SURGERY FOR CONGENITAL HEART DISEASE

GEOMETRIC MISMATCH OF PULMONARY AND AORTIC ANULI IN CHILDREN UNDERGOING THE ROSS PROCEDURE: IMPLICATIONS FOR SURGICAL MANAGEMENT AND AUTOGRAFT VALVE FUNCTION

V. Mohan Reddy, MD[a]
Doff B. McElhinney, MD[a]
Colin K. Phoon, MD[b]
Michael M. Brook, MD[b]
Frank L. Hanley, MD[a]

Background: **There is often substantial mismatch between the diameters of the pulmonary and aortic anuli in young patients with systemic outflow tract disease. To implant the autologous pulmonary valve in the aortic position under such circumstances, it is necessary to adapt the geometry of the systemic outflow tract. The effects of such adaptations on autograft function in children are not well known.** ***Methods:*** **To determine factors predictive of autograft regurgitation, we analyzed 41 cases of children who have undergone the Ross procedure. The diameter of the pulmonary valve was greater (by at least 3 mm) than that of the aortic valve in 20 cases, equal (within 2 mm) in 12 cases, and less (by at least 3 mm) in nine cases, with differences ranging from +10 to –12 mm. In 12 patients with a larger pulmonary anulus, aortoventriculoplasty was used to correct the mismatch. In patients with a larger aortic anulus, the mismatch was corrected by gradual adjustment along the circumference of the autograft, rather than by tailoring of the native aortic anulus.** ***Results:*** **At follow-up (median 31 months), two patients had undergone reoperation on the neoaortic valve for moderate regurgitation. In the remaining 38 cases, autograft regurgitation was as follows: none or trivial in 30, mild in seven, and moderate in one. There was no correlation between regurgitation and age, geometric mismatch, or previous or concurrent procedures.** ***Conclusions:*** **Subtle technical factors that may result in distortion of the valve complex are probably more important determinants of autograft regurgitation than are indication for repair, geometric mismatch, or previous or concomitant outflow tract procedures. Significant mismatch of the semilunar anuli is not a contraindication to the Ross procedure in children. (J Thorac Cardiovasc Surg 1998;115:1255-63)**

At many centers, the Ross procedure has become the preferred option for aortic valve replacement in children.[1-8] The pulmonary autograft has demonstrated long-term functional integrity in the aortic position,[9, 10] and the promise of growth potential appears to be realized.[3, 7, 11] As the merits of this procedure become established, it is being applied in increasingly diverse situations.[4, 5, 11-15] In many children undergoing the Ross procedure for congenital heart lesions, there is a significant discrepancy between the pulmonary and aortic anuli, with differences in diameter as large as 10 mm or more in either direction. There has been no systematic study examining whether such mismatch constitutes a contraindication to the Ross procedure in children. To optimize autograft valve function, any degree of geometric mismatch requires focused surgical attention. Several techniques have been described for

From the Divisions of Cardiothoracic Surgery[a] and Pediatric Cardiology,[b] University of California, San Francisco, Calif.

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Address for reprints: V. Mohan Reddy, MD, 505 Parnassus Ave., M593, San Francisco, CA 94143-0118.

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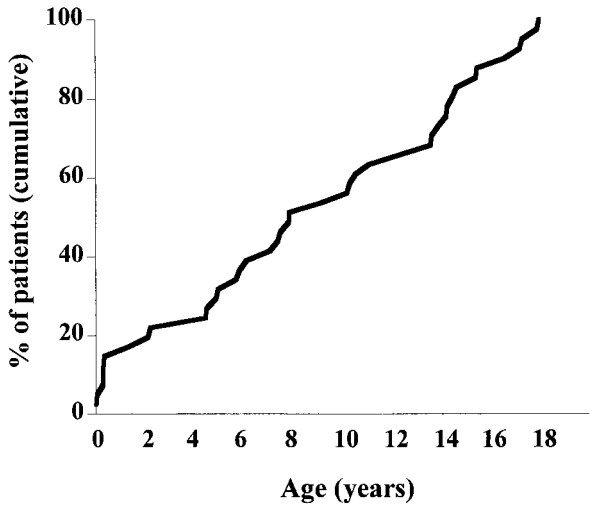

Fig. 1. Age distribution of children undergoing the Ross procedure.

implanting the pulmonary autograft in place of a larger[15-17] or smaller[13, 14] aortic valve. In this study we reviewed our experience with the Ross procedure in children, with a focus on the surgical management of geometric mismatch of the pulmonary and aortic anuli and on the effects on autograft valve function of such mismatch and of other surgical and morphologic variables.

