

Clinical, laboratory , echocardiographic and hemodynamic characteristics in relation to survival in pulmonary arterial hypertension associated with congenital heart disease

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Pulmonary arterial hypertension (PAH) associated with congenital heart disease (APAHCCHD) is classified as a subgroup of PAH that includes four different forms sharing a similar clinical and pathological findings. However, attention have been focused on the Eisenmenger syndrome (ES), and predictors of the survival in other patients (pts) with APAHCCHD remain to be determined.

In this study,we investigated clinical, biochemical, neurohumoral ,echocardiographic and hemodynamic predictors of the survival in 134 pts with APAHCCHD (F72 M 62, age 40.7±17.5 years) out of the 341pts included into the our single-center, prospective study.Subgroups of APAHCCHD were as follows;1.ES (n=71), 2.PAH associated with prevalent systemic-to-pulmonary shunts (n=37), and 4. PAH after defect correction (n=26). Baseline six-minute walk distance (6MWD) was 301.5±127.8 m, and pulmonary arterial systolic and mean pressures (PASP, PAMP, mm Hg) were 99.6±32.9 and 64.7±22.85, and transpulmonary and diastolic pressure gradients (TPG,DPG,mm Hg) were 51.8±22.9 and 29.6±20.3, respectively. Pulmonary and systemic vascular resistance (PVR,SVR,WU) and PVR/SVR ratio were 10.6±9.3, 21.8±9.6 and 0.52±0.37, respectively. Targeted PAH treatments were noted in 100 (75%) pts. Mean follow-up was 30.9 ± 28 (0,5-122,6) months. The APAHCCHD compared with idiopathic PAH(IPAH) and PAH-associated with connective tissue disease (APAHCCTD) was associated with a better 8-year survival (83 vs 54,4 and 61%, p<0,05), and APAHCCHD subgroups showed comparable survival (p>0,05). The age, sex,functional class, haemoglobin, oxyhaemoglobin saturation %, C-reactive protein, uric acid and brain natriuretic peptide were not related with survival(p>0,05). Neither tertile definition (<190,190-330,>331), nor pre-defined cut-off values of 6MWD (m) (<165, 165-440,>440) related with APAHCCHD survival(p>0,05). Similarly, tertiles of TPG,DPG, PVR and PVR/SVR ratio showed comparable survival (p>0,05). However, pericardial effusion(p=0,0001) and the highest tertile of right atrial pressure (RAP,mm Hg) (> 11 vs 9-11 and < 9, p<0.01) were found to discriminate the hihg-risk pts.

In conclusion, APAHCCHD compared with other PAH subgroups is associated with a better survival. However, only pericardial effusion and RAP predict the worst clinical outcome in these pts