

#### S44 ORTHODEOXIA, AND POSTURAL ORTHOSTATIC TACHYCARDIA, IN 165 CONSECUTIVE, UNSELECTED PATIENTS WITH PULMONARY ARTERIOVENOUS MALFORMATIONS

V Santhirapala, JT Springett, H Wolfenden, HC Tighe, JE Jackson, CL Shovlin; *Imperial College London, London, UK*

10.1136/thoraxjnl-2013-204457.51

**Background** Patients with pulmonary arteriovenous malformations are often quoted as displaying orthodeoxia, that is, a fall in oxygen saturation on standing. It is unclear how common this phenomenon is, and how patients would compensate for any acute fall in SaO<sub>2</sub>.

**Methods** Postural changes in oxygen saturation and pulse were examined in a series of 165 prospectively-recruited patients with radiologically-proven PAVMs. Self-reported exercise at presentation was graded using a modified MRC dyspnoea scale. SaO<sub>2</sub> and pulse measurements were made in both erect and supine postures on 1–8 separate occasions through presentation to post-embolisation follow up. These 522 sets of postural measurements displayed very high within-patient reproducibility.

**Results** Age ranged from 17–87 (median 49) ys, 62 (37.6%) were male, and for 159 (96%), PAVMs were attributable to HHT. 18.9% were obese with a body mass index (BMI) >30, including one with morbid obesity (BMI>40). At presentation, the SaO<sub>2</sub> fell by at least 2% on standing in 51(33%) patients, compared to the equivalent average supine reading. A smaller fall of 1–2% was present in a further 28 (17%) of patients. Patients with higher BMI had significantly higher supine SaO<sub>2</sub> for their erect SaO<sub>2</sub>, and significantly lesser falls in SaO<sub>2</sub> on standing. A postural tachycardia consistently exceeded the increment required to sustain oxygen delivery at rest, across all SaO<sub>2</sub>. 12/163 (7.4%) of individuals met the pulse definition for postural orthostatic tachycardia syndrome with an increase on standing of more than 30 beats per minute. Once adjusted for baseline supine pulse and age, the erect pulse was lower in patients with higher BMI (adjusted regression coefficient 0.23 (95% confidence interval 0.026, 0.48), p = 0.011). Using the pre-specified 5 group grading system, exercise tolerance was worse in patients with lesser postural tachycardias (regression coefficient -2.19 (-3.74, -0.65) p = 0.006).

**Conclusions** Orthodeoxia and postural tachycardia are common in PAVM patients. More pronounced postural tachycardias were associated with improved exercise tolerance. Further studies will be required to assess if this is because it is a surrogate of lower BMI, with obesity effectively limiting a further fall in SaO<sub>2</sub> on standing because of obesity-related lower supine SaO<sub>2</sub>.

#### S45 SPECIFIC VENTILATION INEQUALITY AND DEAD SPACE COMPONENTS OF LUNG CLEARANCE INDEX IN PATIENTS WITH ASTHMA AND CYSTIC FIBROSIS

<sup>1</sup>S Gonen, <sup>1</sup>S Natarajan, <sup>1</sup>A Singapuri, <sup>1</sup>Ce Brightling, <sup>2</sup>P Gustafsson, <sup>3</sup>A Horsley, <sup>1</sup>S Siddiqui; <sup>1</sup>Institute for Lung Health, University of Leicester, Leicester, UK; <sup>2</sup>Department of Paediatrics, Central Hospital, Skövde, Sweden; <sup>3</sup>Manchester Adult Cystic Fibrosis Centre, Manchester, UK

10.1136/thoraxjnl-2013-204457.52

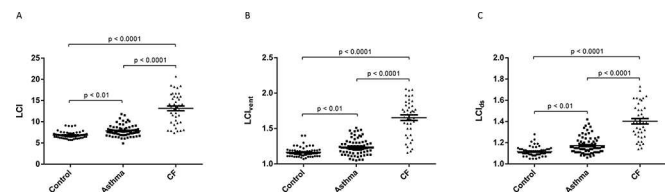
**Background** Lung clearance index (LCI) is a widely reported marker of gas mixing inefficiency within the airways that is derived using the multiple breath inert gas washout (MBW) technique. We developed two novel parameters, LCI<sub>vent</sub> and LCI<sub>ds</sub>,

to reflect the components of increased LCI due to (i) unequal convective ventilation between relatively large lung units, and (ii) increased respiratory dead space, respectively. We hypothesised that these parameters would be repeatable, would effectively discriminate between healthy controls and patients with asthma and cystic fibrosis (CF), and would distinguish between different sub-phenotypes of these diseases.

**Methods** Washout data from sixty-six healthy control subjects, seventy-four patients with asthma, and forty-one patients with CF were fitted to a two-compartment model of gas mixing, and the parameters LCI<sub>vent</sub> and LCI<sub>ds</sub> were calculated.

**Results** LCI<sub>vent</sub> and LCI<sub>ds</sub> were markedly elevated in patients with CF, and mildly elevated in patients with asthma, compared to controls, as illustrated in Figure 1. LCI<sub>vent</sub> and LCI<sub>ds</sub> were weakly correlated in controls (R = 0.36, p < 0.01), moderately correlated in patients with asthma (R = 0.51, p < 0.0001), and strongly correlated in patients with CF (R = 0.89, p < 0.0001). LCI<sub>ds</sub> was significantly raised in CF patients with chronic *P. aeruginosa* colonisation compared to those without chronic colonisation (1.49 vs 1.34, p = 0.004). LCI, LCI<sub>vent</sub> and LCI<sub>ds</sub> were significantly raised in CF patients with a severe genotype compared to those with a mild genotype. No significant differences were observed between any of the asthma sub-phenotypes (severe vs non-severe, poorly-controlled vs not poorly controlled, exacerbator vs non-exacerbator, and eosinophilic vs non-eosinophilic) with respect to any MBW parameter. The intraclass correlation coefficients of LCI<sub>vent</sub> and LCI<sub>ds</sub> exceeded 0.85 in the asthma and CF groups, and 0.60 in controls.

**Conclusion** The novel parameters LCI<sub>vent</sub> and LCI<sub>ds</sub> are repeatable and effectively discriminate between sub-phenotypes of CF, although their utility in asthma is currently unproven. Further studies are required to determine their utility in other airway diseases such as chronic obstructive pulmonary disease, to investigate their role as outcome measures in clinical trials, and to delineate their structural correlates.



**Abstract S45 Figure1 Multiple breath washout parameters across groups. Error bars denote mean +/- standard error of the mean. Groups compared using one-way analysis of variance with Bonferroni correction**

## Clinical studies in pulmonary vascular disease

#### S46 OUTCOME AFTER PULMONARY ENDARTERECTOMY (PEA): LONG TERM FOLLOW-UP OF THE UK NATIONAL COHORT

J Cannon, K Page, M Roots, A Ponnabernam, C Tracy, D Taboada Buasso, K Sheares, C Ng, J Dunning, S Tsui, J Pepke-Zaba, D Jenkins; *Papworth Hospital, Cambridge, UK*

10.1136/thoraxjnl-2013-204457.53

**Introduction** Chronic thromboembolic pulmonary hypertension (CTEPH) is a life threatening condition that historically has a poor outcome with supportive medical treatment. Pulmonary endarterectomy (PEA) is the treatment of choice and offers the