Patients and methods

Patients. Between July 1992 and December 1996, 41 children between the ages of 7 days and 18 years (median 7.8 years) underwent aortic valve replacement with the pulmonary autograft at the University of California, San Francisco Medical Center. The distribution of ages is depicted in Fig. 1. This does not include four neonates with borderline hypoplastic left heart syndrome in whom a Ross procedure was part of a more complex operation performed in an attempt to save the patient after the parents refused a Norwood operation and heart transplantation. Indications for the Ross procedure are summarized in Table I. In 15 of the 41 children the Ross operation was performed as the first surgical procedure; in seven it was the first aortic valve intervention of any kind. Previous left ventricular outflow tract interventions are summarized in Table II.

Preoperative imaging studies. Preoperative cross-sectional echocardiography was performed at our institution for all patients. The aortic and pulmonary anuli were measured at the hinge point of the valves in views obtained from the parasternal long axis window, subcostal window, or both. Off-line measurements were made retrospectively by an echocardiographer who was unaware of operative details and the results of follow-up. Each measurement was made twice, and the average of the two values was recorded for analysis.

No patient was found to have abnormal pulmonary valve morphologic characteristics. The diameter of the pulmonary valve was greater (by at least 3 mm) than that of the aortic valve in 20 cases, equal (within 2 mm) in 12 cases, and less (by at least 3 mm) in nine cases, with differences ranging from +10 to –12 mm. A ratio of annular mismatch (the difference between pulmonary and aortic annular diameters) to pulmonary annular diameter ([pulmonary anulus − aortic anulus]/pulmonary anulus) was calculated from the absolute values. This ratio ranged from –0.7 to +0.6.

Preoperative catheterization (within 2 years) was performed in 25 patients. Eight of these underwent balloon aortic valvuloplasty at that time.

Surgical techniques. Complete root replacement was the technique of pulmonary autograft implantation in all cases. In general the technique we employ is similar to those previously described by others.[1-3] This procedure was performed in 15 of the 41 patients in this study. Large coronary buttons, comprising essentially the entire wall of the coronary sinuses, were mobilized for reimplantation into the autograft so that the autograft sinuses were essentially completely replaced by native aortic wall tissue. The noncoronary aortic sinus was left in situ as a proximally based flap and incorporated into the autograft–to–ascending aorta anastomosis, serving to buttress the noncoronary sinus of the autograft externally.

In only 12 of the 41 patients were the aortic and pulmonary valve diameters within 2 mm in size. Size disparities ranged from a pulmonary valve 10 mm larger than the aortic valve to an aortic valve 12 mm in excess of the pulmonary valve. In the 12 patients in whom the diameters of the aortic and pulmonary anuli were within 2 mm (group I), no specific adaptations were necessary to implant the autograft into the aortic position, and the Ross procedure was performed with a running suture technique of autograft implantation. In cases of geometric mismatch of at least 3 mm, however, technical adaptations were necessary to ensure proper autograft fit and function.

In 12 of the 20 cases in which the pulmonary valve was larger than the aortic valve, a Ross-Konno technique was employed (group II).[14] This operation, which we described elsewhere,[14] consists of implanting the autograft pulmonary valve into the left ventricular outflow tract after the aortic anulus and interventricular septum have been incised in the manner used for a Konno procedure (Fig. 2). The septal defect thus created is incorporated into the autograft suture line, and the defect is patched with either an extension of infundibular muscle harvested with the autograft or a patch of synthetic material. Otherwise, the technique is the same as that employed for a standard Ross procedure.

When the native aortic anulus was larger than the pulmonary autograft by at least 3 mm ($n = 9$), we excised the autograft with a slightly larger than usual cuff of infundibular muscle (Fig. 3). The cuff of muscle attached to the autograft was used to compensate for differences in annular size. With a running suture technique, the larger mass of muscle cuff was incorporated into the anastomosis (group III). With a running suture line, the needle was placed into the infundibular muscle of the autograft

Table I. *Indications for the Ross procedure in children (n = 41)*

Indication	*No. of patients*
Congenital aortic stenosis	29
Now with aortic stenosis and regurgitation	22
Now with aortic stenosis	2
Now with aortic regurgitation	5
Subvalvular aortic obstruction	13
Supravalvular aortic obstruction	4
Congenital aortic regurgitation	3
Rheumatic heart disease	3
Shone's complex	2
Mechanical valve stenosis (growth-related patient-prosthesis mismatch)	1

Numbers of patients do not total 41 because of multiple diagnoses in individual patients.

Table II. *Previous left ventricular outflow tract interventions (n = 41)*

Procedure	*No. of patients*	*No. of procedures*
Surgical valvotomy or valvuloplasty	18	23
Transcatheter balloon valvuloplasty	13	17
Aortic arch repair*	8	11
Subaortic resection	4	4
Aortic valve replacement	1	1
Konno aortoventriculoplasty	1	1
No previous operations	16	
No previous interventions (surgical or catheter)	8	

*Includes coarctation, interrupted arch, and recurrent arch obstruction.

extremely close to the valve leaflets (at the "anulus"), just as in a standard Ross procedure (Fig. 4). The running suture thereby incorporated the entire mass of infundibular muscle into the suture line, allowing this muscle mass to compensate for the larger aortic root diameter without stretching the anulus of the autograft. The native aortic anulus was not plicated or tailored to accommodate the smaller autograft valve. In eight of the nine cases, reduction ascending aortoplasty was also performed by removing a triangular wedge of the anterior aspect of the ascending aorta (Fig. 4).

