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

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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 SaO2.

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. SaO2 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) years, 62 (37%) 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 SaO2 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 SaO2 for their erect SaO2, and significantly less SaO2 on standing. A postural tachycardia consistently exceeded the increment required to sustain oxygen delivery at rest, across all SaO2 groups. Error bars denote mean +/- standard error of the mean. Groups compared using one-way analysis of variance with Bonferroni correction.

Conclusions Orthodeoxia and postural tachycardia are common in PAVM patients. More pronounced postural tachycardias were associated with improved exercise tolerance. 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 Figure 1 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

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

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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...
only chance of cure. Data on the long term survival after PEA are limited.

Method All patients who have undergone a PEA for CTEPH at Papworth hospital were included between January 1997 and November 2012. Patients who had a re-do operation were excluded. Pre- and post-operative data on haemodynamics, exercise capacity, functional class and targeted PAH therapies taken were obtained from our PH database and from other UK PH centres. The long-term survival of patients who returned for follow-up at 3 months post PEA was determined using the NHS spine summary care record tracking system. Overseas patients were censored when last seen.

Results 880 patients underwent PEA over the 15 year period. The mean age was 57 (range 15–84) and 53% were male. The majority (89%) were in WHO functional class 3 or 4 prior to surgery with an average mean pulmonary artery pressure (mPAP) of 47 mmHg and PVR of 795 dyn·sec/cm². 65% of patients were taking at least 1 targeted therapy as a “bridge to surgery”. Post surgery the majority of patients (86%) were in WHO functional class 1 or 2 at the 12 month follow-up with only 17% taking targeted therapy. There was a reduction in the average mPAP to 27 mmHg and PVR to 308 dynes by 12 months. The 10 year conditional survival post PEA of the first 314 patients from the cohort (Freed et al. J Thorac Cardiovasc Surg, 2011;141:383–7) was 74%.

Conclusion The conditional survival of a subset of the cohort at 10 years was 74%. There was a significant functional and haemodynamic improvement in the majority of patients at 12 months post surgery. Only 17% of patients at 12 months post surgery were being treated with targeted therapy.

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Effect of Macitentan on Haemodynamics in Patients with Pulmonary Arterial Hypertension: Results from the Long-Term, Randomised, Placebo-Controlled SERAPHIN Trial

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Introduction Venticulo-arterial (VA) coupling (Ees/Ea) in the right heart is defined by RV end-systolic elastance (Ees) and pulmonary arterial effective elastance (Ea) with Ees/Ea representing the mechanical efficiency of forward flow from the RV. Ees/Ea may influence exercise capacity in pulmonary hypertension (PH) because patients exhibit cardiac limitation at peak oxygen uptake (peak VO2) and suffer impaired exercise cardiac output adaptation. We hypothesised that Ees/Ea in the RV represents a