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

1 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

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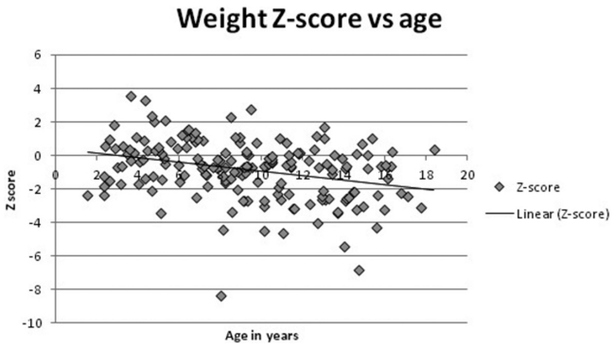
Background Ataxia telangiectasia (A-T) is a rare multisystem disease with high early mortality from lung disease and cancer. Nutritional failure adversely impacts outcomes in many respiratory diseases. Several factors influence nutrition in children with A-T including catabolism during recurrent infections 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.



Abstract P95 Figure 1

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

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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