Transposition Physiology in the Setting of Concordant Ventriculo-arterial Connections

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Abstract

Background and Aim: To review the anatomical details, diagnostic challenges, associated cardiovascular anomalies, and techniques and outcomes of management, including re-interventions, for the rare instances of transposition physiology with concordant ventriculo-arterial connections. **Methods:** We reviewed clinical and necropsy studies on diagnosis and surgical treatment of individuals with transposition physiology and concordant ventriculo-arterial connections, analyzing also individuals with comparable flow patterns in the setting of isomerism. **Results:** Among reported cases, just over two-thirds were diagnosed during surgery, after initial palliation, or after necropsy. Of the patients, four-fifths presented in infancy with either cyanosis or congestive cardiac failure, with complex associated cardiac malformations. Nearly half had ventricular septal defects, and one-fifth had abnormalities of the tricuspid valve, including hypoplasia of the morphologically right ventricle. A small minority had common atrioventricular junctions We included cases reported with isomerism when the flow patterns were comparable, although the atrioventricular connections are mixed in this setting. Management mostly involved construction of intraatrial baffles, along with correction of coexisting anomalies, either together or multi-staged. Overall mortality was 25%, with one-fifth of patients requiring pacemakers for surgically-induced heart block. The majority of survivors were in good functional state. **Conclusions:** The flow patterns produced by discordant atrioventricular and concordant ventriculo-arterial connections remain an important, albeit rare, indication for atrial redirection. The procedure recruits the morphologically left ventricle in the systemic circuit, producing good long-term functional results. The approach can also be used for those with isomeric atrial appendages and comparable hemodynamic circuits.

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