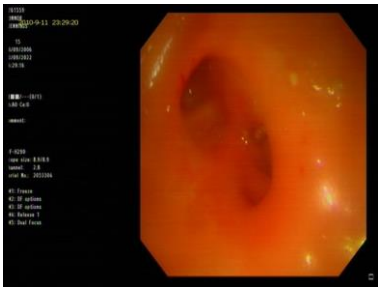
Case Presentation: In this case report we describe a 16-year-old male patient who had capsule retention due to duodenal diverticula. He already had an established diagnosis of Eosinophilic Oesophagitis (EoE) and was on PPI's. On revaluation, microscopy showed neutrophilic infiltration within duodenum and since he had family history of Inflammatory bowel disease (IBD) so was furtherinvestigated for IBD. Management required endoscopic removal of VCE.



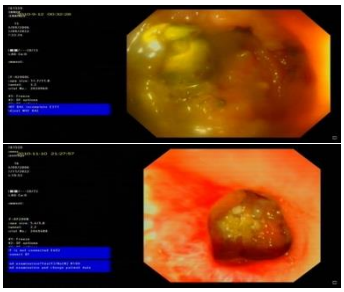
1A.VCE Capsule visible on X Ray Abdomen



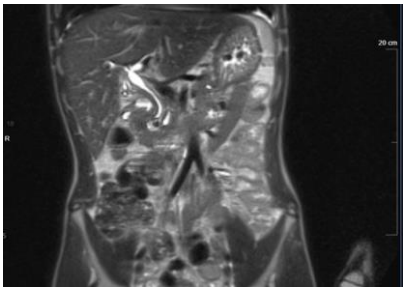
1B. Barium follow-through showing 2 Duodenal diverticula and relative narrowing of D2.



2A. Duodenal diverticulum



2B.VCE capsule
2C. Food bolus in the diverticulum



3B. MRCP showing the larger proximal diverticula (23 mm) impinges on the CBD and causes a fusiform dilatation (11mm) of the proximal CBD. The smaller diverticula (13 mm) ithe confluence of the CBD and pancreatic ducts and causes mild distension of the latter.

Discussion: VCE is capable of evaluation and diagnosis of IBD and various other pathologies especially in the small bowel but retention of VCE capsule remains a major concern. Prolongedcapsule retention has increased potential for intestinal perforation.

Conclusion: Although VCE is very safe and serious complications are rare, but patients should be monitored for anypotential complications especially retention of VCE capsule.

OC71

Reducing the carbon footprint of upper GI endoscopy in paediatric patients

by R. Babiker, M. Furman, D. Crespi, R. Levi, J Koeglmeier, R. Al-araji, O. Borrellii, K. Nikaki, F.Kiparissi, E. Gaynor, K. Jones, K.J. Lindley. *Department of Paediatric Gastroenterology, Great Ormond Street Hospital for Children NHS Foundation Trust, London WC1N 3JH and the Department of Paediatric Gastroenterology, Royal Free Hospital NHS Trust, London, NW3 2QG, UK.*

Upper Gastrointestinal endoscopy in the paediatric population has evolved during the last 30 years with an increasing number of diagnostic and therapeutic applications. However endoscopy is a major contributor to the environmental footprint of the NHS. This study aims to assess the indications, diagnostic yields, concordance between histopathological and endoscopic findings and accuracy of upper endoscopies in two quite different Paediatric Gastroenterology referral centres.

A multicentre retrospective analysis of hospital records of children who underwent their first upper endoscopy in 2022. Demographics, endoscopic details (e.g., indications, findings and any complications), and histopathological findings were collected. The relationship between endoscopic findings and histopathological abnormalities was reported.

The study included 100 patients (age, 1- 17 years) of which 51% were male. The median age of the children was 11 years (10 patients, 10%).

The most common primary indications for endoscopy were generalized abdominal pain 21%, followed by suspicion of inflammatory bowel disease 17%, and then gastro oesophageal reflux disease / dysphagia with each 10% prevalence for each. Normal upper endoscopy was reported in 70 cases in the two centres (70%), comprising 74% in centre A and 66% in centre B. Most of the normal endoscopies were performed in children with abdominal pain 21.4%. The highest rates of endoscopic abnormalities were found in patients with a suspicion of inflammatory bowel disease. Macroscopic findings at endoscopy showed 63.3% sensitivity and 82.8 % specificity to predict histopathological abnormalities. Females had higher rates of abnormal histological abnormalities 56.6%.

There is an extremely high rate of normal upper GI endoscopy in these two centres. Abdominal pain in particular has a low diagnostic yield. Concordance between endoscopic and histopathological findings is high but far from perfect and normal endoscopic findings shouldn`t discourage the endoscopist from obtaining tissue biopsies. The carbon footprint of endoscopy services has scope to be reduced significantly and should be a priority in the next 10 years for Paediatric GI centres.

OC72

Lower gastrointestinal bleeding as a feature of haemophagocytic lymphohistiocytosis: endoscopic therapeutic intervention in a high risk patient

by N. Francis, C. Thomas, S. Loganathan, D. Devadason, S. Kirkham, *Nottingham Children's Hospital, Derby Road, NG7 2UH, UK.*

Haemophagocytic lymphohistiocytosis (HLH) is a rare immunological disorder more commonly seen in children than adults. It results from impairment in the functioning of natural killer and cytotoxic T cells leading to haemophagocytosis presenting with evidence of pancytopaenia, hypertriglyceridaemia and overactive inflammation with or without organ failure. We present a patient with acute massive gastrointestinal bleeding associated with HLH.

A 6 year old girl initially presented with fever and rash following treatment for group A streptococcus tonsillitis, later developing renal impairment. Investigations revealed tubulointerstitial nephritis on renal biopsy and low grade haemophagocytosis on bone marrow biopsy. Serological investigations to include soluble CD25 met the full diagnostic criteria for HLH.

The patient was treated with oral high dose steroids for HLH and tubulointerstitial nephritis. Her renal function failed to improve so she was admitted to commence peritoneal dialysis. During education prior to this she developed frank lower gastrointestinal bleeding requiring several blood transfusions. Initial urgent OGD and unprepped colonoscopy did not identify a bleeding source. CT angiogram showed evidence of hyperaemia in the ileocecal valve area. The patient continued to bleed and the surgeons and lead renal consultant were keen to avoid surgical resection if at all possible due to implications for prospects for future renal transplantation. Repeat colonoscopy following limited bowel preparation identified a large rolled age ulcer encircling the ileocaecal valve with a visible vessel with attached clot in the base. Three modalities of treatment were used to control bleeding – injection with adrenaline, clipping of the visible vessel and topical haemostatic therapy. A single biopsy was taken with caution adjacent to the ulcer. Histology of this ulcer showed a predominance of macrophages with phagocytosed red blood cells within in keeping with a HLH diagnosis. Haemostasis was secured with the treatment modalities used. At the time of writing the patient is awaiting definitive management of her underlying HLH by the oncology team and continues with supportive renal replacement therapy

GI bleeding (GIB) associated with HLH is extremely rare. It is due to transmural lymphohistiocytic infiltration of macrophages resulting in localized ulcers or diffuse mucosal irritation in the gastrointestinal tract. Management of GIB is challenging, owing to difficulty in localizing mucosal bleeding and increased risk of recurrent bleeding upon intervention, and has extremely high mortality. Adult studies estimate a 12.2% rate of having a GIB as a complication of HLH, with an associated mortality of 66%[1]. Early diagnosis of HLH and treatment is the key. Emergency surgery or angioembolization for GIB has been partially successful in a few patients. In our patient the stakes were high, due to the need to avoid emergency right hemi colectomy to preserve the possibility for later renal transplantation and it was agreed that an initial repeat attempt at multi modality endoscopic therapy was justified.

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OC73

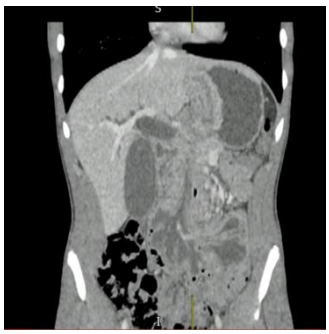
Successful Endoscopic treatment of an unusual case of Gastric outlet obstruction

by D. Basude and A Mallikarjuna. *Department of Paediatric Gastroenterology, Bristol Royal Hospital for Children, Bristol, BS2 8BJ, UK.*

We present a case history where a major surgery was prevented by successful therapeutic endoscopic interventions in an child who presented with gastric outlet obstruction.

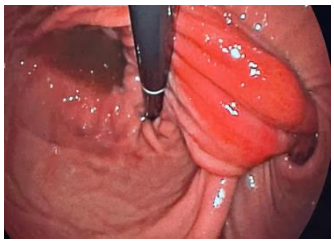
A 15yr old boy presented to Hospital A with a two-week history of acute onset progressive epigastric pain, nausea, vomiting and 15% weight loss. He was previously well and trained at a high level in martial arts but denied any significant injuries.

Fig 1. CT scan abdomen



His blood tests showed liver enzymes raised >3 times the upper limit of the normal, normal synthetic function of the liver and mildly raised amylase. His ultrasound scan of the abdomen showed dilated common bile duct and extrinsic compression of the pancreatic head by jejunum which was confirmed by MRCP. The CT scan of the abdomen suggested invagination of gastric pylorus and antrum into jejunum and external compression of CBD at the ampulla. He was transferred to surgical team in our centre and following a joint review was planned for Gastroscopy followed by Laparotomy with Paediatric Gastroenterologist.

Gastroscopy showed a giant cauliflowler pedunculated polyp atleast 120mm in diameter. This was retrieved into the stomach with alligator forceps. Peicemeal hot snare polypectomy with electrocautery performed and retrieved twelve 20-25mm pieces. Haemostais was achieved and additional hemospray used. Histology showed mucosa with interdigitating smooth muscle bundles characteristic of Peutz-Jegher's polyp. He was admitted in 6 weeks and remaining 8omm poplypectomy performed with endoloop and hot snare polypectomy successfully.



He had no mucocutaneous pigmentation and Video capsule endoscopy did not show any other small bowel polyps. This was a case of sporadic hamartomatous polyp.

We report a successful therapeutic endoscopic treatment of a giant hamartomous polyp presenting with gastric outlet and biliary obstruction.

OC74

Restrospective review of height velocity of male teenagers requiring re-initiation of PN during adolescence

by K. Poulton, T. Johnson, S. Protheroe, W. Haller. *Birmingham Children's Hospital, Steelhouse Lane, Birmingham, B4 6NH, UK.*

Background: During adolescence there is an increased requirement of nutrients to go through puberty and achieve adult height. Nutrition also plays a role in progression of puberty. Malnutrition in chronic disease causes a reduced pubertal growth spurt [1]. Short bowel syndrome (SBS) and enteropathies can result in malabsorption of nutrients and in turn affect their final adult height. Expected linear growth increases from 5cm per year to approximately 9.5cm per year for males during puberty [1].

Methods: We reviewed our home parenteral nutrition (PN) cohort in a tertiary hospital over the last 20 years. Six patients were identified who had successfully weaned off PN, but their growth had become static during adolescence. PN was restarted at this point. Weight and height centile and z-score were documented prior to re-initiation of PN, a year later and at the time when PN was stopped again.

Results: All the patients identified were male, two patients had a diagnosis of trichohepatoenteric syndrome. Four had SBS as a result of neonatal surgery, with varying lengths of remaining bowel at diagnosis (median length was 42.5cm). All six patients had required PN from birth which was stopped at a median age of 5 years and 1 month (range 1 year 4 months to 11 years 9 months). Enteral nutrition was optimised before restarting PN. The median age of restarting PN was 14 years and 7 months (range 13 years 3 months to 15 years 8 months). Five patients stopped PN once adult height was reached. One patient continues on PN and has transitioned to adults. Five had delyaed bone age prior to restarting PN. Other pathological causes of growth failure were ruled out. The median growth rate in the year prior to PN restarting was 4cm per year.

Table 1. Changes in z-score for height

Patient	When reinitiating PN (cm)	When PN stopped and adult height reached (cm)	Z-score change when PN stopped and adult height reached (cm)
A – Trichohepatoenteric syndrome	-1.07	-0.97	+ 0.1
B – Trichohepatoenteric syndrome	-1.22	-1.23	- 0.1
C – SBS	-2.4	- 2.2	+ 0.2
D – SBS	- 2.1	-2.4	- 0.3
E – SBS	-2.22	-1.33	+ 0.89
F – SBS	-2.11	-1.17	+ 0.94

Conclusions: The age at which PN is restarted is important to achieve maximal height velocity. In this cohort the patients who grew the most (patient E and F) started at a median age of 14 years 9 months, the patient (D) with the greatest reduction in z-score was the oldest when restarting PN. In this population of patients with the potential to malabsorb nutrients it is important to monitor their growth closely to ensure their full adult height is achieved.

