Long-term outcome of 117 patients with univentricular heart and common atrioventricular valve

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Introduction: Few studies investigated the long-term outcome of patients with univentricular heart and common atrioventricular valve.

Method: We retrospectively analysed the medical files of all patients univentricular heart with common atrioventricular valve in the setting of heterotaxy or with unbalanced atrioventricular septal defect preventing biventricular repair.

Results: 117 patients were identified during the study period. 89/117 had a postnatal diagnosis. 28/117 patients never underwent surgery, 25/117 underwent one palliation surgery (Blalock-Taussig-shunt (BTS)/ pulmonary banding), and finally, 61/117 patients entered a sequential cavopulmonary connection program: 37/61 had partial cavopulmonary connection at the time of data analysis while 24/61 had total cavopulmonary connection (TCPC). The average age at TCPC was 7.6 years +/-4 years [1.7-16 years]. Three patients were eventually transplanted. The overall mortality was 59 % (69/117): 65% and 30% in heterotaxy and in patients with unbalanced atrioventricular septal defect respectively. Mortality was 85% (24/28) in the subgroup of patients who never underwent surgery, 93% in the subgroup of patients who had a BTS, and 89% after pulmonary banding. In the subgroup planed to have TCPC, 49% died after partial cavopulmonary connection and survival rate was 71.6 % [50.7-100] in patients who had TCPC.

Conclusion: The long-term outcome of univentricular hearts with common atrioventricular valve treated in a tertiary referral center showed a high mortality rate. Patients with this kind of complex congenital heart disease should undergo the sequential univentricular program without delay in order to obtain better long-term survival.