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

old girl who presented to the Emergency Department with non-traumatic right calf pain and swelling.

Case Summary A 4 year old girl presented to Emergency Department (A&E) with a 3 week history of right calf swelling after her usual, non-traumatic dancing. The swelling had worsened over 48 hours. On examination, her weight was 20 kg, just above the 75th centile with a BMI of 17.9. She was unable to fully extend her right leg or weight bear due to her calf swelling. Her right calf measured 28.6 cm compared to the left calf of 26.4 cm. She had been afebrile with no erythema of the joint. There was no demonstrable joint effusion and no issues with joints above and below. There was no family history of clotting disorder or joint arthropathy.

Ultrasound Scan showed a fluid collection in her right gastrocnemius. MRI confirmed a ruptured Baker's Cyst with inflammatory arthropathy alongside an extensive synovial proliferation throughout the knee joint with large joint effusions and associated soft tissue oedema tracking superiorly and inferiorly along the medial head of gastrocnemius and anteriorly along the tibia. Her was ESR of 26 mm, CRP <4 and a normal white cell count. clotting screen and auto-immune panel was negative. investigations revealed bilateral uveitis consistent with a diagnosis of JIA. resolution of joint pain and swelling only occurred after initiation of a short course of oral steroids and steroid injection to the right knee joint.

Clinical Implication This case highlights the atypical presentation of JIA and the clinical manifestations are highly variable in the presence of a ruptured Baker's Cyst. Although, Baker's cysts rare in children. Majority of them are managed conservatively with no other associated comorbidity. To our knowledge, this is the first case of JIA presenting with calf swelling in a toddler

Conclusion We suggest clinicians consider a possible joint inflammatory arthropathy in children presenting with a ruptured Baker's Cyst as early diagnosis and treatment could reduce morbidity associated with this inflammatory arthropathy.

G313 SHOULD RHEUMATIC FEVER AND POST-STREPTOCOCCAL REACTIVE ARTHRITIS BE MANAGED IN THE SAME WAY? A LITERATURE REVIEW

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Aims Although the incidence of rheumatic fever (RF) in the world is declining, the incidence of post-streptococcal reactive arthritis (PSRA) is on the rise. It remains unclear whether both diseases are separate entities or if PSRA is a form of RF. The management of PSRA varies. This review investigates and evaluates the similarities and differences between RF and PSRA, with a specific emphasis on the evidence base for current management guidelines for PSRA in literature.

Methods In this literature review, a focused and methodical search was done on PubMed, Cochrane Library, Google Search, using the words rheumatic fever and post-streptococcal reactive arthritis. Studies which were published between 1999 and 2019 were reviewed with regard to definition, aetiology, epidemiology, clinical presentation, diagnosis and treatment, with the intention of establishing if there is an

evidence-based definitive management for post-streptococcal reactive arthritis.

Results In terms of clinical presentation, RF and PSRA present differently. The age at which paediatric patients are affected by both RF and PSRA is similar, although PSRA has a second peak in adulthood. Additionally, the arthritic presentation in RF is more prolonged (2–3 weeks), whereas in PSRA lasts for 7–10 days. In terms of response to treatment, RF responds better than PSRA, which further indicates the need and importance for defining clear management guidelines for PSRA. There is limited evidence to support antibiotic prophylaxis in PSRA.

Conclusion Current literature supports rheumatic fever and post-streptococcal reactive arthritis as separate entities. However, the management for post-streptococcal reactive arthritis remains unclear. The lack of evidence supporting commonly used management guidelines in PSRA demonstrates the need for further research in this area.

G314(P) A SCOPING REVIEW TO SUPPORT THE DEVELOPMENT OF PGALSPLUS: A MULTI-PROFESSIONAL TOOL AND EDUCATIONAL RESOURCE

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Background Musculoskeletal (MSK) problems in children and young people (CYP) are common. The majority present to healthcare professionals in the community, but it can be challenging to identify those with serious disease. pGALS (paediatric Gait, Arms, Legs and Spine) was developed as a simple MSK clinical assessment to discern abnormal joints, initially within paediatric rheumatology and general paediatrics; pGALS can detect joint and functional problems in CYP with other serious conditions but alone is unlikely to be specific enough to diagnose definitive conditions or signpost referrals to orthopaedics, rheumatology, neurology or general paediatrics. Our aim was to scope the literature about MSK assessments applicable to CYP used in practice, focusing on evidence of validity in the context of diagnosis and assessment of Juvenile Idiopathic Arthritis (JIA), Mucopolysaccharidoses (MPS), Muscular Dystrophy (MD) and Developmental Coordination Disorder (DCD) as exemplar MSK conditions.

Methods Scoping review using a Library search tool (including several databases), Google Scholar, and consulting NICE guidance/pathways. Search terms included dyspraxia, paediatric MSK assessment, screening tools, balance, rheumatology, assessment tools for MD, MPS, JIA. Studies cited within relevant articles were also checked.

Results 32 articles were deemed appropriate, describing specific assessment or screening tools in the context of diagnosis. Within DCD, motor co-ordination batteries aid diagnosis as part of specialist assessment, but are regarded as too lengthy for the purpose of screening; a questionnaire may be a useful first-step diagnostic tool, along with assessment of static balance (found to be significantly worse in children with DCD). In paediatric rheumatology, pGALS is the only validated screening tool to discern normal from abnormal. For