Prognostic Factors In Pulmonary Atresia With Ventricular Septal Defect

Hospital de Santa Cruz, Carnaxide, Portugal (1); Hospital das Caldas da Rainha, Caldas da Rainha, Portugal (2); Hospital da Universidade de Coimbra, Coimbra, Portugal (3)

Objectives: The main purpose of this work is to evaluate the impact of different anatomical and therapeutical factors in the prognosis of patients with pulmonary atresia with ventricular septal defect (PAtr + VSD).

Materials and Methods: Retrospective analysis of 84 patients with PAtr + VSD and balanced ventricles consecutively treated in our center. Demographic data, pulmonary artery (PA) anatomical features, including PA dimensions evaluated by McGoon Index (McGooni), presence of non-confluent PAs, surgical and percutaneous approach required and health condition on follow-up were recorded.

Results: 42 (50%) patients were male. Major aortic pulmonary collateral arteries (MAPCAs) were found in 48 (57.1%) patients, PA hypoplasia with McGooni <1.5 in 50 (59.5%) and non-confluent PAs in 20 (23.8%). There was a late referral to our center (beyond the neonatal period) of 20 patients (24%). Corrective surgery (Corr Surg) was performed in 35 patients (41.7%) between 0.9 and 23 years of age, and all had a McGooni ≥1.5. In 15 patients (17.9%), right ventricle to PA continuity was established without VSD closure as there was pulmonary hypoplasia. The initial McGooni predicted the possibility of corrective surgery with an odds-ratio of 12.1 (p=0.04); the best cut-off was a McGooni >1.3 (by ROC curve analysis). Final McGooni was 1.8 in the group who underwent corrective surgery and 1.4 in the group who was considered unsuitable for correction (p=0.03). Age at the time of first surgery was not different between the group who had Corr Surg and the remaining patients. Presence of non-confluent pulmonary arteries did not influence negatively the possibility of total correction (p NS). The actuarial survival of the whole group was 96.4% at one year of age; 92.8% at 2 years; 90.4% at 5 years and 88% at 10 years. At last follow up, 38 (55.1%) patients were in NYHA class I or II.

Conclusions: In patients with PAtr + VSD and balanced ventricles, the initial McGooni predicted the possibility of corrective surgery with a best cut off value of 1.3. Age at time of initial surgery and presence of non-confluent pulmonary arteries did not influence the possibility of corrective surgery.