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When Reconstruction Fails or is Not Feasible: Valve Replacement Options in the Pediatric Population

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Valvular pathology in infants and children poses numerous challenges to the pediatric cardiac surgeon. Without question, valvular repair is the goal of intervention because restoration of valvular anatomy and physiology using native tissue allows for growth and a potentially better long-term outcome. When reconstruction fails or is not feasible, valve replacement becomes inevitable. Which valve for which position is controversial. The goal of this article is to address valve replacement options for all four valve positions within the pediatric population. We will draw from our institutional experience and review current literature to support our preferences.
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Tricuspid Valve Replacement

Of all the cardiac valves, the tricuspid is the least common to require replacement, making up less than 2% of valve replacements in the adult population. Tricuspid valve replacement (TVR) is even less frequently required in children. Irreparable Ebstein's tricuspid valves and tricuspid valve (TV) endocarditis are the two most frequent indications for TVR in children. In the largest single center report of TVR, the Mayo Clinic (Rochester, MN) has performed more than 323 TVRs.[1] The need to replace the TV in their large Ebstein's experience was greater than 50%. TVR was performed when TV repair was not feasible. The surprising conclusion of this sentinel report was that perhaps older children requiring TVR with Ebstein's anomaly fared better than children requiring TVR for other disease entities. Bioprosthetic valves fared better in the TV position than the same bioprosthesis in other cardiac positions.

Kiziltan et al[1] reported on 158 consecutive patients requiring TVR for Ebstein's anomaly at the Mayo Clinic over a 25-year period. Follow-up of 149 patients who survived 30 days ranged up to 17.8 years, with a mean of 4.5 years. Survival was 92.5% ± 2.5% at both 10 and 15 years, with nine late deaths. Freedom from bioprosthesis replacement was 97.5% ± 1.9% at 5 years and 80.6% ± 7.6% at 10 and 15 years, respectively. Significant differences were noted in regards to freedom from reoperation for bioprosthesis in the tricuspid position than in all other cardiac positions (Fig. 1).[1] In addition, freedom from reoperation was less for bioprosthesis than for mechanical prosthesis in the tricuspid position (Fig. 2).[1]

Further support for bioprosthesis in the tricuspid position has been provided by Guerra et al[2] reported a 14-year follow-up on 45 patients at a single institution. Thirty-eight of these 45 patients also had other valves replaced in other cardiac positions simultaneously with their TVR. Morphologic examination of explanted porcine bioprosthesis showed that those implanted in the tricuspid position had lower degrees of calcification and less severe structural changes than those simultaneously explanted from the mitral position. Overall, actuarial freedom from structural deterioration at 14 years for the bioprosthesis was 68% ± 13% in the tricuspid position.

Our experience with TVR has resulted in some technical points that we recommend.

1. Valve placement should be cephalad to the coronary sinus, the atrioventricular node, and occasionally the right coronary artery, to decrease the risk that the prosthetic sewing ring or struts will compromise these structures.

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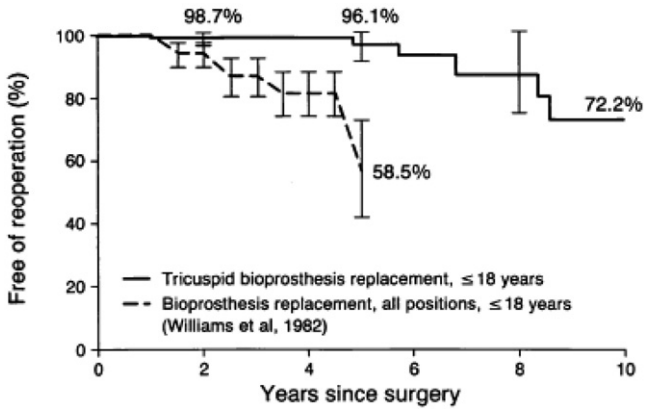

Figure 1 Freedom from reoperation for tricuspid bioprostheses replacement for patients ≤ 18 years in the present series compared with freedom from reoperation for bioprosthesis replacement in all cardiac positions in patients ≤ 18 years previously reported from our institution.

2. Great care should be taken to ensure that the struts of the bioprosthesis straddle the area of the membranous septum and conduction tissue.
3. The valve should be seated into position with the heart beating to observe for rhythm disturbances.
4. Concomitant procedures such as a right-sided MAZE can and should be performed at the time of TVR if indicated.

