people (<29 weeks of gestational age) of whom more than 90% were exposed to both antenatal steroids and postnatal surfactant.

Methods Lung function was assessed in 59 (34 of whom had BPD) young people. Airway function was assessed including by forced expiratory flow at 75% and 25% of the expired vital capacity (FEF₇₅, FEF₂₅₋₇₅), forced expiratory volume in one second (FEV₁) and peak expiratory flow (PEF). Lung volumes were assessed including by total lung capacity (TLC), forced vital capacity (FVC), functional residual capacity by helium gas dilution (FRC_{He}). Gas transfer was assessed by the diffusing capacity for carbon dioxide (DL_{CO}). The results were converted to z-scores, corrected for height, sex and ethnicity using GLI reference ranges.

Results The BPD and no BPD group were of similar height and weight at assessment. The lung function results are expressed as mean z score (standard deviation). Airway function (FEF₇₅ -1.31 (1.35) versus -0.64 (0.89), p=0.035; FEF₂₅₋₇₅ -1.88 (1.42) versus -1.10 (0.84), p=0.020; FEV₁ -1.41 (1.44) versus -0.52 (1.23), p=0.017) and gas transfer DLCO (-1.50 (1.22) versus -0.72 (1.22), p=0.019) were significantly lower in those with BPD compared to those who had not developed BPD. Lung volumes (TLC 0.81 (1.24) versus 1.01 (0.82), p=0.504; FVC -0.54 (1.36) versus 0.13 (1.53), p=0.086 and FRC He 0.30 (1.11) versus -0.16 (1.20), p=0.669) were similar in the BPD and no BPD groups.

Conclusions Airway function was significantly lower in the young people who had BPD, whereas lung volumes did not differ significantly between the two groups. Those results suggest that dysynaptic growth may be more pronounced in those who had BPD.

G450 AN INTEGRATED APPROACH USING QUALITATIVE METHODS TO IDENTIFY PERCEPTIONS OF ASTHMA IN BRITISH SOUTH ASIAN AND WHITE BRITISH CHILDREN

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Background and aims Childhood asthma places a significant physical, financial, and psychological burden on patients, families, communities and the healthcare system. Current research indicates certain minority ethnic groups, especially South Asian children, encounter inequalities in asthma management and outcomes. Furthermore, children's views and aspirations are infrequently evaluated. We report here the process of encouraging children's participation in a multiphase participatory the Management in Asthma (MiA) study. The aim of this study was to explore the perceptions and experiences of asthma and its management in British South Asian (BSA) and White British (WB) children, using adapted qualitative methods.

Methods Two qualitative methods were used: semi-structured interview and prioritisation workshop, adapted to encourage children's participation (table 1). 14 themes for optimising asthma management identified from the semi-structured interviews and were used in the ranking exercises. Children ranked

these themes by linear (LI, non-equal ranking), diamond (DA, equal ranking permitted) and budget pie (BP, ranking by funds allocation).

Abstract G450 Table 1

Qualitative methods	Adaptations
Semi-structured interview	Drawings and text to prompt questions Extended time for creative activities e.g. drawings
Health Prioritisation workshop	Trained 'peer' researchers (14–16 year-old) as facilitators Toy money and board games

Results Semi-structured interviews with children, with or without parents/guardian present (33 BSA and 14 WB children with asthma aged 5–12-years-old) revealed considerable similarities in experiences of asthma, notably inadequate holistic approach to asthma management amongst healthcare professionals and the role of schools and peers in children's asthma perception and management. Barriers to health information provision existed in both groups. Consistently, WB children's perception of asthma aetiology was different from BSA children. In all the ranking methods, children (21 BSA and 1 WB child aged 5–12-years-old) highly prioritised acute attacks according to their perceived health importance. 'Strength of opinion' was demonstrated in BP, as acute attack received £1430 in total, whilst the 2nd place theme only received £610.

Conclusion Future intervention in children with asthma may wish to focus on managing acute attacks by a holistic approach, by involving schools, families and healthcare professionals. Furthermore, ethnospecific divergent beliefs about asthma aetiology can be tackled by a targeted educational programme.

G451 JUVENILE SYSTEMIC LUPUS ERYTHEMATOSUS (JSLE) PRESENTING AS IDIOPATHIC INTRACRANIAL HYPERTENSION

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Background Idiopathic intracranial hypertension (IIH) is a rare but recognised presentation in Juvenile SLE. We describe a child who presented with IIH and was also found to have bilateral thromboembolism.

Case presentation 13 year old girl presented with a 6 week history of fatigue, headache, blurred vision, abdominal pain and painful fingers. Examination showed faint malar rash, small joint arthritis and bilateral papilledema. Diagnosis of IIH was confirmed by elevated CSF opening pressure 27 cm H₂O. Normal CSF sugar, protein and cells and neurological examination. No radiological evidence of underlying pathology including a normal CT and MR venogram. Laboratory workup revealed normocytic normochromic anaemia (9.9), lymphopenia (0.6), low platelets (110), hypocomplementemia, raised ESR (90), positive antinuclear antibody and positive anti ds

DNA. Clinical findings and serological criteria confirmed the diagnosis of Juvenile SLE.