Additional procedures included left ventricular outflow tract myectomy or membrane resection in six cases, attempted aortic valve repair before resorting to the Ross procedure in six cases, a sinus obliteration procedure[18] in 15 cases, mitral valve repair in one case, and internal thoracic artery bypass of the left anterior descending coronary artery in one case.

Data analysis. Perioperative data were collected by retrospective review of patient records. Follow-up was carried out by direct contact with referring physicians and was current and complete in all cases. Statistical analysis was performed with SPSS software version 7.0 (SPSS Inc., Chicago, Ill.). Follow-up autograft valvular regurgitation was the primary dependent variable assessed, and was scored on a scale of 0 to 4, with *0* for none, *1* for a trace, *2* for mild, *3* for moderate, and *4* for severe. For the purposes of analysis, autograft regurgitation was treated as an ordinal variable (0 through 4) and also collapsed into two separate sets of dichotomous variables, with less than mild and mild or greater as one set of categories and mild or less and more than mild as the other. Although the difference between mild and less than mild regurgitation was not considered clinically important, this distinction was made for the purpose of dichotomous analysis because of the small number of patients with more than mild regurgitation.

Analysis of autograft regurgitation as an ordinal variable was performed with the Wilcoxon signed-rank test for categoric independent variables (such as previous or concurrent left ventricular outflow tract procedure, sinus obliteration technique, and attempted valve repair) and with general factorial analysis of variance for continuous variables (such as age, annular mismatch, cardiopulmonary bypass time, and duration of follow-up). When autograft regurgitation was analyzed as a dichotomous variable,[2] analysis was used to compare categoric independent variables and the unpaired *t* test was used to compare the mean values of continuous variables. Time-related analysis was performed with the Kaplan-Meier product-limit method and the Cox proportional hazards model. Separate time-related analyses were performed with outcome events defined as reoperation, more than mild autograft regurgitation, and mild or more autograft regurgitation. Because no significant risk factors for these outcomes were identified, time-related analysis was not performed with autograft regurgitation treated as an ordinal variable. Factors analyzed for correlation with autograft regurgitation and reoperation included age, primary diagnosis, previous or concurrent left ventricular outflow tract operations, sinus obliteration, attempted aortic valve repair, and cardiopulmonary bypass or aortic crossclamp time.

Results

Operative outcomes. No abnormal pulmonary valve morphologic characteristics were found on intraoperative inspection. In six cases aortic valve repair was attempted with an unsatisfactory result before the decision was made to proceed with the Ross procedure at the same operation. We have previously reported on these cases in an article that examined factors favoring a Ross procedure rather than aortic valve repair.[19]

Among 41 children there was one early death, that of a patient with severe subaortic stenosis who underwent the procedure 5 months after repair of interrupted aortic arch with subaortic resection and ventricular septal defect closure. This patient arrested on the table shortly after anesthesia induction, and the heart could not be restarted. Cardiopulmonary bypass was instituted and the Ross procedure was performed, but the patient could not

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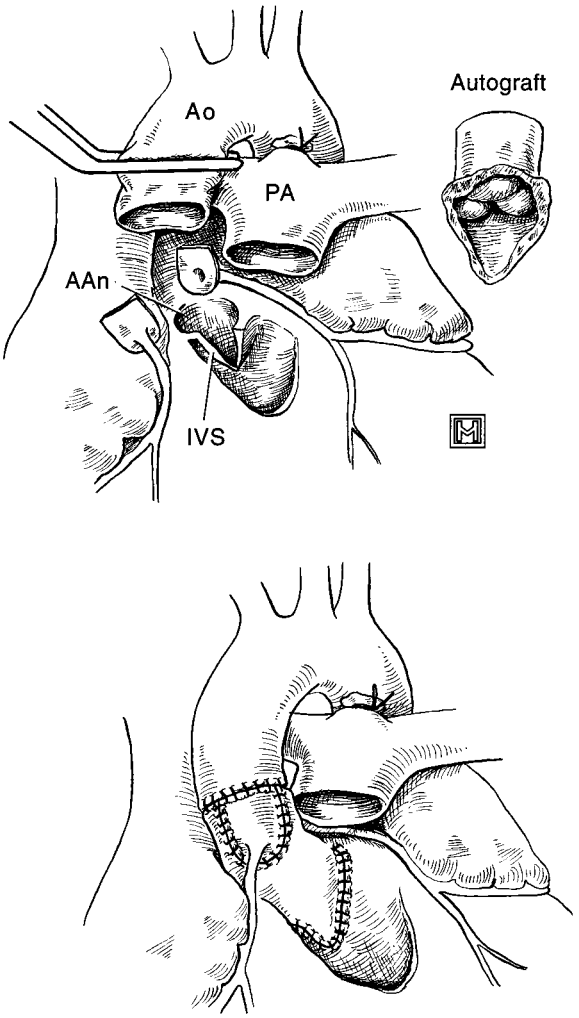

