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Complex Clinical Cases

EBSTEIN ANOMALY - WHEN NOT TO TRUST THE RIGHT VENTRICLE

Moderated Poster Contributions
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Session Title: Complex Clinical Cases: Pediatric and Congenital Heart Disease
Abstract Category: MD/PhD: Pediatric and Congenital Heart Disease
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Authors: Fabio Akio Nishijuka, Izadora Bighetti Brito, Barbara Santos, Luana Callado, Maritza Anzanello, Denoel Oliveira, maria cola, Thaissa Monteiro, Instituto Nacional de Cardiologia, Rio de Janeiro, Brazil, Faculdade Estácio de Sá IDOMED, Rio de Janeiro, Brazil

Background: Ebstein anomaly (EA) is rare, but the most common condition of the tricuspid valve. It is characterized by tricuspid valve leaflet's low implantation, resulting in inadequate coaptation and atrialization of the right ventricle (RV).

Case: 37year-old women with EA and atrial septal defect underwent atrioseptoplasty and tricuspid valve repair in adolescence. Cardiac magnetic resonance revealed right atrium dilatation (155ml/m2), moderate RV dysfunction and torrential tricuspid regurgitation. Ergospirometry showed a notable decrease in O2 consumption over the years, the last test with peak VO2 of 8.8 ml.kg-¹.min-¹. Right heart catheterization indicated normal mean pulmonary artery pressure.

Decision-making: Heart team concluded that the RV would not support tricuspid valve replacement. The decision was made to conduct univentricular correction by means of total cavopulmonary anastomosis (Fontan), which was then executed employing a fenestrated extracardiac tube. It is well-known amines and positive pulmonary pressure are detrimental to Fontan circulation, hence they were cautiously administered in the immediate post-operative period. Long-term, regular exercise is indicated to improve venous return.

Conclusion: Fontan surgery offers greater long-term survival than heart transplantation. In EA with unfavorable anatomy, biventricular correction may be unfeasible, and univentricular correction may be a viable solution.