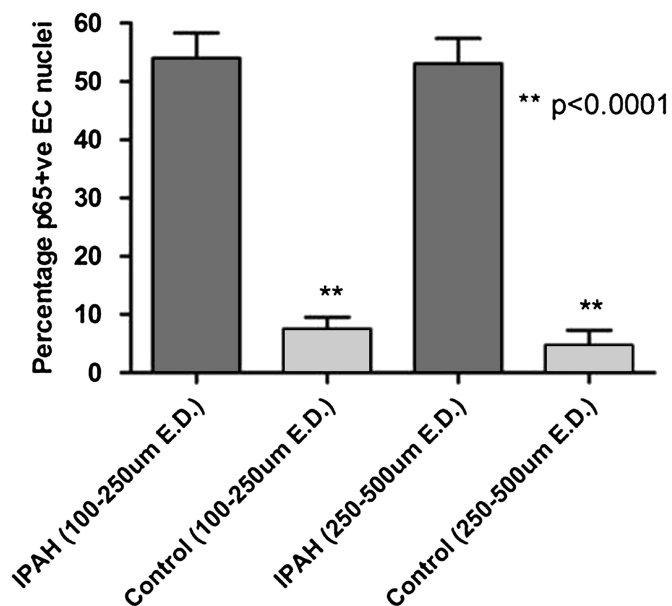


slide performed by a researcher blinded to the groups. Data are expressed as median and IQR, and the groups were compared by the Kruskal–Wallis or Mann–Whitney U test.

**Results** (1) PAH specimens showed co-localisation of p65 within CD68+ macrophages in 75.4 (64.8–84.6)% of samples. Airway epithelium, neutrophils and lymphocytes were also positive for p65. (2) Pulmonary arterial medial thickness was increased in PAH compared to controls, at 33.7 (18.8–67.9)% in vessels 100–250 mm external diameter (E.D.) and 27.2 (14.8–44.2)% in vessels 250–500 mm ED, vs 17.7 (11.2–30.3)% and 14.9 (11.8–17.8)% in controls ( $p < 0.0001$  between groups). (3) Nuclear p65 was present in pulmonary artery endothelial cells (EC) but not other vascular cells including pulmonary artery smooth muscle cells in PAH: 53.9 (0–100)% of vessels 100–250 mm E.D. and 53.1 (0–100)% of those 250–500 mm E.D. scored EC p65 positivity in PAH compared to 7.5 (0–25.0)% in 100–250 mm ED and 4.7 (0–21.1)% in 250–500 mm ED in controls ( $p < 0.0001$  between groups) (Abstract P29 Figure 1).



Abstract P29 Figure 1 Percentage of positive endothelial cell (EC) nuclear p65 immunostaining subdivided according to pulmonary artery external diameter (E.D.).

**Conclusion** NF- $\kappa$ B activation is present in macrophages and pulmonary arterial endothelial cells in pulmonary arteries of 100–500 mm ED in patients with PAH.

### P30 THE CHANGING FACE OF PULMONARY HYPERTENSION: THE ROLE OF HEART AND LUNG DISEASE

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**Introduction** The advent of disease-targeted therapy for pulmonary arterial hypertension (PAH) has led to an increased awareness of this condition within the general cardiology and respiratory communities. More patients are being referred to specialist centres for diagnostic assessment, however there is concern that many will have pulmonary hypertension (PH) due to underlying lung disease or left heart disease.

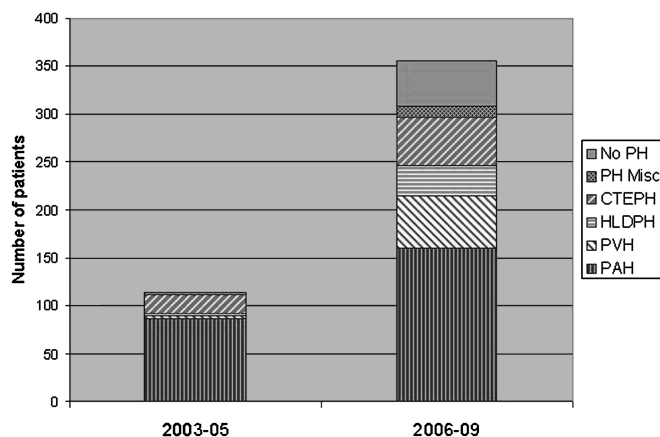
**Aim** The aim of the study was to review the outcome of diagnostic admissions to the Scottish Pulmonary Vascular Unit (SPVU) before

(2003–2005) and after (2006–2009) the introduction of disease-targeted PAH therapy.