Fig. 2. Ross-Konno procedure, performed when the autograft is larger than the native aortic anulus *(AAn).* The autograft is harvested with a an extension of infundibular muscle, which is used to patch the Konno incision in the interventricular septum *(IVS),* thereby enlarging the left ventricular outflow tract. The right ventricular outflow tract is then reconstructed with an allograft conduit, as in the standard Ross procedure (not shown). *Ao,* Aorta; *PA,* pulmonary artery.

be separated from bypass. Echocardiography performed before discharge showed none or trivial autograft regurgitation in 34 of the 40 survivors and mild regurgitation in the remaining six. There was no difference in early postoperative regurgitation according to preoperative annular mismatch or any of the other variables analyzed.

Follow-up. There have been no late deaths. Patients were followed up for a median of 31 months

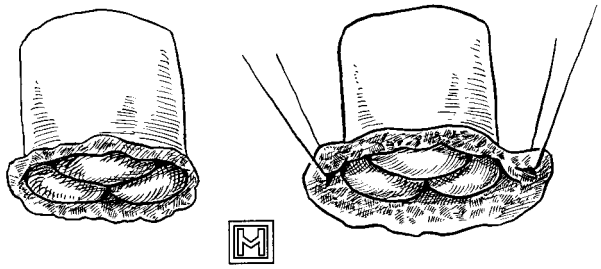

Fig. 3. Usual appearance of the harvested autograft, with a small cuff of infundibular muscle, is shown on the *left.* The autograft is taken with a wider cuff of infundibular muscle, as depicted on the *right,* when the native aortic anulus is larger than the pulmonary anulus.

(range 4 to 46 months)after the Ross procedure, with seven patients followed up for longer than 3 years, 27 for longer than 2 years, and four for less than 1 year. During this time two patients have undergone reoperation. A group II patient (preoperative annular mismatch of 3.5 mm) had recurrent arch obstruction, with resulting progressive autograft regurgitation, and required aortic arch augmentation and commissural resuspension of the autograft valve 19 months after the Ross procedure. This patient had trace autograft regurgitation 13 months after reoperation. A group III patient (annular mismatch of –3 mm) had moderate to severe autograft regurgitation and required mechanical aortic valve replacement. This patient had excellent valve function 12 months after the Ross operation, but moderate to severe autograft regurgitation developed acutely in a short period, and the patient underwent reoperation 11 months later. There was no evidence of neoaortic annular dilatation. Actuarial freedom from reoperation was 97% at 1 year after the operation and 95% at 19 months and beyond.

Among the remaining 38 children, none had autograft stenosis develop, and neoaortic regurgitation at follow-up was trivial or none in 30, mild in seven, and moderate in one. Among the 34 patients with no or trivial autograft regurgitation at discharge from the hospital, three had progression to mild regurgitation at follow-up and one had progression to moderate regurgitation (this was the patient who required reoperation for recurrent arch obstruction). One of the six patients with mild regurgitation at discharge had moderate regurgitation at follow-up. Otherwise, follow-up autograft regurgitation was not different from that in the early postoperative period. Accordingly, early postopera-

tive autograft regurgitation was significantly associated with follow-up autograft regurgitation ($p < 0.001$) by χ^2 analysis. However, mild autograft regurgitation in the early postoperative period did not correlate with likelihood of progression to more than mild regurgitation or with the need for reoperation at follow-up ($p > 0.36$ for both). Actuarial freedom from more than mild autograft regurgitation was 95% at 1 year after the operation and 92% at 2 to 4 years. Freedom from mild or more autograft regurgitation was 90% at 1 year and 80% at 2 years. No patient was found to have evidence of significant autograft root dilatation, and there was no evidence of recurrent subaortic obstruction or septal patch aneurysm in any of the 12 patients who underwent a Ross-Konno procedure.

The distribution of follow-up autograft regurgitation according to the preoperative pulmonary-aortic annular mismatch is depicted in Fig. 5, with group I, II, and II patients indicated separately. As can be seen clearly in the figure, there was no correlation between the degree of annular mismatch and postoperative autograft function. This was also true when regurgitation was analyzed as a dichotomous variable (lowest p value for either of these analyses was 0.57). These results did not change when annular mismatch was analyzed as a ratio of pulmonary-aortic annular diameter difference to pulmonary annular diameter, with regurgitation treated as either an ordinal or dichotomous variable (lowest p value was 0.44). The degree of autograft regurgitation at follow-up (according to both time-related and time-independent analyses) did not correlate with any of the other variables analyzed.

