SURGERY FOR CONGENITAL HEART DISEASE

MORPHOLOGIC DETERMINANTS FAVORING SURGICAL AORTIC VALVULOPLASTY VERSUS PULMONARY AUTOGRAFT AORTIC VALVE REPLACEMENT IN CHILDREN

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The pulmonary autograft is being used with increasing frequency to replace the diseased aortic valve in the pediatric population. Attempted surgical aortic valvuloplasty with an unacceptable result and return to cardiopulmonary bypass for aortic valve replacement with a pulmonary autograft results in prolonged bypass time and increased potential for morbidity. Therefore, the ability to predict an unsuccessful outcome for valvuloplasty would be of significant clinical benefit. This issue is addressed in the present study. *Methods:* Twenty-two patients (median age 5.7 years, range 3 weeks to 14 years) with bicuspid ($n = 11$), tricuspid ($n = 9$), or quadricuspid ($n = 2$) aortic valves underwent valvuloplasty for aortic stenosis ($n = 9$), aortic regurgitation ($n = 7$), or a combination ($n = 6$). Previous related procedures included balloon aortic valvuloplasty ($n = 3$) and open surgical valvotomy ($n = 1$). Median pressure gradient across the aortic valve was 80 mm Hg. Surgical valvuloplasty techniques included thinning of leaflets ($n = 18$), commissurotomy ($n = 15$), suspension of reconstructed leaflet to the aortic wall ($n = 10$), closure of leaflet fenestration ($n = 5$), shortening of free edge of prolapsed cusp ($n = 4$), repair of torn leaflets ($n = 3$), and augmentation of scarred leaflets with autologous pericardium ($n = 3$). Concomitant subvalvular and supra-ventricular stenosis were repaired in nine and four patients, respectively. In five patients, during the same hospital stay, a failed valvuloplasty was converted into a valve replacement with a pulmonary autograft because of residual or resultant stenosis ($n = 3$) or regurgitation ($n = 2$). *Results:* No early or late deaths occurred. At a median follow-up of 16.3 months the median pressure gradient across the aortic valve in the 15 patients with preoperative stenosis or combined stenosis and regurgitation was 16 mm Hg ($p < 0.01$ versus preoperative gradient). Of the 22 patients, the aortic valve functioned normally (defined as $\leq$ mild stenosis or regurgitation, or both) in 14 patients (including five patients with valve replacement); four patients had stenosis (gradients 40, 45, 60, and 60 mm Hg), two patients had regurgitation, and two patients had combined stenosis (gradients 40 and 50 mm Hg) and regurgitation. Three of the patients with recurrent stenosis underwent secondary surgical valvuloplasty without improvement. Outcome after valvuloplasty was examined according to valve structure: six of nine tricuspid valves functioned

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normally, whereas only three of 13 nontricuspid valves functioned normally ($p = 0.07$). Patients with a nontricuspid aortic valve and regurgitation had a high probability of requiring immediate valve replacement ($p = 0.009$). The actuarial freedom from significant native valve stenosis or regurgitation at 24 months was 82% for tricuspid valves and 36% for nontricuspid valves ($p = 0.007$). **Conclusions:** (1) Surgical aortic valvuloplasty should be the preferred approach when the aortic valve is tricuspid. (2) In contrast, aortic valve replacement with a pulmonary autograft should be the preferred strategy in the presence of a nontricuspid aortic valve (especially when the aortic valve is regurgitant) and after failed surgical valvuloplasty. (J Thorac Cardiovasc Surg 1996;111:1149-57)

Awareness of the many drawbacks inherent in aortic valve replacement (AVR) with a prosthesis, as well as the good results of aortic valvuloplasty in adults with aortic regurgitation (AR),^{1,2} has stimulated interest in aortic valve repair rather than prosthetic replacement in the pediatric age group.³⁻⁵ Additionally, it is becoming clear that AVR with a pulmonary autograft (Ross operation), especially in the pediatric population, is the procedure of choice when AVR is necessary.⁶⁻¹² These two developments have led to the need for more precise clinical decision-making for the diseased aortic valve. Attempted open surgical aortic valvuloplasty with resulting residual or new-onset aortic valve stenosis (AS) or AR and return to cardiopulmonary bypass (CPB) for AVR with a pulmonary autograft results in prolonged CPB time and increased potential for morbidity. Because the ability to predict an unsuccessful outcome for surgical aortic valvuloplasty would be of significant clinical benefit, we reviewed our experience with this technique in an attempt to define predictors of unfavorable outcome. This report includes an analysis of the morphologic characteristics of the aortic valve both in patients in whom surgical aortic valvuloplasty was performed successfully and in patients in whom a failed valvuloplasty was converted into an AVR with a pulmonary autograft.

