Results of surgery for aortic root aneurysm in patients with Marfan syndrome

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See related editorial on page 773.

Objectives: This study was undertaken to examine the long-term results of surgery for aortic root aneurysm in patients with Marfan syndrome.

Methods: Forty-four patients underwent aortic root replacement and 61 underwent aortic valve-sparing operations for aortic root aneurysm. Patients who underwent aortic root replacement had more severe symptoms, worse left ventricular function, more severe aortic insufficiency, and larger aortic root aneurysms than did patients who had aortic valve-sparing operations. Two types of valve-sparing operations were performed: reimplantation of the aortic valve in 39 patients and remodeling of the aortic root in 22 patients. Echocardiography was performed annually during follow-up. The mean follow-ups were 75 ± 54 months for the aortic root replacement group and 49 ± 38 months for the aortic valve-sparing group.

Results: There were 1 early death and 7 late deaths; 6 deaths were in the aortic root replacement group and 2 were in the aortic valve-sparing group. Survivals at 10 years were 87% in the aortic root replacement group and 96% in the aortic valve-sparing group ($P = .3$). Freedoms from reoperation at 10 years were 75% in the root replacement group and 100% in the valve-sparing group ($P = .1$). Freedoms from valve-related mortality and morbidity were 65% after root replacement and 100% after valve-sparing operation ($P = .02$). Freedom from aortic insufficiency greater than 2+ after aortic valve-sparing operations was 75% at 10 years and was similar for both types of valve-sparing operations; however, the diameters of the aortic annulus and neoaortic sinuses increased only after the remodeling procedure.

Conclusions: This study suggests that aortic valve-sparing operations provide similar survival but lower rates of valve-related complications than aortic root replacement for patients with Marfan syndrome. Reimplantation of the aortic valve may be more appropriate than remodeling of the aortic root to prevent dilation of the aortic annulus, and for this reason we now use only this technique to treat patients with Marfan syndrome.

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ACD

Until recently, composite replacement of the aortic valve and ascending aorta was the standard operation for aortic root aneurysm in patients with Marfan syndrome.[1] In the early 1990s two types of aortic valve-sparing operations were introduced to repair aortic root aneurysms: reimplantation of the aortic valve and remodeling of the aortic root.[2,3] Although these operations were rapidly incorporated into practice by some surgeons,[4-6] others remained skeptical, particularly for patients with Marfan syndrome because of abnormal fibrillin in the aortic cusps.[1,7] Abnormal fibrillin is also present in the leaflets of myxomatous mitral valve,[7] however, and yet the durability of mitral valve repair in this entity has been excellent.[8]

It remains unknown whether aortic valve-sparing operations are better than aortic root replacement for patients with Marfan syndrome.[1,9] It is also unknown what type of aortic valve-sparing procedure is best suited for patients with Marfan syndrome.[4-6,9-12] It has been suggested that the remodeling procedure may better preserve aortic cusp motion, theoretically enhancing the durability of the repair.[11,12] Conversely, patients with Marfan syndrome often have annuloaortic ectasia, which genetic substrate probably has a temporal expression, and the aortic annulus may dilate after the remodeling procedure, with consequent failure of the repair. This study examines the clinical results of both types of aortic root surgery in patients with Marfan syndrome and compares the early and late echocardiographic findings of two types of aortic valve-sparing operation.

Patients and Methods

Cases of all patients operated on for aortic root aneurysm in our institution with the diagnosis of Marfan syndrome according to the Gent criteria[13] were reviewed. Pediatric patients are not operated on in our hospital, but an exception was made for a 12 year-old boy. There were 105 patients: 44 underwent aortic root replacement, and 61 underwent aortic valve-sparing operations. Aortic root replacement was performed throughout the period of study, which extended from 1979 to 2001, whereas aortic valve-sparing operations were performed from 1988 to 2001.

Table 1 shows the clinical profiles of these two groups of patients. All patients underwent transthoracic echocardiography before surgery. Coronary angiography was performed in older patients.

Operative Procedures

Aortic root replacement with a valved conduit was performed in 44 patients. The coronary arteries were reimplanted with the button technique. Mechanical valves were used in 26 patients, aortic valve homografts were used in 9, and bioprosthetic valves were used in 9. The mitral valve was replaced in 7 patients and repaired in 4. Coronary artery bypass grafting was performed in 5 patients. The transverse aortic arch was replaced in 2 patients.

