

Outcome of atrioventricular septal defects with single ventricular palliation: 50 years of experience

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Introduction: "Single ventricles" with atrioventricular septal defect (AVSD) include a variety of complex congenital heart defects such as unbalanced AVSD and univentricular hearts with common atrioventricular valve, commonly associated with heterotaxy. Our objective was to describe hard outcomes (death or heart transplantation) and their predictors in these defects.

Methods: We conducted a retrospective, monocentric review of children with common atrioventricular valve, who entered a univentricular path repair at our institution.

Results: 161 patients with AVSD were included into the study: 121 (75%) had univentricular hearts with heterotaxy syndrome, and 40 (25%) had unbalanced AVSD that were not suitable for biventricular repair. Median follow-up was 7.3 years (max 34 years). 41.6% patients had a prenatal diagnosis. Overall mortality was 50.3%. 24% (39/161) never had surgery with a mortality of 77% at a median age of 1.7 years. 24% (39/161) had the first stage palliation (Blalock-Taussig shunt 10.5%, banding of pulmonary artery 10.5%, other 3%) with a mortality of 69% at a median age of 3.2 years. 26% of patients reached the second stage palliation (Glenn surgery) (42/161) with a mortality of 48% at a median age of 8 years, and 26% of patients had the third stage palliation (Fontan completion) (41/161) with a mortality of 10% at a median age of 16.2 years. Protective factors were prenatal diagnosis (HR 0.32, $p=0.002$) and to have reached Fontan stage for those who effectively entered the surgical program (HR 0.08, $p<0.0001$). These results were confirmed by multivariate logistic regression analysis. 70% of survivors ($n=51$) were in NYHA functional class I and 78% received at least one cardiac medication.

Conclusion: Mortality and morbidity of univentricular hearts with atrioventricular septal defect remain high. Prenatal diagnosis is a protective factor. Once the total cavopulmonary connection had been performed, the outcome is comparable to that of other univentricular hearts.