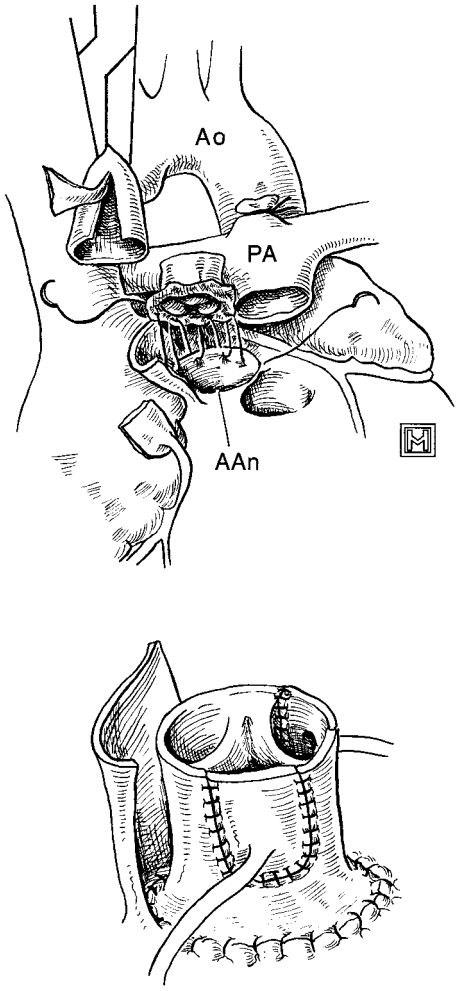

Fig. 4. The autograft with a wide infundibular cuff is sutured to the larger aortic anulus *(AAn)* with a running suture technique, which can be performed to exert a purse-string effect, allowing any residual geometric discrepancy to be compensated for by gradual reduction along the course of the entire suture line. A reduction ascending aortoplasty is also shown, with a wedge of aortic wall tissue removed and closed primarily. The sinus obliteration technique, shown at *bottom,* can also be added. *Ao,* Aorta; *PA,* pulmonary artery.

Discussion

The pulmonary autograft is proving to be an effective and reliable method of aortic valve replacement in children for whom adequate repair cannot be achieved with plastic techniques. It most likely retains growth potential, does not require anticoagulation or induce an immune response, is less prone to the complication of endocarditis than other prostheses, and has demonstrated excellent performance.[1, 6, 7, 9, 13] Because the early mortality rate is generally low,[1-6, 8] the major outcome of interest for patients undergoing the Ross procedure is autograft valve function. To date, functional results have been encouraging, with Gerosa and coworkers[6] and Elkins and associates[10] both reporting approximately 90% freedom from aortic valve reoperation at 5 years and 85% freedom at 8 years. In addition, most series report extremely few cases of significant systemic outflow gradient or autograft regurgitation more than mild in degree.[7, 8, 12, 13, 15, 17] Despite the generally good long-term functional results, there remains considerable room for improvement in both outcomes and patient selection. It is thus important at this juncture to focus on identifying discrete morphologic and technical factors that contribute to autograft regurgitation.

Although growth of the transplanted autograft in

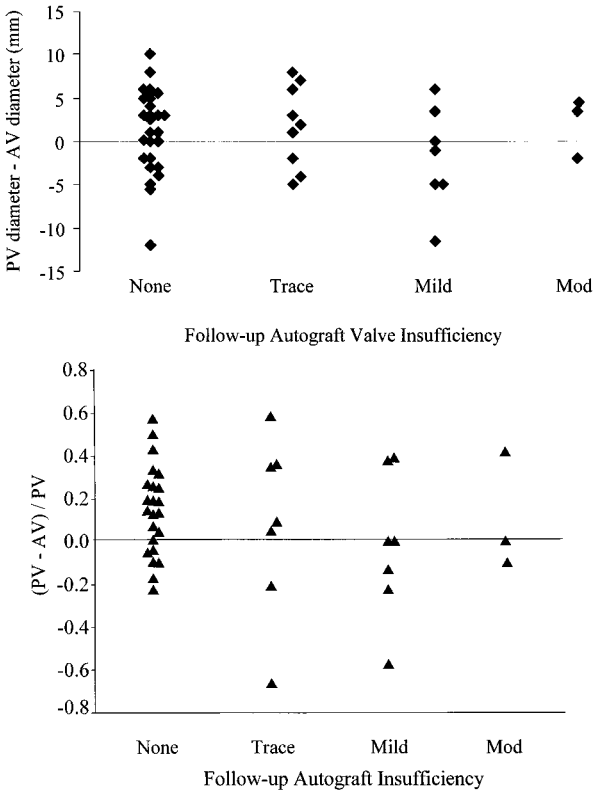

Fig. 5. Distribution of pulmonary valve *(PV)*–aortic valve *(AV)* annular diameter mismatch according to degree of autograft regurgitation at follow-up echocardiography (28 ± 11 months). *Mod,* Moderate.