Since 1988, aortic valve-sparing operations were performed whenever the aortic cusps were normal or amenable to repair. Two techniques, reimplantation of the aortic valve and remodeling of the aortic root, were initially used without any particular selection criterion, but only the first procedure was used during the last 2 years of the study period. Technical details of these operations have been published elsewhere.[14,15] Reimplantation of the aortic valve was used in 39 patients and remodeling of the aortic root was used in 22. Age, functional class, size of the aortic root, severity of aortic insufficiency (AI), and ventricular function were identical in these two subgroups, but all patients with acute type A aortic dissection had the reimplantation technique. Six patients (4 in the reimplantation subgroup and 2 in the remodeling subgroup) underwent repair of elongated aortic cusps with a double layer of a 6-0 expanded polytetrafluoroethylene suture.[15] The mitral valve was repaired in 8 patients (4 in the reimplantation subgroup and 4 in the remodeling subgroup). The transverse arch was replaced in 1 patient who underwent the remodeling procedure, and coronary artery bypass grafting was performed in 4 patients, 2 from each subgroup.

Echocardiography

Intraoperative transesophageal echocardiography was used in all patients who underwent aortic valve-sparing operations. A transthoracic study was also performed before discharge from hospital. The first postoperative and most recent echocardiographic studies were analyzed, and the offline measurements of the aortic annulus, the neoaortic sinuses, and the aortic graft diameters from these two examinations were compared. The diameter of the neoaortic sinuses was measured approximately 1.5 cm above the aortic annulus. All measurements were performed during diastole and are presented as the mean value of two consecutive cardiac cycles. Morphologic assessment of the aortic cusps included extent and degree of calcification and thickening. AI was estimated as none (grade 0), trivial (grade 1), mild (grade 2), moderate (grade 3), or severe (grade 4) on the basis of information from color flow mapping and continuous-wave Doppler echocardiography.[16,17]

Follow-up

Patients were seen annually by the referring cardiologist, and ongoing records were maintained in our Adult Congenital Heart Clinic. Echocardiography was performed annually. For this study follow-up was closed on February 28, 2002, and was 100% complete. The mean follow-ups were 75 $\pm$ 54 months for patients who underwent aortic root replacement and 49 $\pm$ 38 months for patients who underwent aortic valve-sparing operations ($P = .01$). The mean follow-ups were 44 $\pm$ 43 months for patients with the reimplantation technique and 63 $\pm$ 24 months for those with the remodeling procedure ($P = .05$).

Statistical Analysis

Comparisons between the two main groups and subgroups were made with unpaired t tests for continuous variables and χ^2 or Fisher exact tests for categoric variables. Estimates for long-term survival or freedom from morbid events were made by the Kaplan-Meier method. Differences between survival curves were evaluated with the log-rank statistic.

TABLE 1. Clinical profile of patients with Marfan syndrome

Variable	Root replacement	Valve sparing	*P* value
No. of patients	44	61	
Sex (No.)			.2
Male	35 (79%)	46 (75%)	
Female	9 (21%)	15 (25%)	
(Age, y)			.7
Mean ± SD	34 ± 11	35 ± 10	
Range	19-75	12-59	
Timing of surgery (No.)			.02
Elective	28 (64%)	45 (74%)	
Urgent	11 (25%)	3 (5%)	
Emergency	5 (11%)	9 (15%)	
Congestive heart failure (No.)	15 (34%)	2 (3%)	.001
Previous cardiac surgery (No.)			
Total	13 (29%)	1 (2%)	.001
Aortic valve replacement	8 (18%)	0	
Aortic valve repair	2 (5%)	0	
Mitral valve replacement	1	0	
Mitral valve repair	1	0	
Replacement of ascending aorta (No.)	1	1	.001
New York Heart Association functional class (No.)			.001
I	9 (20%)	45 (74%)	
II	9 (20%)	5 (8%)	
III	10 (23%)	4 (7%)	
IV	16 (36%)	7 (11%)	
Echocardiographic and angiographic data			
Left ventricular ejection fraction (No.)			.001
Normal	19 (43%)	52 (85%)	
Mild impairment	9 (20%)	8 (13%)	
Moderate impairment	10 (23%)	0	
Severe impairment	2 (4%)	0	
Unknown	4 (9%)	1	
Active infective endocarditis (No.)	4 (9%)	0	.02
Type A aortic dissection (No.)			.0
Acute	6 (14%)	9 (15%)	
Chronic	4 (9%)	0	
Bicuspid aortic valve (No.)	4 (9%)	0	.1
Aortic root diameter (mm, mean ± SD)	62 ± 14	54 ± 8	.001
Mitral regurgitation (No.)	10 (23%)	7 (11%)	.05
AI (No.)			.001
Trace (1+)	5 (11%)	27 (44%)	
Mild (2+)	6 (14%)	19 (31%)	
Moderate (3+)	13 (29%)	9 (15%)	
Severe (4+)	20 (45%)	6 (10%)	
Coronary artery disease (No.)	5 (11%)	4 (7%)	.4

