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When Repair is not Feasible: Prosthesis Selection in Children and Adults with Congenital Heart Disease

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Congenital heart surgeons face many challenges when dealing with valvular pathology in the pediatric population. Because of the concerns related to growth, repair should be the main goal. However, this is not always feasible and valve replacement becomes the only other alternative. Valve replacement also represents one of the most common procedures performed for adults with congenital heart disease, with several valve options existing including homografts, xenografts, autografts, and other artificial prostheses. The choice sometimes may be difficult because there are advantages and disadvantages to each valve substitute. In this article, we will address the different options of valve replacement in children and adults with congenital heart disease, and review the current literature that supports current practice.
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Introduction

Valvular pathology and the need for reinterventions represent an ongoing challenge to congenital heart surgeons. In infants and children, valve repair should be the top priority because of ongoing growth issues. However, this is not always possible and then replacement will be needed despite the expected need for reoperation because of somatic outgrowth. Adults with congenital heart disease (CHD) are a fast-growing population because of improved survival and surgical outcomes of those who have undergone surgery for CHD during infancy and childhood. Therefore, there is a growing need for reoperation; many patients require numerous surgical procedures or interventions over a lifetime.

It is estimated that there is more than a million adults with CHD in the United States alone.[1] This is the result of the significant improvement in surgical techniques and perioperative care, with at least 95% of newborns with CHD surviving to adulthood.[2] However, despite this improvement, residual or recurrent lesions may progress over years and decades, resulting in an increase in the need for reoperation. It is expected that the number of adults with CHD will continue to increase as the population increases.

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We recently reviewed our experience with reoperation in adults with CHD, which included 984 adults with a mean age of 36.4 years; the majority of these reoperations (85%) were related to valve dysfunction,[3] including 25% with multivalve pathology. The most common valve requiring replacement was the pulmonary valve, followed by the aortic valve. However, among these repairs, the most common was the tricuspid valve followed by the mitral valve (MV).

In this article we will review the current literature related to valve replacement in children and adults with CHD and discuss the different valvular substitutes that are available.

The Ideal Prosthetic Valve

The ideal prosthesis chosen by the surgeon to replace the native valve should possess the following favorable characteristics[4]:

- available in different sizes
- has excellent handing features
- growth potential, especially in the younger population
- low infectious potentials
- low risk of thrombosis and dysfunction
- known long-term function, and
- low cost.

Unfortunately, this so-called 'ideal prosthesis' does not exist. However, the above-listed criteria may not all be required or needed to be applied to every patient and by every surgeon.

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Nonetheless, some of these features are essentials in any conduit, including availability in different sizes, good handling characteristics, and some period of freedom from reinterventions because of prosthetic dysfunction.

Numerous factors should be taken into account when selecting a prosthesis, including age, lifestyle, future pregnancy, ventricular function, previous surgeries, number of resternotomies (including the number of previous valve repairs and if there previous valve replacements), as well as the patient's track record with prostheses and anticoagulation.

Aortic Valve Replacement

Aortic valve (AV) replacement is one of the most common procedures performed for adults with CHD, and AV pathology remains one of the most common congenital cardiac defects. Congenital aortic stenosis caused by bicuspid AV continues to be a common indication for valve replacement in a high percentage of patients over their lifetimes.[5] Other congenital diagnoses that may require AV replacement include tetralogy of Fallot, truncus arteriosus, transposition of the great arteries, and aortic aneurysmal disease.

Management of AV pathology in children and young adults is a complex problem, despite the presence of several substitutes to replace the AV, including pulmonary autograft (Ross procedure), homografts, xenografts, and mechanical prostheses. The freedom from reoperation after initial AV replacement according to the prosthesis type has been studied (Fig. 1) and, overall, autografts have superior longevity compared with homografts and tissue valves.[8]

Ross Procedure (Pulmonary Autograft)

The Ross procedure was introduced by Donald Ross in 1967[6] and is considered the substitute of choice for AV replacement in some institutions, with several centers reporting excellent results with this procedure.[5,7] Several advantages support the use of pulmonary autografts, including excellent hemodynamic profile, freedom from anticoagulation and hemolysis, superior longevity, and growth potential.[8] However, it is a technically demanding procedure with reoperation risk to both aortic and pulmonary valves.