Patients and methods

Definitions. AS was defined as mild when the left ventricular-aortic systolic gradient was less than 40 mm Hg, moderate when the gradient was between 40 and 75 mm Hg, and severe when the gradient was more than 75 mm Hg.¹³

The degree of AR was graded as mild when the regurgitant jet reached 25% of the distance from the valve to the left ventricular apex, moderate when the jet reached 50% of the distance, and severe when the jet reached more than 50% of the distance.

Aortic valve function was defined as normal in the absence of AS or AR, when AS was mild or negligible, or when AR was mild or negligible.

Tricuspid aortic valve was defined as an aortic valve with three well-developed leaflets of approximately equal size, with three well-developed commissures, and with three normally developed interleaflet triangles.¹⁴⁻¹⁶ Bicuspid aortic valve was defined as an aortic valve with either two leaflets or three leaflets with a raphe representing the fused commissure in the conjoint leaflet and with only two well-developed commissures and interleaflet triangles.¹³⁻¹⁷

Patients. Between September 1991 and December 1994, 22 patients (14 boys and eight girls) with a median age of 5.7 years (range 3 weeks to 14 years) underwent surgical aortic valvuloplasty for valvular AS or AR at the University of California at San Francisco. These patients are the basis for this report. During the same period 30 other patients underwent a primary AVR with a pulmonary autograft. These 30 patients had either severely dysmorphic aortic valves not amenable to successful repair or complex left ventricular outflow tract obstruction.

All 22 patients in the study had two-dimensional echocardiograms with Doppler studies and cardiac catheterization before the operation. Fourteen patients had associated cardiovascular anomalies, including subvalvular AS ($n = 9$), ventricular septal defect ($n = 4$), and supravalvular AS ($n = 4$), which are summarized in Table I. Seven patients had previous operations (some performed at our institution and some elsewhere) related to the left ventricular outflow tract: surgical aortic valvotomy ($n = 1$), balloon aortic valvuloplasty ($n = 3$), resection of subaortic membrane without additional myectomy ($n = 2$), and placement of a stent for subvalvular AS ($n = 1$). In the four patients who had a primary balloon aortic valvuloplasty or surgical aortic valvotomy, the intervals between these procedures and surgical aortic valvuloplasty were 3 hours (in a patient who underwent emergency operation for severe AR owing to a torn aortic leaflet), 15 months, 8 years, and 14 years. Six patients had previous surgical procedures not related to the left ventricular outflow tract, including repair of ventricular septal defect ($n = 3$), tetralogy of Fallot ($n = 2$), atrial septal defect ($n = 2$), aortic coarctation ($n = 1$), and ligation of a patent ductus arteriosus ($n = 1$).

Indications for operation included AS with a peak systolic gradient across the aortic valve greater than 50

mm Hg in nine patients, moderate or severe AR in seven patients, and a combination of both in six patients. The preoperative mean pressure gradient between the left ventricle and ascending aorta in the 15 patients with isolated AS or AS combined with AR was 80 mm Hg (range 30 to 150 mm Hg). Of the seven patients with isolated AR, two had severe AR and five had moderate AR. Aortic regurgitation in all six patients with combined AS and AR was moderate.

At discharge and at regular intervals thereafter, all patients were examined for assessment of their condition on the basis of history, physical examination, and two-dimensional echocardiography with Doppler studies.

Surgical techniques. The operation was performed with cardiopulmonary bypass with cannulation of the ascending aorta and both venae cavae. A left ventricular vent was introduced through the right upper pulmonary vein, and systemic moderate hypothermia was used in all patients. After aortic crossclamping, the aorta was opened with an oblique incision extending into the noncoronary sinus of Valsalva. In all patients, cold blood cardioplegic solution was infused selectively into the coronary ostia.

Commissurotomy was performed with a No. 11 blade knife, extending the opening to within 1 to 2 mm of the anulus. Because of the dysmorphic nature of some of these valves, discrete commissures often cannot be discerned; therefore, only those commissures with adequate leaflet suspension to the aortic wall were opened because of concern that division of rudimentary commissures would likely produce significant incompetence.

