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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: 57/61 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 planned 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.

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Toward better ventricular pacing in patients with a systemic right ventricle

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Introduction: Patients with transposition of the great arteries (TGA) and atrial redirection, have an important risk of heart failure caused by dysfunction of the systemic RV. Conventional non-systemic ventricular pacing (nonsystVP) may even further increase this risk. We investigated whether these patients may benefit from biventricular pacing (BiVP) and/or single-site systemic ventricular pacing (systVP).

Methods: During clinically indicated catheterization in 9 patients with TGA and status post Senning procedure, endocardial ventricular stimulation (overdrive DDD, 80-90 bpm) was applied with temporary pacing leads at the non-systemic ventricle and the systemic ventricle. Acute changes in dP/dtmax and systolic pressure of the systemic ventricle, as induced by non-systVP, systVP and BiVP compared to reference, were assessed with a pressure wire (Radi Medical Systems®) within the systemic ventricle. Reference was AAI pacing with similar heart rate (n=7; filled squares), or nonsystVP at a lower heart rate than during stimulation at experimental sites (85 vs. 90 bpm; n=2).

Results (Figure): Systemic dP/dtmax and systolic ventricular pressure were significantly higher during systVP (+5.6% and +5.1%) and BiVP (+14.3% and +4.9%) when compared with non-systVP. In 6 out of 7 patients, dP/dtmax was even higher during BiVP and systVP than during AAI pacing.

Conclusions: Patients with systemic RV, such as patients with TGA and atrial redirection, may benefit from biventricular or systemic ventricular pacing, especially when ventricular pacing is indicated.

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Is valvulation of Fontan circulation of clinical value?

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Objectives: Fontan circulations conduct to complications such as heart dysfunction, arrhythmia, exsudative enteropathy. The circulation largely depends on breathing and gravity. A part of the caval flow comes back in the abdominal compartment. We propose to study the insertion of valve in the systemic ventricle in patients with refractory dysfuncation. We hypothesized that the valve will reduce the effects of the respiration and gravity improving the net flow in the lungs, the exercise capacity and the enteropathy. The Melody valve is the valve of choice because it is fine and harvested from the bovine jugular vein.

Methods: All patients who received a valve in a Fontan circulation were eligible.

Results: four patients were included. All had a Fontan circulation with clinical and hemodynamic alterations: 2 patients had refractory exsudative...