Botulinum toxin despite her anorectal manometry not showing high resting pressures.

Results SRUS is an important comorbidity in children presenting with constipation on a background of rectal dyssynergia/dysfunctional elimination. Our patient's significant anxiety has contributed to her dysfunctional elimination and she has ongoing CAMHS involvement so that biofeedback can be initiated. The absence of conclusive histology perhaps delayed a diagnosis being reached and her initial non-compliance with treatment was likely to be a significant contributing factor too. The key to diagnosis here was reviewing her histology in the context of her initial presenting features including passage of blood and mucus alongside straining and tenesmus.

Conclusions Whilst SRUS is a relatively uncommon diagnosis seen in the paediatric population, good history taking and consideration of clinical features is key to its diagnosis, even if histology is not confirmatory.

British Association of General Paediatrics

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INVESTIGATION AND MANAGEMENT OF VITAMIN B12 DEFICIENCY: EXPERIENCE IN A TERTIARY PAEDIATRIC CENTRE

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Background Vitamin B12/Cobalamin (Cbl) is a water soluble vitamin which is found in animal products (liver, salmon, beef, egg, chicken), milk products, and fortified cereals. Breast milk is sufficient to meet the demands of 0–6 month olds as long as the mother is not herself deficient. Causes of deficiency include dietary (low protein or protein restricted diet), pernicious anaemia, small bowel surgery, inflammatory bowel disease, deficient intrinsic factor, disorders in cobalamin transport and inborn errors of metabolism. Patients can present in a variety of non-specific manifestations such as pancytopenia, lethargy, chest pain, developmental delay, and weakness thus will be seen across different specialties. B12 deficiency can have significant long-term neurological effects, and the finding mandates appropriate further aetiological investigations and immediate treatment.

Objectives This audit will review current practice across all specialties at a tertiary paediatric hospital in the United Kingdom to inform further changes required to ensure optimal practice.

Methods Our study period spanned May 2018 to May 2020. We obtained data from our biochemistry department which showed 221 results with low B12 levels. We audited a random selection of the data to see if key investigations for vitamin B12 deficiency had been conducted including: Plasma homocysteine, plasma and urine methylmalonic acid (MMA), folate, full blood count (FBC), auto antibodies, intrinsic factor (IF), genetics, cobalamin, endoscopy as well as specialist investigations such as MRI and nerve conduction studies (NCS). We also documented the main diagnosis of the patient, suspected cause of B12 deficiency, treatment plan, and if a repeat vitamin B12 level was normal.

Results Main documented cause of low Vitamin B12 include a metabolic diet (20%) mainly those on protein restriction. Other causes include gastroenterological causes (8%) and low maternal levels in infants (5%).

49 patients (52%) had no documented cause for their vitamin B12 deficiency, of these 15 (30%) were treated with either IM or enteral vitamin B12 and of these patients 73% had repeat blood tests. Of those who were not treated, only 21% had repeat samples taken. Of all the patients, only 43% had documentation of treatment either as supplemental drop/tablets, or an IM injection.

Haematological presentations included persistent neutropenia, low vitamin B12 with variable blood sugars, and haemolytic anaemia. Homocysteine, and MMA were sent along with FBC and folate. In 1 patient, specialist investigations were sent (autoantibodies, intrinsic factor, and genetics).

Conclusions Vitamin B12 deficiency spans multi-disciplines and is an important investigation to consider for many presentations and currently there is no set local guideline; the majority of patients are not being fully investigated or treated. There is also inadequate documentation and follow-up for these patients. Metabolic patients in particular who are on dietary restrictions such as a low protein diet should have regular levels checked and treated as appropriate due to their higher risk of deficiency. Recommendations from this audit suggest a trust guideline for the initial investigation of low vitamin B12 levels, appropriate and timely management and follow-up.

British Association for Paediatric Nephrology

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A STATE OF THE ART: USING 3D MODELS TO ALLOW BETTER PLANNING OF COMPLEX VASCULAR SURGERIES AND ENHANCING SAFETY IN CHILDREN

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Background 3D organ printing allows better surgical planning and enhances safety for complex cardiac and renal transplant surgeries in adults. We present the use of 3D printing of abdominal organs and vessels for two paediatric patients with complex renovascular disease who underwent aortic graft bypass and bilateral renal auto-transplant. To our knowledge, this is the first use of 3D printing for this cohort of complex patients.

Objectives To explore ways in which complex surgeries in children can be improved allowing for optimal multidisciplinary surgical planning, contributing to better consent process and enhancing patient safety.

Methods We used 2D abdominal MRI to create a 3D model of kidneys, abdominal aorta, IVC and pancreas. Segmentation, the process of converting 2D slice data CTA scans to 3D surface models, was completed in Materialise Mimics. This surface model was exported as an STL file and then printed using a Stratasys Objet260 Connex3 polyjet printer. Soluble support was used for minimal post-print clean up. All of this in the laboratory at Great Ormond Street Hospital for Children.

