Abstract P92 Figure 1 Spectrum for m/z 59 (1a) and capnography trace (1b). ncps = number of counts per second

Discussion The comparability of the traces for the spectrum of m/z 59 and capnography suggests that real-time PTR-ToF-MS is detecting eVOC which relate to end-tidal breathing. The traces are likely offset due to the lag between end-tidal exhalation and the sample reaching the apparatus. This feasibility study has demonstrated that real-time analysis of eVOC is possible in preschool children in an acute setting. Further work is needed to determine the most accurate way of analysing the spectra to be able to apply this novel, non-invasive method of investigating airway inflammation and pathology in preschool children.

REFERENCE

- 1 van de Kant KD, van Berkel JJ, Jöbsis Q. *et al.* Exhaled breath profiling in diagnosing wheezy preschool children. *Eur Respir.* 2013;**41**:183–8

P93 THE PRACTICALITIES OF USING ALLERGEN-IMPERMEABLE BED COVERS IN CHILDREN WITH MITE ALLERGIC ASTHMA

¹H Sumner, ²H Begum, ¹A Simpson, ¹A Custovic, ¹CS Murray. ¹University of Manchester, Manchester, UK; ²University of Salford, Salford, UK

10.1136/thoraxjnl-2015-207770.230

Objective As part of a randomised double-blind placebo-controlled study in children with mite-allergic asthma, mite-impermeable bedding encasings were evaluated in terms of reduction in allergen exposure and patient/parent acceptability.

Methods Physician diagnosed asthmatic children (3–17 years) were recruited following an exacerbation of asthma. Mite-sensitised children were randomised to either mite-impermeable encasings for the mattress, pillow and duvet (Astex Pristine; ACP solutions, Gloucestershire, UK) or placebo encasings (100% polycotton; Musbury fabrics, Rossendale, UK), in a double-blind manner. Vacuumed dust samples were collected from the child's mattress prior to fitting the encasings and at 12 months, stored at -20°C, then analysed for mite allergen content (Der p1) by enzyme-linked immunosorbent assay (Indoor Biotechnologies, Cardiff, UK). Questions aimed at assessing the practicalities of using the encasements were asked of parents by an interviewer blind to their allocation, 8–12 months later.

Results 284 children (mean age 7.7 years; 65.8% male) were randomised (146 active; 136 placebo). There was an 84% decrease in Der p 1 levels in child's mattress in those using the mite-impermeable encasings, which was not seen in the Placebo group ($p < 0.001$). Data on 'use of bedding encasements' were obtained from 232 participants. Significantly more families in the active group reported that the duvet slipped within its cover, compared to the placebo group (32.2% vs 5.3% respectively, $p < 0.001$) and that it was "noisy" (14.4% active vs 0.9% in placebo, $p < 0.001$). Some reported the extra covers made them too warm (3.4% active vs 1.8% placebo, $p = 0.64$). Overall 31 (26.3%) using the mite-impermeable encasings rated them as "uncomfortable" compared with 2 (1.8%) in the placebo group ($p < 0.001$). Furthermore, 30 (25.4%) children in the mite-impermeable group said they would prefer to have the encasings removed, compared to 3 (2.6%) in the placebo group ($p < 0.001$). There was no difference in the numbers of families who stated that they would continue to use the encasings if it were of benefit to the child, (87.3% mite-impermeable vs 89.35% placebo $p = 0.68$)

Conclusions Mite-impermeable encasings can significantly reduce mite allergen levels in the bed. Despite some practical issues, most families are willing to use this intervention if it is of benefit to their child's asthma.

P94 EFFECT OF HYDROXYUREA ON NOCTURNAL AND AWAKE OXYGEN SATURATION IN CHILDREN WITH SICKLE CELL DISEASE

¹L van Geyzel, ²B Singh, ¹M Akthar, ¹G Ruiz, ²B Inusa, ¹D Rees, ¹A Gupta. ¹King's College Hospital, London, UK; ²Evelina London Children's Hospital, London, UK

10.1136/thoraxjnl-2015-207770.231

Introduction Sickle cell disease (SCD) causes lifelong morbidity and reduced life expectancy. Resting hypoxaemia and intermittent nocturnal oxygen desaturation are often seen in children with SCD, which may contribute to morbidity associated with vaso-occlusive episodes. Treatment with hydroxyurea reduces the frequency and severity of vaso-occlusive episodes¹ but the impact of hydroxyurea on oxygen saturation and sleep apnoea is unknown.

Objective To look for any difference in baseline oxygen saturation asleep and awake and the frequency of intermittent nocturnal desaturation after starting hydroxyurea in children with SCD.

Methods A retrospective review of children who were commenced on hydroxyurea between March 2006 and July 2014 attending two UK sickle-respiratory clinics. Data was collected from overnight sleep studies and averaged pulse oximeter spot check recordings in clinic notes when awake from a) 6 months before starting hydroxyurea and b) up to 2 years after. Lung function and haemoglobin changes were also noted over the same time periods.

Results Forty six children (25 male) with a median age of 10 years (range 5–19 years) were started on hydroxyurea. Haemoglobin and HbF rose significantly on hydroxyurea as expected (Table 1). After starting hydroxyurea the average overnight oxygen saturation increased from median of 93.5% to 95.2% ($p = 0.01$) and the median daytime spot oxygen saturation rose from 93.5% to 96.3% ($p = 0.001$). There was no significant change in the median intermittent nocturnal 3% oxygen desaturation index (ODI), nocturnal PCO₂ or spirometry.