She was treated with intravenous pulse methylprednisolone, oral prednisolone acetazolamide and hydroxychloroquine. Headache and papilledema resolved however she was represented 2 weeks later with chest pain and breathlessness but on review of history she admitted to breathlessness going back several weeks, CT pulmonary angiography showed bilateral acute pulmonary embolism. Although she had thromboembolism her anticardiolipin, lupus anticoagulant, Beta 2 glycoprotein 1 antibodies were negative. Diagnosis of antiphospholipid syndrome could not be made as it did not fulfil the criteria. Anticoagulation treatment was commenced. She was also given 2 doses of Rituximab. She remains well with clinical improvement as well as improvement in her inflammatory markers.

Discussion Our patient illustrates SLE can present with idiopathic intracranial hypertension though very rare and exact pathophysiology remains unclear. Various mechanisms have been proposed including venous thrombosis, immunological or inflammatory but none have been proven. There have been few case reports in paediatric population as first presentation of lupus.

Conclusion SLE should be considered as a possible cause in a child presenting with idiopathic intracranial hypertension and a connective tissue screen should be included in the work up. In a known patient with SLE, IIH may be part of the spectrum of neuropsychiatric manifestation and should be considered as a possible cause

G452(P) A COMPLEX CASE OF CYSTIC FIBROSIS AND COFFIN-SIRIS SYNDROME

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We describe a 4-year-old girl with advanced Cystic Fibrosis (CF) and Coffin-Siris syndrome.

Antenatally, there were concerns about Down Syndrome. She was born at term weighing 3388 g. Dysmorphic with micrognathia, broad nasal bridge, (P)iorly rotated ears, short fingers, almond-shaped eyes with thick eyelashes and fixed bilateral talipes. Genetics for Down Syndrome negative. Admitted to Neonatal Unit with stridor and obstructive breathing requiring Vapotherm.

On newborn screening, identified as having CF (Delta F508/3849+10kbc >T) but pancreatic sufficient. Respiratory polysomnography confirmed partial obstruction with mixed events and microlaryngoscopy and bronchoscopy identified a retroverted larynx and laryngomalacia. In view of micrognathia, abnormal upper airways, obstructive breathing, and ventilatory support, required tracheostomy at 1 month. Successfully decannulated at 6 months. Significant global developmental delay and hypotonia apparent in the first few months and entered into the Decipher Developmental Disorders (DDD) study.

Frequent respiratory exacerbations and poor growth, remaining an inpatient for the first 6 months. Port-a-cath inserted at 4 months for intravenous (IV) antibiotics. High resolution computed tomography (HRCT) chest showed significant volume loss in both lower lobes, with possible associated traction bronchiectasis. *Burkholderia cepacia* isolated in sputum at 7 months and *Pseudomonas aeruginosa* at 22 months. Aged 2

years, she required home oxygen at 1 L/min. At 3 years, bronchoscopy showed thick, copious secretions throughout inflamed airways. Bronchoalveolar lavage showed no evidence of lipid-laden macrophages, but *Pandorea* isolated. HRCT chest significantly progressed with extensive widespread bronchiectasis, multifocal consolidation, mosaic attenuation and peripheral tree in bud opacities.

Other multisystem problems include severe gastro-oesophageal reflux, feed aversion and poor growth (weight SDS -3.09, height SDS -2.52) requiring laparoscopic fundoplication and gastrostomy at 8 months. Despite fundoplication, vomiting remains problematic. Left renal and ureteric calculi incidentally found on annual review abdominal ultrasound at 2 years requiring lithotripsy. Results from the DDD study confirmed ARID1A mutation and a diagnosis of Coffin-Siris Syndrome.

Discussion

- In a child with CF and respiratory disease out with that expected for genotype, it is important to actively seek further diagnoses.
- Coffin-Siris Syndrome is a rare multisystem disorder manifested by craniofacial abnormalities, recurrent respiratory tract infections, failure to thrive, vomiting, hypotonia, developmental delay and renal or genitourinary abnormalities.

G453(P) A SURVEY OF PAEDIATRIC BRONCHOSCOPY DECONTAMINATION PRACTICE IN UK REGIONAL CENTRES

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Aims Flexible bronchoscopes are difficult to decontaminate due to their intricate internal structure. The British Thoracic Society (BTS 2013) and the Department of Health England (Heath Technical Memorandum 01-06 2016) have published guidelines on flexible endoscope decontamination.

'The Health and Social Care Act 2008: Code of Practice on the prevention and control of infections and related guidance' states that 'decontamination policy should demonstrate that it complies with guidance establishing essential quality requirements and a plan is in place for progression to best practice'. The objectives of the study were to review the practice of bronchoscope decontamination in accordance with BTS (2013) or HTM 01-06 (2013) guidelines, in UK tertiary paediatric centres and to assess for related adverse events.

Method A 10 question web-based survey was emailed to 26 paediatric tertiary centres which perform bronchoscopy in the UK. Questions considered adherence to BTS (2013) or HTM 01-06 (2013) guidelines and associated nosocomial infection between 01/01/2015-01/01/2016.

Results 11 of the 26 trusts contacted returned a completed survey.

5/11 trusts followed HTM 01-06 guidelines and 3/11 followed BTS guidelines, total 8 of 11 responding trusts.

4 (0.247%) paediatric bronchoscopies performed between 01/01/2015-01/01/2016 resulted in nosocomial infection. These were all from the same centre. These infections resulted in either delayed discharge or readmission.

2/11 of the responding centres, reported sampling from the suction channel of the bronchoscope prior to each procedure