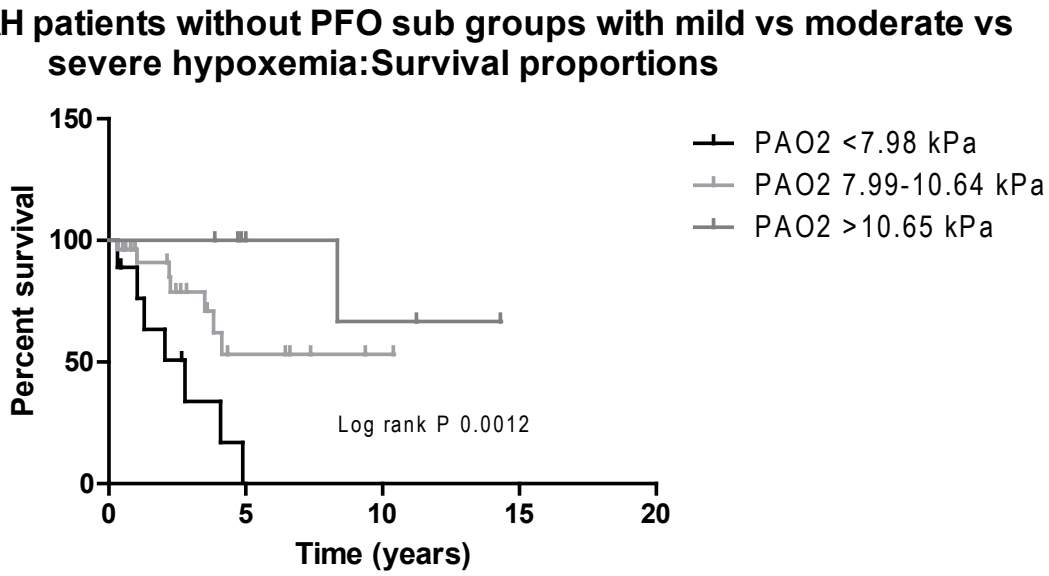

Abstract P138 Figure 1 Survival of IPAH patients without PFO based on severity of hypoxemia. Log rank comparisons

P139 SURVIVAL OF IDIOPATHIC PULMONARY ARTERIAL HYPERTENSION BMPR2 MUTATION CARRIERS VS BMPR2 NON CARRIERS

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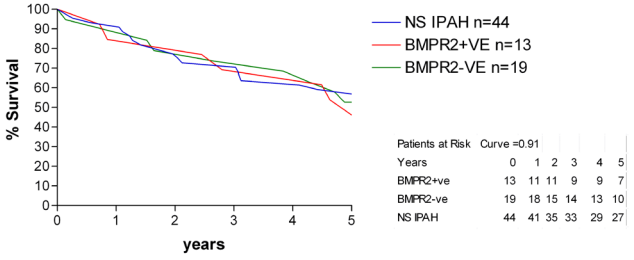
Introduction Idiopathic pulmonary arterial hypertension (IPAH) is a devastating condition charactericed by the narrowing and obliteration of small pulmonary arteries, leading to elevated pulmonary arterial pressure and ultimately to right heart failure and death. Even with treatment, IPAH survival is poor with 3 year survival reported to be 63% on epoprostenol (Mclaughlin, Circulation 2002). Pulmonary arterial hypertension (PAH) can be heritable, with mutations found in the bone morphogenetic protein type 2 receptor (BMPR2) gene found in > 70% of familial cases, (Lane, Nat Genet 2000). There are previous conflicting reports of worse survival in heritable PAH, but this has not previously been determined in the UK patient population.

Methods This study examined the retrospective database of 76 IPAH cases diagnosed between 2001–2006. 32 patients were screened for BMPR2 mutation. 13 were BMPR2+ve. Non-screened (NS) IPAH were included in the survival analysis (n=44). Survival time was taken from the time of initial diagnostic right heart catherisation. Demographics, treatment, 6MWT, time to transplant and World Health Organization (WHO) classification were compared. Physicians were blinded to the BMPR2 status at time of treatment.

Results All three cohorts presented at baseline with no significant difference in functional class, 6MWT, hemodynamics except age, cardiac index (CI), right atrial pressure (RAP) and treatment modalities. Patients with BMPR2 mutation were significantly younger. Of the BMPR+ve group, 6 patients were transplanted compared to zero in the BMPR-ve group. The NS IPAH cohort had 2 patients transplanted and fewer patients were treated with prostanoids. Time to transplant was shorter in the BMPR2 mutation carriers, 2.65 years, vs. 3.1years in the NS IPAH group.

Conclusion Survival for the first 5 years from diagnosis was similar in IPAH and heritable PAH. The BMPR2 +ve patients presented younger with severe disease and were treated more aggressively with 6 patients undergoing transplant in the 5 year period. BMPR2 mutation frequency was 41% from our cohort of 33 which is higher than previously reports of 20–30% (Morrell, Proc Am Thorac Soc, 2006). Survival for IPAH continues to be poor even with improvements in treatment. Patients with BMPR2 mutations present with severe disease earlier and time to lung transplant is shorter.



Abstract P139 Figure 1

P140 HIGH-DOSE OESTROGENS COMMENCED FOR GENDER REASSIGNMENT SURGERY MAY PREDISPOSE TO THE DEVELOPMENT OF CHRONIC THROMBOEMBOLIC PULMONARY VASCULAR DISEASE

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Background Chronic thromboembolic pulmonary hypertension (CTEPH) often occurs as a sequela of venous thromboembolism (VTE) however, there appears to be marked differences in laboratory risk-profiles between CTEPH and VTE. Clinically, exogenous oestrogens (EO) are known to increase the risk of acute DVT in a dose-dependent manner[2] but a pathological link with CTEPH is less clear. We describe a case-series of three patients with CTEPH and one with chronic thromboembolic disease without evidence of pulmonary hypertension diagnosed following administration of high-dose EO for gender re-assignment surgery (GRAS). This constitutes a long-term component of treatment, always commenced in males preoperatively.[3]