Results Data were collected for 51 patients (63% female; mean age at surgery 14.7 years). Diagnoses included adolescent (62.8%), juvenile (7.8%) and infantile (5.9%) idiopathic scoliosis, congenital (7.8%) and syndromic scoliosis (2%) and Scheuermann's kyphosis (13.7%). Mean ($\pm$ SD) thoracic curvature was $61.3 \pm 15.2^\circ$, and mean kyphosis in those with Scheuermann's was $92.4 \pm 7.3^\circ$. No correlation was elicited between FEV1 or FVC (%predicted) and VO2peak(%predicted). Greater thoracic curves were associated with lower FEV1(%predicted), $r = -0.45$, $p = 0.02$, smaller total lung capacity ($r = -0.47$, $p = 0.002$) and lower BR at the end of exercise ($r = -0.57$, $p < 0.0001$). There was a positive correlation between FEV1(%predicted) and BR ($r = 0.65$, $p < 0.001$) and a negative correlation between FEV1 and VO2 at AT ($r = -0.37$, $p = 0.008$). Those with higher VO2peak measures also had higher VO2 at AT ($r = 0.78$, $p \leq 0.001$). SRS-22 scores correlated significantly with VO2peak(%predicted) (total SRS-22 vs VO2peak(%predicted), $r = 0.50$, $p = 0.006$).

Conclusion Contrary to previously published data, those with larger thoracic curves and therefore poorer lung function show improved exercise capacity and later onset AT, likely as a consequence of improved physical conditioning. These patients also report better quality of life, which may be due to maintenance of normal exercise levels. These findings suggest that physical adaptation occurs in scoliosis with impaired lung function, and that this may be protective to patients' mental health.

G446

HIGHLY ELEVATED FERRITIN LEVELS ARE ASSOCIATED WITH HAEMOPHAGOCYTIC LYMPHOHISTIOCYTOSIS – ARE WE MISSING TREATABLE DIAGNOSES? A RETROSPECTIVE SERVICE EVALUATION OF DIAGNOSIS IN PATIENTS WITH FERRITIN $>10,000 \mu\text{g/L}$

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Background Haemophagocytic lymphohistiocytosis (HLH) is a hyperinflammatory syndrome which can complicate sepsis, malignancy or autoimmune disease and may lead rapidly to critical illness and death. Early treatment reduces mortality but diagnosis requires a high index of suspicion and correct interpretation of laboratory results. Highly elevated ferritin levels (HEF) $>10,000 \mu\text{g/L}$ are highly specific for HLH and should prompt consideration/exclusion of hyperinflammation. Diagnostic guidelines for HLH were published in 2004 requiring the presence of $\geq 5/8$ criteria.

Aim To assess recognition of HLH in a paediatric population with HEF.

Methods This retrospective study was conducted at 8 UK centres. Biochemistry databases identified patients ≤ 16 years with serum ferritin $>10,000 \mu\text{g/L}$ between 01.04.2014 and 31.03.2017. A standardised proforma was used to collect data. Cases were assessed against the 2004 HLH criteria. Due to limited access to some laboratory tests, modified criteria using

a threshold of $\geq 4/5$ (excluding tissue haemophagocytosis, decreased natural killer cell function, increased soluble interleukin-2 receptor) were also applied.

Results Ninety-four patients (60.6% male) were identified. 36% were aged <1 year, 16% 1–5 years, 34% 6–12 years and 14% 13–16 years. Management was in neonatal/paediatric intensive care in 49.4%. Diagnoses included: infections (29.8%), malignancies (21.3%), rheumatological (13.8%), immunological (9.6%) and cardiac surgery (6.4%). A diagnosis of HLH was made by the treating clinical team in 30.9% and considered in the differential in a further 18.1%. Just 5 patients had complete data available to score against all 8 2004 HLH criteria whereas 49 patients had data to score against the modified criteria. Using all available data:

- 14/94 (14.9%) met $\geq 5/8$ criteria and all these patients were diagnosed with HLH by the treating team.
- 33 (35.1%) met $\geq 4/5$ criteria and 17 (51.5%) of these were diagnosed with HLH by clinicians. HLH was not documented as being considered in the differential in 11 (33.3%)

Overall mortality was 33.0% (31/94) but was 17.2% (5/29) in those patients diagnosed with HLH during their admission.

Conclusion Although HEF is highly specific for HLH, the diagnosis was made or considered in just half of paediatric patients with this laboratory result. Increased awareness of this potentially-lethal condition is likely to lead to earlier treatment and reduced mortality.

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G447

THE CROSS-SECTIONAL AND LONGITUDINAL ASSESSMENT OF THE EFFECTS OF NURSERY ATTENDANCE AND BREASTFEEDING ON RESPIRATORY SYMPTOMS AND THEIR IMPACT ON QUALITY OF LIFE IN THE FIRST 22 MONTHS OF LIFE- RESULTS FROM THE LIVERPOOL BABY BREATHING STUDY

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Aims To determine the effects of breastfeeding and nursery attendance on respiratory symptoms and quality of life related to respiratory symptoms in infants from Liverpool over the first 22 months of life.