Aortic valve reconstruction involved numerous techniques. Myxomatous nodularities and areas of leaflet thickening were thinned out to decrease the bulk obstructing the aortic orifice and to improve leaflet mobility and coaptation. Tears in the free edge of the leaflet were reapproximated with 6-0 or 7-0 polypropylene sutures (Prolene, Ethicon, Inc., Somerville, N.J.). Small leaflet perforations were closed directly and larger ones with fresh autologous pericardial patches. Scarred and retracted leaflets were augmented with a strip of pericardium, which was sutured to the free edge of the leaflet; reconstructed leaflets were subsequently suspended to the aortic wall with pledget-supported sutures. Similarly, prolapsed cusps were shortened by resuspension of the cusps at the commissures with pledget-supported sutures.

In patients with associated discrete or tunnel subvalvular AS, the fibromuscular membrane was accessed retrogradely through the aortic valve and removed by sharp dissection.¹⁸ Extensions of the membrane onto the aortic valve leaflets were peeled off. An additional generous myectomy was performed, extending from the nadir of the right aortic cusp to the anterior leaflet of the mitral valve. In patients with associated supravulvular AS, a Y-shaped incision was made into the noncoronary and right sinuses of Valsalva (to the left of the ostium of the right coronary artery).^{19, 20} The stenotic ridge at the sinotubular junction was excised as much as possible, thereby releasing and lengthening the involved aortic leaflet(s) and relieving coronary ostial obstruction. Subsequently, a generous Y-shaped collagen-impregnated knitted Dacron patch (Hemashield, Meadox Medicals, Inc., Oakland, N.J.) was used to close the incision.

Table 1. Associated cardiovascular anomalies (14 patients)

Anomaly	Patients (No.)
Subvalvular aortic stenosis*	9
Ventricular septal defect	4
Supravulvular aortic stenosis†	4
Tetralogy of Fallot	2
Atrial septal defect	2
Patent ductus arteriosus	2
Supravulvular mitral ring*	1
Coarctation of aorta*	1
Supravulvular pulmonary stenosis†	1
Double-chambered right ventricle	1
L-Malposition of aorta	1
Juxtaposed left atrial appendages	1

*In one patient supravulvular mitral ring, subaortic stenosis, and aortic coarctation were part of Shone's syndrome.

†Part of Williams-Beuren syndrome.

AVR with a pulmonary autograft was performed by the root replacement technique with coronary implantation and allograft reconstruction of the right ventricular outflow tract.¹¹ The quality of repair was assessed by transesophageal echocardiography once the patient had been weaned from CPB.

Statistical methods. Data are expressed as mean $\pm$ standard deviation. The relation between aortic valve structure and outcome of operation was analyzed with Fisher's exact test. The actuarial freedom from significant residual or recurrent AS or AR was calculated by means of the Kaplan-Meier method (SPSS for Windows Software, SPSS, Inc., Chicago, Ill.).

Results

The operative findings with regard to the number of aortic valve leaflets in the 22 patients were as follows: bicuspid ($n = 11$) (Figs. 1A and 1B), tricuspid ($n = 9$), and quadricuspid ($n = 2$) (Fig. 1C). The valve leaflets displayed various degrees of dysplasia, thickening, and myxomatous nodularities (Figs. 1A to 1C). Various degrees of commissural fusion were present in 18 patients. Lack of cusp coaptation was the predominant factor contributing to AR in 13 patients. In two patients with bicuspid aortic valves, the free edge of the right or left cusp was torn near the commissure; the tear was the result of balloon valvuloplasty, necessitating emergency operative intervention, in one of these two patients. Three patients with bicuspid aortic valves had large fenestrations of the right cusp. In two patients the fenestrations were iatrogenic after repair of a ventricular septal defect and tetralogy of Fallot elsewhere; the third was due to bacterial endocarditis.

The various techniques of surgical aortic valvulo-

Fig. 1A. Bicuspid aortic valve. *Left*, Valve in open position. *Right*, Valve in closed position. Note commissural fusion and poor coaptation of leaflets caused by relatively annular arrangement of attachment of leaflets with resultant reduced height of interleaflet triangles and reduced depth of cusps.

Fig. 1B. Bicuspid aortic valve with raphe (*arrow*) representing rudimentary fused commissure.

