

Aortic Valve Interventions in Children: Still Only Scratching the Surface



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In this issue of the Seminars, Bouhout et al provide a timely and detailed systematic review of aortic valve interventions in pediatric patients.¹ Aortic valve disease in children is a chronic disease, which entails multiple operations in most patients. Interventions include balloon valvuloplasty, surgical valvotomy, complex valve reconstructions, and valve replacement using either autologous (Ross) or prosthetic valves. Bouhout et al admirably summarize the available data and evidence, addressing how strategies should evolve depending on the age of the patient.

They screened no less than 3349 abstracts and included 73 studies in their systematic review. This included 5499 pediatric patients in total, 1808 with balloon aortic valvuloplasty, 1206 with aortic valvotomy or reconstruction, and 2485 with a Ross procedure. This review presents all of the available data in a succinct, well summarized, and usable format that should be of keen interest to congenital cardiac surgeons and pediatric cardiologists. This review should form the basis for a guideline or scientific statement on aortic valve interventions in pediatric patients by our scientific societies.

The main recommendations can be summarized with the following:

- In the neonates and infants, surgical valvotomy should be the favored approach, given the high-operative risks associated with neonatal Ross procedures and lower freedom from AR and less possibility of aortic valve repair after balloon dilatation, with the aim of postponing aortic valve replacement while optimizing valve function. Avoiding patch material and complex reconstructions, if possible, may allow for growth and a more definitive Ross procedure later in childhood.
- In children and adolescents, aortic valve repair yields excellent survival, although with freedom from reoperation of just 50–80% at 10 years. Predictors of reoperation include preoperative stenosis or unicuspid valves, lack of coaptation symmetry, and use of patch materials. The Ross procedure is safe in this age group, with mortality of



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Central Message

The systematic review on aortic valve interventions in pediatric patients by Bouhout et al is an outstanding step in improving the care of our patients, but it scratches at the surface of evidence. It is our duty to undertake rigorous trials and improve the level of evidence in this field.

0–3% in recent studies, with a pooled annualized risk of autograft reintervention of 1.38% per patient-year.

- Mechanical prostheses are used as a last resort option. Younger age and smaller devices were risk factors for late death.
- There aren't enough data on bioprostheses or homografts.

A systematic review can only be as good as the data it is based on. A major issue in our field, as is often the case in uber-specialized congenital cardiac surgery, is the limited sample size. Despite the large sample size of the review, there was an average of just 75 patients per study. Almost all of these studies were retrospective chart reviews. Few had a comparison group. As clinician-scientists, our patients deserve better evidence to help choose the optimal strategy to treat their disease. Although done with the best of intentions, all of this data do little to improve the level evidence to guide our decision-making. As a coauthor of one of the included studies, a meta-analysis of individual patient data would be of interest.²

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Plagued by a low sample size, we have organized into the Pediatric Heart Network in North America and the ECHSA study groups in Europe to produce several multicenter studies.³ These retrospective studies provide important data on a larger sample size and are generalizable to a larger population. However, they are subject to multiple biases, and randomized controlled trials (RCT) remain necessary. Recent landmark RCT have put into question practices that were thought to be unquestionable in other fields, from arthroscopic partial meniscectomy⁴ to stenting in stable coronary artery disease⁵ or to appendectomy for acute appendicitis.⁶

Within congenital cardiac surgery, there have only been a handful of prospective randomized trials, such as the Single Ventricle Reconstruction trial,⁷ the Atenolol vs Losartan in Marfan study,⁸ or the Enalapril for Single Ventricle Fontan Surgery trials.⁹ Multiple reasons are provided to justify why RCTs aren't feasible in surgical fields,^{10,11} such as the small sample size in our specialized field, or the fact that each one of our patients is unique, and we are often obliged to tailor our repair of congenital cardiac defects to the operative findings. Despite these arguments, it is our duty to our patients to study these questions. The highly controversial ORBITA trial⁵ is particularly informative to our specialty: the investigators conducted a rigorous evaluation of intensive medical treatment with or without coronary stenting in stable coronary artery disease, using a sham procedure as control. They found that stenting didn't improve stress test results, symptoms or maximal oxygen consumption, garnering a New York Times notification of "Unbelievable: heart stents fail to ease chest pain." This trial included just 200 patients, with precise planning and power calculations, while there are an estimated 500,000 percutaneous coronary interventions performed each year world-wide. The ethical justification for this trial, including a sham coronary procedure, comes from the lack of credible evidence to show that coronary stenting improves outcomes in these patients, and that hundreds of thousands of patients were put at risk of a procedure that wasn't proven to benefit them. It is even more interesting that the investigators came under fire from their colleagues, while they are interventional cardiologists convinced that the trial would be positive!

Given studies including 5499 pediatric patients with aortic valve interventions have been published over the last 10 years;

it appears feasible to design and conduct RCTs in these patients. Two main study questions that come to mind are:

- Neonatal aortic stenosis: balloon dilatation vs surgical valvotomy.
- Aortic valve reconstruction in older children: repair vs Ross procedure.

In conclusion, the review by Bouhout et al is an outstanding step in improving the care of our patients with congenital aortic valve disease, but it scratches at the surface of evidence. It is our duty to undertake more rigorous trials.

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