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OC75

Acid-base status of children on home parenteral nutrition

by L. Abdelgadir, H. Littlechild, S. Montgomery-Stuart, R. Al-Araji, S.M. Hill, J. Köglmeier. *Department of Paediatric Gastroenterology, Unit of Nutrition and Intestinal Failure Rehabilitation, Great Ormond Street Hospital for Children NHS Foundation Trust London, UK.*

The 2018 ESPGHAN guidelines state that only those children receiving long term parenteral nutrition (PN) at high risk to develop acid-base imbalance should have bicarbonate and chloride levels regularly monitored. In June 2022 we introduced regular screening for all patients on home PN (HPN) over concerns that some children may be missed. Our aim was to establish if routine measurement of bicarbonate and chloride status had a clinical impact.

All children attending the intestinal failure (IF) rehabilitation clinic of a large tertiary IF centre who started HPN before December 2022 and had bicarbonate and chloride level checked at least once 3 years before June 2022 till December 2022 were included. 37 patients fulfilled the inclusion criteria (48.6% male, 51.4% female; mean age 11.2 years). Data were collected retrospectively 3 years until June 2022 and prospectively from June to December 2022. Demographics, indication for home PN, bicarbonate and chloride levels, the cause for monitoring and clinical consequences (change in HPN prescription, start of oral supplements, investigations for renal tubular acidosis (RTA) and nephrology consultation) were reviewed.

Indications for HPN: Motility disorders (35.1%), short bowel syndrome (29.8 %) and intestinal mucosal disorders (35.1%). Out of the patients who started home PN before June 2022, 42.2% had levels monitored 3 years prior to June 2022, the rest started monitoring after June 2022 either due to no monitoring at all before or start of HPN after June 2022. Half of these patients had random checks, the remainder were monitored due to renal disease, and gastrointestinal fluid loss. Only 4 (10.8%) patients had low bicarbonate and 3 (8.1%) high chloride levels. 3 patients were referred for renal review, 3 patients had an increase in acetate in HPN, 2 started oral bicarbonate supplements, 1 had their PN fluids increased (some patients underwent multiple interventions).After June 2022 (24.3%) patients had low bicarbonate and 1 patient had a high chloride. Patients developed acid base imbalance due to, gastrointestinal and renal losses, small intestinal bacterial overgrowth (SIBO). 2 patients received antibiotics for SIBO, 2 patients had increased in PN volume made, 2 patients increased in acetate and 1 reduction in PN chloride done. 2 patients received sodium bicarbonate supplements and the remainder had no change in management as minimal abnormal levels only or known RTA. Regular monitoring made a clinical difference in 9 patient (24.3%) and no differences in 28 (75.7%) Only 3 patients, who were previously not routinely screened, had low bicarbonate after start of regular monitoring.

Our data confirm that regular screening of all children PN has a low diagnostic yield and should be done only in those at risk of developing acid base imbalance.

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Hill, S., Ksiazyk, J., Prell, C.(2018). ESPGHAN/ESPEN/ESPR/CSPEN guidelines on pediatric parenteral nutrition: Home parenteral nutrition. Clinical Nutrition, 37(6), 2401–2408.

OC76

Prevalence of diversion colitis in children with gastrointestinal dysmotility: a single centre experience

by T. Bildstein, M. G. Puoti, L. Abdelgadir, E. Margau, K. Lindley, O. Borrelli and K. Nikaki. *Great Ormond Street Hospital for Children, Department of Paediatric Gastroenterology, Division of Neurogastroenterology and Motility, London, WC1N 3JH, UK.*

Diversion colitis (DC) can develop in the defunctioned colon segment. Prevalence of DC in children is unknown. We aim to identify the prevalence of DC in a paediatric population with lower gastrointestinal (GI) motility disorders and/or paediatric intestinal pseudo-obstruction (PIPO) after ileostomy formation.

Data on children seen in our Joint Gastro-Surgical GI Dysmotility clinic between January 2014 and December 2022 for diagnosis of intractable constipation or PIPO who underwent ileostomy formation were obtained retrospectively. Patients with diagnosis of Hirschsprung`s disease, anorectal malformations and inflammatory bowel disease were excluded. Demographics, prevalence of DC, age at DC onset, time from ileostomy formation to DC diagnosis, colonic manometry (CM) result and treatments were reviewed.

Out of 56 patients (mean age 12.1±5.7 years; 27 male), 37 (66%) developed diversion colitis. Mean age at DC onset was 10±4.9 years. Mean time from ileostomy formation to onset of DC was 37.5±30.9 months with shortest period of 3.1 and longest of 116 months. Thirty-nine patients had diagnosis of intractable constipation and colonic dysmotility on CM and 17 patients had PIPO. There was no difference between the two groups in the prevalence of DC (24/61.5% vs 13/74.5%; p=0.2). As treatment, short chain fatty acids was given in 5 patients, topical steroids in 4, systemic steroids in 2, probiotics in 1 and immunosuppressant in 1.

DC in children with lower GI dysmotility or PIPO presents in two-thirds of children undergoing an ileostomy formation. Onset of DC can be present as early as 3 months post-operatively. Further studies are needed to define predictive factors for development of this condition and preventative treatments.

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OC77

The additional burden of having a developmental behavioural disability (DBD) in children with chronic functional constipation

by E.P. Athanasakos, C. McLaughlin, P. Handley and S. Cleeve. *Barts Health NHS Trust and Queen Mary University of London. The Royal London Hospital, c/o Paediatric Surgery Secretary 7th Floor, Royal London Hospital, Whitechapel, E1 1FR, UK.*

Introduction: Developmental behavioural disabilities (DBD) including autism spectrum disorder (ASD), attention and other learning difficulties is well known to be associated with significantly high rates of gastrointestinal problems, including constipation and faecal incontinence (CCFI), imposing a significant burden on child and the family (1). A child living with DBD has both impact on the child and their families' quality of life (QoL) (2). It is also well recognised that having a child with CCFI has an impact on the child and families QoL (3,4). Yet, understanding the impact of having a child both with CCFI and DBD compared to a child without DBD, remains rudimentary (1). We aim to compare bowel and QoL outcomes for patients and their families with DBD and CCFI, compared to patients without DBD.

Methods: Patients with CCFI who presented to our specialised service from the period of September 2016 and September 2022 were included. Measures included: demographics, bowel scores: St Marks Incontinence Scores (SMIC) and Cleveland Constipation Scores (CCS), risk of distress: Paediatric Index of Emotional Distress (PI-ED), quality of life for patient and family (Pediatric Quality of Life Inventory) and patient perception of severity of their condition using Wong Baker smile faces: scale of 0-10 (10 being the worst).

Results: Out of 341 patients with functional constipation, 29% (100/341) had DBD. There was a male predominance of males with DBD and CCFI compared to patients without DBD. There were no significant differences between the groups regarding bowel scores and perception of severity of their condition. Patients with DBD and CCFI, significantly demonstrated lower Ped-QL for the patient and family impact module ($p<0.05$) for all parameters, except 'worry'. Patients with DBD and CCFI were also significantly higher risk of distress compared to patients without DBD ($p<0.001$). Refer to Table 1.

Conclusion: This study demonstrates the additional burden of having DBD and CCFI in patients and their families:

- QoL and risk of distress is significantly worse in patients with both CCFI and DBD.
- It is impossible to separate the relative contribution CCFI and DBD to poor QoL.
- A novel approach is necessary to intervene in the perpetual deterioration in symptoms and QoL
- .Earlier recognition and clinical assessment of DBD and CCFI is urgently necessary.A pathway that identifies and manages children at risk of poor QoL related to DBD and CCFI should be explored.

Table 1: Outcomes Measures

MEASURES	CCFI n =241 M, ± *(range)*	CCFI with DBD n = 100 M, ± *(range)*	Significance (p value)
Sex	117 males; 124 females	72 males; 28 females	**p<0.001**
Age	M: 9 ± 3.75 *(1-18)*	M: 10 ± 4.23 *(1-27)*	NS
Perception of Severity	M:8 ± 2.09 (1-10)	M: 8 ± 1.85 (2-10)	NS
Bowel Scores			
SMIC	M: 12 ± 6.52 (0-24)	M: 13 ± 6.04 (0-24)	NS
CCS	M: 16 ± 5.33 (1-30)	M: 16 ± 4.98 (0-27)	NS
Abdominal pain severity	M: 7 ± 2.49 (0-10)	M: 7 ± 2.41 (0-10)	NS
PI-ED	M: 15 ± 7.45 (1-35)	M: 18 ± 7.01 (4-42)	**p<0.001**
Peds-QL (patient)			
Total QoL	M: 67 ± 20.48 (7-100)	M: 51 ± 18.97 (1-93)	**p<0.001**
Emotion	M: 57 ± 25.79 (0-100)	M: 44 ± 22.47 (0-90)	**p<0.001**
Social	M: 76 ± 24.66 (0-100)	M: 56 ± 31.53 (0-100)	**p<0.001**
School	M: 65 ± 23.36 (0-100)	M: 47 ± 31.09 (0-100)	**p<0.001**
Physical	M: 66 ± 23.36 (0-100)	M: 52 ± 26.08 (0-100)	**p<0.001**
Psychological	M: 69 ± 23.09 (0-100)	M: 52 ± 24.32 (0-100)	**p<0.001**
Peds-QL (Family Impact)			
Total QoL	M: 62 ± 22.31 (0-100)	M: 51 ± 19.48 (0-100)	**p<0.001**
Parent QoL	M: 65 ± 22.31 (0-100)	M: 53 ± 20.99 (5-91)	**p<0.001**
Family QoL	M: 70 ± 23.40 (12-100)	M: 59 ± 25.27 (3-89)	**p<0.001**
Physical	M: 62 ± 25.94 (11-100)	M: 51 ± 25.60 (0-100)	**p<0.001**
Emotion	M: 56 ± 27.91 (3-100)	M: 48 ± 25.97 (0-100)	**p<0.001**
Social	M: 73 ± 25.60 (0-100)	M: 53 ± 28.88 (0-100)	**p<0.001**
Cognitive	M: 70 ± 28.17 (0-100)	M: 62 ± 28.11 (0-100)	**p<0.05**
Communication	M: 66 ± 33.35 (0-100)	M: 47 ± 26.85 (0-100)	**p<0.001**
Worry	M: 38 ± 23.95 (0-100)	M: 35 ± 24.21 (0-100)	**NS**
Family Daily Impact	M: 58 ± 31.80 (0-100)	M: 42 ± 33.68 (0-100)	**p<0.001**
Relationships	M: 78 ± 24.57 (0-100)	M: 70 ± 29.86 (1-100)	**p<0.05**

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OC78

Awake high resolution anorectal manometry: does age affect successful testing?

by C. Bingham, E. White, M. Papadopoulos and M. Mutalib. *Evelina London Children's Hospital, Westminster Bridge Rd, London, SE1 7EH, UK.*

High resolution anorectal manometry is useful tool for determining anorectal function, but can be difficult to perform the full awake procedure in paediatric settings due to the stigma surrounding intimate procedures on children and anxiety on the part of the young person, and many centres mainly investigate anorectal function under sedation. At our tertiary centre, we routinely perform awake high resolution anorectal manometry on children and young people of all ages with only rare occasions where no data at all is collected.

We retrospectively reviewed the last 100 patients to attend our centre for awake high resolution anorectal manometry and collected data on whether each of the six tests in the protocol (from the BSPGHAN working group consensus [1]) were able to be performed and compared across five age groups.

The resting pressure test was almost universally successful across all age groups. As expected, almost all other tests were more successful as the children got older and able to better follow instructions and understand what was being requested of them. Of note however, the recto-anal inhibitory reflex (RAIR) showed a lower success rate in older children. On investigation, it appears that all RAIR test failures were a result of hyper sensitivity to the inflation of the balloon and active contraction of the sphincter in response to the sensation. These children has a subsequent positive RAIR under sedation. We theorise that older children are more likely to be aware of the urge to pass stool triggered by the RAIR testing and therefore more concerned about having an accident during the procedure than a younger child might be.

The relative success rates of each test will be of interest to referrers to predict what information they are likely to get out of a request for awake high resolution anorectal manometry – older children are more likely to be compliant for the entire protocol, but a majority of under 5s can produce meaningful data for at least half of the tests. Endurance squeeze and sensation testing proved the most challenging for our youngest group, likely due to a lack of understanding of the test requests, but can still be attempted where previous compliance with the testing protocol has been demonstrated.