Fig. 1C. Quadricuspid aortic valve. Note redundancy of two leaflets with consequent regurgitation caused by sub-optimal coaptation.

plasty that were performed are summarized in Table II. In four patients (all with a bicuspid aortic valve) surgical aortic valvuloplasty failed and was converted into AVR with a pulmonary autograft at the

same operation. Two of these patients with a pre-operative diagnosis of combined AS and AR had residual gradients of 40 and 50 mm Hg; one patient with severe AR had moderate residual AR and one

patient with severe AR had a new AS (gradient 40 mm Hg). In one patient with severe AR and a tricuspid aortic valve, an initial surgical aortic valvuloplasty that consisted of shortening and suspension of all three leaflets resulted in mild residual central AR; however, serial postoperative echocardiographic studies demonstrated progressive AR, necessitating AVR with a pulmonary autograft at 1 week after valvuloplasty.

Nine patients underwent concomitant repair of associated anomalies other than subvalvular or supravulvar AS, namely, repair of ventricular septal defect ($n = 1$) and residual ventricular septal defect ($n = 1$), insertion of right ventricle–main pulmonary artery conduit and patch augmentation of both branch pulmonary arteries ($n = 2$), repair of double-chambered right ventricle ($n = 1$), patch augmentation of supravulvar pulmonary stenosis ($n = 1$), resection of supramitral ring ($n = 1$), resection and plication of left ventricular aneurysm ($n = 1$), ligation of patent ductus arteriosus ($n = 1$), and pacemaker implantation ($n = 1$).

No early or late deaths occurred. The median hospital stay was 7 days (range 5 to 11 days). One patient had seizures on the second postoperative day that resolved spontaneously after 48 hours.

Follow-up ranged from 4 to 36 months (median 16.3 months) and was available in all 22 patients. The postoperative pressure gradient in the 15 patients with preoperative isolated AS or combined AS and AR, assessed by the most recent continuous wave Doppler study, was 16 mm Hg (range 10 to 60 mm Hg) ($p < 0.01$ versus the preoperative value). Echocardiographic evaluation in the 17 patients in whom surgical aortic valvuloplasty was the final procedure demonstrated an increase in valve orifice in all patients and improvement in cusp coaptation in 13 patients. The aortic valve functioned normally in nine of these 17 patients, was stenotic in four patients (gradients 40, 45, 60, and 60 mm Hg), regurgitant in two patients (severe AR in one and moderate in one), and stenotic (gradients 40 and 50 mm Hg) and moderately regurgitant in two patients. Three of the 17 patients underwent reoperation for recurrent AS at 6, 11, and 16 months after surgical aortic valvuloplasty. All three of these patients had a bicuspid aortic valve.

At a median follow-up of 17 months in five patients who underwent AVR with a pulmonary autograft for failed surgical aortic valvuloplasty, the new aortic valve continues to function normally; the allograft valve in the pulmonary position was com-

Table II. Various reconstructive surgical techniques on left ventricular outflow tract at primary operation (22 patients*)

Surgical technique	Patients (No.)
Resection of fibrous tissue from aortic valve	18
Commissurotomy	15
Suspension of aortic valve leaflet to aortic wall	10
Resection subaortic membrane and myectomy†	9
Closure of fenestration of aortic valve leaflet	5
Shortening of free edge of prolapsed cusp	4
Patch enlargement of aortic root (supravulvar aortic stenosis)	4
Repair of torn aortic leaflet	3
Pericardial patch augmentation of scarred and retracted aortic leaflet	3
Resection of vestigial cusp of quadricuspid aortic valve	1
Removal of fractured and partially embolized subaortic stent	1

*Includes the five patients in whom a failed surgical aortic valvuloplasty was converted into an aortic valve replacement with a pulmonary autograft.

†Resection of subaortic membrane and myectomy was always combined with removal of fibrous tissue from the aortic valve.

petent in four patients and mildly regurgitant in one patient.

