

**Clinical characteristics of pulmonary arterial hypertension associated with congenital heart disease: baseline results from the Turkish Congenital Heart Disease-associated Pulmonary Arterial Hypertension (THALES) Registry**

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**Introduction:** Turkish Congenital Heart Disease-associated Pulmonary Arterial Hypertension Study (THALES) is a national multicenter prospective observational registry which aims to provide information regarding demographic and clinical characteristics, laboratory data, prognosis and treatment patterns in pulmonary arterial hypertension associated with congenital heart disease (APAH-CHD). The registry is designed to record data at baseline and follow-up at 1-year intervals thereafter; the baseline clinical characteristics of patients enrolled to date are presented in this study.

**Methods:** Pediatric and adult patients (>3 months of age) with APAH-CHD (mean pulmonary artery pressure [mPAP]  $\geq 25$  mmHg, pulmonary capillary wedge pressure  $\leq 15$  mmHg, and pulmonary vascular resistance index [PVRI]  $> 3$  Wood units.m<sup>2</sup>), were enrolled at 61 centers over the country.

**Results:** Between May 2009 and October 2011, 1034 patients (female:male 1.38:1) aged 3 months-79 years (mean 16.91 $\pm$ 17.91, median 11 years) were enrolled. 249 patients (24.1%) were <2, 416 (40.2%) were 2-18, and 369 (35.7%) were >18 years of age. Mean mPAP was 54,67 $\pm$ 22,15mmHg, PVRI 10,23 $\pm$ 10,84 WU.m<sup>2</sup>. 55.4% of patients had Eisenmenger's syndrome due to unrepaired or partially repaired CHD with significant residual defects (Group I), 34.5% had PAH associated with systemic-to-pulmonary shunts due to unrepaired or partially repaired CHD with significant residual defects (Group II), 1.2% had PAH with small defects (Group III), 7.3% had PAH after repair of CHD in the absence of significant residual defects (Group IV), and 1.6% had PAH associated with unrepaired or partially repaired complex cyanotic CHD and increased pulmonary blood flow (Group V). Dyspnoea was the most frequent symptom (89.8%), followed by chest pain (10.4%), and haemoptysis (6.9%). Syncope/pre-syncope was reported in 5.9%, the highest percentage recorded in group IV (16.2%). Functional class was predominantly III in Group I (48.2%), and II in groups II-V (75%, 83.3%, 57.5%, 66.7%, respectively). 40.1% received targeted PAH therapies (monotherapy in 80.7%, combination in 19.3%).

**Conclusions:** This study seeks to define characteristics of APAH-CHD according to age groups and type of underlying CHD. In due course, we hope to gain additional information regarding the course, prognosis, impact of surgical/interventional procedures on outcome, planning and timing of targeted therapies in this group of patients with PAH.