**Methods** 470 new patients with suspected PH were admitted between January 2003 and December 2009 for diagnostic assessment including right heart catheterisation (RHC). Demographic and haemodynamic data from these patients were retrospectively reviewed. Following RHC patients were diagnosed with:

- Group 1. PAH;
- Group 2. Pulmonary venous hypertension (PVH) due to left heart disease;
- Group 3. PH due to hypoxic lung disease (HLDPH);
- Group 4. Chronic thromboembolic PH (CTEPH);
- Group 5. PH due to unclear multifactorial mechanisms (PH Misc);
- No PH.

**Results** In 2003–2005, 114 patients underwent diagnostic assessment, and 112 had PH. Of these 77.7% had PAH and 18.8% had CTEPH. Only 2.7% of patients had PVH and 0.8% had HLDPH. In 2006–2009, 356 patients underwent diagnostic assessment, and 308 had PH. Of these only 51.9% had PAH and 15.9% had CTEPH. However, now 17.9% of patients had PVH, and 10.4% had HLDPH. 48 patients had No PH at the time of RHC (Abstract P30 Figure 1).



Abstract P30 Figure 1 Outcome of diagnostic assessment in the pre and post-treatment era.

**Conclusion** Prior to admission all referrals are screened by an SPVU consultant, but despite this there are a significant number of patients proceeding to RHC who will have PH due to left heart or lung disease. This highlights the importance of fully assessing all patients with suspected PH, as per ESC/ERS 2009 guidelines, before instituting expensive and potentially dangerous PAH therapy. We need to improve our non-invasive screening methods so that fewer patients proceed to RHC who do not have PAH.

### P31 DISEASE TARGETED THERAPIES AND EFFECT ON SURVIVAL IN IDIOPATHIC, HERITABLE AND ANOREXIGEN-ASSOCIATED PULMONARY ARTERIAL HYPERTENSION (PAH)

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**Introduction** The median survival of idiopathic PAH was 2.8 years before the availability of modern therapies. Substantial progress has been made over the past 20 years. Currently available disease targeted therapies have been shown in clinical trials to improve symptoms, exercise capacity and survival. Combination therapies

should also be considered in patients with inadequate clinical response to monotherapy (ESC/ERS 2009). We aim to assess the use of modern advance treatments in ‘real-life’ patients and their impact on survival.

**Methods** A retrospective review of all incident cases of idiopathic, heritable and anorexigen-associated PAH diagnosed between 1st January 2001 and 31st December 2009 in our unit. Baseline and follow-up data were collected from case notes and local pulmonary hypertension database.

**Results** 94 incident cases were diagnosed (62% female) with median age of 63 years. 36% (n=34) of patients were on combination therapy. All except one patient were started on monotherapy, with second agent added at later stage. The median time from initiation of monotherapy to introduction of the second agent was 244 days. 25% had prostanoids; 51% endothelin receptor antagonists and 81% phosphodiesterase type-5 inhibitor at some point of their treatment. The median survival is 5.63 years. We performed further analysis on the 28 patients who have died by censor date. Only seven patients were on prostanoids and eight patients on combination therapy. Possible explanations for low proportions of dead patients on prostanoids and/or combination therapy are outlined in Abstract P31 Table 1.

Abstract P31 Table 1 Possible explanations why prostanoids or combination therapies were not started

	Number of dead patients (total=28)
Already on prostanoids	7
Died from unrelated causes	6
Prostanoids considered but felt inappropriate, for palliation only	5
No documented explanation	3
Treatment optimised less than 4 months before death	2
Clinically improving at last clinic review before death	2
Sudden unexpected death	1
Lost to follow up	1
Poor compliance, frequent non-attendance of clinics	1

**Conclusion** Our results demonstrate improved survival compared to pre-advance therapy era. Apart from proven efficacy of various PAH specific treatment, several other factors may also influence the treatment decision for individual patient.

