treatment. Further research is required to explore the possible mechanisms behind these findings.

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British society for paediatric and adolescent rheumatology

G308 COMBINED RHEUMATOLOGY AND ORTHOPTIC-LED SCREENING CLINICS FOR JUVENILE IDIOPATHIC ARTHRITIS

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Background Uveitis in Juvenile idiopathic arthritis (JIA) is prevalent in 8–30% cases. BSPAR and the RCPOphth (2006) jointly has recommended guidelines for Screening for Uveitis (SfU) in Juvenile Idiopathic Arthritis (JIA). However, concerns with compliance with screening remain (*East of England audit of service standards 2012*).

Aims This review aims to study the effectiveness of uveitis screening following implementation of local changes to improve compliance to the recommended SfU guidelines.

Method Following a local review of uveitis screening against set SfU standards between 01/06/2014 – 31/05/2016, the paediatric rheumatology multidisciplinary team was expanded to include trained orthoptists to do slit lamp examination on children with JIA in the same outpatient paediatric rheumatology clinic appointment session. The orthoptist performs VA testing and basic orthoptic assessment. Slit lamp was purchased and installed in the paediatric outpatients department. The orthoptic-led clinics commenced in 01/12/2017. Uveitis screening compliance on children with JIA between 01/12/2017 – 30/06/2019 against set SfU standards was then analysed.

Results 41 children (12 males, 29 females M: F = 0.4:1) with age range 2–16 years (mean 9.2 years) were reviewed across 108 attended appointments. 9 new referrals were received. Prior to the implementation of the orthoptic-led clinics, 35.4 percent children were seen within 6 weeks of the diagnosis of JIA; in addition, there were significant delays (2.1% compliance in the first year of diagnosis) in follow up appointments against SfU standards. Following the orthoptic-led clinics 75% children were seen within 6 weeks and follow up compliance was achieved in 67% children. 33% of new patients were seen on the day of referral. 7.3% children were referred back to paediatric ophthalmology clinics.

Conclusion Significantly better adherence to the recommended SfU standards was achieved with the implementation of the orthoptic-led clinics. Installation of slit lamp in paediatric outpatients and a trained orthoptist as a part of multidisciplinary assessment of children with JIA should lead to continued improvement in achieving uveitis screening standard, avoid extra hospital visits and improve patient experience.

G309 A TALE OF TWO BROTHERS – CASE REPORT AND LITERATURE REVIEW OF TREATMENT FOR ADENOSINE DEAMINASE 2 (DADA2)

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Background Two brothers, both with past history of early-onset strokes (bilateral thalamic infarcts aged 6 years in one and left mid brain infarct aged 4 in other) were referred to the neurology and genetics service. Investigation revealed iron refractory anaemia (subsequently managed by haematology) and isodisomy for chromosome 22. Both made a good neurological recovery and were commenced on daily aspirin. Gene CECR1 was considered but dismissed due to no reported myalgia, fever or skin rash.

Five years after initial referral whole genome sequencing revealed a homozygous mutation in the adenosine deaminase 2 gene (CECR1). Both brothers were diagnosed with adenosine deaminase 2 deficiency (DADA2) and referred to paediatric rheumatology. Review of previous investigations revealed persistently raised inflammatory markers. Hepatosplenomegaly was present in both and lymphadenopathy and ear symptoms in younger brother on initial rheumatology review.

Mutations in the CECR1 gene prevent it correctly encoding the enzyme Adenosine Deaminase 2. This is a growth factor for endothelial cells and promotes the differentiation of M2 macrophages. Deficiency results in decreased vascular integrity, a predominance of proinflammatory macrophages and perivascular inflammation mediated by tumour necrosis factor (TNF).

Methods A literature review was carried out to determine evidence of treatment for this rare condition.

Results DADA2 is currently extremely rare with only 200 cases reported in the literature. Small studies have found anti-TNF medication to be beneficial in reducing glucocorticoid use and preventing further neurological events. There are case reports of successful allogeneic hematopoietic stem-cell transplantation but concerns remain that associated vascular problems may increase the risk of serious side effects.

Conclusion Following literature review an internal funding request for Adalimumab was accepted and both boys commenced anti-TNF treatment.

We present a case of two brothers with this recently discovered monogenic systemic vasculopathy, now recognised to show huge clinical diversity even among family members with identical mutations. Immunodeficiency and haematological features are commonly reported but awareness across all specialties is needed. It is currently unknown how defects in a single gene results in these heterogeneous presentations. It is hoped that gene therapy will provide a cure for this condition in the future.

G311 A RARE CASE OF JUVENILE IDIOPATHIC ARTHRITIS FOLLOWING A RUPTURED BAKER'S CYST IN A TODDLER

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Introduction A Baker's Cyst is usually an incidental finding in adults being investigated for a joint arthropathy and its rupture preceding the diagnosis of Juvenile Idiopathic Arthritis (JIA) is rare in children. Here we describe a case of a 4 year