Results

Clinical Outcomes

There was 1 operative death, that of a patient who underwent aortic root replacement. He was in cardiogenic shock secondary to end-stage AI before the operation and died of intractable heart failure.

Nine patients required reexploration of the mediastinum for bleeding, 4 with aortic root replacement and 5 with aortic valve-sparing operations (3% in the reimplantation subgroup and 18% in the remodeling subgroup, $P = .01$). Two patients who underwent aortic root replacement had strokes, with complete neurologic resolution. Blood transfusion or blood products were needed in 32% of patients who underwent aortic root replacement and in 23% of those who underwent aortic valve-sparing operations. There were no cases of wound infection, myocardial infarction, renal or respiratory failure, or any other postoperative complication.

There were 7 late deaths, 5 in the aortic root replacement group and 2 in the aortic valve-sparing group. The deaths in the aortic root replacement group were due to rupture of the false lumen in 2 patients, congestive heart failure in 1 patient, prosthetic valve endocarditis in 1 patient, and com-

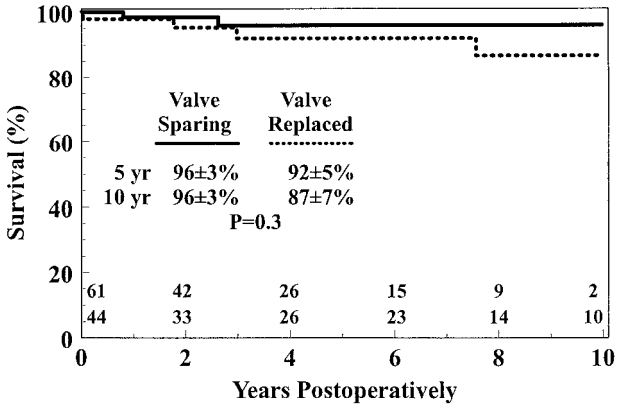

Figure 1. Kaplan-Meier estimates of survival in patients with Marfan syndrome after aortic valve-sparing operations *(solid line)* or aortic valve replacement *(dashed line).*

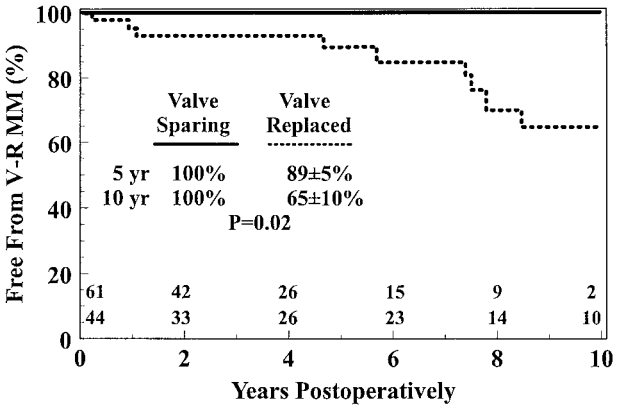

Figure 3. Freedoms from any valve-related mortality or morbidity in patients with Marfan syndrome after aortic valve-sparing operations *(solid line)* or aortic valve replacement *(dashed line).*

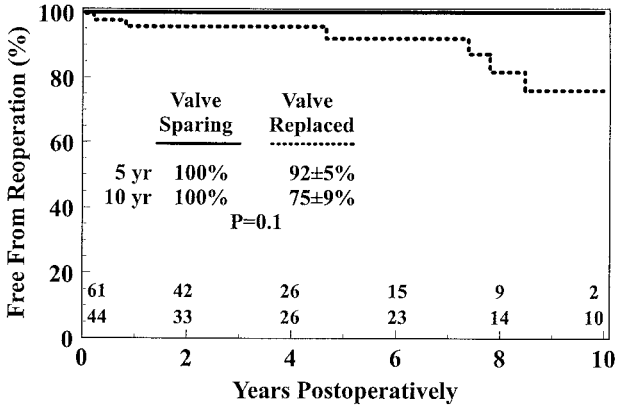

Figure 2. Freedoms from reoperation on aortic valve in patients with Marfan syndrome after aortic valve-sparing operations *(solid line)* or aortic valve replacement *(dashed line).*