In summary, we recommend porcine valve replacement for TVR in the majority of patients and feel the risks to conduction and the right coronary artery can be minimized with attention to technical details.

Mitral Valve Replacement

The evolution of mitral repair techniques has revolutionized surgical management of the mitral valve. As a result, the need for mitral valvular replacement (MVR) is relatively uncommon in children. The most common indications for MVR in children include rheumatic disease, endocarditis, mitral stenosis in Shone's complex, or failed AV canal repair. MVR carries the highest mortality for any pediatric valve replacement and has a much poorer long-term prognosis than any other valve replacement in children. The reported operative mortality for MVR in infants is 10% to 30%. The 5- and 10-year survival for these patients has been reported at 50% to 80%.[3,4] Because of these concerning statistics, alternatives to MVR should include aggressive attempts at valve repair, conversion from biventricular to a single ventricle repair, or even cardiac transplantation.[5]

MVR is common in small children who have a small mitral annulus. Unfortunately, annular enlargement options are sparse. Attempting to oversize the prosthesis at the time of MVR can produce subaortic obstruction and should be avoided. Prosthetic leaflet entrapment and conduction block after MVR pose significant postoperative morbidity and mortality. Common reasons for reoperation include prosthetic stenosis, thrombosis, and endocarditis. In comparison to initial MVR, the mitral annulus can usually be upsized 2 to 3 mm in diameter at the time of redo MVR. Low-profile, bileaflet pyrolytic carbon valves are the most popular prostheses for MVR; however, all mechanical valves require lifelong anticoagulation. Bioprosthetic xenografts and mitral homograft valves do not require anticoagulation but have limited durability of 3 to 5 years in the mitral position.

Perhaps the largest review of MVR in the young (under 5 years of age) pediatric population was published by Caldarone et al[5] who analyzed data gathered by the Pediatric Cardiac Care Consortium (45 centers, 1982–1999). MVR was performed 176 times in 139 patients all under 5 years of age. Median follow-up was 6.2 years with age at initial operation being 1.9 ± 1.4 years. Operative morbidity in these patients included heart block requiring pacemaker implantation (16%), endocarditis (6%), thrombosis (6%), and stroke (2%). Patient survival was reported to be 79% at 1 year, 75% at 5 years, and 74% at 10 years. Early mortality was quite high, but late mortality was acceptable. The diagnosis of complete atrioventricular septal defect, Shone's syndrome, and an increased ratio of prosthetic size/weight were all found to be statistically significant predictors of early mortality.

Gunther et al[6] reviewed and published their experience from a single institution for children under 6 years of age requiring MVR. They reported a retrospective analysis of 35 consecutive children requiring MVR with a mean age of 1.9 ± 1.7 years. Actuarial survival at 20 years was 51.2 ± 13.3%. In this series, 29 mechanical and six biologic prostheses were implanted. Surgical mortality was 17.1% with seven late deaths. Overall, eight patients (23%) have thus far required re-operation. Freedom from re-operation at 10 years was 50% ± 22% (Figs. 3, 4).

In an attempt to find a more durable tissue valve that does not require lifelong anticoagulation, our institution has used the pulmonary autograft mitral valve replacement (PA-MVR) or Ross II technique. We have used this strategy in selected older children and young adults whose pulmonary valve was

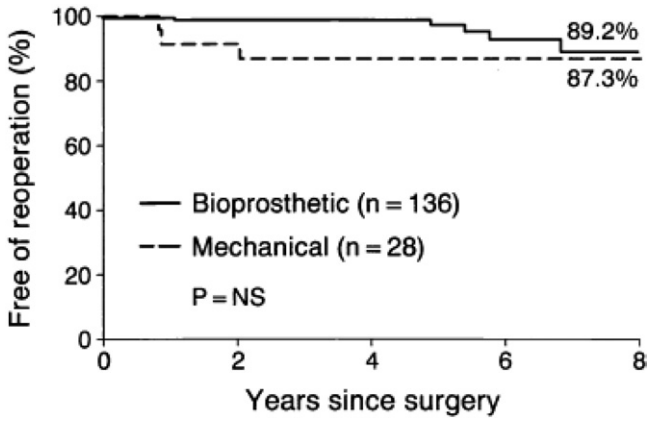

Figure 2 Freedom from reoperation after tricuspid valve replacement with a bioprosthesis compared with freedom from reoperation after tricuspid valve replacement with a mechanical valve.