The need for reconstruction of the right ventricular outflow tract (RVOT) in the Ross procedure has been considered by some surgeons as placing two valves at risk. The reported freedom from RVOT replacement for children is reported to be about 90% at 12 years; this has been similar among most authors.[9]

Other indications for reoperation include progressive dilatation of the pulmonary autograft and autograft valve dysfunction. Raja and Pollock[10] reported 75% to 100% freedom from autograft dysfunction including severe autograft insufficiency, but this is dependent on the duration of follow-up. The following factors have been identified as risk factors for pulmonary autograft insufficiency: Ross procedure performed for aortic insufficiency, aortic annular dilatation, bicuspid AV, pulmonary valve pathology, and technique of insertion.[11]

The initial implantation technique was the subcoronary free-hand replacement of the AV with the pulmonary valve. However, currently, the most common implantation technique is the aortic root replacement. This places the pulmonary root and its sinuses under the effect of systemic blood pressure and can be a risk factor for future autograft dilatation.[12] Despite early reports that considered this autograft dilatation beneficial, it can be associated with progressive neo-AV regurgitation (Fig. 2).[13] Histologic studies have demonstrated fracture of elastin fibers of the pulmonary artery wall when placed at high pressure.[14]

Recently, several technical modifications to minimize future autograft dilatation have been suggested, including annular reinforcement with felt or Dacron, reinforcement of the entire autograft root (Fig. 3), and resection and replacement of the

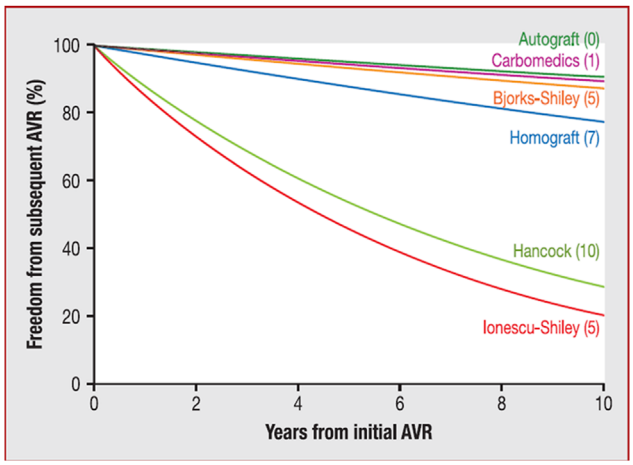

Figure 1 Freedom from reoperation after initial aortic valve replacement, notice the superior longevity of autografts compared with the tissues valves and homografts. The numbers in parentheses represent the total number of AVR episodes for each prosthesis type. (Reprinted from *Circulation*, Vol 122, Karamlou T, Jang K, Williams WG, et al. Outcomes and associated risk factors for aortic valve replacement in 160 children: a competing-risks analysis; pp 3462-3469; 2005, with permission from Wolters Kluwer.[8])

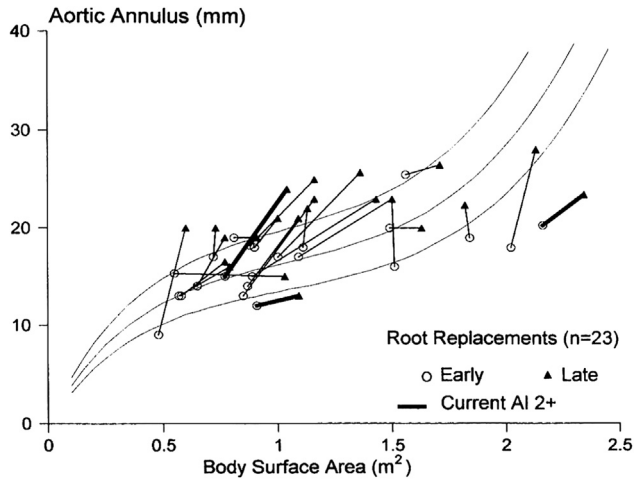

Figure 2 Excessive dilatation of the neoaortic root following the Ross procedure as has been observed when it is done as root replacement. (Reprinted from *Seminars in Thoracic and Cardiovascular Surgery: Pediatric Cardiac Surgery Annual*; Vol 8, Jonas RA. The Ross procedure is not the procedure of choice for the teenager requiring aortic valve replacement; pp 176-180; 2005, with permission from Elsevier.[12])