Methods The Liverpool Baby Breathing Study (LBBS) uses a parent completed respiratory symptom questionnaire, to assess respiratory symptoms in infants from four to twenty-two months of age. Infants were recruited from the Liverpool Women's Hospital between 23/01/2013 and 03/11/2014. Questionnaires were sent to participants at four, ten, sixteen and twenty-two months of age via post or an automated emailing system. The cross-sectional analysis of the LBBS was weighted to ensure the cohort's index of multiple deprivations was comparable to the eligible births. χ^2 , Mann-Whitney U and

Fisher's Exact Test were used to compare questionnaire scores and symptom prevalence between those who were breastfed and attended nursery and those who were not. The longitudinal analysis of the LBBS was performed using multi-level mixed effects models.

Results At four, ten and sixteen months of age infants who were breastfed had a lower symptom scores and a smaller prevalence of a variety of respiratory symptoms than those who were not. Some of these findings showed a dose-response relationship. This relationship was no longer present at twenty-two months of age. Longitudinally, breastfeeding was shown to reduce symptom scores over the first twenty-two months of life.

At four months infants who attended nursery had a higher prevalence of snoring and were more likely to attend the GP with respiratory symptoms. At ten, sixteen and twenty-two months of life nursery attenders had higher symptom scores and a higher prevalence of various respiratory symptoms than non-attenders. Longitudinally over time, nursery attendance increased symptom scores over the first twenty-two months of life. Nursery attendance reduced the quality of life of both the infant and their families due to respiratory symptoms at ten and sixteen months of age.

Conclusion Breastfeeding was protective of respiratory symptoms over the first 22 months of life. Contrastingly, nursery attendance increased respiratory symptoms and reduced the quality of life of both infants and their family over the first 22 months of life.

G448 JUVENILE DERMATOMYOSITIS ASSOCIATED WITH INTERSTITIAL LUNG DISEASE: WOULD ANTI-MDA5 ANTIBODY DETECTION IMPROVE EARLY DIAGNOSIS AND MANAGEMENT?

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Aims We describe a case of anti-MDA5 antibody positive Juvenile Dermatomyositis(JDM) associated with interstitial lung disease(ILD) and propose early antibody detection may guide management and improve outcome.

Methods A 15-year-old male presented with a 3 month history of pain and stiffness in his right ankle, left knee and hand joints associated with mild weakness. A blistering rash on his extremities developed into classical JDM facial rash with peri-orbital swelling, Gottron papules and paronychia ulcers. Initial investigations included: EMG study showing myopathy; MRI buttock and skin biopsy both consistent with dermatomyositis (see table 1). He started hydroxychloroquine, weekly methotrexate injections and high dose prednisolone including fortnightly 3 day intravenous pulses.

Results Within one month, he developed a knee staphylococcus aureus arthritis which required prolonged antibiotics. Despite minimal respiratory symptoms, his lung function test showed a restrictive pattern with reduced diffusion(FVC-7.57 predicted, TLC-4.18, DLC -2.2). ILD with patchy consolidation was confirmed on imaging. IV immunoglobulins(IVIG) were added. Two weeks later, he developed chest pain and shortness of breath, with clinical signs of surgical emphysema confirmed radiologically, with apical pneumothoraces, pneumomediastinum and pneumopericardium. The air leaks were

secondary to bronchial perforations secondary to JDM related ILD. Although the rash, arthritis and myositis responded to treatment, his ILD progressed. He required admission to PICU for respiratory support and was covered for infection including pneumocystis pneumonia. Further IVIG and IV methylprednisolone was given and mycophenolate commenced. However, his deterioration necessitated ventilation and transfer for ECMO. His bloods showed anti-MDA5 positive antibodies. Plasmapheresis, cyclophosphamide and rituximab were given but despite best efforts, he died from ILD secondary to JDM.

Abstract G448 Table 1 Laboratory results

	Diagnosis	PICU admission
Hb(g/L)	116	103
WCC($10^9/L$)	6.9	9.2
Neut($10^9/L$)	3.4	7.34
Platelets($10^9/L$)	211	256
ALT(U/L)	131	122
GGT(U/L)	85	-
ALP(U/L)	-	60
ESR(mm)	2	88
ANCA	Negative	-
ANA	Negative	-
Myositis-antibodies	Negative	-
CK(IU/L)	164	462
Ferritin(ug/L)	-	2367
LD(U/L)	-	74

Conclusion Anti-MDA5 antibodies have been reported in rapidly progressive ILD associated with JDM. Despite the usual manifestations responding to treatment, his lung disease was unremittingly progressive. The clinical features associated with this rare form included the unusual vasculitic lesions periungually and on the palms/soles. At diagnosis, we recommend requesting full myositis-specific antibodies including anti-MDA5 which may help guide management. The advent of aggressive staphylococcus aureus arthritis delayed the use of additional immunosuppressants once ILD was diagnosed and IVIG was used whilst treating the infection. Early aggressive immunosuppression is recommended.

G449 LUNG FUNCTION AT FOLLOW-UP OF VERY PREMATURELY BORN YOUNG PEOPLE – IMPACT OF BRONCHOPULMONARY DYSPLASIA

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Aims Bronchopulmonary dysplasia (BPD) is a common adverse outcome of very premature birth. Studies have given conflicting results as to whether BPD is associated with long-term lung function impairment, but few have included populations exposed to routine use of antenatal steroids and postnatal surfactant. To determine, according to BPD status, the lung function of 16 to 18 year old, very prematurely born young

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