

Results Four critically ill children (aged 1–14 years), requiring resection of ischaemic colon following sudden cardiovascular collapse, presented to our institutions over a 2-year period.

Three had a preceding history of recent illness; the other had been well prior to out of hospital cardiac arrest. The 3 who were unwell experienced: headache (1), cough (1) and polyuria and polydipsia (1) for up to 2 weeks prior to hospital attendance. None had abdominal or gastrointestinal (GI) symptoms in their initial symptoms, although all but 1 developed GI upset during their rapid deterioration phase. Three had cardiac arrest before colectomy. All developed abdominal distension after resuscitation. All had significant derangement of blood sugar on monitoring. All 4 received inotropic support before surgery. All 4 had total colonic ischaemia diagnosed during surgical intervention and on histology report. There was no other underlying disease on histology (bowel was ganglionic). No infective organism was isolated (specifically all were negative for clostridium difficile). All 4 died due to multi organ failure.

Summary and Conclusion From this case series we could not identify any specific condition that predisposed these 4 children to develop non occlusive mesenteric ischaemia and colonic injury. Therefore we could not clearly identify means of prevention.

All 4 had a sudden deterioration over less than 24 hrs, and all received inotropes before developing abdominal distension. In addition, 3 out of 4 had cardiac arrest and return of circulation before colectomy. These would support the hypothesis of hypoperfusion/reperfusion injury.

Even though all 4 patients had colonic resection as part of the resuscitation the outcomes were very poor leading to multi organ failure or cerebral ischaemia and death.

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SHOULD CHILDREN WITH COELIAC DISEASE BE SCREENED FOR TYPE 1 DIABETES MELLITUS IN ANNUAL BLOODS? AN AUDIT OF FOUR YEARS SCREENING ACROSS FOUR PAEDIATRIC CENTRES IN ENGLAND

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10.1136/flgastro-2021-bspghan.58

Introduction The genetic association of Coeliac Disease (CD) and Type 1 Diabetes Mellitus (T1DM) is well known. Although NICE does not include annual screening for CD in the T1DM guidance,¹ children with T1DM are routinely screened for CD in England. The incidence of children with CD on gluten free diet (GFD) developing T1DM appears to be small.² The value of screening for T1DM in CD patients is thus not known.

Methods A 4-year retrospective review was conducted of a case series of children with known CD attending outpatient clinic across 4 centres. Patients were diagnosed as per ESPGHAN guidelines. In these centres HbA1c is tested as part of the CD annual review to detect impairment of glucose metabolism (> 41 mmol/mol). Abnormal HbA1c was documented in patients with CD.

Results 345 children with CD who had HbA1c screening were identified. Children with T1DM diagnosed prior to developing CD were excluded from analysis. Six of the 345 patients

(1.7%) were identified with an abnormal HbA1c. Only 2/6 were confirmed as having T1DM (1 within 4 months of diagnosis); 2/6 had subsequent glucose tolerance tests that were normal, 1 patient had Turner's syndrome and was taking growth hormone, which has an impact on glucose metabolism, and 1 patient is undergoing further investigations for Type 2 Diabetes Mellitus (high BMI).

Discussion The findings confirm the conclusion of previous studies that showed that a new diagnosis of T1DM in known CD children is uncommon. The annual screening for T1DM in children with CD who have developed impaired glucose tolerance is questionable. There is no standard screening test for T1DM, and HbA1c as a screening test for T1DM is also not routinely used. The exact mechanism for expressing co-existing autoantigens that generate both autoimmune conditions is poorly understood, and it is not clear whether the GFD in CD plays a role.