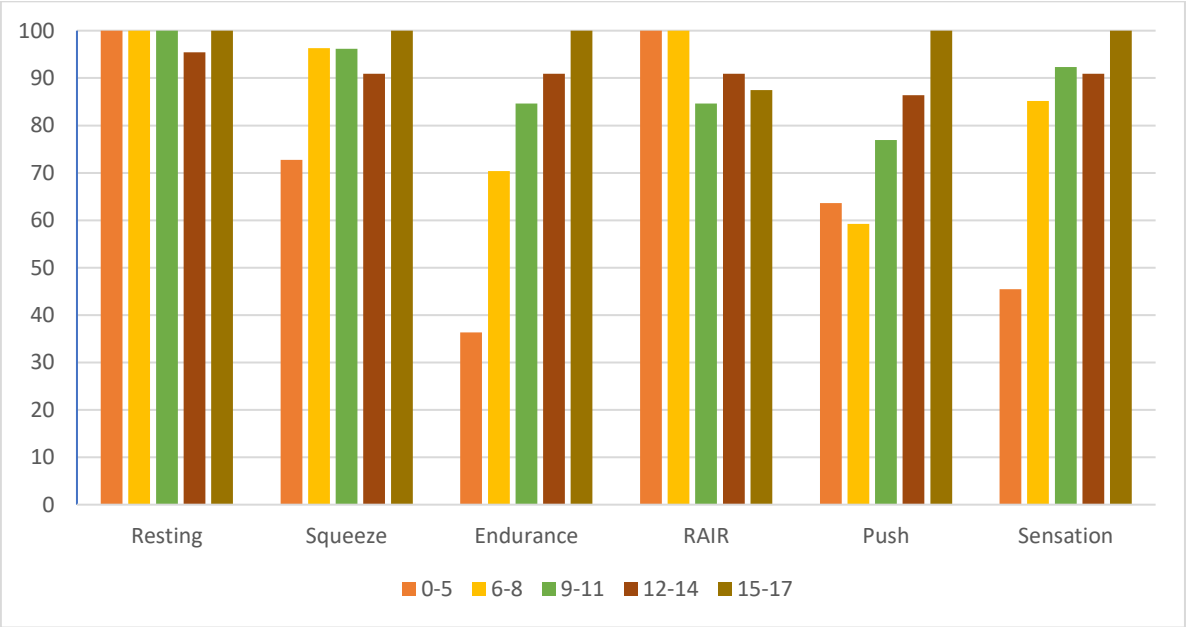

Figure 1: Percentage success of each test in the ARM protocol by age

References:

[1] Athanasakos E, Cleeve S, Thapar N, et al. Anorectal manometry in children with defecation disorders BSPGHAN Motility Working Group consensus statement. Neurogastroenterol Motil. 2020; 32:e13797. https://doi.org/10.1111/nmo.13797

OC79

Case Report: Gastro-oesophageal junction outlet obstruction with subsequent rumination syndrome

by C. Bingham, E. White, M. Papadopoulos and M. Mutalib. *Evelina London Children's Hospital, Westminster Bridge Rd, London, SE1 7EH, UK.*

An 11 year old child was referred to our institution with a 3 year history of persistent and distressing regurgitation episodes. A liquid barium study was normal, but a modified, solid based study showed hold up at the lower oesophageal sphincter (LOS). An endoscopy was normal and a dilatation of the LOS to 20mm provided 8 months of symptomatic relief. However, the regurgitation then reoccurred and a repeat dilatation did not result in any symptom improvement.

At our institution, a high resolution oesophageal manometry (HROM) showed intact peristalsis and median integrated relaxation pressures (IRP) of 37.6 mmHg for wet swallows and 34.9 mmHg for solid swallows suggestive of gastro-oesophageal junction (GOJ) outlet obstruction.

An EndoFLIP Topography showed normally propagated contraction, but non relaxing LOS and a repeat LOS dilatation produced no symptomatic relief. A botulinum toxin injection into the LOS resulted in a good symptomatic improvement. A repeat HROM three months later showed normal peristalsis and median IRP had improved to 19.8 mmHg for wet swallows and 14.7 mmHg for solid swallows. There was no bolus hold up in the distal oesophagus.

Nine months after the botulinum toxin injection, the child reported recurrence of regurgitation and a further HROM, 11 months post botulinum toxin injection, showed median IRP values of 6.0 mmHg for wet swallows and 10.5 mmHg for solid swallows. Abdominal contractions with retrograde bolus movement were seen, a feature consistent with rumination syndrome.

In conclusion, we present a partial response to pneumatic dilatation in child with idiopathic GOJ outlet obstruction, but good symptomatic response to botulinum toxin injection and subsequent development of typical clinical and manometric features of rumination syndrome.

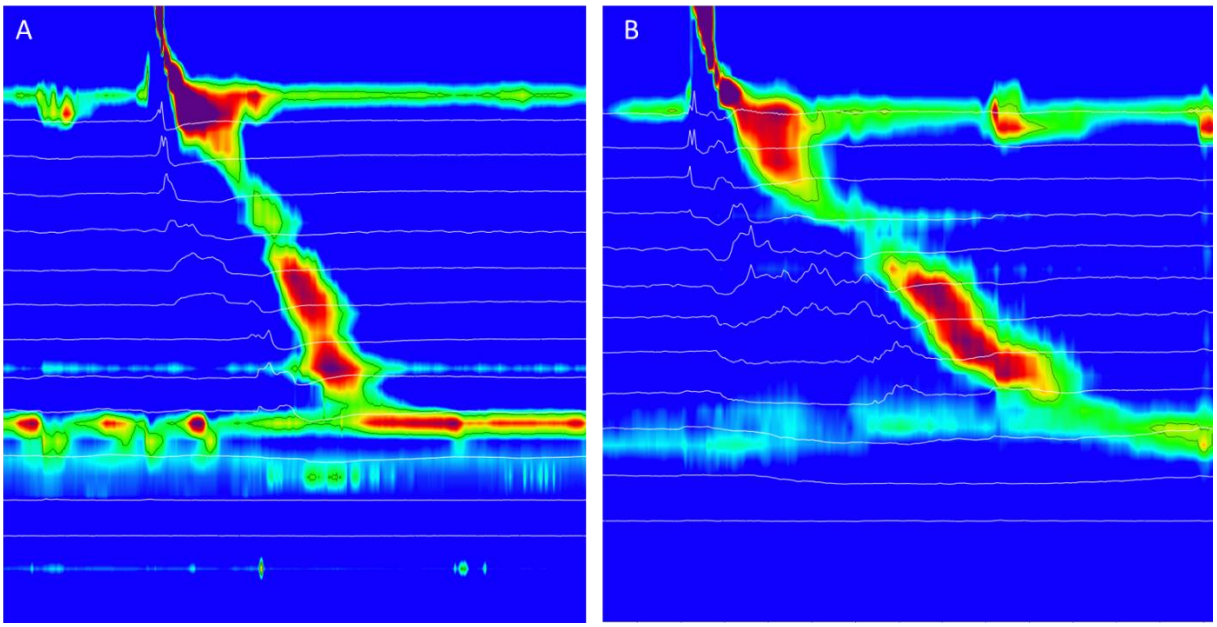

Figure 2: A: Manometry pre-intervention, IRP 38mmHg, B: Manometry post-Botox, IRP 15 mmHg

OC80

High resolution oesophageal manometry post pneumatic dilatation in children with achalasia, a single centre experience

by C. Bingham, E. White, M. Papadopoulos and M. Mutalib. *Evelina London Children's Hospital, Westminster Bridge Rd, London, SE1 7EH, UK.*

Achalasia is a rare motility disorder of the oesophagus, with an incidence rate of 0.38/100,000 children per year in the UK [1]. The characteristics of achalasia are a non-relaxing lower oesophageal sphincter (LOS) and failure of peristalsis. High resolution oesophageal manometry (HROM) is the gold standard for diagnosis and allows achalasia to be divided into three sub-types dependant on the nature of the peristaltic failure. There are a variety of treatment options available for achalasia, including medications, pneumatic dilatation, injection of botulinum toxin and myotomy - all focused on reducing the pressure at the LOS. There is no standard consensus of evidence based for the optimum treatment modality in paediatric achalasia.

Pneumatic dilatations have high success rate on Type I (absent peristalsis) and Type II (pan-oesophageal pressurisations) achalasia [2]. In our institution, children with achalasia (type I&II) will undergo a series of 3 dilatations (rigid balloons), 4 weeks apart by the interventional radiologist and a regular clinical assessment. A repeat HROM will be considered after the last dilatation to assess the response to the intervention.

We present a series of three patients (2 type I and 1 type II, mean age 9.5 years) who underwent HROM pre and post dilatation. Following dilatation, both Type I achalasia patients reported symptomatic relief (reduction in Eckardt Score) and showed significant reduction in both wet and solid IRP values. The patient with Type II achalasia reported only mild symptomatic relief for a short period and showed progression of their condition on repeat HROM.

It is not uncommon for patients with achalasia to report symptoms post interventions (dilatations or myotomies), so it is salient to ensure the appropriate assessment is undertaken prior to escalation or change of therapy.

In conclusion, we report a small case series of HROM findings in children with achalasia before and after pneumatic dilatation enabling tailor made management intervention.

				Pre-dilatation IRP		Post-dilatation IRP	
Age	Type	No. Dilatations	Max Size	Wet	Solid	Wet	Solid
5	2	4	22mm	28	25.2	48	60.1
10	1	4	30mm	44.5	58.8	18.1	16.4
14	1	5	30mm	21.8	17.6	2.9	7.3

Table 1: Manometric data pre and post dilatation

References:

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OC81

Oesophageal function test in children with intractable vomiting with no organic cause – is it beneficial?

by A. Mallikarjuna, L. Griffin, J. Porter. *Bristol Royal Hospital for Children, Bristol, BS2 8BJ, UK.*

Stationary or ambulatory high-resolution manometry (HRM) and intraluminal pH-impedance monitoring can evaluate oesophageal function and monitor gastroesophageal reflux. These tests can be used to diagnose gastroesophageal reflux disease, achalasia, rumination syndrome, aerophagia, and supragastric belching[1].

We did a retrospective review of case notes of the patients diagnosed with functional vomiting or rumination who were referred for High-Resolution Oesophageal Manometry (HRM) from December 2021 until December 2022 to understand the role of oesophageal function tests. We collected data on patient demographics, results of the Oesophageal function test (OFT) and clinical outcomes.

During this period, 19 patients were referred for HRM and 10 of them were for functional vomiting or rumination. These patients were needing intense medical input in the form of prolonged hospital stays, recurrent presentations in the emergency department, or significant weight loss to need enteral feeding support. The age of these patients ranged between 10-16 years (mean 12.5 years), 3 were boys and 7 were girls.

4/10 patients had abnormal manometry results in the form of increased gastric pressure (3/4) and increased pressurisation in the mid-oesophagus (1/4). 5/10 patients had abnormal results on the pH/impedance study. Among them, 2/5 patients showed increased supine acid exposure, 1/5 patients showed raised acid exposure with sipping of low pH fluid and 2/5 patients showed aerophagia before vomiting.

3/10 patients showed symptom co-relation with the results. 4/10 patients who were diagnosed with rumination syndrome either clinically or with OFT had biofeedback. On follow-up over variable periods, 3 patients continued to have ongoing vomiting, 3 patients were on nasogastric feeds, 2 patients were on nasojejunal feeds and 2 patients had recovered completely with no requirement of nutrition support.

To the best of our knowledge, no studies are looking at the benefits of OFT in children with functional vomiting. Although rumination is a clinical diagnosis, high-resolution oesophageal impedance manometry is considered as a gold standard to support the diagnosis but not in clinical management[2]. Our study shows that OFT can help to reassure patients/parents and also provide useful techniques via biofeedback to overcome their symptoms. It also guided our clinical psychologists in their approach to these patients. We conclude that these tests are beneficial if used in the right context. However, we understand there are limitations in our study being a retrospective study with a low sample size. A larger prospective study can help to better understand the role of OFT in this group of patients.

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OC82

Service evaluation of paediatric pH and combined multiple intraluminal impedance – pH studies in Maidstone and Tunbridge Wells NHS Trust, UK

by E. Ajayi, S. Fayed, S. Kanchan, S. Khan, R. Gowda, B..Bhaduri, V. Kolimarala, H. Race, C. Taylor, L. Fuentes. *Maidstone and Tunbridge Wells NHS Trust, UK.*

Gastro-oesophageal reflux disease (GORD) is fairly common in the paediatric population with approximately 10-15% of children affected by it, with predominance in infancy1. Gastro-oesophageal reflux (GOR) can be objectively assessed by ph and/or combined multichannel intraluminal impedance (MII) monitoring which allows detection of most gastroesophageal reflux episodes, acid as well as non-acid reflux.

Objectives:

1. The aim of this evaluation was to measure the current practice and to provide recommendations for improvement.
2. Determine the diagnostic yield of this technique in children with suspected reflux symptoms.

Patients and Methods:

The recordings of 245 children from 2007 to 2022 were collected, including whether endoscopy was performed and if a chest x-ray was done to detect the placement of the catheter. Reflux index of more than 4.2% was used as the cut-off for a presence of GOR in a patient. For combined MII studies, a cut off 70 episodes in less than 1 year age and 100 episodes in more than 1 year age was taken as a positive finding. The temporal relationship between symptoms and reflux episodes was analysed with symptom index (SI) >50% and a symptom association probability (SAP) > 95% considered indicative of a positive association.