plications of vertebral column surgery in 1 patient. The 2 deaths in the aortic valve-sparing group were due to acute type B aortic dissection with rupture in 1 case and chronic obstructive lung disease in 1 case. Figure 1 shows the survivals of both groups of patients; at 10 years the survivals were 87% ± 7% in the aortic root replacement group and 96% ± 3% in the aortic valve-sparing group ($P = .3$).

There were 2 late strokes in patients who underwent aortic root replacement; both patients recovered completely. There were no strokes or transient ischemic attacks in patients who underwent aortic valve-sparing operations. The 10-year freedoms from thromboembolic events were 93% ± 5% after aortic root replacement and 100% after aortic valve-sparing operations ($P = .1$).

There were 4 episodes of prosthetic valve endocarditis in 3 patients who underwent aortic root replacement. One patient, whose condition was deemed inoperable, died. He was an elderly man who had undergone aortic root and mitral valve replacement twice before and had multiple comorbid conditions. The other 2 patients underwent reoperative aortic root replacement (1 patient had 2 reoperations 4 years apart) and survived. The 10-year freedom from prosthetic valve endocarditis after aortic root replacement was 88% ± 7%. One paraplegic patient who underwent an aortic valve-sparing operation acquired infective endocarditis with an aortic root abscess with *Streptococcus faecalis* 11 years after the operation. He underwent aortic root replacement with a homograft and survived.

Five patients required reoperative aortic root replacement, and 2 of them underwent this twice. Three reoperations (in 2 patients) were because of prosthetic valve endocarditis and 4 reoperations (in 3 patients) were because of biologic valve failure. Only 1 patient who underwent an aortic valve sparing-operation needed aortic root replacement for an aortic root abscess 11 years after the operation. This patient had undergone the remodeling procedure. Figure 2 shows the freedom from aortic root reoperation. The 10-year freedoms from reoperation on the aortic root were 75% ± 9% for patients who underwent aortic root replacement and 100% for those who underwent aortic valve-sparing operations ($P = .01$). For the 26 patients who underwent aortic root replacement with mechanical valves, the freedom from reoperation was 92% ± 6%.

Figure 3 shows the freedoms from valve-related mortality and morbidity in both groups of patients. At 10 years, 65% ± 10% of patients who underwent aortic root replacement and 100% of patients who underwent aortic valve-sparing operations were free from any complications ($P = .02$). Among the 26 patients who underwent aortic root replacement with mechanical valves, the freedom from valve-related mortality and morbidity was 87% ± 7%.

Four patients, 2 from each group, with chronic type A

aortic dissections required replacement of the entire thoracic and abdominal aorta because of expansion or rupture of the false lumen. One patient died, and 1 (the same who had endocarditis after an aortic valve-sparing operation) became paraplegic; the other 2 did well. A fifth patient, this one in the aortic root replacement group, needed repair at another institution of a false aneurysm at the right coronary artery reimplantation site.

Echocardiography After Aortic Valve-Sparing Operations

Table 2 shows AI grades early and late after the operation in patients who underwent aortic valve-sparing operations. Figure 4 shows freedom from AI greater than 2+ for all patients who had aortic valve sparing. Figure 5 shows the freedoms from AI greater than 2+ in the two subgroups through 8 years only because of the small number of patients at risk.

The early and late diameters of the aortic annulus (25 ± 3 and 26 ± 3 mm, respectively), neosinuses of Valsalva (31 ± 4 and 31 ± 5 mm, respectively), and sinotubular junction (28 ± 4 and 29 ± 4 mm, respectively) remained unchanged in patients who underwent reimplantation of the aortic valve. The diameters of the aortic annulus and neoaortic sinuses increased in patients who underwent remodeling of the aortic root, as shown in Table 3. As expected, the diameter of the sinotubular junction did not change. Although there was an increase in annular diameter in the entire subgroup from 23.1 to 24.8 mm, annular diameter did not change in 11 patients and increased by 10% or more in 10 patients. Aortic annuloplasty had no effect on annular dilation. Ten patients underwent aortic annuloplasty, and the annulus was dilated by 10% or more in 6 of them. Neoaortic sinus diameter increased in all patients, but it increased by more than 20% in 10 of them; in those 10 patients the aortic annulus was also dilated. We could not establish a relationship between the degree of postoperative dilation of the aortic annulus and the severity of AI in patients who underwent remodeling of the aortic root. In all patients, regardless of the type of aortic valve sparing-operation, the aortic cusps remained thin and pliable.

