

# Short and Midterm Results of Aortic Valve Cusp Extension in the Treatment of Children With Congenital Aortic Valve Disease

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**Background.** We evaluated our experience with aortic valve cusp extension techniques to identify predictors of successful intraoperative repair and subsequent durability.

**Methods.** Twenty-two children (ages 5–18 years) underwent aortic cusp extension with autologous pericardium between 1999 and 2005. Sixteen children had previous surgical or percutaneous intervention. Ten children had bicuspid aortic valves. Cusp extensions were performed on 1 cusp in 3 patients, 2 cusps in 3, and 3 cusps in 16. Serial echocardiographic measures ( $n = 81$ ) were obtained during a 5-year period and underwent blinded review. Longitudinal trajectories of ventricular and aortic valve function were modeled using mixed linear regression analysis.

**Results.** There was no hospital or late mortality. Five-year freedom from valve replacement was 75%. Comparison of preoperative and post-repair echocardiograms demonstrated reductions in aortic insufficiency (decreased in jet-width/aortic valve diameter ratio from  $0.39$

$\pm 0.12$  to  $0.22 \pm 0.11$ ;  $p < 0.0001$ ), aortic stenosis (decreased in peak aortic valve gradient from  $41 \pm 25$  mm Hg to  $29 \pm 15$  mm Hg;  $p = 0.04$ ), and left ventricular end-diastolic dimensions Z-score (decreased from  $1.39 \pm 0.38$  to  $1.16 \pm 0.34$ ;  $p < 0.001$ ). During the follow-up period, post-repair jet-width and aortic valve diameter increased nonlinearly ( $p < 0.001$ ). Patients with postoperative peak aortic gradients greater than 30 mm Hg had progression of aortic stenosis, whereas those with lesser postoperative peak gradients tended to regress during follow-up ( $p < 0.001$ ). The decrement in Z-score of the left ventricular end-diastolic dimensions remained stable during the follow-up period.

**Conclusions.** Aortic valve cusp extension can result in acceptable hemodynamic results with stabilization of left ventricular geometry. However, residual lesions are common and progression and regression of these lesions can be predicted based on echocardiographic data.

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Surgical treatment of aortic insufficiency (AI) and combined aortic stenosis and aortic insufficiency is problematic in children. Replacement options are limited and are associated with major drawbacks. Mechanical prosthetics require long-term anticoagulation therapy and are often limited by the size of the aortic annulus, whereas homografts and bioprosthetic valves have a high rate of early calcification and failure [1–4]. The Ross procedure using a pulmonary autograft is frequently chosen for infants and small children as it allows for growth [5, 6]. Emerging reports of late dilatation of the neo-aortic root and autograft insufficiency, along with the risk of future right ventricular

outflow tract homograft re-intervention, have contributed to decreased enthusiasm for the Ross procedure in many centers [7–9].

Improved understanding of aortic valve anatomy and the mechanism of AI and the refinement of surgical repair techniques have prompted surgeons to consider repair of incompetent aortic valves in children as an attractive alternative to valve replacement. Several groups have reported encouraging early and mid-term results for cusp extension in the treatment of adult patients with rheumatic aortic valve disease [10–13]. However, few of these studies have attempted to identify intraoperative predictors of early successful repair or evaluated predictors of the long-term durability of the repair. Consequently, we evaluated the short-term and mid-term results of aortic cusp extension at our institution with the objective of assessing the relationships between operative techniques and intraoperative results, as well as attempting to identify predictors of the durability of this repair strategy.

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Table 1. Clinical Profile of All Patients

|                                             |                |
|---------------------------------------------|----------------|
| No. of patients (n)                         | 22             |
| Age range                                   | 5.0–17.6 years |
| Median age                                  | 11.4 years     |
| Male gender                                 | 17 (77%)       |
| Previous percutaneous aortic valvuloplasty  | 8 (36%)        |
| Previous cardiac surgery                    | 6 (27%)        |
| Endocarditis                                | 1 (5%)         |
| Valve type                                  |                |
| Bicuspid                                    | 10 (45%)       |
| Tricuspid                                   | 12 (55%)       |
| New York Heart Association functional class |                |
| I                                           | 0              |
| II                                          | 12 (55%)       |
| III                                         | 8 (36%)        |
| IV                                          | 2 (9%)         |
| Grade of aortic insufficiency               |                |
| None or trace                               | 0              |
| Mild                                        | 0              |
| Moderate                                    | 4 (18%)        |
| Severe                                      | 18 (82%)       |
| Left ventricular ejection fraction          |                |
| EF > 60%                                    | 22 (100%)      |
| EF ≤ 60%                                    | 0              |