Abstract P94 Table 1 Change in haematological and respiratory indices with hydroxyurea

	Before Median (Interquartile range)	On Hydroxyurea Median (Interquartile range)	*P value
Hb (g/L)	76 (69.5–86.5)	83 (72.7–87.7)	0.04
HbF (%)	6.1 (3.7–12.9)	8.8 (6–16)	<0.001
Average overnight SpO ₂ (%)	93.5 (88–97)	95.2 (93–98)	0.01
Nadir overnight SpO ₂ (%)	84 (77.4–89)	87 (83–91)	0.009
3% ODI overnight (events/hour)	3.0 (1.5–5.2)	2.8 (1.1–4.6)	0.08
Mean overnight PCO ₂ (kPa)	5.7 (4.7–6.2)	5.5 (5.2–6.0)	0.3
Spot daytime SpO ₂ (%)	93.5 (91–97)	96.3 (94–98)	0.001
% FEV ₁	70 (61.5–83.5)	73 (68–88)	0.6

*P values based on Wilcoxon matched-pairs signed rank test.

Conclusion In children with SCD, the use of hydroxyurea was associated with a significant increase in awake and nocturnal baseline oxygen saturation, but no change in intermittent nocturnal desaturation indices or lung function. This preliminary data suggests that improving oxygen saturation may be an important outcome of hydroxyurea therapy with potential benefits in reducing not only vaso-occlusive crises but future respiratory morbidities. This hypothesis would need to be tested by a prospective multicenter trial.

REFERENCE

- Charache S, Terrin ML, Moore RD, *et al.* Effect of hydroxyurea on the frequency of painful crises in sickle cell anemia. *Investigators of the Multicenter Study of Hydroxyurea in Sickle Cell Anemia. N Engl J Med* 1995;**332**:1317–1322

P95 GROWTH AND NUTRITION IN ATAXIA TELANGIECTASIA

¹El Stewart, ²A Tooke, ²S Pasalodos, ²M Suri, ³A Bush, ²J Bhatt. ¹University of Nottingham, Nottingham, UK; ²Nottingham University Hospitals NHS Trust, Nottingham, UK; ³Royal Brompton and Harefield NHS Foundation Trust, London, UK

10.1136/thoraxjnl-2015-207770.232

Background Ataxia telangiectasia (A-T) is a rare multisystem disease with high early mortality from lung disease and cancer. Nutritional failure adversely impacts outcomes in many respiratory diseases. Several factors influence nutrition in children with A-T including catabolism during recurrent infections and inadequate oral intake (fatigue, difficulties with chewing or swallowing, poor appetite, and nausea due to medications). We hypothesised that children with A-T have progressive growth failure.

Methods Data was collected prospectively on weight, height and body mass index (BMI) at the national paediatric A-T specialist clinic in Nottingham. Adequacy and safety of oral intake was assessed. Nutritional advice was given at each multidisciplinary review.

Results 92 children (46 girls) (33 once, 37 twice, 20 thrice, 1 child four times and 1 child 5 times) had 176 measurements since 2009. Median (range) age was 9.2 (1.5 to 18.4) years. Weight, height and BMI Z-scores were respectively -0.84 (-8.34 to 3.58), -0.98 (-5.85 to 3.66) and -0.24 (-4.45 to 2.75). Weight, height and BMI Z-scores inexorably declined over time. 10

children had a gastrostomy, with longitudinal data available for 8. 87.5% of these children improved their BMI Z-score with time. 18.5% (17) children were considered wasted (BMI Z-score ≤ -2). All of these children were above 8 years old. Longitudinal data was available for 14 wasted children. 6 of these children had a gastrostomy inserted and 5 then improved their Z-score. Of the remaining 8 children without gastrostomy, 7 (87.5%) continued to decrease their BMI over time despite dietary advice to fortify food or add in supplements.

Conclusions There is a remorseless decline in growth over time. There is an urgent need for new strategies, including an understanding of why growth falters. Undernutrition adversely affects acute and chronic lung health. Outcomes for late gastrostomy insertion in AT are poor (*Lefton Greif OJRD* 2011;210). We suggest early proactive consideration of gastrostomy from age 8 years upwards in order to prevent respiratory deterioration.

Weight Z-score vs age

Abstract P95 Figure 1

P96 INTERSTITIAL LUNG DISEASE CAUSED BY STING-ASSOCIATED VASCULOPATHY WITH ONSET IN INFANCY (SAVI)

¹EJ Pellowe, ²SLN Clarke, ¹TN Hilliard, ²AV Ramanan. ¹Department of Paediatric Respiratory Medicine, Bristol Royal Hospital for Children, Bristol, UK; ²Department of Paediatric Rheumatology, Bristol Royal Hospital for Children, Bristol, UK

10.1136/thoraxjnl-2015-207770.233

Introduction An increasing number of monogenic auto-inflammatory conditions known as primary type I interferonopathies are being recognised. We present a case of the newly described condition, stimulator of interferon gene (STING) associated vasculopathy with onset in infancy (SAVI), with significant respiratory involvement.

Presentation A male infant, born to healthy non-consanguineous parents, presented to his local hospital with tachypnoea, intermittent fever and failure to thrive from 5 weeks of age. Over the following months, episodes of increased respiratory effort, cough and fever were attributed to infection. He subsequently developed a papular rash in discrete clusters over his back. At presentation to our centre at 7 months of age, he was tachypnoeic but not hypoxaemic. A chest radiograph showed extensive airspace shadowing with chest computed tomography demonstrating interstitial changes. Bronchoalveolar lavage was negative for infection. Laboratory tests revealed microcytic anaemia, raised inflammatory markers, positive anti-nuclear antibody, raised IgA and IgG and abnormal lymphocyte proliferation. A lung biopsy showed a mixed pattern of inflammation, with type 2 pneumocyte hyperplasia and endothelial tuboreticular