Results:

A total of 245 studies from 2014 to 2022 were evaluated. A total of 198 pH studies and 47 MII were recorded. There were 157 boys and 88 girls with an age range of 0.1-18.0 years. More studies were done in 2015 and 2019, 44 each and the least was performed in 2007. The average duration of a study was 22 hours. Majority of studies (81), about 33% were performed in the 6 to 12 years category and about 17% (42 patients) were done in children less than 1 year of age. In 149 (60.8%) children an endoscopy was also performed. In the 245 studies, only 89 (36.3%) had x-rays to confirm the position of catheters. A significant presence of GOR, using Reflux index >4.2%, was found in 67 of the 245 studies (27.3%) evaluated. 24 patients (57%) in the less than 1 year age group had more than 70 episodes documented while there were 21 patients (10%) in the more than 1 year age group who had more than 100 episodes. A positive SAP was found in 48 (19.6%) patients including acid reflux in 42 (17%) and non-acid reflux in 6 (2.4%). A positive SI (>50%) was found in 50 (20.4%) patients, including acid reflux in 46 (18.8%) and non-acid reflux in 4 (1.63%).

Conclusion:

Our analysis found a significantly higher incidence (27%) of GOR in the children that we studied and perhaps this was a selected group being seen in a specialist paediatric gastroenterology clinic. There was also a significant proportion (57%) of children in the less than 1 year age group to be having increased number of reflux episodes and the perhaps we are capturing a normal physiological process. We also recommend that confirming tip position during endoscopy is a relatively safe alternative to performing a chest x-ray to confirm catheter placement. Combined pH-MII studies are recommended for a better diagnostic yield and symptom correlation, especially in the younger age group.

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OC83

The association of interoception in children with developmental behavioural disability (DBD) and chronic functional constipation

by E.P. Athanasakos, C. McLaughlin, P. Handley, S. Cleeve. *Barts Health NHS Trust and Queen Mary University of London. The Royal London Hospital, c/o Paediatric Surgery Secretary 7th Floor, Royal London Hospital, Whitechapel, E1 1FR, UK.*

Introduction: Developmental behavioural disabilities (DBD) including autism spectrum disorder (ASD), attention deficit disorder and other learning difficulties have been linked to chronic constipation and faecal incontinence (CCFI) (1-3). In children, ASD have impairments in interoception (4-5). Interoception is the ability to sense the internal state of the body—for instance, to accurately identify sensations such as hunger, thirst, pain, and internal temperature. Knowledge about the function (specifically rectal sensation) of the anorectum in children with DBD and CCFI, using diagnostic investigations such as high resolution anorectal manometry (HRAM) is unknown. This study aims to investigated the symptomatologic and physiological outcomes in children with DBD and CCFI, compared to children without DBD.

Methods: Patients with CCFI who presented to our specialised service from the September 2016 and September 2022 were included. Measures included: demographics, bowel scores: St Marks Incontinence Scores (SMIC) and Cleveland Constipation Scores (CCS), HRAM parameters and colonic transit x-rays.

Results: Out of 341 patients with functional constipation, 29% (100/341) had DBD. There was a male predominance of males with DBD and CCFI compared to patients without DBD. There were no significant differences between the groups regarding bowel scores and HRAM parameters ($p>0.05$). However, one rectal sensation parameter ('urge to defaecate) was significantly delayed in patients with DBD and CCFI compared to those patients without DBD ($p< 0.005$). Refer to Table 1.

Conclusion: This is the first study, to demonstrate the possible link between interoception and impaired physiological outcomes.

- The interaction of interoception and DBD with CCFI is poorly understood
- Patients with DBD and CCFI have delayed rectal sensation to defaecate compared to patients without DBD.
- Further original work on interoception and DBD and CCFI is required.
- We will pilot the Multidimensional assessment of interoceptive awareness (MAIA) questions (5) in patients with DBD and CCFI.
- Specialist and experienced multi-disciplinary input are likely to be necessary to improve prognosis in children with DBD and CCFI

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Table 1: Outcomes Measures

MEASURES	CCFI n =241 M, ± *(range)*	CCFI with DBD n = 100 M, ± *(range)*	Significance (p value)
Sex	117 males; 124 females	72 males; 28 females	**p<0.001**
Age	M: 9 ± 3.75 *(1-18)*	M: 10 ± 4.23 *(1-27)*	NS
Urinary Incontinence	(38%)	(43%)	NS
Perception of Pain			
HRAM	M:3 ± 2.16 (0-10)	M: 3 ± 2.64 (0-10)	NS
Venipuncture	M:6 ± 3.44 (0-10)	M: 6 ± 3.87 (0-10)	NS
Perception of Severity	M:8 ± 2.09 (1-10)	M: 8 ± 1.85 (2-10)	NS
HRAM			NS
Anal Canal (cm)	M: 2.20 ± 0.37	M: 2.13 ± 0.34	NS
Resting pressure	M: 57.40 ± 14.95	M: 58.11 ± 13.60	NS
Squeeze increment	M: 112.53 ± 37.83	M: 109.75 ± 33.43	NS
Maximum squeeze increment	M: 145.17 ± 55.68	M: 142.57 ± 51.20	NS
Enhanced squeeze	M: 116.82 ± 60.48	M: 108.25 ± 37.29	NS
Rectal Sensatioin			
First sensation	M: 41.82 ± 40.08	M: 50.58 ± 43.56	NS
Urge to defaecate	M: 74.82 ± 48.13	M: 93.88 ± 65.81	**p <0.005**
Maximum Tolerable Volume	M: 153.31 ± 64.58	M: 164.04 ± 66.80	NS
RAIR	97% * present	98% * present	NS
Dyssynergia	(54%)	(53%)	NS
Colonic Transit x-ray	47% Normal 35% RED 21% STC	47% Normal 39% RED 14% STC	NS
Bowel Scores			
SMIC	M: 12 ± 6.52 (0-24)	M: 13 ± 6.04 (0-24)	NS
CCS	M: 16 ± 5.33 (1-30)	M: 16 ± 4.98 (0-27)	NS
Abdominal pain severity	M: 7 ± 2.49 (0-10)	M: 7 ± 2.41 (0-10)	NS

OC84

Efficacy of Pranic Healing-Energy Therapy in the Management of Chronic Pain and Associated Medical Condition in Children and Adolescents

by L. Flitcroft, B. Hope, H.M. Lee and A. Bhatia. *Paediatric Liver, GI and Nutrition Centre, King's College Hospital, London, SE5 9RS, UK.*

Pranic Healing is a form of complementary therapy that is used to help alleviate symptoms of chronic pain and illness. Our study aims to evaluate the efficacy of Pranic Healing as a natural complementary treatment for symptoms of chronic pain and clinical symptoms associated with Disorder of Brain-Gut Interaction (DGBI) and Inflammatory Bowel Disease (IBD) in paediatric and young adult patients.

This is a prospective, longitudinal, qualitative and quantitative experimental study. Inclusion criteria: 5-24 years old, chronic pain, patients with a diagnosis of DGBI or IBD. Patients underwent 8 weekly Pranic Healing sessions. Pranic Healing protocols synonymous with patients' physical and psychological medical diagnosis were applied. Routine medical care was continued during treatment. Data collected included Pain–Visual Analogue Scale (P-VAS), medication dosage and frequency, and symptoms journal.

The primary outcome was the improvement in the intensity and frequency of pain. Secondary outcomes were improvements in the intensity and frequency of nausea, diarrhoea, constipation, vomiting, and sleep disturbance due to symptoms. The tertiary outcome was the improvement in patient anxiety.

38 participants were recruited (32 DGBI, 6 IBD) between July - Nov 2022. M: F ratio was 14:24. Median age was 13 years (range 7– 20 years). At the end of the 8-week treatment, patients' clinical responses to treatment were evaluated and categorised as Complete Clinical Response, Partial Clinical Response or No Clinical Response (data available for Phase 1 and 2 patients). At Week 16-20 (8-12 weeks after the last session), patients' clinical responses were further evaluated (data currently available for Phase 1 patients).

Table 1 Results

PHASE 1 Patients (8-week treatment from Jul-Sept 2022) and Week 16-20 review (Oct-Nov 2022)										
Review	**N**	**PRIMARY OUTCOME**			**SECONDARY OUTCOME**			**TERTIARY OUTCOME**		
Week 8		CCR	PCR	NCR	CCR	PCR	NCR	CCR	PCR	NCR
DGBI	16	11	5	0	11	5	0	11	5	0
IBD	3	1	2	0	1	2	0	1	2	0
Week 16-20		CCR	PCR	NCR	CCR	PCR	NCR	CCR	PCR	NCR
DGBI	16	14	2	0	14	2	0	14	2	0
IBD	3	2	1	0	1	2	0	2	1	0
PHASE 2 Patients (8-week treatment from Sept - Nov 2022)										
Review	**N**	**PRIMARY OUTCOME**			**SECONDARY OUTCOME**			**TERTIARY OUTCOME**		
Week 8		CCR	PCR	NCR	CCR	PCR	NCR	CCR	PCR	NCR
DGBI	16	11	5	0	11	5	0	11	5	0
IBD	3	2	1	0	2	1	0	2	1	0

CCR (Complete Clinical Response) – Complete relief of symptom(s)
PCR (Partial Clinical Response) – Discernible improvement in symptom(s)
NCR (No Clinical Response) – No change in symptom(s) **N**= Number of patients

Our preliminary study shows that whilst continuing with medical care, Pranic Healing appears to enhance the relief of symptoms associated with DGBI and IBD. Further review at 8-12 weeks after Pranic Healing treatment completion showed sustained or further improvement of symptoms. More studies are needed to provide sufficient scientific evidence of its efficacy as a complementary therapy.

OC85

Use and abuse of oral rehydration solutions in a hospital setting: an urgent need for re-education of clinicians to prevent iatrogenic illness and promote cost savings

by. G. Yan, V. Suri, J. Koeglmeier and K. Lindley. *Department of Gastroenterology, Great Ormond Street Hospital for Children, Great Ormond Street, London, WC1N 3JH, UK.*

Oral rehydration solutions (ORS) utilise the electrochemical gradient for sodium to drive electrogenic sodium coupled glucose and water absorption across the small intestinal epithelium. These solutions have proven to be a lifesaving therapy worldwide in acute diarrhoeal diseases. The precise composition of these solutions varies depending upon clinical indication with higher sodium solutions being used where secretory diarrhoeas, such as cholera, are prevalent and hypo-osmolar lower sodium solutions being used in developed countries where other forms of diarrhoea are prevalent.

In the UK, Dioralyte is the most common ORS used in paediatrics. It is a hypo-osmolar solution with an illustrative composition of sodium (Na) 60mmol/L, potassium 20mmol/L, glucose 111mmol/L, chloride 40mmol/L and citrate 10mmol/L with an osmolality of 240 mosm/Kg. The composition of this solution is optimised for rehydration rather than electrolyte replacement, where other solutions are more appropriate.

There is a current national shortage of Dioralyte expected to last until the end of 2022. This has prompted us to review current use of Dioralyte in clinical practice and consider alternative solutions.

We undertook a point prevalence audit of the use of ORS (Dioralyte) in a tertiary paediatric referral hospital. We evaluated the most recent prescriptions of Dioralyte in 50 patients across the Trust (August-September 2022).

Only 6% (3/50) patients were being treated by the Gastroenterology Department. 90% (45/50) required feeding support via nasogastric tube, nasojejunal tube, gastrostomy, jejunostomy and/or parenteral nutrition. Only 18% (9/50) patients had acute diarrhoea. 34% (17/50) received Dioralyte whilst fasting for a procedural anaesthetic. Dioralyte was used to build up feed volume over time in 16% (8/50) and to meet fluid target requirement in 8% (4/50). Dioralyte was also prescribed for constipation, dystonia, hypokalaemia, high nasogastric tube losses and as part of bowel prep for colonoscopy. No intravenous access was stated as the rationale for starting Dioralyte in 14% (7/50). In 3 patients the ORS was being used to replace secretions with high Na content (typical Na content of losses was 120-150 mmol/L).

Overall we found 82% of ORS prescriptions were for non-diarrhoeal illnesses and thus widespread overuse of ORS.

Clinicians need to be educated about the indications for ORS and the choice of appropriate ORS for each clinical situation. This point prevalence audit has highlighted the potential substantial cost savings that could be made within the NHS and acknowledges the significant risks of a “one glove fits all” strategy for the use of ORS in healthcare settings.