Analysis of outcome of surgical aortic valvuloplasty by aortic valve structure (tricuspid versus nontricuspid) revealed that six of the nine patients with a tricuspid valve had “normal” valve function (according to our definition) after valvuloplasty, whereas only three of 13 patients with a nontricuspid valve had normal valve function after valvuloplasty ($p = 0.07$). In the nine patients with a tricuspid valve, the three valvuloplasty failures were due to AS ($n = 1$), AR (converted to AVR with a pulmonary autograft in the early postoperative period) ($n = 1$), or combined AS and AR ($n = 1$). In the 13 patients with nontricuspid valves, the 10 valvuloplasty failures were due to AS ($n = 5$), AR ($n = 2$), or combined AS and AR ($n = 3$); four of these valvuloplasties were converted to AVR with a pulmonary autograft at the same operation. In the 17 patients who underwent surgical aortic valvuloplasty, the actuarial freedom from significant residual or recurrent AS or AR at 24 months was 82% for those with tricuspid valves and 36% for those with nontricuspid valves ($p = 0.007$) (Fig. 2). The combination of a nontricuspid aortic valve and more than mild AR had a high probability of necessitating a primary AVR with a pulmonary autograft ($p = 0.009$). Preoperative function of the aortic valve

Fig. 2. Freedom from significant residual or recurrent AS or AR.

alone (predominant AS, predominant AR, combined AS and AR) did not have a significant effect on the outcome.

Discussion

Current therapy for congenital critical AS involves either balloon or surgical valvotomy early in life.²¹ Both procedures leave residual lesions that often necessitate subsequent therapy. Less severe congenital aortic valve disease may not necessitate early intervention; however, ultimately surgical therapy will likely be necessary. In both of these settings the surgeon is confronted with a congenitally malformed aortic valve that may be stenotic, regurgitant, or both. Awareness of the drawbacks of prosthetic AVR in young patients has contributed to the great interest in both valve reconstruction techniques and AVR with a pulmonary autograft. Although excellent results can be achieved with valve reconstruction techniques, the inherent tissue limitations and wide morphologic variability of congenital aortic valve disease make it an unpredictable procedure. AVR with a pulmonary autograft is a proven and reliable procedure; however, it has the drawbacks of complexity, violation of the otherwise normal right side of the heart, and use of allograft material. Logic would dictate that valve reconstruction would be the procedure of choice over AVR

with a pulmonary autograft if an excellent hemodynamic result were to be achieved. This unfortunately will not always be the case. The surgeon may be faced with performing an AVR with a pulmonary autograft, with the associated prolonged CPB and aortic crossclamp times required, after a failed attempt at valve reconstruction, which itself often requires an extensive period of CPB. If the surgeon is working within a decision tree such as the one described, it would be very useful to be able to predict those valves in which reconstruction is likely to be successful and those in which it is likely to fail. In the latter, morbidity is likely to be minimized by performing primary AVR with a pulmonary autograft.

Multivariate logistic regression analysis was not performed in this study because of the small number of patients and the potentially large number of variables. Valve structure (tricuspid versus nontricuspid) and preoperative valve function (presence or absence of associated AS, AR, or combined AS and AR) were chosen as easily recognizable and relatively simple variables that are likely to have an impact on the ability to repair the valve. These variables were examined alone and in all possible combinations to determine predictability of surgical valvuloplasty failure. The combination of nontricuspid valve structure and associated AR was highly

predictive of the need for immediate AVR with a pulmonary autograft. In essence, the bicuspid regurgitant valve with or without associated stenosis is not amenable to repair. Analysis based on morphologic considerations alone, that is, tricuspid versus nontricuspid, also showed important differences in postrepair function. However, analysis based on function of the aortic valve alone did not predict the outcome.

The results of this study support the concept that surgical aortic valvuloplasty should be attempted in tricuspid aortic valves that generally have well-developed commissures, especially when the valve is stenotic or when it is regurgitant because of a circumscribed lesion, such as a torn or perforated leaflet. Surgical aortic valvuloplasty is also an option in the purely stenotic nontricuspid aortic valve, with the expectation that AVR with a pulmonary autograft may be needed several years later. Valvular extensions of a subaortic fibrous membrane or leaflet nodularities can be peeled or shaved off the leaflets. Release of any tethering tissue in the subcommissural area improves cusp mobility and further enlarges the orifice. Torn or perforated leaflets (often iatrogenic, infectious, or traumatic) can be repaired primarily or with autologous pericardium. In long-existing AR, however, repair may be suboptimal because of cusp retraction and secondary annular dilatation.^{22, 23}

In contrast, as supported by this study, it is generally inadvisable to primarily repair a regurgitant nontricuspid aortic valve. Such valves invariably have poorly developed commissures and interleaflet triangles and consequently shallow and dysplastic cusps, often resulting in both AS and suboptimal leaflet coaptation.¹⁴⁻¹⁷ Although leaflet augmentation with autologous pericardium has been suggested in such cases,⁵ we have observed that such repair is generally not durable. Therefore, we believe that patients with regurgitant nontricuspid aortic valves should undergo AVR with a pulmonary autograft directly, rather than repair. In patients with recurrent AS after previous surgical aortic valvuloplasty, secondary valvuloplasty may generally lead to an unsuccessful outcome; therefore, AVR with a pulmonary autograft is also generally indicated in this situation.