REFERENCE

1. National Institute of Health and Care Excellence (NICE) (2020) 'Management of children and young people with type 1 diabetes'. Available at: <https://cks.nice.org.uk/topics/diabetes-type-1/management/management-children-young-people/>
2. Ludvigsson JF, *et al.* 'Celiac disease and the risk of subsequent type 1 diabetes'. *Diabetes Care* **29**:2483–2488, 2006

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SINGLE CENTRE EXPERIENCE OF ENDOSCOPIC BALLOON DILATATION FOR LUMINAL STRICTURES IN PAEDIATRIC PATIENTS WITH CROHN'S DISEASE

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10.1136/flgastro-2021-bspghan.59

Introduction/Background Intestinal strictures are a complication of Crohn's disease despite optimal medical management. Endoscopic balloon dilatation is frequently used for management of simple strictures in adults in comparison to the paediatric population. Therapeutic endoscopy is rarely performed in paediatric gastroenterology centres in the UK. Strictures in the duodenum, jejunum, ileal and colonic area are accessible by endoscopy and enteroscopy. Endoscopic balloon dilatation is a less invasive treatment option for management of short strictures and can defer surgical intervention.

Aim We aimed to evaluate the outcome of paediatric patients undergoing stricture dilatation over a 10-year period.

Subjects and Methods We retrospectively reviewed all paediatric patients with Crohn's disease who underwent endoscopic balloon dilatation at a tertiary paediatric gastroenterology centre in the last 10 years (2010 to 2020). Strictures were identified using magnetic resonance enterography (MRE) and also during endoscopy. Patients were booked for endoscopic balloon dilatation if they were symptomatic and had pre-stenotic dilatation on MRE or inability to pass colonoscopy into stenosed lumen at previous endoscopy. Both paediatric colonoscopy and single balloon enteroscopy was used for endoscopic balloon dilatation which was done under fluoroscopy guidance. Clinical and endoscopic data were collected from electronic patient records.

Results During the 10-year period 20 patients with Crohn's disease underwent endoscopic balloon dilatation. The mean age of diagnosis of Crohn's disease was 12.45 years (5–16.4 yrs) and the mean age at the time of the occurrence of first stricture was 14.2 years (10.9– 17.9 yrs). 65% patients were on biologics and 85% were on an

immunomodulator (azathioprine, methotrexate or mycophenolate mofetil). Multiple strictures were noted in 25% of patients. Location of strictures included ileal, ileocaecal, sigmoid and caecal.

A total of 32 dilatations were performed in the 20 patients and 8 patients underwent multiple endoscopic balloon dilatations (7 patients underwent 2 dilatations and 1 patient had 6 dilatations). 85% of patients were symptomatic (abdominal pain, vomiting) from the stricture and after endoscopic balloon dilatation in 70.5% the symptoms had improved.

There were 2 procedure related complications 0.06% (1 perforation requiring surgery and 1 perforation was managed conservatively). Mean follow-up since the first stricture dilatation was 2.67 years (0.1- 6.11 yrs). During the follow-up of these 20 patients; 4 underwent stricture related surgery and 80% have not undergone any surgical intervention.

Summary and Conclusion Our experience has shown that endoscopic balloon dilatation is a relatively safe procedure for the treatment of luminal strictures. Endoscopic balloon dilatation results in symptomatic relief and delays surgical intervention in Crohn's patients with luminal strictures.

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SODIUM-DEPENDENT MULTIVITAMIN TRANSPORTER DEFECTS – A RARE CAUSE OF CYCLICAL VOMITING AND FALTERING GROWTH

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10.1136/flgastro-2021-bspghan.60

Case A previously fit and well 13-month-old boy of South Asian descent born to consanguineous parents presented acutely with persistent diarrhoea, vomiting and a perioral rash. He was febrile, developed bloody diarrhoea and clinically deteriorated with significant weight loss (50th centile to 9th centile). He continued to have episodes characterised by cyclical vomiting and feed intolerance, often with associated febrile illness with no microorganisms found from multiple septic screens.