children has not been demonstrated definitively, several clinical studies[3, 7, 9] and experimental work[11] suggest that growth potential is indeed maintained. Both growth and function of the autograft are probably influenced by technical factors. For example, it has been proposed that growth potential may be optimized by use of a root replacement technique, which we employ in all cases, rather than freehand or inclusion cylinder approaches.[3, 9] Moreover, as both physiologic and morphologic studies show, the semilunar valve root is a complex structure, and structural integrity of the entire root is important in valve function.[20, 21] In cases of size discrepancy between the pulmonary and aortic anuli, the importance of implanting the autograft without distortion of the root is obvious. Because the aortic valve dysfunction in most children undergoing the Ross procedure is either congenital or associated with other congenital anomalies, size differences between the semilunar valve roots are common. Techniques to accommodate and neutralize geometric mismatch will thus be especially important in the pediatric population. The issue of whether annular size mismatch should be considered a contraindication to the Ross procedure in this population has not been previously addressed.

When the native pulmonary valve is larger than the aortic valve, the left ventricular outflow tract can be enlarged by performing an autograft aortic valve replacement with a Konno-type ventriculoplasty. We previously described our experience with the Ross-Konno procedure in some of the cases included in this series.[14] Others have reported similarly promising outcomes.[5, 13] This approach is an attractive alternative for young patients with aortic valve disease and subaortic stenosis because it provides relief at both levels of obstruction. We typically use infundibular muscle for our septal patch, because it fits naturally with the curve of the septum and because it is native tissue and presumably allows for growth of the patch along with the autograft. On occasion we use synthetic material for the patch, which is the primary technique employed in other reports.[5, 13] None of our patients have had aneurysms develop at the patch site, and it remains unclear whether one approach will prove substantially better than the other.

In cases of an aortic root wider than the autograft, our approach is to excise the autograft with a slightly larger than usual cuff of infundibular muscle, which increases the diameter of the autograft sufficiently to compensate for the discrepancy in annular size. A running suture technique is used for implanting the autograft, which tends to have a slight purse-string effect on the circumference of the aortic root, further aiding in adjustment for annular size mismatch. In addition, we often remove a wedge of ascending aortic tissue in these patients to reduce the diameter, and hence wall tension, of the autograft–to–ascending aorta anastomosis. This practice is based on biomechanical studies that have shown the sinotubular junction of the pulmonary root to be significantly more distensible than that of the aortic root or the anulus of either semilunar valve,[22] which raises the concern that the autograft–to–ascending aorta anastomosis may be the most vulnerable point in the reconstructed left ventricular outflow tract with respect to the development of autograft regurgitation. Our approach differs from previously reported tailoring procedures, in which the root diameter is reduced either by removing a wedge of tissue from the aortic anulus and repairing the defect[17] or by plicating the fibrous tissue at the

base of one or both commissures of the noncoronary aortic sinus.[16] Techniques of reinforcing the oversized aortic anulus with pericardium or synthetic material have proved of use in older patients, but they cannot be applied in young children because they are likely to impinge on autograft growth potential. Regardless of the approach selected, full root replacement (as opposed to freehand valve or inclusion cylinder implantation) allows maintenance of autograft root integrity and technically easier adjustment for annular size.

For children with and without annular mismatch, no significant predictors of autograft regurgitation were found in this study. In both patients who required reoperation, there was a mild degree of preoperative annular mismatch, with the pulmonary valve 3 mm larger in one patient and 3.5 mm smaller in the other. Although geometric mismatch is almost certainly a potential cause of autograft dysfunction, surgical techniques described here and by others[15-17] can satisfactorily neutralize this mismatch in patients of all ages. Sinus obliteration was not found to correlate with better autograft valve function, but we consider this technique an important component of the Ross procedure, especially for older patients with relatively thin-walled pulmonary arteries. Although our results are limited by the relatively short follow-up and thus may not be predictive of long-term outcome, they are nevertheless encouraging.

The Ross procedure is an important advance in the treatment of aortic valve disease in neonates and infants with congenital aortic stenosis or regurgitation and in other young patients with aortic valve dysfunction. Even when there is significant geometric mismatch between the pulmonary and aortic anuli, techniques can be used to allow implantation with maintenance of autograft competence. Geometric mismatch of the semilunar valve anuli is not a contraindication for the Ross procedure in children.

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Discussion

Dr. Jan M. Quaegebeur *(New York, N.Y.).* I share your enthusiasm about the Ross operation in terms of its effectiveness in the treatment of children with severe left ventricular outflow tract obstruction. I believe that its performance easily surpasses that of any alternative. The results, as you have shown and also in our experience, are excellent both in terms of reduction of gradients and in terms of survival. Despite the complexity of the operation, it yields excellent results when done in institutions who are prepared to undertake such an operation.