In summary, primary surgical aortic valvuloplasty may generally lead to acceptable results in aortic valve disease when the aortic valve is tricuspid. It is also a reasonable option in the purely stenotic bicuspid aortic valve. In contrast, when the bicuspid

aortic valve is regurgitant, either alone or in combination with AS, surgical aortic valvuloplasty is generally less effective. In this setting, our preferred strategy is to perform primary AVR with a pulmonary autograft.

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Discussion

Dr. John J. Lamberti (*San Diego, Calif.*). The authors have retrospectively reviewed their experience with 22 patients referred for reparative surgery on the aortic valve. The patient population ranged in age from 3 weeks to 14 years and there were multiple associated abnormalities. This heterogeneous group of patients is separate from 30 other patients operated on during the same period at the University of California at San Francisco. Those 30 patients were judged to have no chance for successful repair of the aortic valve, and they underwent a primary pulmonary autograft procedure. The 22 patients who were examined in the manuscript were thought to have valvular lesions amenable to surgical correction. The authors used an aggressive array of techniques known to be useful in the treatment of both AS and AR. Indications for operation were not completely defined. We do not know how many of the patients had symptoms, we have no data regarding left ventricular size and function, nor do we have data regarding aortic valve area. The issue regarding indication for operation is a real one because the traditional approach to the management of a child with asymptomatic moderate AS and AR has been to delay surgery as long as possible.

We completely agree with the authors' conclusion that patients with a bicuspid aortic valve and AR will have a suboptimal result from valve repair. Whether this subset of patients should undergo the pulmonary autograft procedure before symptoms develop or before they exhibit any evidence of left ventricular dilatation or dysfunction is an unresolved question. The liberal use of the pulmonary autograft procedure in patients without symptoms implies that the surgeon believes that the pulmonary autograft is a curative operation. The literature suggests that autograft valve survival at 5 and 10 years is not 100%. In addition, larger series with longer term follow-up reveal trivial to mild AR in a significant subset of patients undergoing the pulmonary autograft procedure. There are still some unanswered questions regarding the pulmonary autograft in infants and young children.

This paper confirms our clinical impression that some

patients with congenital aortic valve disease are poor candidates for aortic valve repair. It seems that the pulmonary autograft is the ideal operation for infants, children, and young adults with aortic valve disease when AVR is necessary. What is unclear to us is the appropriate timing of that intervention. In our series of pulmonary autografts, most patients are operated on for objective indications, but some patients and their families choose an early autograft procedure in order to avoid lifestyle limitations inherent in the "medical" management of mild to moderate aortic valve disease.

I have some questions for the authors: Can you clarify your indications for operation? Do you perform the pulmonary autograft procedure on patients with asymptomatic moderate valve disease and little evidence of ventricular dysfunction?

Dr. van Son. In general, our clinical decision-making regarding indication for operation is based on physiologic parameters such as degree of AS or AR or a combination of both. This information is augmented with clinical parameters such as the development of left ventricular hypertrophy or dilation and mitral regurgitation. Severe AS or AR, almost always associated with marked left ventricular hypertrophy and/or dilation, generally is a clear-cut indication for operation. In the presence of moderate AS or AR we generally would only operate on the patient who has symptoms or who has signs of progressive left ventricular hypertrophy, dilation, and importantly, mitral regurgitation.

With regard to the second question, our indications for performing a primary Ross operation have evolved over time. As we became more comfortable with this procedure, our tolerance for accepting residual lesions has become drastically less. As a result, our threshold for performing a Ross operation currently is lower than early in our experience. At present, we would not perform a Ross operation on an asymptomatic patient with moderate aortic valve disease and absence of left ventricular dysfunction or mitral regurgitation. However, if future long-term follow-up studies demonstrate that the pulmonary autograft holds up extremely well, then this may change, even to the extent that we may operate on the patient with AS or AR who is without symptoms and still has normal left ventricular function.