OC86

Differential superior mesenteric artery and coeliac axis flow demonstrated by doppler ultrasound in median arcuate ligament syndrome

by P. Richardson, L. McDonald, N. Davidson, D.I. Campbell, *Great North Children's Hospital, Newcastle Upon Tyne, NE1 4LP, UK.*

Median arcuate ligament syndrome (MALS) is secondary to compression of the coeliac artery by the median arcuate ligament[1] with secondary critical reduction in blood flow to the proximal duodenum and stomach. The diagnosis of MALS is problematic due to the frequent overlap with functional symptoms.[2] Doppler sonography may be used as an effective/ primary imaging modality in the initial assessment of MALS which may remove the gold standard examination of CT angiography and limit the use of ionising radiation.[3]

A 14 year old girl presented with post-prandial abdominal pain, nausea and associated weight loss. She was under outpatient follow-up for a diagnosis of Postural Orthostatic Tachycardia syndrome and hypermobility (non-genetic Ehler's Danlos). Investigations for her abdominal pain were normal (bloods, stool FCP, abdo ultrasound, barium meal). Upper GI endoscopy was also normal. She required a prolonged inpatient stay post endoscopy (pain and parasyncope) and was established on nasogastric feeds but reported ongoing pain and escalating vomiting and weight loss. CT angiography of the abdomen and pelvis was performed demonstrating an ostial stenosis (1mm orifice) at the origin of the coeliac axis, a characteristic hooked appearance at the location of the median arcuate ligament and post stenotic dilatation confirmed on axial slices and visually augmented using 3D reconstruction. Abdominal doppler USS, comparing SMA to CA flow, showed high velocity post stenotic flow in the coeliac axis (400cm/s); double the value measured at the origin of the SMA (150cm/s). She has now been established on jejunal feeds with excellent improvement in symptoms and no current plan for any surgical intervention. If symptoms persist she will have division of the MAL.

Merging the CT findings with the Doppler appearances suggest US may have a role in the early imaging assessment of a rare condition that is often a diagnostic conundrum.

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OC87

Intestinal lymphangiectasia- a diagnostic challenge in a child with complex gastroschisis

by S. Rajani, K. Green, S. Mathai, M. Cohen, A. Urs, M. Thomson, F. Shackley, R. Lindley, S. Sharma.
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Intestinal lymphangiectasia is a rare congenital/acquired malformation where impaired lymph drainage leads to lymph leakage in the intestinal lumen and/or peritoneum. Epidemiology is unknown. There are no well-established management guidelines. A review of clinical notes was undertaken along with review of literature.18-month-old female, born at 34 weeks with complex gastroschisis, duodenal and jejunal atresia needing silo reduction followed by laparotomy and duodenal jejunal anastomosis. She required parenteral nutrition (PN) for 2 months and was discharged at 3 months of age. She was lost to follow up and presented with anasarca, chronic diarrhoea, electrolyte imbalance, metabolic acidosis, failure to thrive and persistent hypoalbuminemia at 18 months. She received PN and attempts to feed her led to diarrhoea and hypoalbuminemia. Endoscopy revealed a no-fibrotic colonic stricture 50 cm from anal verge requiring dilatation. She received immunoglobulins for hypogammaglobulinemia. She was also noted to have persistent norovirus in her stools. A clinical diagnosis of protein losing enteropathy was made based on hypoalbuminemia and raised faecal alpha-1 antitrypsin (7.47 mg/g ww) however gastro-duodenoscopy was normal. She was trialled on modular feeds with low fat and medium chain triglyceride. Her genetic screen for R-15, R-33 panel were normal. Whole genome sequencing revealed GLMN variant associated with glomerular malformation. In view of persistently raised alpha-1 antitrypsin she underwent Magnetic resonance lymphangiogram which did not suggest any lymphatic anomalies. A double-balloon balloon enteroscopy revealed mid-jejunal lesions consistent with lymphangiectasia approximately 40 cm from the pylorus. She was then commenced on sirolimus with a favourable response.

Intestinal lymphangiectasia should be considered in context of diarrhoea, hypoalbuminemia and raised alpha-1 antitrypsin. Majority of the cases respond well to dietary modification. Small bowel resection may be needed. Radiological investigations alone cannot refute the diagnosis and luminal view is probably the best modality to rule out lymphangiectasia.

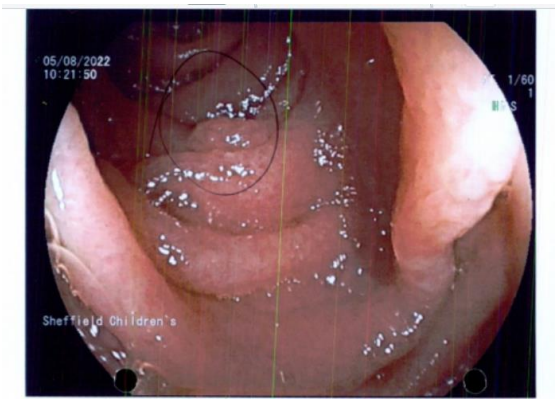

Figure1: Endoscopic appearance of lymphangiectasia

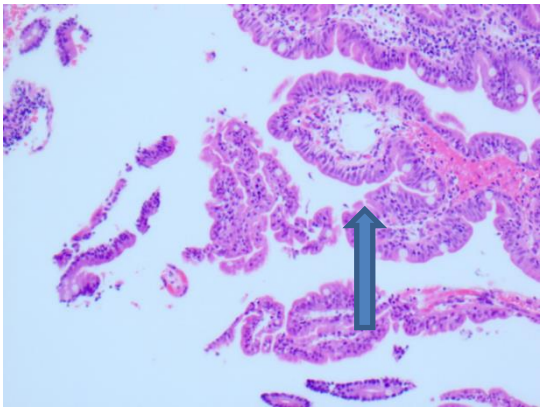

Figure2: Microscopic appearance of intestinal lymphangiectasia

OC88

An evaluation of clinical communication methods between centres in a shared-care setting

by S. Gulam Khader, H. Kannappan. *University Hospitals Coventry and Warwickshire NHS Trust, Clifford Bridge Road, Coventry, CV2 2DX, UK.*

After each of the 5 million outpatient clinic appointments that occur every month in the National Health Service (NHS), a letter containing a summary and management plan is written by the clinician and sent to the patient's general practitioner (GP); a copy to the patient and to the tertiary care team if the patient is under a shared-care agreement.(1) Outpatient clinic letters remain the primary means of correspondence across the interface between primary, secondary and tertiary specialists and are crucial in ensuring that both patients and clinicians receive up-to-date essential information to ensure continuity of care. However, every so often these letters may not meet the information needs of the recipients.

Our study aims to address the issue of variability in the method of communication between the secondary and tertiary care specialists to improve the quality and standards of care. In the context of paediatric inflammatory bowel disease, we analysed 99 unselected clinic letters of Crohn's disease patients presenting in the year 2022 in a university paediatric gastroenterology clinic. The sample included clinic letters from the secondary-care hospital and the local tertiary centre that shared the clinical responsibility of these patients. The letters were analysed and the following items were rated as essential information: diagnosis, latest blood results, latest endoscopy and colonoscopy reports current medications with dose, anthropometry, examination findings, current management and the follow-up plan. The most obvious finding to emerge from this study is that although the letters commonly contained details on diagnosis and examination findings, around one-third of the letters lacked information on recent blood results and the latest biopsy reports. Secondly, around 10% of the letters did not contain updated information about the current medication and anthropometry of the child. Taken together, it can be suggested that the inconsistency in the information provided in the outpatient letters can result in confusion and difficulty in understanding the contents of the letter both for the patients and general practitioners.

A reasonable approach to tackle this issue is to introduce a common structured approach to the content of outpatient clinic letters between the secondary and tertiary care units that include the use of headings such as diagnosis, previous investigations, anthropometry, current treatment, recent blood results and forthcoming follow-up plan. We believe that this change has several benefits including improving continuity of care, avoiding the need to check previous letters for important information and enhancing the patient-centredness of clinicians' communications. Alternatively, a shared-care digital database could be established that provides a common platform for patient records to be accessed by all clinicians involved in patient care. However, the different software systems used by various hospitals and the technical difficulties in setting up a digital database can prove to be quite challenging in implementing the above changes.

Nonetheless, greater efforts are needed to implement targeted interventions ensuring appropriate systems and services for clinicians dictating clinic letters. We conclude that this is likely to improve the quality of communication and the comprehensiveness and value of these letters to both patients and clinicians.

References:

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OC89

Medically resistant solitary rectal ulcer syndrome: a case report

by O. Lark, J. Chan. *Noah's Ark Children's Hospital for Wales, Cardiff and Vale University Health Board, Cardiff, CF14 4XW, UK.*

Solitary rectal ulcer syndrome (SRUS) is a rare disorder associated with abnormal defaecation. It typically presents with rectal bleeding, mucorrhoea, tenesmus and an altered bowel habit, often mirroring symptoms of ulcerative proctitis. SRUS can be identified on endoscopy by macroscopic findings of: solitary or multiple ulcerated or polypoid lesions 4-10cm from the anal margin. Mucosal prolapse syndrome has recently been proposed as a more accurate diagnostic description. Adult incidence is 1 per 100,000 with the condition thought to be rarer in children. No guidelines exist on managing SRUS.

A 15-year-old boy with a background of autism, anxiety and ADHD was referred to our rapid access clinic with a 7-month history of rectal bleeding and mucous discharge. His main symptoms were altered bowel habit, weight loss and on some days extreme faecal urgency and frequency, resulting in toileting 40 times in a 24-hour period. His inflammatory markers, albumin and haemoglobin were within normal range and his faecal calprotectin was <20ug/g. His MRI enterogram showed rectal wall thickening suggestive of proctitis. An urgent ileocolonoscopy was performed with macroscopic findings (Figure 1) and histology confirming SRUS. The histology showed focal superficial necrosis, patchy mild fibrosis with moderately dilated crypts, thickening of muscularis mucosa with extension of smooth muscle fibres into lamina propria.

He was commenced on prednisolone suppositories and subsequently foam enemas which did not alleviate his symptoms. Over the next few months, he lost 10% of his bodyweight despite regular community dietetic review and increased nutritional supplements. Inpatient admission was required to establish nasogastric tube feeding which he currently continues.

His ileocolonoscopy was repeated after 3 months due to limited symptomatic improvement, alongside an OesophagoGastroDuodenoscopy (OGD). His SRUS remained unchanged macroscopically and histologically. Oesophageal histology showed eosinophilic abscesses, 23 eosinophils per high powered field and basal cell hyperplasia. He was commenced on orodispersible budesonide to treat unrecognised Eosinophilic Oesophagitis (EoE). Our gastroenterology psychologist trialled defaecation behavioural therapy which had limited success due to the patients existing autism and ADHD.

Despite his EoE symptoms improving his lower gastrointestinal symptoms have worsened and he is now experiencing faecal stress incontinence. He no longer leaves the house and the family's quality of life is poor. He has been referred to adult surgical colleagues to consider surgical approaches including laparoscopic rectopexy, mucosal resection and stoma formation.

In summary, first line approaches for SRUS include treating any existing constipation and modifying defaecation behaviour. We present a particularly challenging case not amenable to medical or psychological behaviour modifications. Dramatic weight loss and an association with EoE has not been previously described. The prognosis for this case remains unclear and we thank the patient and their family for allowing us to raise awareness, to help inform clinicians and families dealing with this rare and disabling condition.

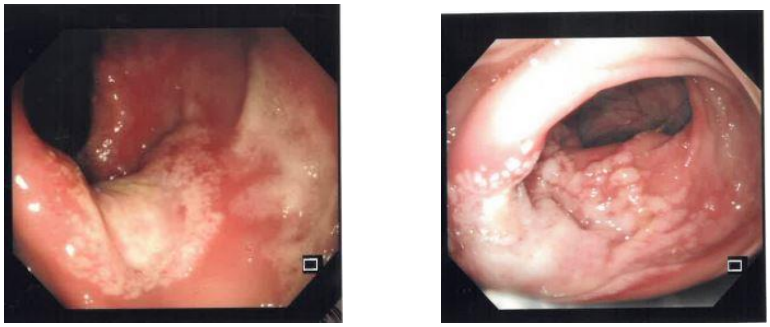


Figure 1 - Endoscopic views of the Rectum)

OC90

Gastro-sphincteric pressure gradient as new parameter for diagnosis of rumination episodes in children

by M.G. Puoti, O. Borrelli, A. Rybak, K. Lindley, K. Nikaki. *Great Ormond Street Hospital for children, Department of Paediatric Gastroenterology, Division of Neurogastroenterology and Motility, Great Ormond Street, London, UK.*

In children an intragastric pressure rise >25 mmHg associated with retrograde bolus flow into the proximal oesophagus has been proposed as diagnostic cut-off for rumination episode, but this can still fail to diagnose rumination events[1]. Gastro-sphincteric pressure gradient (GSPG), the difference between intragastric pressure (IGP) and oesophago-gastric junction (EGJ), is a new parameter for diagnosis of rumination syndrome during high resolution oesophageal impedance manometry (HRIM) in adults[2]. A difference ≥2mmHg prior to retrograde oesophageal bolus flow is diagnostic. We aimed to evaluate its use in diagnosis of paediatric rumination.