Investigations Investigations showed persistently raised inflammatory markers, anaemia and thrombocytopenia, with hypogammaglobulinaemia. Ultrasound abdomen showed evidence of generalised enteritis initially, but he failed to improve on a course of triple antibiotics. Stool cultures and viral PCR were negative. Faecal calprotectin was raised (>2000 mg/kg). Repeat ultrasound abdomen showed fluid filled, actively peristalsing small and large bowel, with very mild wall thickening and inflammatory mesenteric change. Subsequent upper and lower GI endoscopy was macroscopically normal, and histology revealed chronic active oesophagitis/gastritis with a normal colon. Ophthalmology assessment due to vision concerns revealed bilateral optic atrophy. MRI brain showed lack of supratentorial, infratentorial and parenchymal bulk with thinning of the corpus callosum and optic nerves.

Progress The rash spread to include the peri-oral, peri-auricular, sacral and genital areas. He was initially treated for acrodermatitis enteropathica due to clinical presentation and borderline low zinc levels, however zinc supplementation did not resolve symptoms. Feed intolerance persisted despite switch to amino acid formula. Parenteral nutrition was commenced after failed enteral (gastric and jejunal) feeding trials.

Whole exome sequencing revealed two missense mutations in the SLC5A6 gene.

Management and Discussion The SLC5A6 gene produces sodium-dependent multivitamin transporters (SMVT) which are expressed in various tissues including the intestine, brain, liver, lung, kidney, cornea, retina and heart. It plays a major role in the uptake of biotin, pantothenate and lipoate in the digestive system and transporting B-group vitamins across the blood brain barrier.

This case was only the fourth to ever be described in literature. The first case described a 15-month-old with failure to thrive, microcephaly, developmental delay, severe immune deficiency and severe gastroesophageal reflux.¹ A subsequent series described two siblings with profound neurodevelopmental, progressive truncal ataxia and refractory cyclical vomiting.²

Our patient was managed on vitamin replacement therapy: Biotin (10 mg, intramuscular), Dexpantenol (250 mg, intramuscular) and α -lipoic acid (300 mg, intravenous) given weekly. With treatment he has shown significant improvement. Cyclical vomiting has settled, his rashes are quiescent, and bloods have normalised. He is now 2 years old, fully enterally fed with his weight on the 70th centile.

This case highlights how defects in multi-vitamin transporters can lead to multi-systemic disease. It also demonstrates the diagnostic role of whole exome sequencing, and with growing genetic databases it will only increase its future potential.

REFERENCES

1. Subramanian VS, Constantinescu AR, Benke PJ, Said HM. Mutations in SLC5A6 associated with brain, immune, bone, and intestinal dysfunction in a young child. *Hum Genet.* 02 2017;136(2):253–261.
2. Byrne AB, Arts P, Polyak SW, et al. Identification and targeted management of a neurodegenerative disorder caused by biallelic mutations in SLC5A6. *NPJ Genom Med* 2019;4:28.

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SUCCESSFUL SALVAGE OF CENTRAL VENOUS CATHETER AFTER > 75% OF CATHETER-RELATED BLOOD STREAM INFECTION (CRBSI) IN CHILDREN ON LONG-TERM HOME PARENTERAL NUTRITION (PN)

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10.1136/flgastro-2021-bspghan.61

Objectives and Study Preserving venous access in children with intestinal failure (IF) requiring long-term parental nutrition (PN) can be critical for patient survival. Data regarding salvage of central venous catheter (CVC) after a catheter-related blood stream infection (CRBSI) in children are limited. We aimed to determine the incidence of CRBSI and rates of CVC salvage in children receiving home PN for IF.

Methods We searched our prospective PN database for the records of all CRBSI in children receiving home PN from January 2015 to April 2019. All the patients were at home with care by parents formally trained to connect, disconnect and manage PN. They all had shared care set up between our IF rehabilitation service and their local hospital. Data abstracted from the medical records included demographics, underlying disease, CRBSI number per patient, microorganism(s) isolated and CRBSI outcome. The CRBSI incidence and rates of catheter salvage were determined. Children with immunodeficiencies were excluded. Diagnosis of CRBSI was based on clinical