

Prenatal diagnosis predicts respiratory failure in patients following absent pulmonary valve repair.

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Background: Absent pulmonary valve syndrome (AVPS) is a rare congenital heart disease. Mortality and morbidity after repair is mainly related to acute or chronic respiratory failure due to the dilated pulmonary arteries.

Objective: To describe outcome and risk factors for mortality and respiratory morbidity for AVPS.

Methods: Retrospective analysis of all consecutive patients undergoing surgical repair from 1995-2015 in a single center. Patients' and procedure-related variables were analyzed by means of cox analysis and logistic regression.

Results: 31 patients underwent surgery at a median weight of 4.5 kg (range: 2-63) and a median age of 4.5 months (range: 0.4-184). 58% (n=18/31) had a prenatal diagnosis of APVS and 26% (n=8/31) had Di George syndrome. 23% (n=7/31) had to be operated in the neonatal period. 39% (n=12/31) required preoperative mechanical ventilation. Mortality before hospital discharge was 13% (n=4/31). All deaths were due to respiratory failure. Lower weight at surgery, younger age, and larger left pulmonary artery were univariate risk factors for death. Prenatal diagnosis of APVS and type of right ventricular outflow tract (RVOT) repair, e.g. valved conduit, transannular patch or monocusp patch, did not impact mortality. 23% (n=7/31) needed more than 15 days of mechanical ventilation after surgery. Multivariate risk factors for mechanical ventilation > 15 days were prenatal diagnosis (p=0.0001), younger age (p=0.0001) and Di George syndrome (p=0.021). 1, and 10-year survival rate after repair was 87% (CI95%: ±12%), and 83% (CI95%: ±14%), respectively. 5-year survival without surgical RVOT reintervention was 60% (CI95%: ±20%). Mean follow-up was 6.3 years (range: 0.3-16.8). At last visit, all survivors were in good clinical condition without need for chronic cardiac medication in 92%. Four had undergone catheter interventions for RVPA stenosis. 19% (n=5/26) had a recent history of respiratory symptoms or repetitive infections.

Conclusion: Hospital mortality from respiratory failure is significant. Prenatal diagnosis identifies patients at risk for postoperative respiratory failure. Hospital survivors have a good midterm outcome with absence of symptoms in the vast majority of the patients.