Children with confirmed rumination syndrome who had a multichannel intraluminal pH-impedance monitoring (pH-MII) and HRIM between January 2015-December 2021 were identified. Diagnosis of rumination syndrome was based on HRIM conventional criteria with rise of IGP prior or during a retrograde bolus flow. All children with rumination syndrome had a normal pHMII study. An age and sex-matched gastroesophageal reflux disease (GERD) group was included. Children in the GERD group had pathological oesophageal acid exposure time (>6%) and negative HRIM for rumination syndrome. All episodes of rumination, reflux, straining without regurgitation and transient lower oesophageal sphincter relaxation (TLESR) were analysed to calculate the GSPG.

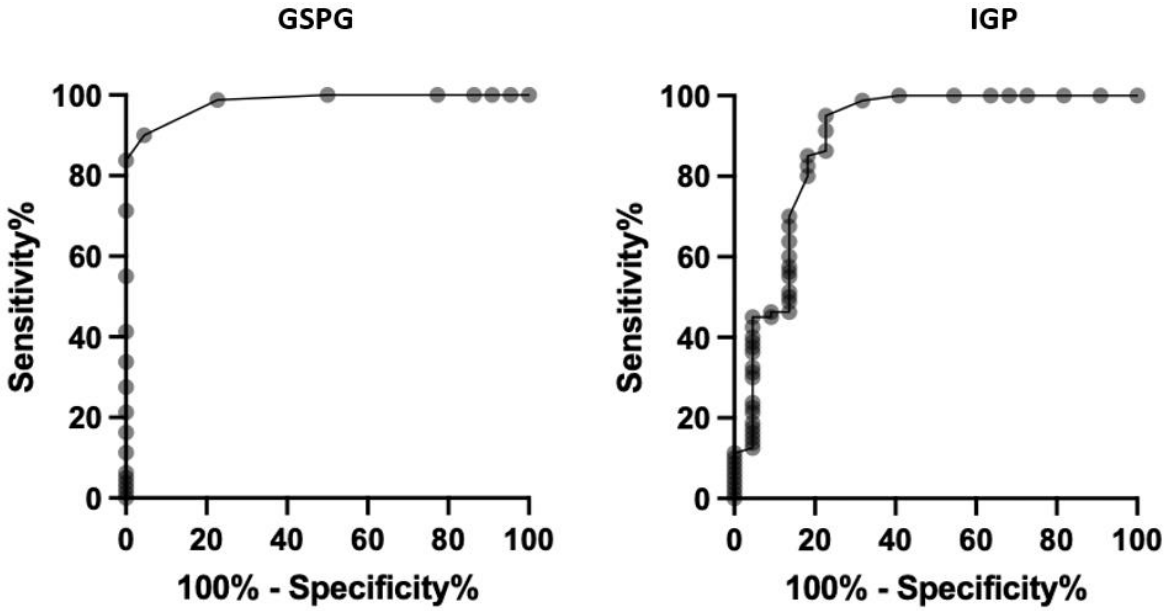
Thirty patients were included; 15 children with rumination syndrome (6 male; mean age 12.7±2.5 years) and 15 with GERD (7 male; mean age 12±2.9 years). No difference was found in lower oesophageal sphincter mean resting pressure between the two groups (26.1±11.1 vs 25.9±11.8; p=0.9). In total, 80 rumination episodes, 23 reflux events, 22 straining events without regurgitation and 14 TLESRs were detected. GSPG was significantly higher in rumination event compared to all other events (Table 1). In 31 of the 80 rumination episodes IGP peak did not exceed 25 mmHg which occurred in 9 out of 15 children with rumination syndrome. Based on ROC analysis we found that a GSPG >1.5 mmHg can identify rumination episodes with sensitivity of 90% and specificity of 95% compared to IGP >25 mmHG which has 51% sensibility and 86% specificity (Figure 1).

GSPG can be used to diagnose paediatric rumination syndrome and reliably differentiate rumination episodes from reflux events, abdominal straining without regurgitation and TLESRs.

Tab 1. HRIM parameters in the different types of episodes

HRIM parameters (median, IQR)	**Rumination**	**Reflux**	**Straining**	**TLESR**	***p* value**
Maximal IGP	32.5 (23-54)	13 (10.5-13.8)	41 (32.5-46)	12 (2.5-14.8)	<0.00001
IGP preceding an episode	11 (7-16.3)	7 (3-9)	12 (10-16)	6.5 (1.5-9.8)	<0.00001
EGJ pressure preceding an episode	5 (2-8.3)	7 (4-8)	18 (13.5-23)	5.5 (2.5-90	<0.00001
Intra-oesophageal pressure 5cm above EGJ	1 (-4-6)	3 (-2.5-7.5)	1 (-4.5-5)	-4.5 (-7-{-3})	<0.005
GSPG	5 (3-8)	0 (-1-0)	-7 (-10-{-2})	0 (-1-0)	<0.00001

Figure 1. Predictive value of GSPG and IGP for rumination episodes



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1. Singendonk MMJ, Oors JM, Bredenoord AJ et al. Objectively diagnosing rumination syndrome in children using esophageal pH-impedance and manometry. *Neurogastroenterol Motil*. 2017; 29(5).
2. Geysen H, Michielsen S, Rommel N et al. The gastro-sphincteric pressure gradient: A new parameter to diagnose a rumination episode. *Neurogastroenterol Mot*. 2021; 33(6):e14068.

OC91

Bloody diarrhoea in a paediatric male – inflammatory bowel disease or not?

by S. Emmitt. *Advanced Clinical Practitioner in Paediatric Gastroenterology. Sheffield Children's Hospital NHS Foundation Trust, UK.*

A 9-year-old male without significant previous medical history was transferred to our centre for persistent frequent bloody diarrhoea and minor abdominal pain with no reports of pyrexia, which had developed whilst on a local holiday 4 weeks prior. A stool sample returned positive for a verotoxin producing Escherichia coli (0154) not the 0157:H7 variant which is associated with varying symptoms ranging from mild diarrhoea to haemorrhagic colitis (Rahal, Kazzi, Nassar et al 2012). A further two samples taken afterwards returned negative results. During the initial presentation of symptoms, he was placed nil by mouth and commenced on intravenous fluids, however antibiotics were not administered to reduce the risk of HUS occurring and the patient wasn't pyrexial or displaying signs of infection. Prior to transfer the patient's haemoglobin decreased to 73 g/L where he was given a blood transfusion and started on octreotide.

On admission the patient was clinically stable, but bowel motions did not respond to gut rest, and he continued to drop his haemoglobin. A Direct Coombs Test was performed which ruled out haemolysis and a decision was made for endoscopic assessment to distinguish between post infectious enterocolitis or Inflammatory Bowel Disease (IBD). This showed severe colitis on 70cm of the descending colon alongside pus/fibrin deposits which bled easily and why the endoscopy was only performed up to the transverse colon. Erythema and oedema were present in D1 & D3. Following the macroscopic picture and concerns of inadequate nutrition during the previous 4 weeks, the patient was maintained on gut rest and total parental nutrition (TPN). Methylprednisolone 40mg was started to reduce inflammation alongside esomeprazole 20mg. This regime continued for 2 weeks.

The patient's symptom's improved with decrease of stool frequency and bleeding, however, did not resolve completely. Before escalation of treatment endoscopic reassessment was performed, this time including a complete ileocolonoscopy and small bowel assessment via wireless capsule. Findings demonstrated mayo 2 grade ulceration and the small bowel assessment was unremarkable. During the latter stages of admission, the patient's histology report became available which interestingly suggested the presence of an underlying IBD. However, at a histology MDT the following week it was unable to be confirmed whether there was an actual IBD present or not. Meanwhile, the patient came of TPN and gradually worked up to his usual diet with no exacerbation of previous symptoms. He was subsequently discharged home and continued oral steroids on a weaning plan. It was decided to observe for symptoms of IBD once he finished steroids and treat if required.

Faecal calprotectin post steroids was 55, and in summary the patient presented with an unusual case that was difficult to distinguish between IBD or infection, and whether this infection exacerbated an underlying IBD.

References:

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OC92

Protein losing enteropathy without GI symptoms – Is cow's milk the culprit?

by S. Matthai[1], A. Soni[1], [1]*Department of Medicine, Sheffield Childrens Hospital NHS Foundation Trust, S10 2TH, UK.*

Protein losing enteropathy (PLE) is a complication of various intestinal disorders characterised by an excessive loss of protein into the gastrointestinal tract. It can be a rare complication of severe iron deficiency anaemia due to excess cow's milk intake. There is a lack of GI symptoms with oedema as main presenting feature. Therefore diagnosis may be missed if clinical suspicion is low and stool Alpha1 Antitrypsin (A1AT) levels are not sent. We describe a case series of 3 toddlers who presented with oedema and were found to have severe iron deficiency anaemia due to excess cow's milk in the diet. Case 1: 17 month old girl presented with 3-week history of tiredness and pallor. She had periorbital oedema a week before presentation. There was no history of diarrhoea. Dietary history revealed excess milk intake of 800mls a day. Serum albumin was low without any proteinuria and normal liver function. Case 2: 13 month old girl presented with 4-day history of oedema. She was breast fed till 12 months of age. She was drinking cow's milk for the previous few weeks and was taking 900 mls per day. She had low serum albumin with no proteinuria. Case 3: 13-month-old, presented with periorbital swelling for 2 weeks and was found to be anaemic. Bloods parameters were not typical of iron deficiency but she had low serum albumin. She had normocytic, hypochromic anaemia with pencil cell and acanthocytes in blood film. Because of concerns regarding malabsorption and diagnostic uncertainty, micronutrients and vitamin levels were checked along with stool A1AT. Copper and Zinc levels were found to be low. She was given supplementation for a month. The blood film then became typical of Fe deficiency. All 3 children needed a blood transfusion and were started on iron supplements. It was remarkable that none had diarrhoea. Stool A1AT was high in all patients indicating a PLE. All 3 children had low immunoglobulins. Cow's milk intake varied from 700-900 mls a day. One child had a micronutrient deficiency (Cu and Zn) which masked the Fe deficiency and caused concerns about the diagnosis. However, a typical picture of Fe deficiency evolved once micronutrient deficiency was treated. PLE resolved within 8 weeks of treatment with iron supplements and reducing milk intake. In children presenting with oedema and severe Iron deficiency, dietary history of excess cow's milk intake must raise a clinical suspicion of PLE. Stool A1AT should be sent in these cases to avoid more invasive investigations. Dietary history is crucial as literature suggests that more than 16 oz of Cow's milk per day (480ml) can cause Iron deficiency and PLE. Micronutrient deficiency may be seen in severe PLE and may mask Fe deficiency. The exact mechanism of PLE is not known in Fe deficiency but it is not associated with hypersensitivity to cow's milk protein.

OC93

Delayed diagnosis of paediatric eosinophilic esophagitis: A case report

by S. Gulam Khader, H. Kannappan. *University Hospitals Coventry and Warwickshire NHS Trust, Clifford Bridge Road, Coventry, CV2 2DX, UK.*

Eosinophilic esophagitis (EoE), a condition identified just over 30 years ago, has become one of the most common esophageal pathologies in both adults and children. The disease progression ranges from an initial eosinophilic esophageal inflammation progressing to fibrosis and strictures. A multinational European study of children with EoE demonstrated that younger children were more likely to present with failure to thrive and diarrhoea (median age 6–7 years) and older children with abdominal pain and dysphagia (median age 10 years) or food bolus impaction (median age 12 years). [(1)]

We report a case with a delayed diagnosis of EoE highlighting the importance of clinical awareness, missed opportunities for early screening and diagnosis potentially leading to the development of complications.

A 5-year-old child presented to the paediatric clinic with difficulty swallowing certain types of food, particularly meat. He had no history of reflux, vomiting or any other gastrointestinal symptoms. His systemic examination was within normal limits and he was thriving along the 25th centile. His bloods showed a low ferritin level and a normal coeliac screen. He was commenced on iron supplements and referred to the dietician who advised the inclusion of iron-rich and softer foods like minced meat. Six months later, his diet had improved and he was better at tolerating lumpier food. After four years, he represented to the paediatric clinic with occasional episodes of dysphagia where he was reassured and discharged as he remained well. He reattended his GP surgery four years later for hoarse voice and choking episodes. Subsequently, he underwent a nasolaryngoscopy which showed edematous vocal cords. A barium swallow was requested and he was referred to paediatric gastroenterology and speech therapy. In the gastroenterology clinic, he was found to be systemically well, growing along the 25th centile with a history of asthma, eczema and hay fever. He was started on lansoprazole 15 mg once daily. The barium meal showed a focal narrowing of the esophagus at the level of the C6 vertebra; however, there was no holdup to the flow of contrast and appearances were not typical of a stricture. There was an apparent linear filling defect on the lateral views at the C4/5 intervertebral disc space level, initially thought to be secondary to an oesophageal web. However, endoscopy showed significant concentric narrowing of the upper oesophagus (15 centimeters from incisors) with the inability to pass a paediatric or neonatal-sized gastroscope. Although proximal oesophageal biopsies were normal, these could not be obtained from the stricture site. This child is scheduled to undergo oesophageal balloon dilatation.

