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

for surveillance. These centres did not report any nosocomial infections during the survey timeframe.

Conclusions Response to the survey and adherence to BTS (2013) and HTM 01–06 (2016) guidelines were lower than expected. Nosocomial infection was rare but associated with significant morbidity, and occurred only where water channel sampling was not practiced. It is recommended that steps are taken to improve adherence to decontamination guidelines and include suction channel sampling for surveillance.

G454(P) FLAGELLATE ERYTHEMA ASSOCIATED WITH JUVENILE DERMATOMYOSITIS

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Aim Flagellate erythema was first reported in association with bleomycin treatment and since then has been linked to chemotherapeutic agents, dermatomyositis, adult-onset Still disease and Shiitake mushroom dermatitis. To date, only one patient with juvenile dermatomyositis and flagellate erythema has been reported in the literature. Hence, we would like to present this case and highlight importance of this uncommon dermatological condition in children.

Method A 9-year-old girl presented in 2016, with a 2 month history of new onset proximal muscle weakness, heliotrope rash, Gottron's papules, leg erythema and soft tissue restriction at elbows, hips and knees. Muscle biopsy and MRI confirmed myositis and fasciitis. Juvenile dermatomyositis was diagnosed.

She commenced steroid therapy, initially intermittent monthly pulsed intravenous methylprednisolone, with oral prednisolone, in conjunction with subcutaneous methotrexate and oral hydroxychloroquine. Two months later, due to lack of significant improvement and deterioration in muscle function, intermittent pulsed intravenous cyclophosphamide was substituted for methotrexate, with improvement in muscle weakness.

Three months into cyclophosphamide therapy, she developed a florid, widespread eruption, with violaceous, urticated erythema of the face and upper trunk with areas of sparing. There were also erythematous, linear flagellate areas on her proximal limbs and back. Dermatographism was absent. There was no associated deterioration in muscle strength or elevation of inflammatory markers or muscle enzymes at that time. No relation to medication or potential allergic exposures was noted.

Intravenous immunoglobulin therapy was commenced but unfortunately caused an anaphylactic reaction. Methotrexate was restarted and she has been maintained since on a combination of this with hydroxychloroquine, sun protection, topical steroids and calcineurin inhibitors with improvement in her skin and muscle strength and oedema on MRI.

Result Skin biopsy showed a marked vacuolar interface reaction with colloid bodies and epidermal atrophy associated with a mild perivascular lymphocytic infiltrate in the superficial dermis with absence of eosinophils and increased dermal oedema and mucin. The features were in keeping with a flagellate erythema secondary to dermatomyositis.

Conclusion The pathogenesis is not completely clear, although physical injury, sun exposure or minor trauma could be

incriminated. The significance of this rash in relation to the prognosis or underlying neoplasm in adults remains unclear.

G455(P) LOWER RESPIRATORY TRACT INFECTION IN PATIENTS WITH NEUROLOGICAL IMPAIRMENT: A NARRATIVE REVIEW ON MICROBIOTA AND ANTIMICROBIAL INTERVENTIONS

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Background Lower respiratory tract infections (LRTI) are a major cause of morbidity and mortality in patients with neurological impairment. Prophylactic antibiotics and eradication of colonisers are approaches employed in the prevention of recurrent LRTI.

Aims To determine the evidence base for this practice in children with neurological impairment.

Methods An electronic database search identified studies reporting on outcomes of efficacy for antimicrobial interventions and microbial findings relevant to LRTI in our population of interest. Results were synthesised into a narrative review.

Results Our search revealed a small case-series suggesting nebulized tobramycin may be effective in reducing the frequency of pneumonias and associated hospitalisations. We identified 5 papers focusing on microbial findings – there was significant variability in the frequency of bacterial isolation, and in the species identified. These studies were heterogeneous and subject to individual biases which limits the wider applicability of results.

Conclusions The direct evidence for antimicrobial interventions as a prophylactic strategy against LRTI in patients with neurological impairment is lacking and warrants further research. In addition, we still lack a clear understanding of the microbiota contributing to respiratory pathology, which could inform therapeutic targets.

G456(P) WAVING THE RED FLAG: CHARACTERISTICS OF CHILDREN WHO REQUIRE CRITICAL CARE FOR LIFE-THREATENING ASTHMA AND WHEEZE

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Aim The RCPCH report 'State of Child Health' (2017) reported the prevalence of deaths due to asthma in the UK as among the highest in Europe. Local differences in these indices suggest varying standards of care may be an underlying factor.

As part of an evaluation of the hospital-wide asthma service, we aimed to identify characteristics of children who required critical care in order to determine if the needs of this high-risk group are being met.

Method The PICU database was interrogated to identify children who had required critical care for wheeze or asthma. Characteristics of the critical care stay were retrieved from the clinical record to identify characteristics locally of children

who experience life-threatening episodes of acute respiratory distress.