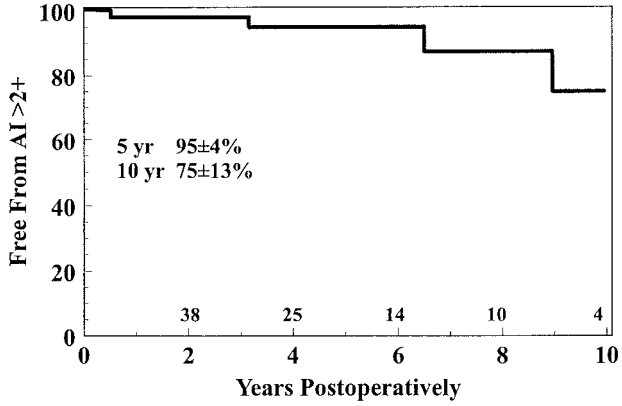

Figure 4. Freedom from AI greater than 2+ in patients with Marfan syndrome after aortic valve–sparing operations.

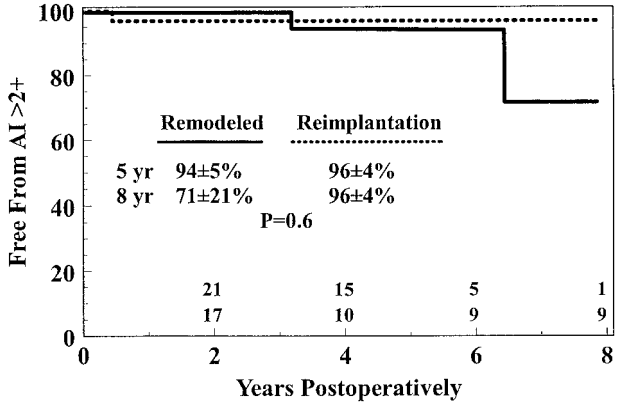

Figure 5. Freedoms from AI greater than 2+ in patients with Marfan syndrome after reimplantation of aortic valve *(dashed line)* and remodeling of aortic root *(solid line).*

Discussion

Without surgery most patients with Marfan syndrome die in the third decade of their lives from complications of aortic root aneurysm, such as aortic rupture, aortic dissection, and AI.[18,19] Aortic root replacement dramatically improves the survival of these patients.[1] In a recent report by Gott and associates[20] on the results of aortic root replacement in 271 patients with Marfan syndrome, there was no operative mortality among 235 patients operated on electively, and operative mortality was only 5.6% among 36 patients operated on urgently. Twenty-four patients who underwent aortic valve-sparing operations were included in that study. The 5- and 10-year survivals was 89% and 81%, respectively. One suspects that most patients received mechanical heart valves and required anticoagulation with warfarin sodium. Only 2 patients died as consequence of oral anticoagulation therapy. At 10 years the freedom from thromboembolic complications was 93%. These rates of thromboembolic and hemorrhagic events are certainly lower than in other reports on aortic valve replacement with mechanical valves.[21,22] The differences could be due to the young age of patients with Marfan syndrome and to other anatomic factors, such as the absence of suture knots on the sewing ring of commercially available valved conduits. Eleven patients had endocarditis develop, and the freedom from prosthetic valve endocarditis was 94% at 10 years.[20] The results of aortic root replacement in our series were similar to those reported by Gott and associates.[20] Dysrhythmias (possibly sudden death), dissection or rupture of the remain-

TABLE 2. AI after aortic valve-sparing operations

	Remodeling (n = 22)		Reimplantation (n = 39)	
	Early	Late	Early	Late
None or trivial	16 (73%)	7 (32%)	27 (70%)	25 (64%)
Mild	6 (27%)	11 (50%)	12 (20%)	12 (31%)
Moderate	0	3 (14%)	0	2 (5%)
Severe	0	1 (4%)	0	0