This case demonstrates that occasionally children with EoE may elude diagnosis for long periods of time, even under the scrutiny of multiple physicians. It highlights that accurate and timely diagnosis of EoE relies on early endoscopy based on clinical suspicion. There remains a need for increased awareness about risk stratification, management strategies, facilitating early investigations and ensuring appropriate follow-up for children that present with symptoms of EoE in order to minimize complications, including oesophageal strictures as in our case.

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OC94

Inflammatory cloacogenic polyp as a cause of rectal bleeding in a teenager

by S. Rajani, M. Cohen, D. Scluckebier, A. Urs, P. Narula, M. Thomson. *Sheffield Children's Hospital, Clarkson Street, Sheffield S10 2TH, UK.*

Inflammatory cloacogenic polyp (ICP) is a rare lesion arising in the region of the anorectal transitional zone. It is considered to be a part of mucosal prolapse, which includes solitary rectal ulcer syndrome (SRUS), rectal prolapse, intussusceptions and rectocele. The etiopathogenesis is mucosal prolapse, which produces local trauma and ischemic injury followed by inflammation, repair and regenerative changes. The vast majority (85%) are located above the anal border and predominantly in the anterior lateral wall. The polyps vary in size from 3-4 cm in diameter and have a sessile appearance [(1)]. The estimated annual incidence of ICPs is 1 to 3.6 per 1, 00,000 among all solitary rectal ulcers [(2).] It is rare in children. A review of clinical notes and investigations was undertaken along with review of literature.

13-year boy, a Syrian refugee presented with history of pr bleeding for over 2 years which had increased over the past few weeks. There was no other significant medical history. He had a normal general and systemic examination on presentation. His weight and height were on the 25th centile. On presentation he had raised calprotectin(>4000)microgram/miligram. Haemoglobin of 128 g/dl on presentation, erythrocyte sedimentation rate-17 mm/hr. The MRI abdomen was normal.

He underwent colonoscopy which revealed multilobulated polyp needing piecemeal resection (Figure 1& 2). Histopathology (Figure 3) was suggestive of a polylobulated polyp in the rectum. There was smooth muscle passing up between the glands and the stroma appeared to be a mixture of smooth muscle and fibrotic tissue. Ulceration and inflammatory slough were noted on the surface. This was diagnostic of inflammatory cloacogenic polyp. Despite being on laxatives he required three piecemeal resections over a year . He continues to present with bleeding per rectum and is planned for endoscopic submucosal dissection (ESD).

Awareness of this entity in children is important because of both the propensity for recurrence and persistence of the polyps. If the underlying etiology is not corrected, the long-term implications are of internal intussusception: procidentia, descending perineum syndrome, and ultimately, incontinence. Rectal bleeding is the most common presenting clinical symptom.

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2. 2. Ewertsen C, Svendsen CB, Svendsen LB, [Inflammatory cloacogenic polyp]. Ugeskr Laeger. 2008 Aug 25;170(35):2708.

Figure 1

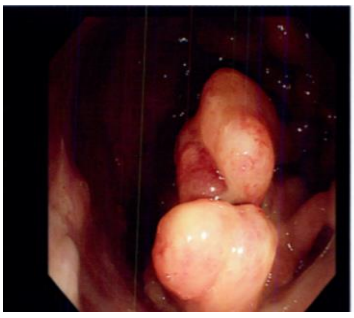

Figure 2

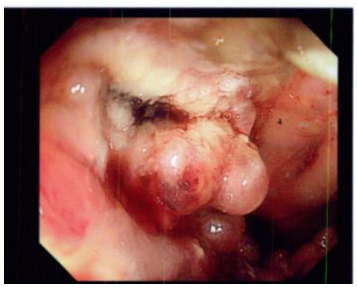

Figure 3

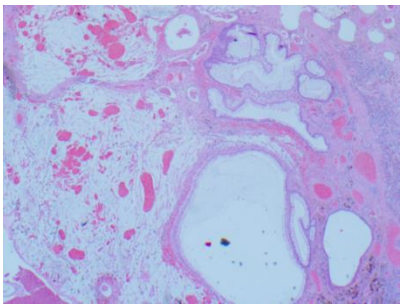

Figure 1 and 2: Endoscopic appearance of inflammatory cloacogenic polyp

Figure 3: Histology appearance of inflammatory cloacogenic polyp

OC95

Perceptions on the scale of Fabricated or Induced Illness (FII): presentations and guidance within Paediatric Gastroenterology

by H. Howard[1], F. Campbell[1,2], D. Campbell[3]. [1]University of Sheffield, S10 2TN, UK. [2]Newcastle University, NE1 7RU, UK. [3]Great North Children's Hospital, NE1 4L, UK.

Fabricated or Induced Illness (FII) is perhaps, professionally, the most complex form of child abuse. Data on the prevalence and presentation of FII is sparse, making it difficult to scale the problem and resource the solutions, yet if managed poorly, could lead to death and serious injury to the child. A 2018 survey by the RCPCH showed 92% of the 216 surveyed paediatricians recalled seeing at least one perplexing presentation in the previous 12 months, 30% recalled seeing over five[1]. We have conducted an opportunistic and purposive National survey (3 UK Nations) of general paediatricians (and safeguarding specialists), paediatric gastroenterologists and AHPs.

Informed consent was obtained, along with ethical approval from University of Sheffield Ethics forum, to conduct a survey. This was constructed of 10 questions with mixed qualitative and quantitative data. The survey was promoted via national meetings and fora, and regional leads were approached to enhance participation.

A response was received from 28 individuals (10 gastroenterologists, 7 general paediatrics/safeguarding, 3 allied healthcare professionals, 8 undeclared):

1. How many suspected FII cases per year do you see? 64% 1-5 with median 2 cases, 21% 6-10 cases.
2. How many confirmed FII cases in last 5 years? 68% 1-5, median 2, 25%, 6-20 cases.
3. Is FII overrepresented in paediatric gastroenterology? 75% Yes, 4% no, 21% unsure.
4. Has FII in paediatric gastroenterology increased in last decade? 82% yes, 14% unchanged, 4% unsure.
5. Have you been trained in FII in paediatric gastroenterology or given adequate guidance? 89% yes, 11% no.
6. Who provided training? 54% RCPCH, 4% BSPGHAN, 21% NHS employer, 21% other.
7. Is that training adequate to enable you to manage PP/FII? 48% yes, 30% no, 22% unsure.
8. What would help with FII? Narrative responses focus on enhanced safeguarding support, acceptance by the general population that FII is a fact, approaches to second opinions.

Provisional data suggests a professional view of rising frequency of FII that is disproportionately higher frequency in paediatric gastro than other specialities.

References:

1. RCPCH. Perplexing Presentations (PP) / Fabricated or Induced Illness (FII) in Children - RCPCH guidance: 2.2, p9. Published February 2021, Accessed 5 January 2023, at https://childprotection.rcpch.ac.uk/wp-content/uploads/sites/6/2021/03/Perplexing-Presentations-FII-Guidance.pdf.

OC96

A tertiary centre review of outcomes of paediatric patients with gastrointestinal dystonia associated with neurodisability

by H. Atta, K. Elesnawy, E. Effandie, L. Whyte. *Gastroenterology, Birmingham Children's Hospital, Birmingham, UK.*

Objectives and Study: Patients with severe symptoms of gastrointestinal dystonia (GID) associated with neurodisability are increasingly referred to gastroenterologists for investigations and symptom management. In this study, we review the reasons for referral, investigations recorded and effectiveness of interventions.

Methods: Retrospective review for patients referred to the gastroenterology team between April 2019 to March 2022. Data gathered includes demographics, primary diagnosis, enteral feeding plan, medical and surgical management.

Results: 70 patients were included, mean age at the start of the symptoms = 2.4years; while the mean age at referral was 6.5years. The main aetiology were genetic/chromosomal abnormalities (51.4%).

Main presenting symptoms were upper gastrointestinal i.e.vomiting (54.2%), reflux (55.7%), delayed gastric emptying (32.8%), constipation (57.1%), and bloating (15.7%).

Patients had been on variable feed types prior to referral that includes 29% who were on blended/solid food. 15.7% of patients had restricted feed intake due to symptoms.

Not all patients needed investigations, but when barium meal was done it showed reflux in 11/34 patients and delayed gastric emptying in 2/34 patients. Abdominal ultrasound showed renal anomalies in 2/16 patients.15patients had endoscopies, 9 of them had normal results while the rest has different pathologies. PH impedance study showed variable degrees of reflux in 11 out of 13. Table 1 shows various modalities of medical management before and after the consultation.

Table 1

Management	Before consultation (%)	After consultation (%)
Blended diet (partial/complete) or solid	29	30
Proton Pump Inhibitor	42	7
Prokinetics	6	20
Laxatives (started/optimized)	18	9
Stimulants for constipation	6	8
Pain relief	4	12

Out of 30 patients who were started on a blended diet, 27 patients had significant improvement. 24.3% needed to change the route of feeding. Nine cases needed surgical intervention; 3 had gastrostomy,3 had fundoplication,2 had gastrojejunal-tube insertion, 1 had gastrojejunal-tube insertion and fundoplication.

Conclusion: Patients with GID have a variety of symptoms, with multiple symptoms in some patients. Investigations were not necessary in most patients. Patients who received blended diet showed most improvement.Medical management of symptoms was offered based on history. Many of the interventions offered could be managed by a local dietician and paediatrician with expertise in Neurodisability.Only those children with ongoing or red-flag symptoms should be referred to a gastroenterologist.

OC97

Impact of a Paediatric Gastroenterology Out-of-Hours Service- a Service Evaluation

by K.Y. Lee, D. Basude, A. Wiskin, E. Volonaki, L. Selvarajan, C. Spray. *Department of Paediatric Gastroenterology, Bristol Royal Hospital for Children, BS2 8BJ, UK.*

Aligned with recommendations from the 2021 paediatric gastroenterology, hepatology and nutrition national census[1], our tertiary paediatric gastroenterology department implemented an out-of-hours (OOH) service in January 2022. This included provision for 24/7 specialist telephone advice, consultant-led weekend ward rounds and OOH emergency endoscopy.

We sought to evaluate the OOH service one year post-introduction. We collected admissions data and endoscopy logs from a 5-year period (1 January 2017- 31 December 2021) representing our service pre-OOH service and compared this with the one-year period (1 January 2022-31 December 2022) representing our service after introduction of the OOH service. We also sent an online survey to general paediatricians, subspecialty paediatricians and paediatric surgeons in our tertiary centre and our ten regional network hospitals for feedback on the OOH service.
From 2017-2021, there was an average of 124 admissions/year, compared with 170 in 2022. Those patients admitted electively for procedures and infusions were excluded. Median length of hospital stay for both epochs was 7 days. There was a significant increase in weekend discharges (12.9% vs 20%, $p<0.05$). There was no significant difference in weekend admissions (14.3% vs 15.9%, $p=0.6$).

In 2022 alone, 25 patients underwent endoscopy out-of-hours (Figure 1). 17/25 (68%) occurred during the weekend, the remainder during weekday evenings. The most common indication for endoscopy was inflammatory bowel disease assessment (n=14, 56%). In comparison, 20 patients underwent endoscopy out-of-hours in the five years preceding this.

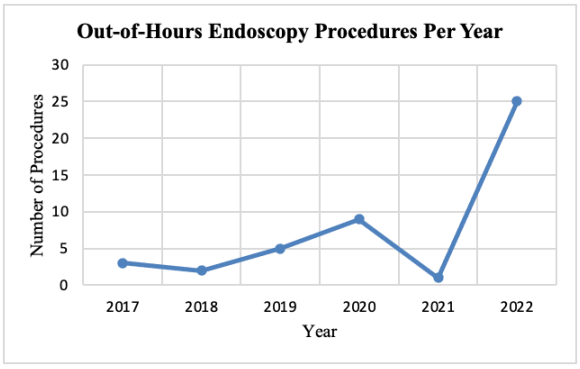

Figure 3: OOH Endoscopy Procedures Per Year

We received 37 survey responses, of which 24 (65%) came from general paediatricians, 11 (30%) from subspecialty paediatricians and 2 (5%) from paediatric surgeons. 10 respondents (27%) contacted the OOH service for specialist advice in 2022. All found it easier/ much easier to access timely specialist advice, compared to before. In terms of qualitative feedback, respondents were generally complimentary of the OOH service, and found its availability reassuring. In terms of constructive feedback, one respondent recommended better advertisement of the service; another respondent noted an increased challenge with coordinating the timing of different weekend ward rounds with the available trainees.