In terms of the annular mismatch problems in those patients, most patients in our experience, as in yours I believe, have aortic stenosis. I agree that the aortic anulus is usually much smaller, and the pulmonary anulus can be as much as 300% larger than the aortic anulus. We have been using the procedure you described, the "glorified Ross procedure" as I call it, for many years with the same excellent results.

Regarding the mismatch in the other direction, I am a bit puzzled, because we do not seem to find that type of difference, with the pulmonary anulus smaller than the aortic anulus. In analyzing our data, we have been more and more concerned about the long-term performance of the neoaortic root in terms of incompetence and dilatation. My first question is therefore whether you find an influence of age among patients who have smaller pulmonary anuli than aortic anuli? Does this happen in older patients rather than younger patients?

Second, if you use a larger amount of infundibulum underneath the autograft, you have a floppy area underneath the aortic anulus. I have always stressed the fact that the proximal suture line should almost be inserted inside the left ventricular outflow tract to support it. Does that influence the stability of the autograft?

Third, in the intriguing case in which severe incompetence developed after initial valve competence, have you determined whether this patient had rheumatic disease to start with? Some patients with rheumatic disease would have autograft destruction earlier than seen in other patients. It may be that this is a group of patients who should still undergo another form of treatment.

Dr. James H. Oury *(Missoula, Mont.).* I compliment you and your coauthors on an excellent series and to support your conclusions. In the International Ross Registry Data, a database now available through the Internet and housed at our hospital in Missoula, Montana, we have more than 2500 cases listed. In looking at the incidence of postoperative aortic insufficiency, if you take what I would call the worst-case scenario—that is, patients who have significant degrees of preoperative aortic insufficiency—and who have all had echocardiographic follow-up, we have data available for 1656 patients. The incidence of moderate to severe aortic insufficiency is 4% in this group. I stress that these are all patients who have had postoperative echocardiographic evaluation. Keep in mind that in a normally functioning mechanical bileaflet valve, the regurgitant fraction is in the 6% neighborhood. I think this puts the incidence of postoperative aortic insufficiency with the Ross procedure into some perspective.

We have analyzed a subgroup of athletes who have been exposed to severe exercise. All these athletes have undergone the Ross procedure. In this series the incidence of aortic insufficiency did not change with severe exercise. These athletes went to between 6 and 8 in the modified Bruce protocol, so they were exercising maximally. My conclusion from this is that significant postoperative aortic insufficiency is not inherent in the Ross procedure; however, I believe that it is due to technical factors in the operation. In addition, these athletes had normal (physiologic) gradients at maximal exercise.

I have two questions. First, at what age would you consider some form of external annular support? I note that the age range of your patients was 4 to 18 years. Second, at what annular ratio or aortic-to-pulmonary annular mismatch would you consider altering your technique, either interrupting the proximal suture line associated with an external form of annular support, doing an annuloplasty, or possibly even abandoning the procedure?

Dr. Thomas L. Spray *(Philadelphia, Pa.).* You mentioned that you decrease the size of the aorta distal to the autograft implantation when there is that sort of size discrepancy. In most of these cases, however, the size discrepancy is in the opposite direction. In our own experience of about 48 such pediatric Ross operations, a significant number of these children have had an extremely small ascending aorta but a good-sized proximal autograft. Would you comment on the techniques you use to make up this size discrepancy? At least in our experience, distal obstruction has been associated with early insufficiency of the autograft, fortunately not severe enough to require reintervention.

Would you also comment on the magnitude of pulmonary valve abnormality that you would consider acceptable for use in a Ross operation? Many of these patients have undergone previous operations, including ventricular septal defect closure, for example, and there may be scarring underneath the pulmonary valve leaflets. Again, in our experience, the only patients who have needed reoperation had an abnormal pulmonary valve at the time of the Ross operation.

Dr. Vaughn A. Starnes *(Los Angeles, Calif.).* I just wanted to comment on one subgroup of patients with the case presentation that you had, the early infants, younger than 3 months. Would you comment on the presence or absence of endocardial fibroelastosis in that group of patients and also on the impact it possibly has on the other valve, the mitral valve? We have about 12 patients in that group, and we have unmasked severe mitral pathology in three cases. It is as though this is a whole left ventricular outflow tract syndrome.

Dr. Hanley. I appreciate all the interest. Dr. Quaegebeur's first question addressed the influence of age on valve mismatch. I agree completely with his observations that in the extremely young infants, the group with critical aortic stenosis, it is essentially unheard of for the pulmonary valve to be smaller than the aortic valve. The aortic valve is always smaller than the pulmonary valve.

We did have a number of patients in the series in whom the pulmonary valve was smaller, but all were in the older

age range and had primary aortic insufficiency as their pathology. I do not know that there is any real difference between our data and Dr. Quaegebeur's observations. To reemphasize, we saw no neonates or young infants in whom the pulmonary valve was smaller than the aortic valve.

