

He described 9 days of breathlessness, chest tightness, vomiting and abdominal pain. His chest was clear and heart sounds normal with a soft murmur. He appeared very anxious. There were two positive findings of significant concern; a 3 centimetre palpable liver and persistent hypertension of more than 160/120. A chest radiograph showed a large heart and fluid in the horizontal fissure. More focused history taking revealed the shortness of breath was worse on exertion and on lying down, and he had to prop himself up at night to sleep.

The history and examination was consistent with heart failure. An ECG showed right ventricular hypertrophy and widespread T-wave inversion. An echocardiogram showed a structurally normal heart, mild mitral and aortic regurgitation, severely impaired left ventricular function (fractional shortening 13%) and bilateral pleural effusions. A provisional diagnosis of viral myocarditis was made. However, in addition to persistent hypertension, his urea was 30 and creatinine 523.

He was admitted to a Paediatric Intensive Care Unit where acute management included oxygen, fluid restriction, furosemide and a milrinone infusion. In view of the deranged renal profile he was discussed with the renal team. Renal ultrasound revealed small, scarred kidneys and calyceal dilatation. The diagnosis was end stage renal failure due to undiagnosed grade 4 vesico-ureteric reflux, hypertension secondary to chronic kidney disease, and dilated cardiomyopathy secondary to hypertension.

It is rare for a teenager to present in heart failure, having never been unwell before. Originally we thought this was a primary cardiac problem, but the learning point here is that despite antenatal screening and good health care, congenital renal disease can remain asymptomatic, and that end stage renal failure can present with heart failure.

G40(P) IS THERE ROLE FOR ECULIZUMAB IN THE MANAGEMENT OF C3 GLOMERULOPATHY?

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Introduction The new classification, and increasing recognition, of dense deposit disease (DDD) as a C3 glomerulopathy and an alternative complement pathway disorder suggests that eculizumab, an anti-C5 monoclonal antibody, may be beneficial in treatment. However, the use of eculizumab as an off-label and expensive treatment in paediatrics is limited with variable clinical response in literature.

Method We present the outcomes of 2 paediatric patients in Scotland who have received eculizumab for the management of DDD.

Results Case 1: A 10-year-old male presented with steroid-resistant proteinuria and biopsy confirmed DDD. He was managed symptomatically with an angiotensin converting inhibitor and diuretics. Four years later, due to worsening symptoms (oedema, lethargy, poor growth), renal function deterioration, and elevated terminal complement complex (TCC) levels, eculizumab was commenced. Repeat biopsy demonstrated chronic irreversible damage but 25% glomerular architecture preservation. Symptomatic improvement was seen along with a reduction in proteinuria and renal function stabilisation. After 24 months, eculizumab was discontinued with progressive renal function deterioration seen in the final

months of therapy. Six months later, due to worsening of symptoms and increasing proteinuria, eculizumab was recommenced with symptomatic improvement noted.

Case 2: A 5-year-old girl presented with steroid-resistant haematuria and proteinuria and biopsy confirmed DDD. Sixteen months following diagnosis, she rapidly progressed towards end-stage renal disease. Haemodialysis was commenced following no benefit from immunosuppression (mycophenolate mofetil or plasmapheresis). TCC level was elevated and eculizumab treatment was commenced 18 months after diagnosis. Renal biopsy prior to treatment showed marked pathological changes in all visible glomeruli. However, due to lack of clinical response, eculizumab was discontinued after 2 months. The patient remaining dialysis dependent.

Conclusions Our experience with eculizumab has demonstrated variable results. Only one patient exhibited a positive clinical response. Eculizumab was administered to both patients at varying points in their clinical journey with differing progressive biopsy features. Our experience adds to the limited evidence in literature, suggesting that a role for eculizumab in DDD may be guided by clinical and pathological features. However, further trials of its use in paediatric DDD are clearly needed.

British Inherited Metabolic Diseases Group and British Society of Paediatric Gastroenterology, Hepatology and Nutrition

G41 A JOINT COLLABORATIVE REGIONAL NETWORK APPROACH TO OVERCOME DIFFICULTIES IN DIAGNOSIS OF PAEDIATRIC COELIAC DISEASE

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Aims Diagnosis of paediatric coeliac disease (pCD) continues to be a challenge despite the new NICE, BSPGHAN and ESPGHAN guidance. To understand a 'true abnormal serologic result' and the need for a duodenal biopsy, we aimed for a quality improving exercise in a regional network setting.