**P32 DELIVERING PULMONARY HYPERTENSION SERVICES—5 YEAR EXPERIENCE FROM A SATELLITE CENTRE**

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**Introduction** There are currently no National Commissioning Group (NCG) designated Pulmonary Hypertension (PH) centres in the South West. As a result patients can travel up to 300 miles to access specialist care. In 2005, a satellite service was developed at the Royal United Hospital (RUH), Bath in conjunction with the Royal Free, London. Initially the service simply provided a follow up service for patients known to the Royal Free. Since 2007 the service has fully investigated and managed new referrals locally, with supervision from the Royal Free. Patients have typically only needed to travel to an NCG centre for initiation of parenteral prostanoid therapy or for pulmonary endarterectomy (PEA).

**Aim** This abstract describes the first 5 years of activity by the RUH satellite PH service.

**Results** As of 1st July 2010, 147 new referrals have been received, increasing steadily from 7/year in 2005–2006 to 57/year in 2009–2010. Referrals originated from 7 specialities across 12 PCTs, with 48.6% from outside the immediate catchment area. 43 patients were diagnosed with Pulmonary Arterial Hypertension (PAH) (including 34 with Connective Tissue Disease-related PAH), 21 with Chronic Thromboembolic Pulmonary Hypertension (CTEPH) (including 15 referred for PEA surgery), 13 with PH associated with lung disease, 24 with PH associated with left heart disease. PH was excluded in 45 cases. 62 have received PH-specific medication; 38 monotherapy and 24 combination therapy (including 7 intravenous, 1 subcutaneous and 2 nebulised prostanoid therapy). Survival data for the 38 treated WHO Group 1 PAH patients compares favourably with published data for NCG designated centres; 89.5% at 1 year (95% CI 74.2% to 96.6%), 82.3% at 2 years (95% CI 65.9% to 92.2%) and 77.2% at 3 years (95% CI 60.3% to 88.6%). 489 journeys were made to the RUH and only 60 to NCG centres, saving approximately 56 000 patient miles. A recent patient survey indicated high levels of satisfaction, with 94.4% stating they would rather continue follow-up locally.

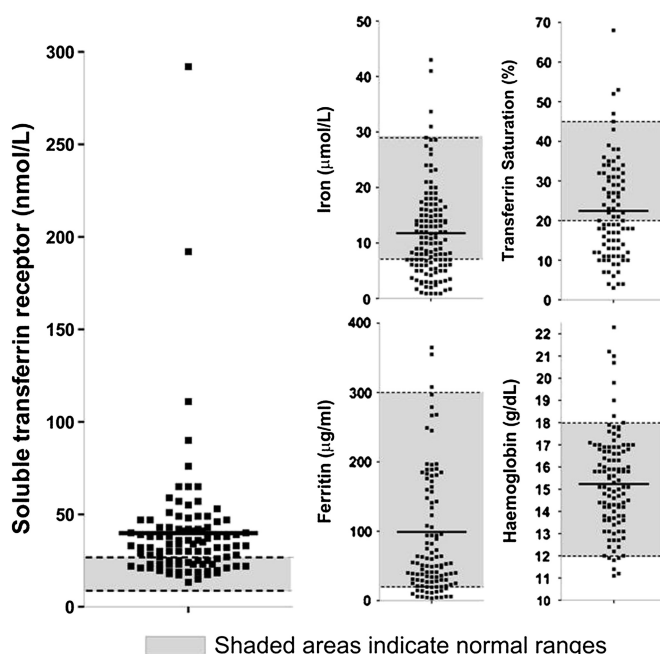
**Discussion** These results suggest that a comprehensive PH specialist service can be delivered locally in a way that is more convenient for patients whilst still maintaining national standards of care.

**P33 IRON DEFICIENCY INDEPENDENTLY PREDICTS SURVIVAL IN IDIOPATHIC PULMONARY ARTERIAL HYPERTENSION**

doi:10.1136/thx.2010.150961.33

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**Objectives** Characterise the iron status of patients with idiopathic pulmonary arterial hypertension (IPAH) and its clinical importance.



Abstract P33 Figure 1