Finally, I want to stress that, regardless of the degree of aortic valve disease as determined by preoperative echocardiography, we always carefully assess the aortic valve intraoperatively by direct inspection before embarking on the Ross procedure. Whenever feasible we try to save the native aortic valve.

Dr. Lamberti. My final question is related to the adverse influence of AR on subsequent reparative operations on the aortic valve. It is my opinion that a properly done surgical procedure on the aortic valve will minimize AR and that a creative aortic valvuloplasty will achieve better relief of obstruction than blind balloon dilatation. Do you think that balloon dilatation of the aortic valve should be limited in its application because it produces uncontrolled AR, which hastens the need for AVR and negates the possibility of re-repair?

Dr. van Son. There is no question that the experienced cardiac surgeon with the aortic valve under direct vision

can generally achieve a better result in terms of relief of gradient across the aortic valve than can be done with a balloon. However, we work in an institution where we have excellent interventionalists and they generally achieve good results with balloon valvuloplasty. It is our philosophy that in neonatal AS, in which the aortic valve is almost always highly dysmorphic, gentle balloon dilation may achieve results that are comparable with open surgical valvotomy. Beyond the neonatal period, however, the aortic valve generally is less dysmorphic, in which setting use of the knife may lead to better results.

Careful attention should be given to avoiding an over-extensive aortic valvotomy, because the valvular insufficiency that may develop is poorly tolerated. Resistance at the valve level is inversely proportional to the fourth power of the radius of the aortic valve orifice; therefore, a seemingly conservative aortic valvotomy usually produces a satisfactory clinical result.

Dr. Gregory Misbach (*San Bernadino, Calif.*). My question concerns the bicuspid aortic valve without significant AR. I think your data confirm that valvotomy would be the preferred approach for the tricuspid valve, and I think that a unicuspid valve would be best served by a Ross operation. However, many years ago Dobell reported a series from Montreal Children's Hospital of more than 50 patients receiving surgical aortic valvotomy for AS. Their follow-up indicated about one third of their patients would require reoperation within a decade and virtually all other patients would need an operation eventually. The majority of their patients had bicuspid aortic valves with pure AS. Are you expecting at this time that the Ross operation provides better long-term results for the patient with a bicuspid aortic valve with pure AS than what Dobell already reported?

Dr. van Son. At the present time it is difficult to answer your question with certainty. As we become more familiar with the Ross procedure and gather more data regarding its long-term results, we may be able to extend the indications for this operation. At the present time, we would still proceed with an initial valvuloplasty in the setting of a bicuspid aortic valve and pure AS. In several

clinical series it has been shown that the median freedom from important recurrent AS or new-onset AR, or a combination of both, after valvuloplasty is about 5 years. However, detrimental sequelae of this strategy, among others, consist of the development of variable degrees of left ventricular dysfunction at the time of the second intervention. To avoid this complication, it may well be that in the future the patient with a bicuspid aortic valve and pure AS may be offered a primary Ross operation.

Dr. Jorge Wernly (*Albuquerque, N.M.*). Dr. van Son and his associates have demonstrated that the bicuspid regurgitant valve with or without stenosis is not amenable to surgical repair. I agree that these patients should probably be treated directly with AVR. My question concerns the neonate with critical AS who needs surgical intervention after unsuccessful balloon dilatation. Virtually all these patients have a grossly deformed and frequently unicuspid valve which in many instances is insufficient. Are you prepared to extend the conclusions of your study to the neonate with critical AS and suggest that some of them should undergo primarily a pulmonary autograft AVR? If so, what are your criteria for this specific group of patients? I noticed that the youngest patient in your series was 21 days of age.

Dr. van Son. In case of neonatal critical AS, if a biventricular repair is feasible based on echocardiographic (and sometimes catheterization) data, we generally prefer to perform a balloon valvuloplasty. However, in the presence of severe valve dysfunction including regurgitation, we generally would favor operative intervention and decide intraoperatively between a valvuloplasty or a primary Ross operation. Another situation where we would select the surgical option is in the select subgroup of neonates with borderline hypoplastic left heart syndrome and adequate left ventricular function where a Ross-Konno procedure, if necessary extended with resection of endocardial fibroelastosis, offers a two-ventricle alternative to the Norwood procedure. We have performed Ross or Ross-Konno procedures in babies a few days of age with good results.