The introduction of our OOH service has improved patient care. Increased weekend discharges reflect the impact of senior decision-making during the weekend. There is greater access to OOH endoscopy, reducing treatment delays. Professional feedback is also highly positive. There has been an increased OOH workload, and therefore an OOH service needs to be carefully job-planned. We hope our experience will be helpful to other specialist centres planning to introduce an OOH service.

References:

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OC98

Rifaxamin in gastrointestinal decontamination in suspected small intestinal bacterial overgrowth: A single centre experience

by G. Rivlin[1], K. Hugh[2], M. Nair[1], E. Renji[1], M. Auth[1], R. Parmar[1], F. Cameron[1], S. Tamhne[1], H. Doble[1] and J. Cheng[1]. *[1]Department of paediatric gastroenterology, Alder Hey Children's Hospital, Eaton Road, Liverpool, L12 2AP, UK. [2]Leeds Medical School, University Of Leeds, Worsley Building, Leeds, LS2 9NL, UK.*

Small intestinal bacterial overgrowth is a condition in which the small bowel is colonised by excessive bacteria. A tertiary hospital guidelines recommend three cycles of fortnightly Rifaximin and Biokult.

This retrospective study aims to review the indications and outcomes of Rifaximin in gut decontamination. Patients were identified through electronic patient records over a five-year period. Data was collected from clinical notes and letters.

74 patients identified with positive documentation to initiate 'gut decontamination' and received either Rifaximin [8%], Rifaximin with Biokult [88%] and Rifaximin with over-the counter probiotics [4%]. Background included, short gut [20%], genetic disorders [8%], irritable bowel syndrome (IBS) [43%] and neurodisability [27%]. Analysis of symptoms based on background; Short gut: Abdominal pain [15], Loose stools [9], Flatus [5], Nausea/Vomiting [4], Dystonia [1], Constipation [1], Feed intolerance [1]; Neurodisability: Abdominal pain [15], Loose stools [11], Flatus [5], Dystonia [4], Constipation [3], Nausea/vomiting [2], Feed intolerance [2], Offensive stools [1]; Genetic: Abdominal pain [8], Loose stools [4], Flatus [3], Dystonia [2], Constipation [2], Nausea/Vomiting [1], Feed intolerance [1]; IBS: Abdominal Pain [35], Loose stools [17], Flatus [10], Constipation [5], Nausea/Vomiting [3], Feed intolerance [3]. Treatment response based on follow up physician global assessment. No improvement [36%], partial response [26%], full response [26%], Unknown [12%]. Responses for background diagnoses; Short gut: full response [4], partial [9], no [2]; Neurodisability: full response [7], partial [5], no [6], unknown [2]; Genetic: full response [2], no [3], unknown [1]; IBS: full response [6], partial [5], no [16], unknown [6].

40%- 60% of neurodisability or genetic condition patients had symptomatic improvement (either full or partial) post treatment. However, only 33% of IBS patients responded (either full or partial). Given these results and Rifaximin cost; investigations (plasma d-lactate and hydrogen breath test) should be done prior to therapy to avoid over-treatment.

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(2) Grace, E., Shaw, C., Whelan, K. and Andreyev, H.J.N. (2013), Review article: small intestinal bacterial overgrowth – prevalence, clinical features, current and developing diagnostic tests, and treatment. Aliment Pharmacol Ther, 38: 674-688. https://doi.org/10.1111/apt.12456

(3) Koo HL, DuPont HL. Rifaximin: a unique gastrointestinal-selective antibiotic for enteric diseases. Curr Opin Gastroenterol. 2010 Jan;26(1):17-25. doi: 10.1097/MOG.0b013e328333dc8d. PMID: 19881343; PMCID: PMC4737517.

OC99

Investigation and management of Helicobacter pylori in children and adolescents in a Regional London area

by E. Hinde and A. Ocholi, *St George's Hospital, SW17 0QT, UK.*

Introduction: The aim of this study was to review management of *H. pylori* in a regional London area including symptoms and patterns of presentation, investigation routes prior to treatment (stool antigen detection or diagnostic oesophago-gastro-duodenoscopy (OGD) and treatment outcomes through 2020- 2021. It was hypothesised that the pattern and presentation for *H. pylori* in this regional London area will be incongruous with some of the recommendations with regards to investigations recommended in the joint ESPGHAN/NASPGHAN guidelines from 2016.

Methods: This retrospective audit comprised of 598 patients who had either had a *H. pylori* stool test or OGD in 2020-2021.

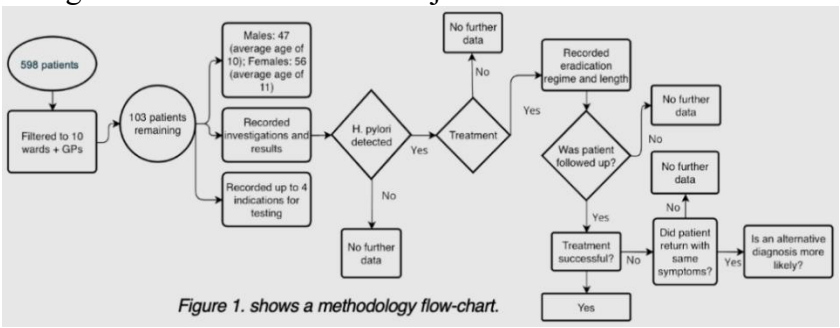

Figure 1. shows a methodology flow-chart.

Results: Demographic Data:

- 598 *H. pylori* stool antigen tests were requested Dec 2020 – Dec 2021 in the regional area - 14.9% (89) of these samples were positive.
- 103 samples (out of 598) were requested after assessment in one of the paediatric areas of our tertiary paediatric unit after assessment by a member of the paediatric team - Of the 103, 37% (38) were positive.
- *H. pylori* stool antigen test was requested following history and examination by senior paediatricians.
- Adolescents and older children had the highest positivity rates compared with prepubertal children in our data set.
- 50% of treated patient (out of 103) were successful (negative *H. pylori* stool test) - In 47% of patients, follow up data was not available.

Patient Information	Median (Range), n(%)
Male	47 (45.6%)
Female	56 (54.4%)
Age (years)	11 (0-18)
0-6 years	16 (15.5%)
7-12 years	42 (40.8%)
13-18 years	45 (43.7%)

Table 1. shows the median (range) and percentage total for patient information variables.

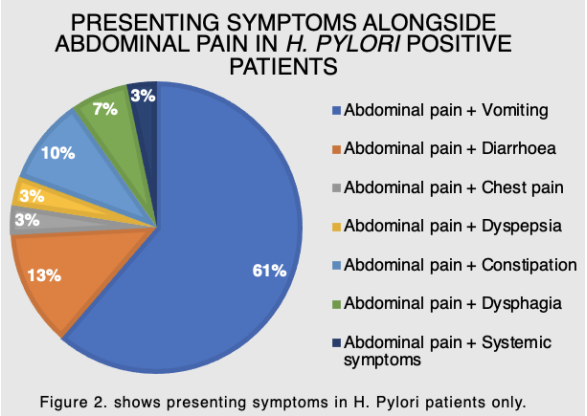

Figure 2. shows presenting symptoms in H. Pylori patients only.

Conclusions: In this regional London centre, *H. pylori* stool testing is a viable and sensible way to manage *H. pylori* infection in contrast to current ESPGHAN/NSPGHAN *H. pylori* guidance. Education of primary care colleagues is needed to prevent excessive number or tests. Advice with regards to primary *H. pylori* stool testing should be directed at adolescents and not prepubertal children, suggesting that adolescents could follow a modified 'test and treat' strategy. Education is also required to ensure follow up testing after eradication is performed.

Age Group	*H. pylori* positive	*H. pylori* negative	*H. pylori* positive (n, %)
0-6 years	3	13	3%
7-12 years	12	30	12%
13-18 years	23	22	22%
Grand Total	**38**	**65**	**37%**

Table 2. shows H. Pylori positive and negative values compared with age and gender.

Age Group	Investigation	*H. pylori* positive	Received Treatment	Treatment Success (%)	Treatment Failure (%)	Treatment Outcome Unknown (%)
0-6 years	OGD	2	2	100.0%	0.0%	0.0%
	Non-invasive	1	1	0.0%	0.0%	100.0%
7-12 years	OGD	3	3	66.7%	0.0%	33.3%
	Non-invasive	9	6	50.0%	0.0%	50.0%
13-18 years	OGD	13	12	50.0%	8.3%	41.7%
	Non-invasive	9	9	33.3%	11.1%	55.6%
	Grand Total	**37**	**33**	**50%**	**3%**	**47%**

Table 3. shows how many H. Pylori patients received treatment and whether it was successful., Unsuccessful or unknown.

References:

1. **Aguilera Matos I, Diaz Oliva S, Escobedo A et al.** Helicobacter pylori infection in children. BMJ Paediatr. Open. 2020. *Online journal article*
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OC100

Chronic pain is common in Paediatric Inflammatory Bowel Disease and impacts quality of life

by R. Mardare, L. Eskell, J. Machta, J. M. E. Fel, D. Thangarajah. *Chelsea and Westminster Hospital, London, SW10 9NH, UK.*

Prevalence of chronic pain (CP) is reported as 38% in adult inflammatory bowel disease (IBD), with a significant impact on quality of life (QOL), functional and social outcomes (1). Prevalence and attributes of CP are unknown in paediatric IBD. Our aim was to evaluate the prevalence, disease related factors, and impact on QOL of CP.

The prospective, cross-sectional single centre study included 41 children, (8-18 years), of a predominantly Crohn's Disease (CD) cohort 29/41, (70%). Patients and their parents completed validated age-appropriate pain and IMPACT III QOL questionnaires. CP was defined as per von Korff scale (2). CP and No pain (NP) medians were compared using Mann-Whitney tests and proportions using Chi squared test ($p<0.05$).

	Chronic pain						**CP vs NP**
	Mild		*Moderate severe*		*None*		*p*
Number (Male)	19	(10)	14	(4)	8	(4)	
Age (range), yr	14.9	(12.5-17.7)	14.8	(13.4-17.3)	14.9	(10.7-17.7)	0.69
Disease duration (range), yr	3.1	(0.5-14.9)	2.2	(0.8-5.2)	3.7	(0.7-5.2)	0.47
Disease activity (active)	11	(58%)	9	(64%)	4	(50%)	0.59
Faecal Calprotectin (FC) (n=31)	469	(37-13250)	1084	(20-4700)	1890	(88-7892)	0.30
CRP	0.8	(0.2-94.5)	3.2	(0.2-139.6)	1.75	(0.8-47.5)	0.29
CD	13	(68%)	11	(78%)	5	(63%)	**<0.05**
PCDAI	13	(5-40)	11	(5-40)	25	(5-50)	0.22
IBD Unclassified					2	(25%)	
Ulcerative colitis	6	(31%)	3	(21%)	1	(12%)	
PUCAI	10	(5-30)	60	(25-60)	10	(10-20)	0.60
Current therapy (%)							
Steroid use	4	(21%)	2	(14%)	0		
Immunomodulator	8	(42%)	6	(42%)	1	(12%)	
Biologic use	17	(89%)	9	(64%)	5	(62%)	
QOL domains median							
Wellbeing score	75	[54-83]	41	[17-71]	87	[80-94]	**<0.05**
Emotional functioning	67	[54-82]	51	[39-67]	85	[68-85]	**<0.05**
Social functioning	75	[68-88]	72	[53-82]	92	[85-97]	**<0.05**
Body image	63	[56-75]	65	[41-75]	59	[44-84]	0.86
Total score QOL	69	[61-76]	53	[46-68]	81	[79-90]	**<0.05**

***Table 1**. Comparison between CP and NP: percentages or medians (range), [IQR] are presented.*

CP was prevalent in 33/41 (80%) patients. CP subjects were adolescents with extensive and established disease, predominantly on biologics 26/33 (79%). Analgesia use was low 15/33 (45%), with no opiate use. There was no difference in disease activity CP vs NP, defined by PCDAI, PUCAI, CRP and FC. QOL score was significantly lower in CP, with no difference in body image domain score in CP vs NP.

CP is common in paediatric IBD and is not associated with disease activity or related biomarkers. CP significantly impacts emotional, wellbeing and social functioning in children with IBD. This is the first study to report on CP in children with IBD, strategies should target psychosocial interventions. We recommend screening for CP in children with IBD as it is common.

References:

1. Morrison G, Van Langenberg DR, Gibson SJ, Gibson PR. Chronic pain in inflammatory bowel disease: characteristics and associations of a hospital-based cohort. Inflamm Bowel Dis. 2013 May;19(6):1210-7.
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