Results The first patient was a 7 year old girl with a background of neurofibromatosis type 1 who underwent left nephrectomy and had 7 angioplasties on the remaining right kidney. An aortic bypass was performed and the kidney was successfully auto-transplanted onto the right iliac vessels.

The second patient was a 2 year old boy with middle aortic syndrome in which a bypass was performed from the thoracic aorta to the aortic iliac bifurcation with successful bilateral renal auto-transplantation. 3D models allowed the team for better planning of surgical approach and in the first case, initially planned surgical approach was amended once the model was reviewed.

Patients and families commented that seeing the model made them better understand the operation and it contributed to better patient information and consenting pathway.

Conclusions This is the first report on the use of the 3D printing for planning of complex vascular surgeries involving major arteries in children with renovascular hypertension and associated severe middle aortic syndrome. Both cases were successfully performed and 3D models benefitted both the surgical and medical planning, enhances safety of complex procedures, as well as improving family understanding of the complexity of the procedure and adding to the quality of informed consent.

British Association for Paediatric Nephrology

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A CASE SERIES OF PAEDIATRIC KIDNEY TRANSPLANTS DURING THE ONGOING COVID-19 PANDEMIC

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Background During the current COVID-19 pandemic, multiple lockdowns and a high incidence of cases severely impacted many European transplant programs. During the first wave of the pandemic, there was a need to stop both the living donor (LD) and deceased donor (DD) paediatric kidney transplant (KT) programs in our centres, subsequently reopening them fully. In contrast, increased confidence in our processes and outcomes led us to keep our deceased donor paediatric programme open during the UK's second COVID-19 wave.

Objectives We report our experience with 25 children who received a KT during the current ongoing COVID-19 pandemic.

Methods From May 2020 to February 2021, all paediatric KT recipients were recorded and followed up in Evelina London Children's Hospital, Southampton Children's Hospital and Great Ormond Street Hospital. Covid-secure pathways were established for both LD and recipients following the UK guidelines on COVID-19. We prospectively recorded transplant outcomes and instances of SARS-CoV-2 infection in KT recipients.

Results There was 100% patient and 100% renal allograft survival in all 25 (12 (48%) female) KT recipients aged 2 to 17 (median 11) years of whom 18 were from LD (72%) and 7 from DD (28% [5 transplants from donors after brain death and 2 from donors after cardiac death]). Two patients (8%) developed COVID-19; one of them 5 weeks post-KT presenting with low grade fever and high CRP for one week without acute kidney injury during admission for surgical complication. The second one 4 month's post-KT, presenting with low grade

fever without any further complication. Four (16%) transplants were intraperitoneal, with 21 (84%) extraperitoneal, including an en-bloc KT. There were no vascular complications and two ureteric complications requiring surgical intervention. All of the patients shielded as per local guidelines.

Conclusions During the COVID-19 pandemic, different strategies had to be taken on the transplant programme to enable paediatric KT programmes to continue. This enabled safe and effective transplantation options from both living and deceased donors. In our experience, two transplant recipients acquired COVID-19 post-transplant without renal allograft dysfunction and did not require any changes to the immunosuppression.

Association of Paediatric Emergency Medicine

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DE-LABELLING FALSE PENICILLIN ALLERGY IN ACUTE PAEDIATRIC SETTINGS: EVALUATING EVIDENCE OF PRACTICABILITY

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Background Many children present to emergency departments (EDs) with a reported allergy to penicillin.¹ There is growing data to support key historical features to accurately stratify patients into low and high-risk groups which is crucial in application of direct provocation test (DPT).² Non-immediate mild cutaneous reactions to penicillin antibiotic are classed as low risk symptoms where DPT with penicillin can be applied. It is apparent that removal of penicillin allergy labels reduces undesirable health care risks, antibiotic resistance, and increased costs.¹

Objectives To answer a 3-part question: In [children presenting with history of non-immediate benign skin rash to penicillin therapy], [can direct provocation test] with a penicillin antibiotic if clinically indicated [safely de-label the allergy] in acute settings?

Methods Search was carried out independently by the authors in PubMed, EMBASE, Cochrane Library, and BestBETs Database from inception to November 2020 using search strategy: (('penicillin allergy' OR 'provocation test') OR ('beta lactam allergy' OR challenge) OR ('penicillin allergy' AND 'de labelling')). The search included studies with reaction to DPT as their main outcome. Studies of mixed adult-children population, and methodology involving prior or concurrent skin testing were excluded.

Results Seven studies were identified to answer the clinical question. The studies aimed to de-label penicillin allergy with oral penicillin-based antibiotics in outpatient settings. Six were prospective observational studies while one was a retrospective study. Notably, a study performed in a US paediatric emergency department was not included because of prior skin prick testing. One of the reviewed studies provided the largest sample size (818 children) in the literature demonstrating the safety and accuracy of DPT with amoxicillin in children with reported non-immediate benign skin rashes. Results of negative DPT in the reviewed studies ranged from 86% to 100%. No severe reaction was reported. However,