tracheobronchitis. She was commenced on Rituximab and a six-month course of Cyclophosphamide.

CT Neck and thorax showed focal obliteration of the airway at the level of hypopharynx and narrowing of left main bronchus. CT bronchogram showed the narrowing improved on a PEEP of 12 cm H₂O. She was trialled on BiPAP overnight which helped her symptoms. She was referred to Great Ormond Street Hospital for a review; no further changes in management were suggested.

The patient had recurrent relapses needing intensive care, therefore she was started on methotrexate and adalimumab, and had a balloon dilatation for her subglottic oedema and stenosis, which she tolerated well. She has been discharged home on overnight BiPAP with a plan for monthly balloon dilatations with ENT and to continue on methotrexate and adalimumab with gradual wean of steroids.

Conclusion RP is rare with estimated incidence of 3.5 per million, only 10% cases are seen in paediatrics. Without aggressive and prompt treatment, life-threatening complications can develop rapidly, and outcomes are poor. Managing children with RP with significant airway involvement with NIV and repeated monthly balloon dilatations can help prevent life threatening exacerbations and the need for a tracheostomy. This case demonstrates the importance of multidisciplinary management of this complex condition.

G305(P) ABSTRACT WITHDRAWN

G306(P) A RARE CASE OF CONGENITAL LEFT LUNG APLASIA DIAGNOSED ANTENATALLY

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Introduction Pulmonary agenesis was first described in 1673 by De Pozze in a female cadaver biopsy. The condition is characterised by undeveloped pulmonary vessels, bronchi, and parenchyma. It can present bilaterally or unilaterally and, in the latter, the remaining lung contains twice as much alveoli as normal, but has normal bronchi. Although the aplastic side does not have the same structures, it has a rudimentary bronchus. It is extremely rare, occurring in 1:100,000 births. Nearly a third of patients have associated congenital heart diseases. We report a case of unilateral left lung aplasia which was diagnosed antenatally.

Case Report A female baby was born by vaginal delivery at term, weighing 3700 grams. Antenatally the baby was noted to have mediastinal shift on 20 week scan, raising suspicion and leading to a diagnosis of complete left lung aplasia, left pulmonary artery hypoplasia, with normal right lung on subsequent MRI scan at 28 weeks gestation. Foetal ECHO showed normal cardiovascular anatomy.

Baby cried immediately at birth and did not require any resuscitation but started to show signs of respiratory distress at about 15 minutes of age. Baby was transferred to the neonatal intensive care unit and managed on 6 litres of high flow until 12 hours of age. Chest x-ray shows mediastinal shift to the left, with possible right lung

hyperplasia and a small undeveloped left lung. She was subsequently breathing by herself with no signs of respiratory distress and discharged home with paediatric respiratory follow-up.

Conclusion Unilateral lung aplasia is a rare congenital anomaly, usually diagnosed later in life as an incidental finding. It is very rare for antenatal diagnosis, especially if not associated with congenital heart defects. This case was identified because of the mediastinal shift. Children with unilateral lung aplasia are at an increased risk of respiratory tract infections and need regular follow up with respiratory team. Right sided lung aplasia might be associated with Scimitar syndrome. Finally, cases from the literature show poorer outcomes associated with right sided lesions (30% survival into adulthood) compared to left sided lesions (50% survival into adulthood).

G307(P) DOES RAISED FRACTIONAL EXHALED NITRIC OXIDE ALWAYS MEAN POOR COMPLIANCE? A CASE BASED OBSERVATION

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Background Fractional exhaled nitric oxide (FeNO) is a well-established inflammation marker in asthma monitoring. It has a diagnostic and prognostic value, helpful in predicting exacerbation rates.¹ The expectation with initiation of treatment is for lung function to improve and FeNO to decrease as inflammation resolves. In some patients, FeNO rises further despite treatment, which some suggest is due to poor compliance. However, this is inconsistent for patients where lung function improves.

Objective Demonstrate that raised FeNO after treatment initiation does not necessarily equate poor compliance.

Method Retrospective retrieval of full spirometry results of pediatric patients started on asthma treatment in the last 2 years.

Results 5 children/young people have incongruent improvement in FEV1 in parallel to an increase in FeNO after initial treatment (table 1).

Conclusion The results question whether increased FeNO is always due to poor compliance. Perhaps the initial low FeNO is due trapped gas, which then escapes as airways open with

Abstract G307(P) Table 1

Patient	FEV1% predicted (%)	FeNO (ppb)	Time interval (days)
1	86	51	0
	94	92	216
2	67	31	0
	69	139	175
3	78	14	0
	93	50	350
4	83	17	0
	97	38	259
5	92	12	0
	93	29	77

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

REFERENCE

1. Petsky HL, et al. Tailoring asthma treatment on eosinophilic markers. *Thorax* **73**, (2018).

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