EF = ejection fraction

## Patients and Methods

From July 1999 to July 2005, 22 children underwent aortic cusp extension with autologous pericardium at the Hospital for Sick Children in Toronto. Median age at time of the operation was 11.4 years (range, 5.0 to 17.6 years). Clinical, operative, and outcome data were collected retrospectively. Institutional review board approval was obtained prior to chart reviews, and individual patient consent was waived. The original pathology was congenital aortic valve stenosis in the majority of patients ( $n = 18$ ). Eight patients had undergone previous percutaneous aortic balloon valvuloplasty, whereas 6 patients had undergone previous cardiac surgery, including open aortic valvuloplasty ( $n = 2$ ), ventricular septal defect repair ( $n = 1$ ), arterial switch operation ( $n = 1$ ), resection of left ventricular outflow tract rhabdomyoma ( $n = 1$ ), and right ventricular to pulmonary artery homograft conduit ( $n = 1$ ). Although the majority of patients had aortic stenosis as their original diagnosis, at the time of repair, the predominant hemodynamic lesion was AI in 18 patients, whereas 4 patients had a significant component of aortic stenosis (ie, combined AI and aortic stenosis). The clinical profile of the entire cohort is summarized in Table 1.

## Echocardiography

Serial transthoracic echocardiographic data were reviewed by a single blinded cardiologist (TB). Measurements were repeated times 3 and were averaged. There were 81 total repeated measures in 22 subjects, 59 of

which were postoperative values. The median number of studies per subject was 3.5 (range, 2 to 5 per subject), and these occurred in an interval of 8.7 months prior to aortic valve repair and 5 years after repair.

Intraoperative transesophageal echocardiography was also recorded in all patients ( $n = 22$ ) at the time of aortic valve repair. All echocardiographic studies included the aortic jet width that was measured using color Doppler and was indexed to the size of the aortic valve diameter and to the body surface area. Left ventricular end-diastolic dimensions (LVEDD) were converted into Z-scores of the left ventricular end-diastolic dimensions (Z-LVEDD) using regression equations based on previously published nomograms [14, 15].

## Operative Technique

All procedures were performed through a midline sternotomy and are similar to previously published techniques [12]. Autologous pericardium was harvested and treated with buffered glutaraldehyde solution (0.625%) for 5 minutes. Cardiopulmonary bypass was established through a standard aortic and dual-stage venous cannulation. The left ventricle was decompressed by venting through the right superior pulmonary vein. Mild hypothermia (32°C to 34°C) was used with a combination of antegrade and retrograde cold blood cardioplegia. Antegrade cardioplegia was initially administered through the root and then by direct coronary artery cannulation. The aorta was opened through a transverse incision that was extended downward toward the noncoronary sinus. The aortic valve was then inspected and the pathology was analyzed. In patients in whom a cusp was found detached from the aortic wall ( $n = 3$ ), the cusp was reattached with a running monofilament nonabsorbable suture (polypropylene). Cusp perforation was repaired with fresh autologous pericardial patch ( $n = 1$ ). Extended commissurotomy was done in patients with fused commissures. The subvalvular area was evaluated and myectomy or subaortic fibrous membrane excision was performed if indicated. Thickened and fibrotic areas of the cusps were thinned out by sharp dissection. Each leaflet was inspected with respect to the height of the cusps and the commissures, and the length of the free edge. Glutaraldehyde-treated pericardial patches were trimmed and fashioned into the proper shape to fill the defective length and depth of each cusp. The length of the patch was determined by the diameter of the aorta supplemented with an additional 15% length to account for a reduction in the pericardial cusp free edge width associated with pericardial shrinkage. The height of the pericardial cusp was tailored based on the height of the most normal cusp so as to extend the line of coaptation of the repaired cusps 5 mm longer than the highest cusp, and to bring the extended aortic cusps into a coaptation point in the center of the aorta. The cusp extensions were sutured to the free edges of the native cusps with a running suture of 5-0 polypropylene. The extensions were also attached to the aortic wall creating neo-commissures at the level of the sinotubular junction. With all the patch extensions in place, the newly con-

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