Results Fifty-four children were admitted on 72 occasions with wheeze. 32 children with asthma, 19 children with wheeze. Three children, admitted on 4 occasions, were excluded for alternative diagnoses. Unsurprisingly children with asthma were older (mean 92.54, sd 53.8 months) than children with wheeze (mean 24.79, sd 17.52 months).

Children with asthma were each admitted to PICU more frequently than children with wheeze (mean 1.97, sd 1.43 vs mean 1.26, sd 0.56). For the 68 included episodes, respiratory support was needed on 26 occasions (formal ventilation on 12).

In 49% of cases children lived in a household where a family member smoked (15 asthma; 5 wheeze). On 15 occasions household smoking status was not documented. In 16 cases of asthma there were documented social concerns (compliance, clinic attendance, formal safeguarding) compared to only 1 case of the 19 children with wheeze. Children in the asthma group came from larger families with 3.5 children (IQR 2,4 max 11) per family compared to 2.4 children (IQR 1,3 max 7) in the wheeze group.

Conclusion Children with the most severe episodes requiring critical care often present on multiple occasions and come from large families with household smoking exposure. Concerns regarding the inappropriately low priority of the children's health by their families are common.

G457(P) INVESTIGATING THE EFFICACY OF HOME RESPIRATORY SURVEILLANCE IN THE MANAGEMENT OF RESPIRATORY CYSTIC FIBROSIS DISEASE IN PAEDIATRICS

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Background Cystic Fibrosis (CF) is an autosomal recessive condition in which much of the morbidity and mortality is attributed to the respiratory component of the disease. Microbiological surveillance of respiratory flora is vital in CF management. In addition to traditional clinic-based sampling, Wythenshawe hospital's paediatric CF department has been training parents to take cough swabs at home.

Aims To assess how effective home sampling is in the monitoring of respiratory flora of CF paediatric patients.

Methods Data on respiratory samples were collected for the year 01/01/2016- 31/12/2016. Patients notes, electronic patient records and Lorenzo were accessed. Kappa testing was used to determine the level of agreement home and clinic samples.

Results 21 patients were identified for this study.

- A total of 172 home samples were collected
- On average, each patient had 8 home samples collected
- Absolute agreement between home and clinic samples was 91.92%
- Number of home and clinic samples having excellent agreement was 10 (52.63%)
- There were several bacteria which were grown only in home or clinic samples. These bacteria are not commonly known to be contaminants
- On average home sampling lead to antibiotic treatment occurring 44.17 days before a clinic attendance

Conclusion The strong level of agreement between samples collected at home and in clinical settings suggests that the bacteria being detected at home is similar to what is detected in the clinics. The differences seen in bacteria grown in these two settings is likely to be a result of differences in the respiratory flora and not due to contamination of samples taken at home.

On average, home sampling resulted in antibiotic treatment 44.17 days before a clinic attendance would have occurred. This reduces the amount of time for inflammatory damage and antibiotic resistance to occur.

This study recommends the continuation of home sampling to boost sampling rates, thereby increasing the clinician's confidence in the each patient's microbiological profile.

Moving forward research should focus on gathering more information on the health benefits to patients receiving home sampling and on cost-benefit analysis.

G458(P) ASSESSMENT OF THE SHORT-TERM CLINICAL OUTCOME OF CHILDREN WITH POLYARTICULAR AND SYSTEMIC ONSET JUVENILE IDIOPATHIC ARTHRITIS: A RETROSPECTIVE COHORT STUDY IN A TERTIARY CENTRE SAMPLE

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Objectives To determine the early clinical improvement and short-term outcome of patients with polyarticular and systemic onset Juvenile Idiopathic Arthritis (JIA) achieved with current therapeutic agents and treatment strategies by analysing the early disease course and one-year outcome of a tertiary centre cohort of patients.

Methods All patients with a diagnosis of polyarticular or systemic onset JIA seen at a paediatric tertiary hospital were identified from the rheumatology clinic registry. Inclusion criteria was a time period of at least one year since the diagnosis. Patients were excluded if they were discharged or transferred to adults' service. Clinical records from the hospital database and medical charts were reviewed for data collection. The collected data was analysed using qualitative measures and results were drawn from observational findings. The outcomes reported in this study include: number of joints with active inflammation, systemic disease features, C-reactive protein (CRP), and functional ability of the child as recorded in the clinical letters.

Results A total of 45 patients were identified, of which 32 were eligible. The frequency of JIA subtypes: 19 patients with RF-negative polyarthritis, 7 with RF-positive polyarthritis and 6 with systemic onset disease. Out of the total, 9 (28%) were completely asymptomatic and did not show any sign of active joint inflammation or residual joint changes at one year from diagnosis. Also at one year, 15 (47%) had no signs of active disease on physical examination but musculoskeletal pain seems to persist in some. For functional ability, 81% did not have any limitation to their physical ability, while the rest 19% had variable degrees of ongoing restrictions to their mobility or activity.

Conclusion Significant improvement in disease activity was noted in all patients with the current therapeutic strategies in clinical practice. However, low levels of disease activity persisted in many