Aortic Valve Repair for Adult Congenital Heart Disease
A 22-Year Experience

Vivek Rao, MD, PhD; Glen S. Van Arsdell, MD; Tiron E. David, MD; Anthony Azakie, MD; William G. Williams, MD

Background—Aortic valve–preserving procedures have resulted in excellent outcomes in selected patients, particularly those with normal aortic valve leaflets and dilated aortic roots. However, several congenital heart lesions are associated with abnormal aortic valve leaflets. The long-term results of aortic valve repair for these lesions are not well defined.

Methods and Results—We reviewed the clinical records of 54 adult (age >18 years) patients who underwent repair of congenital abnormalities of the aortic valve between 1976 and September 1999. Follow-up data were available on 52 (96%) patients (mean 50±67 months, range 1 to 266). Patients underwent repair at a mean age of 34±14 years with associated diagnoses of subaortic stenosis (n=10), ventricular septal defect with prolapsing aortic valve (n=17), bicuspid aortic valve (n=23), sinus of Valsalva aneurysm (n=10), and bacterial endocarditis (n=2). There was 1 operative death (1.9%) and 3 late deaths. Survival at 5 and 10 years was 98±2% and 74±12%, respectively. Freedom from reoperation was 74±9% and 51±15% at 5 and 10 years, respectively. The presence of a ventricular septal defect predicted failure of valve repair (59% versus 22%, P=0.01). A bicuspid aortic valve, subaortic stenosis, or the requirement for mitral valve surgery did not affect outcomes.

Conclusions—Aortic valve repair in adult patients with congenital heart disease can be performed with minimal morbidity and mortality rates. The medium-term results of repair are acceptable, regardless of valvular or associated pathology. However, only 31 patients (57%) demonstrated long-term competence of the aortic valve, suggesting that most adult patients with congenital aortic valve disease will eventually require aortic valve replacement. (Circulation. 2000;102[suppl III]:III-40-III-43.)

Key Words: valves • heart defects, congenital • surgery

Improvements in surgical technique have allowed for aortic valve preservation in selected groups of patients. The results of aortic valve repair in patients with abnormalities of the ascending aorta and/or sinotubular junction are excellent.1 2 However, the aortic valve cusps are usually normal in these instances. Several congenital diseases of the heart are associated with abnormal aortic valve cusps. The results of aortic valve repair in this population are not well described. We review the results of surgery in adult patients with congenital abnormalities of the aortic valve.

Methods
The clinical records of 54 consecutive adult (age >18 years) patients who underwent surgical repair of congenital heart lesions involving the aortic valve were reviewed. Patients underwent surgery from 1976 to August 31, 1999, at the Toronto General Hospital. Follow-up care was provided by the Toronto Congenital Cardiac Center for Adults or the referring cardiologist. Data were available for 52 (96%) patients (mean 50±67 months, range 1 to 266).

Valvular Pathology and Indications for Surgery
Most patients in this series had symptoms of aortic insufficiency. There were 2 patients with asymptomatic bicuspid aortic valve stenosis. There were 13 patients with an incompetent bicuspid aortic valve. Two of these patients had a concomitant aortic coarctation. Eight additional patients had a bicuspid aortic valve in association with abnormalities of the aortic root.

Seven patients had an asymptomatic ascending aortic aneurysm and were advised to undergo repair when the size of the ascending aorta was >55 mm. All 7 patients had a functionally competent, nonstenotic bicuspid aortic valve. Ten patients had a ruptured or dilated sinus of Valsalva associated with aortic insufficiency. One of these patients had an incompetent bicuspid valve; the remaining patients had prolapse of either the noncoronary cusp (n=3) or the right coronary cusp (n=6).

A ventricular septal defect (VSD) was present in 17 patients. Five had a sinus of Valsalva aneurysm and the associated aortic cusp abnormalities described above. Patients with an isolated VSD had aortic insufficiency caused by a prolapsing right or noncoronary cusp. One patient had subacute bacterial endocarditis of the tricuspid valve and underwent tricuspid valve replacement in addition to repair of the VSD and prolapsing noncoronary cusp.

Subaortic stenosis was present in 10 patients. In 6 patients, a prolapsing aortic valve cusp resulted in concomitant aortic insufficiency. Two patients had a bicuspid aortic valve (1 competent, 1 insufficient). Two patients had a fenestration in the noncoronary cusp. One of these patients had a remote episode of acute bacterial endocarditis.
Surgical Technique
A wide variety of surgical techniques were used, depending on the underlying valvular pathology. Two surgeons performed 44 of 54 operations in this series (81%). Regardless of the underlying pathology, no patient was discharged from the hospital after aortic valve repair with more than mild aortic regurgitation.