TABLE 3. Echocardiographic measurements after remodeling of the aortic root

Diameter (mm)	Early	Late	Change	*P* value
Aortic annulus	23.1 ± 1.6	24.8 ± 1.9	1.7 ± 0.4	.0001
Neoaortic sinuses	30.2 ± 4.1	35.3 ± 5.0	5.1 ± 0.7	.0001
Sinotubular junction	26.8 ± 1.5	27.7 ± 2.9	0.9 ± 0.6	.1121

ing aorta, and valve-related complications were the principal causes of death in Gott and colleagues' study[20] as well as in our patients.[1]

The results of aortic valve-sparing operations in patients with Marfan syndrome have been excellent in our institution. Although the long-term survival of these patients was similar to that of patients who underwent aortic root replacement, there was a higher freedom from valve-related morbidity and mortality after aortic valve-sparing operations than after aortic root replacement, as shown in Figure 3. It is unlikely that the preoperative differences in clinical profile accounted for these differences in outcomes. The lack of anticoagulation and the lower incidence of endocarditis may account for some of the differences. We firmly believe that the aortic valve, as well as the mitral valve, should be preserved whenever possible in patients with Marfan syndrome. We consider aortic root replacement and aortic valve-sparing operations not as competitive procedures but as complementary procedures. Aortic valve-sparing procedures should be reserved for patients who have normal aortic cusps, whereas aortic root replacement should be used for those who have abnormal aortic cusps.

We began to perform aortic valve-sparing operations in patients with aortic root aneurysms in 1988. Since then we have replaced the aortic root only if the aortic cusps are abnormal. We started with the reimplantation technique but soon began to use the remodeling procedure because of the theoretic importance of recreating aortic sinuses.[4,12] Later we added an aortic annuloplasty to the remodeling procedure in patients with annuloaortic ectasia or Marfan syndrome in the hopes of preventing future dilation of the aortic annulus.[14] Although the results of both procedures have been satisfactory, we believe that the aortic annulus dilates in a large proportion of patients with Marfan syndrome who had an annulus of normal diameter at the time of surgery. Indeed, we documented a significant increase in the diameter of the aortic annulus in half of 22 patients who underwent remodeling of the aortic root. To our surprise, aortic annuloplasty had no effect on the late dilation, probably because the connective tissue between suture lines dilated or the annuloplasty sutures slowly cut through the abnormal fibrous tissue of the left ventricular outflow tract. We could not establish a relationship between dilation of the aortic annulus and the development of AI in our patients, probably because of a small sample size. As shown in Figure 5 and in Table 3, however, reimplantation of the aortic valve appears to be more stable as far as the lack of AI is concerned.

In a report by Birks and coworkers[23] on 82 patients with Marfan syndrome who underwent the remodeling procedure, the 10-year survival was 84%, and the freedom from reoperation on the aortic root was 83%. At the latest follow-up of patients who were alive without reoperation, 22% had moderate AI. Although no information was given regarding the mechanism of AI, it is conceivable that most failures were due to late dilation of the aortic annulus, because only 2 patients had moderate AI soon after the operation.

The technique of reimplantation of the aortic valve prevents annular dilation because the entire aortic valve is secured inside a tubular Dacron graft. The main shortcoming of this technique is the elimination of the sinuses of Valsalva, which may be important for normal cusp motion and durability.[4,12] This may be true, but it has not become evident during the first decade of follow-up. It is possible to create neoaortic sinuses with the reimplantation technique. All that is required is to use graft larger than needed and tailor it as the operation is done. The diameter of the graft is first reduced at the level where it is secured to the left ventricular outflow tract. Then, after the aortic valve is resuspended inside the graft, the spaces between commissures are plicated to reduce the diameter of the sinotubular junction as desired at the same time as the neoaortic sinuses are created. One may also use a graft with neoaortic sinuses,

which has been developed by De Paulis and associates[24] and is now commercially available. We prefer, however, to tailor our own.

Limitations of the Study

Both aortic root replacement and aortic valve-sparing operations are complex procedures, and probably numerous variables play a role in their outcomes. The sample sizes in this study are too small for a more comprehensive analysis. In addition, because of the small number of patients who underwent aortic valve-sparing operations who were at risk at late follow-up, we may have underestimated the incidence of late AI.

Conclusions

The results of aortic valve-sparing operations in patients with Marfan syndrome have been excellent during the first decade. Valve function appears to deteriorate with time in some patients, which may reflect imperfect techniques or incorrect surgical adjustments of the various components of the aortic root during these reconstructive procedures. Reimplantation of the aortic valve may allow a more stable aortic valve function than does remodeling of the aortic root. Creation of neoaortic sinuses during aortic valve-sparing operations may enhance function and durability of the aortic valve.