Methods A two staged quality approach was adopted:

- An online regional survey was conducted to investigate the currently used serologic screening tests for diagnosis of pCD in the region. Regional paediatricians and pathology laboratories were contacted by phone and email followed by an online survey conducted via the Network Website (AH, NA).
- A summary presentation of the findings, were presented at the annual regional network meeting followed by a rigorous discussion with active participation of immunologists, paediatric consultants and dieticians from the region.

Results 9/13 hospitals in the regional network participated in the survey.

For coeliac screening IgA Anti-tTG is used by all centres (IgA EMA in 7/9 centres (77.8%). 5/9 (55.6%) centres routinely check IgA levels in all cases. 5/9 centres (55.6%) combined

IgA and IgG anti-TTG to increase sensitivity. There is a huge variation in the normal range of IgA TTG with the upper normal limit varying from 2.9 to 19.

The survey results were presented (AH) at the annual regional network meeting generating an active discussion amongst the attending immunologists, paediatricians, dieticians and specialist nurses, with these conclusions.

- The huge variation in normal levels, with lack of standardisation in TTG testing, is an issue in the region, as highlighted nationally.
- The Immunologists highlighted variations even within the same serological test, when using different batches requiring labs to conduct regular quality control checks.
- A network consensus followed with regards to provision of dietary review for all patients (particularly <10X serologic levels), to ensure adequate gluten intake before consideration of biopsy.

Conclusions pCD is a lifelong condition requiring an accurate diagnosis. The clinicians and immunologists continued to be challenged despite following the correct pathways and guidance. This study highlighting the strengths and benefits of a joint regional collaborative approach to help overcome some of these difficulties.

G42

LONG TERM SAFETY AND EFFICACY OF SINGLE DOSE PARENTERAL IRON IN CHILDREN WITH INFLAMMATORY BOWEL DISEASE IN A LARGE TERTIARY CENTRE

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Background Iron-deficiency anaemia is a common complication amongst children with inflammatory bowel disease (IBD). Although single-dose parenteral iron (PIN) preparations are an easily available treatment for children, there are still concerns surrounding its adverse reactions.

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

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

Results A total of 29 patients were identified. The median age was 14 and median weight was 33.4 kg. Two patients did not have the full data set. Repeated-measures ANOVA conducted on 27 patients showed that mean haemoglobin differed significantly between time points [F (4, 104)=29.416, p<0.001]. Residuals were approximately normally distributed. Post-hoc tests using the Bonferroni correction revealed that mean

haemoglobin increased significantly by 6 weeks and remained stable thereafter (p<0.001).

Only one patient had an acute type 1 allergic reactions (not anaphylaxis). Two patients had hair loss at 3 months post-infusion which were unlikely to be secondary to iron overload. None of the patients had dysmetabolic iron overload syndrome (DIOS). All children had normal LFTs and GGTs with no evidence of diabetes, chronic fatigue or hepatosplenomegaly in their follow up.

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

G43(P)

DIFFICULTIES IN COMPLYING WITH THE BSPGHAN GUIDELINES FOR DIAGNOSIS AND MANAGEMENT OF CELIAC DISEASE (2013)

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Aims To audit our paediatric department's practice of diagnosis and management of celiac disease, comparing it to the BSPGHAN and Coeliac UK guidance published in 2013, to review and consider improvements to our current practice.

Method Collection of data from 2015–2017 including all patients aged <18 years old with a positive tissue transglutaminase (tTG). Electronic records were used to gather information regarding other serological tests (endomysial antibodies (EMA)), genetic tests (HLA DQ2/DQ8), and biopsy results; as well as time-to-follow up with both the dietician and clinician.

Results Of the 55 children with a positive tTG there were sufficient data to include 40. 10/40 were asymptomatic; 30/40 symptomatic, and the ratio of females to males was 3.4:1. This audit demonstrated that 67.5% were diagnosed according to the national guidelines, and for those cases where the guidance was not adhered to it was most commonly because a biopsy had not been performed (see table 1 below).

Abstract G43(P) Table 1

No genetic investigations	2
No genetic investigations or inadequate serology	1
Inadequate serology and no biopsy where indicated	2
No biopsy performed where indicated	6
Investigated on a gluten-free diet	2

Only 17% of patients were seen by the dietician within 2 weeks following diagnosis, and only 40% within 6 months. 77% were seen by a clinician at 12 months but only 47% had a repeat tTG at this appointment. Finally, only 20% of those eligible were transitioned to adult services.

Conclusion Clinicians face many difficulties when diagnosing celiac disease. Most notably convincing families to return to a gluten-containing diet prior to further investigations; and