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Predictors of outcomes in pulmonary atresia with intact ventricular septum

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Background

Pulmonary atresia with intact ventricular septum (PA-IVS) is a congenital heart disease with a spectrum of severity that can lead to univentricular strategy or biventricular normal physiology. Predicting outcomes during the neonatal period remains challenging.

Objectives

To identify the predictors of biventricular repair versus 1.5 ventricle repair in PA-IVS and the risk factors for early mortality.

Methods

We retrospectively reviewed all neonates with PA-IVS over a period of 30 years. We characterized the outcomes as univentricular heart (1V) without attempt to decompress the right ventricle, 1.5 ventricle (1.5V) and biventricular repair (2V). The outcomes were the final type of repair and death. Right ventricle morphological and functional characteristics were analysed and the cut-off values of each measurement to predict the type of repair was evaluated.

Results

248 patients were identified. 49 entered the 1V path (median z-score of tricuspid valve (TV) -3.42) and 199 had an attempt to decompress the RV (median Z-score of TV -1). Of those, 143 had a tripartite RV and 56 a bipartite RV. 16/57 patients with bi-partite RV and who had RV decompression finally entered the 1V path (median z-score of TV -3.25). 183 had a RV decompression with need for additional pulmonary blood flow in 82 (median z-score of TV -1.2) or without additional procedure in 101 (median z-score of TV +0.46). The predictors of 2V repair vs. 1.5 V were Z-score of TV > -1 (OR 5.1, CI95% 1.7-15.1), and no need for additional pulmonary blood flow (OR 7.6; CI95% 3.4-16.7). Risk factors for mortality were Z-score of TV > +1 (OR 5.4; CI95% 1-35.6), and severe tricuspid regurgitation (OR 10.4; CI95% 2.4-41.7).

Conclusion

The size of the inlet of the RV estimated with the z-score of the TV predicts the type of repair in PA-IVS as well as the need for additional pulmonary blood flow after RV decompression. Patients with high z-score of the TV and more than moderate tricuspid regurgitation have the highest mortality. They should potentially have a rapid closure of their arterial duct after RV decompression to avoid reverse flow from the aorta to the right atrium.