

**Characteristics of PAH associated with pretricuspid shunts in the registry of the French PAH network**

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Background: Pulmonary arterial hypertension (PAH) associated with pretricuspid shunts is commonly delayed until adulthood but PAH may reveal the underlying unknown congenital heart defect (CHD).

Objective: To review and analyze patients with PAH and pretricuspid shunts enrolled in prospective French PAH registry.

Patients and results: CHD-PAH accounted for 9.8% of PAH in this registry. 145 patients (101 female-44 male) had a pretricuspid shunt (80% of CHD-PAH diagnosed after 30 years of age). Thirty had their shunt previously closed. Mean age at first symptom and at diagnosis of PAH were respectively 37.2 years and 40 years, males being much older at time of diagnosis 48.0 vs. 34.3 for females. In the majority of cases, the diagnosis of the shunt was made concomitantly with PAH diagnosis but the diagnosis of the CHD was posteriorly made in 8 cases. Functional class at entry was I (2%), II (33%), III (60%), and IV (5%). Resting oxygen saturation was below 92% in 7%. Mean 6 minutes walk distance at entry was  $358 \pm 109$  m. Total lung capacity (TLC) was below 80 % of the predicted value in 38%, and the median value of the ratio of the forced expiratory volume in one second to the forced vital capacity (FEV1/FVC) was below 70% of predicted value in 40%. Mean pulmonary artery pressure at RHC (n=136) was  $55 \pm 16$  mmHg, cardiac index  $2.9 \pm 1.1$  L/min/m<sup>2</sup> and PVR were  $1565 \pm 978$  dynes.s.cm<sup>-5</sup>. At entry, 91% received a monotherapy, 8% a combined oral therapy, and 1% a tritherapy including prostacyclin. At last follow-up (mean  $3.7 \pm 2.2$  years), functional class was improved by + 1 in 71% of class IV, 28% of class III, and 13% of class II; 6 minutes walk distance increased by +35 m; and control RHC (n=66) showed increase in cardiac index (p<0.05) and decrease in PVR (p<0.05). Survival at 6 years was 76%. No difference could be found between open and closed shunts in this series.

Conclusion: PAH at diagnosis was only moderately less severe than in idiopathic PAH and outcome was not as favorable than expected in CHD-PAH.