I would also like to make the point that the pulmonary valve does not really vary much except in the patients Dr. Spray mentioned, in whom there may have been a previous large ventricular septal defect. This is a very small subpopulation of patients undergoing the Ross operation, although they do exist. Generally speaking, we are looking at a relatively normal pulmonary valve with the aortic valve either small, normal, or very, very large. This is what determines the mismatch in this ratio.

Dr. Quaegebeur's second question is an extremely pertinent question related to our technique, in those cases in which the aortic valve is larger, of using the slightly bulked up pulmonary infundibulum to bridge that mismatch. I agree completely with all his concerns. It is important to bring the suture line right up to the nadir of the cusps of the three leaflets of the autograft. To leave a large section of infundibular muscle would invite disaster.

I did not want to focus the presentation on the specific details of the technique; however, the purpose of that extra tissue is not to bring the infundibulum up higher off the new aortic root but rather to provide some bulk to "stuff" into the larger aortic anulus. The sutures still do come all the way up to the autograft cusps, but we take wide running sutures to bulk up all this infundibular muscle. This muscle takes up space in the aortic root. The suture line, to reemphasize, is not further away from the anulus than in the normal technique.

The patient with normal function a year after the operation who then had sudden development of severe aortic insufficiency was evaluated extremely carefully. There was no evidence of rheumatic disease and no clinical event of endocarditis that we could determine. When we took the valve out, the valve did not appear to be destroyed. There is no good explanation for the development of aortic insufficiency in this case.

Dr. Oury asks at what age we would think of providing external annular support. I do not know that I have given that question much thought. According to Robert Gross's theories about aortic coarctation, when a child reaches about 8 to 10 years old, the distal aorta is about 50% of normal adult size. If it never grows further, there will probably never be any hemodynamic stenosis at that point. On the basis of this, I would probably not even consider it in anyone younger than 8 years, and I do not think it would be too much of a growth-inhibiting factor in anybody older than 12 years.

Another question asks at what ratio limit of aortic valve and pulmonary valve we would abandon the procedure. That is a difficult question. With the Ross-Konno technique for the tiny aortas, we have shown, as Dr. Quaegebeur mentioned, a 300% difference. I do not think that the ratio gets much larger than that. If the ratio is in the other direction, I do not know that I can come up with an absolute number. We have not yet encountered a case in which we believed that we could not deal with the mismatch.

With regard to Dr. Spray's comments about the distal arch augmentation, I agree completely. In most young patients one is dealing with a hypoplastic aorta and it is in fact necessary to augment the distal aorta. We have done that in quite a number of these cases. In a couple of patients with hypoplastic arches, a Norwood type of extension all the way around the hypoplastic arch was performed. In some of our Ross-Konno cases we have performed this procedure. Whenever necessary, we make an anterior incision and place a triangular patch for isolated ascending aortic hypoplasia. Usually it is a piece of the homograft that we are using for the right ventricular outflow tract reconstruction.

In terms of pulmonary valve abnormalities and size, again, if the abnormality is only a size difference, we have not yet encountered a valve that has made us think that we should not do this procedure. In terms of abnormalities, we have had a couple of patients with bicuspid pulmonary valves and have not done the Ross procedure under those circumstances. We usually accept trivial central physiologic pulmonary insufficiency before the operation, but we have not encountered any other pulmonary valve abnormalities per se.

Dr. Spray. We have actually seen one valve that had a separation of the commissural attachments of the cusps by about 5 mm with an otherwise normal valve. It was extremely tempting to try to bring that commissure back together again, so we did that, implanting it as an autograft. It worked fine for about 3 days and then tore out and had to be replaced. That is the instance in which it failed, and I have been reluctant to consider using any abnormal pulmonary valves since that experience.

Dr. Hanley. I have not had that exact experience, but in general I think it is prudent to be extremely rigid in examining the pulmonary valve and, if any abnormality of concern is present, to fight the tendency to move ahead with the operation as planned. I would agree to abort the Ross procedure if there were significant abnormalities.

To briefly address Dr. Starnes' comment about endocardial fibroelastosis, many of these patients do have it, usually the neonates and small infants. I am sorry that I did not bring a slide of a specimen that looks like the shell of a hard-boiled egg, in which we resected a 1 to 2 mm thick endocardial fibroelastosis all the way down to the apex, sparing the papillary muscles. That is an extraordinary example of a relatively common problem. When we do the Konno incision, which is quite common in these small infants, we resect an extensive amount of the endocardial fibroelastosis. We agree that mitral valve dysfunction, when one sees these sort of mummified papillary muscles, is of great concern; I do not think that there is anything you can do with the papillary muscles. When one resects the endocardial fibroelastosis, I think that it is important to stay away from the base of the papillary muscle even if you go beyond it down into the apex, which we have done on a number of occasions. This is a common scenario.