In patients with a prolapsing cusp (n=26), repair was accomplished by either plication of the abnormal leaflet as described by Trusler et al or by a modification described by David. In the former repair, the leaflets are approximated by placing a central suture in the corpora Arantii. The prolapsing cusp was then inspected, and if the free margin was severely elongated, the cusp was plicated with pledgeted mattress sutures at both of its commissures. In less severe prolapse, the abnormal segment of the cusp was plicated at one commissure in a manner that eliminated the weak, thin portion. Alternatively, for mild prolapse, a double layer of 6-0 Gore-Tex suture was passed along the free margin of the leaflet from commissure to commissure and secured to the outer aortic wall.

There were 23 patients (43%) who had a bicuspid aortic valve. In the 2 patients with stenosis, the valve was repaired with a simple commissurotomy. In the remaining patients who had varying degrees of aortic insufficiency, the prolapsing cusp was repaired with a triangular resection and reapproximation of the free margin with 5-0 polypropylene sutures. The raphe was excised in all patients. In patients with mild annular dilation, pledgeted mattress sutures were placed underneath both commissures to increase cusp coaptation.

In patients with ruptured or aneurysmal sinuses of Valsalva (n=10), patch repair of the aortic sinus was accomplished with either autologous pericardium or glutaraldehyde-preserved bovine pericardium. VSDs were present in 5 patients who had concomitant repair of a sinus of Valsalva aneurysm. Patch repair of the defect was performed with previously described techniques.

Ten patients had subaortic stenosis and concomitant aortic insufficiency. Repair techniques included cusp plication (n=7), patch repair of a perforated cusp caused by remote endocarditis (n=1), plication of the sinotubular junction (n=1), and suture closure of a large fenestration (n=1). All 10 patients underwent resection of a subaortic membrane or a formal myectomy.

Seven patients underwent repair of an aneurysmal aortic root in the presence of a bicuspid aortic valve. After inspection of the aortic valve cusps to ensure normal coaptation and lack of obstruction, an aortic valve-sparing operation was performed with previously described techniques.

Postoperative transthoracic echocardiograms were performed annually, or if clinically indicated, by symptomatology. The degree of aortic regurgitation was estimated by means of standard techniques and graded as none, trivial, mild, moderate, or severe. Similarly, left ventricular function was estimated and graded as normal (ejection fraction [EF] >60%), mildly hypokinetic (EF 40% to 60%), moderate dysfunction (EF 20% to 40%), and severe dysfunction (EF <20%).

Statistical Analysis
All data were analyzed with the SAS software program (SAS Institute). Continuous data were compared by means of the Student’s t test and are expressed as mean ± SD. Categoric data were compared by means of χ² or Fisher’s exact test as appropriate and are expressed as absolute and percentage frequency values. Survival curves were constructed according to the method described by Kaplan and Meier. Cox regression analysis was used to determine the multivariable predictors of survival, freedom from reoperation, and a combined outcome of reoperation/severe aortic insufficiency.

Results
The Table reveals the associated pathology present in this series of patients. The most common abnormality was a bicuspid aortic valve (n=23, 43%). A single prolapsing cusp associated with either subaortic stenosis or a VSD was present in 26 patients. A wide variety of more complex congenital heart lesions was present in a small minority of patients.

Figure 1 illustrates the prevalence of aortic valve repair over time. There is an increasing trend toward valvular preservation in recent times. Almost half of all procedures in this cohort occurred during the past 4 years. There was 1 operative death (1.9%) that occurred in a 21-year-old woman with severe hypertrophic cardiomyopathy. This patient had midcavitary obliteration of the left ventricle and was found to have a fenestration of the noncoronary cusp. She underwent myectomy and suture repair of the aortic valve. Unfortunately, she had severe postoperative low cardiac output syndrome and underwent emergency heart transplantation on the fourth postoperative day. She eventually died of multiorgan failure 2 days after transplantation.

There were 3 late deaths. Two sudden, unexplained deaths occurred at 8 and 10 years after repair of aortic insufficiency and VSD. The third late death occurred at reoperation in a patient who underwent repair of a ruptured sinus of Valsalva, closure of an outlet VSD, aortic valvuloplasty, and double coronary artery bypass at the age of 50 years. Seven years after his initial repair, he returned with moderate aortic insufficiency, recurrent angina, and a dehiscence of his VSD patch. Attempted aortic valve replacement and redo coronary revascularization was unsuccessful.

![Figure 1. Prevalence of aortic valve repair over time.](http://circ.ahajournals.org/content/17/5/1110.full)
Figure 2. Overall survival in 54 adult patients after aortic valve repair for congenital heart disease.

Figure 2 illustrates overall survival in this study population. Survival is 98±2% and 74±12% at 5 and 10 years, respectively. Because of the small number of adverse events, multivariable analysis for the predictors of either early or late survival did not yield meaningful results.

Figure 3 reveals freedom from reoperation and freedom from failure (reoperation or recurrent moderate or severe aortic insufficiency). Freedom from reoperation is only 74±9% and 51±15% at 5 and 10 years, respectively. Freedom from the combined outcome of reoperation or recurrent aortic insufficiency is only 55±10% at 5 years and 42±12% at 10 years. Figure 4 illustrates the univariate predictors of successful repair. There was no effect of a bicuspid valve, subaortic stenosis, or concomitant mitral valve surgery on the results of aortic valve repair; however, patients who underwent concomitant repair of a VSD were more likely to undergo reoperation or to have recurrent aortic insufficiency. Cox regression analysis revealed only the presence of a VSD to be predictive of reoperation or recurrent aortic insufficiency ($P=0.04$, OR 2.53; 95% CI 1.58 to 4.06).

Discussion

There has been an increasing trend toward valvular repair versus replacement for a variety of acquired and congenital heart diseases.\textsuperscript{8–12} The results of aortic valve repair in patients with Marfan’s syndrome or other abnormalities of the aortic root are generally excellent.\textsuperscript{1,2} These patients have dilated sinotubular junctions or annuloaortic ectasia as a cause for their aortic insufficiency and usually maintain normal aortic valve leaflets. However, the decision to repair or replace a structurally abnormal aortic valve remains controversial. Bicuspid aortic valves represent a main cause of isolated aortic insufficiency, which usually presents at a young age.\textsuperscript{9} A majority of patients with a bicuspid aortic valve remain asymptomatic and enjoy a normal lifespan.\textsuperscript{13,14} Therefore, it is reasonable to attempt valve repair in an effort to restore a competent aortic valve and return patients to a normal lifespan. Several recent studies have demonstrated that valve repair can be accomplished with low perioperative morbidity and mortality rates and reasonable medium-term results.\textsuperscript{9,15} Unfortunately, recurrent aortic insufficiency occurs in 20% to 30% of patients at 5 years, and given the young age at initial repair, most of these patients will eventually require aortic valve replacement.

The results of aortic valve repair in adult patients with other forms of congenital heart disease are not well described. As the results of congenital heart surgery improve, there will be an increasing number of patients who, as adults, will have a wide variety of aortic valve pathology. In this series, there were 8 patients (15%) who had complex forms of congenital heart disease ranging from tetralogy of Fallot to atrioventricular septal defect.

Similar to the reports from Germany\textsuperscript{15} and the Cleveland Clinic,\textsuperscript{9} we observed poor durability of valve repair in this group of patients. Surprisingly, the presence of a bicuspid aortic valve did not influence the need for subsequent valve replacement nor the recurrence of aortic insufficiency. In a previous study, we demonstrated that the aortic wall, leaflets, and pulmonary artery were all abnormal histologically in patients with bicuspid aortic valves.\textsuperscript{16}

We found that the presence of a VSD at the time of valve repair significantly increased the risk of repair failure. In contrast to the 85% freedom from reoperation at 20 years reported by Trusler et al\textsuperscript{8} in a pediatric population, we found
that 56% of patients required reoperation or had recurrent aortic insufficiency at a mean follow-up of ≈5 years. The poorer results observed in adults may be due to the chronicity of the disease in this population. Over time, the turbulent flow from a VSD may cause more profound changes in the affected leaflet and adjoining annulus than occurs in children who undergo repair at a relatively early stage in their disease.

Our study is clearly limited by the small sample size. Despite reviewing a >20-year experience with aortic valve repair, the mean follow-up was only 50 months because almost one half of the procedures took place in the past 4 years (Figure 1). As mentioned previously, we are observing that an increasing number of adult patients have congenital abnormalities of the aortic valve. Many of these patients have potentially repairable aortic valves. With increasing experience and continued follow-up, we hope to more accurately describe the results of valve repair in this challenging population.

If the durability of valve repair for congenital aortic abnormalities remains poor, patients must be presented with surgical alternatives. These patients may not wish to undergo mechanical valve replacement because of the lifestyle restrictions imposed by chronic anticoagulation. The results of bioprosthetic aortic valve replacement also may be poor because of the relatively young patient population and the accelerated rates of tissue calcification in this group. Similar concerns exist for both homograft and autograft replacement of the aortic valve. Pulmonary autografts may be at particular risk in patients with bicuspid aortic valves because of their abnormal tissue pathology.

We hope that further studies will define the appropriate surgical strategy for adult patients with congenital abnormalities of the aortic valve. On the basis of the data from this study, we conclude that most patients can undergo successful aortic valve repair with minimal morbidity and mortality rates. However, these patients remain at risk for the development of recurrent aortic insufficiency, and most will eventually require definitive valve replacement.

References

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_Circulation_. 2000;102:iⅲ-40-iⅲ-43
doi: 10.1161/01.CIR.102.suppl_3.Iⅲ-40
_Circulation_ is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/102/suppl_3/iⅲ-40

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