References

1. Gott VL, Greene PS, Alejo DE, Cameron DE, Naftel DC, Miller DC, et al. Replacement of the aortic root in patients with Marfan's syndrome. *N Engl J Med.* 1999;340:1307-13.
2. David TE, Feindel CM. An aortic valve-sparing operation for patients with aortic incompetence and aneurysm of the ascending aorta. *J Thorac Cardiovasc Surg.* 1992;103:617-22.
3. Sarsam MA, Yacoub M. Remodeling of the aortic valve annulus. *J Thorac Cardiovasc Surg.* 1993;105:435-8.
4. Cochran RP, Kunzelman KS, Eddy AC, Hofer BO, Verrier ED. Modified conduit preparation creates a pseudosinus in a aortic valve-sparing procedure for aneurysm of the ascending aorta. *J Thorac Cardiovasc Surg.* 1995;109:1049-57.
5. Schafers HJ, Fries R, Langer F, Nikoloudakis N, Graeter T, Grundmann U. Valve-preserving replacement of the ascending aorta: remodeling versus reimplantation. *J Thorac Cardiovasc Surg.* 1998;116: 990-6.
6. Harringer W, Klaus P, Hagl C, Meyer GP, Haverich AI. Ascending aortic replacement with aortic valve reimplantation. *Circulation.* 1999;100(19 Suppl):II24-8.
7. Fleishcer KH, Nousari HC, Anhalt GH, Stone CD, Laschinger JC. Immunohistochemical abnormalities of fibrillin in cardiovascular tissues in Marfan's syndrome. *Ann Thorac Surg.* 1997;63:1012-7.
8. Fuzellier JF, Chauvaud SM, Fornes P, Berrebi AJ, Lajos PS, Bruneval P, et al. Surgical management of mitral regurgitation associated with Marfan's syndrome. *Ann Thorac Surg.* 1998;66:68-72.
9. Tambeur L, David TE, Unger M, Armstrong S, Ivanov J, Webb G. Results of surgery for aortic root aneurysm in patients with the Marfan syndrome. *Eur J Cardiothorac Surg.* 2000;17:415-9.
10. Birks EJ, Webb C, Child A, Radley-Smith MH. Early and long-term results of a valve-sparing operation for Marfan syndrome. Circulation 1999;100(19 Suppl):II29-35.
11. Leyh RG, Schmidtke C, Siever HH, Yacoub MH. Opening and closing characteristics of the aortic valve after different types of valve-preserving surgery. *Circulation.* 1999;100:2153-60.
12. Grande-Allen KJ, Cochran RP, Reinhall PG, Kunzelman KS. Recreation of sinuses is important for sparing the aortic valve: a finite element study. *J Thorac Cardiovasc Surg.* 2000;119:753-63.
13. De Paepe A, Devereuz RB, Dietz HC, Hennekam RC, Pyeritz RE. Revised diagnostic criteria for the Marfan syndrome. *Am J Med Genet.* 1996;62:417-26.
14. David TE. Remodeling of the aortic root and preservation of the native aortic valve. *Op Tech Cardiac Thorac Surg.* 1996;1:44-56.
15. David TE. Aortic valve surgery. *Curr Probl Surg.* 1999;36:421-504.
16. Hatle L, Angelsen B. Aortic regurgitation. In: Hatle L, Angelsen B. Doppler ultrasound in cardiology. 1st ed. Malvern (PA): Lea & Febiger; 1982. p. 154-62.
17. Perry GJ, Helmcke F, Nanda NC, Byard C, Soto B. Evaluation of aortic insufficiency by Doppler color flow mapping. *J Am Coll Cardiol.* 1987;9:952-9.
18. Murdoch JL, Walker BA, Halpern BL. Life expectancy and causes of death in the Marfan syndrome. *N Engl J Med.* 1972;286:804-8.
19. Silverman DI, Burton KJ, Gray J. Life expectancy in the Marfan syndrome. *Am J Cardiol.* 1995;75:157-60.
20. Gott VL, Cameron DE, Alejo DE, Greene PS, Shake JG, Caparrelli DJ, et al. Aortic root replacement in 271 Marfan patients: a 24-year experience. *Ann Thorac Surg.* 2002;73:438-43.
21. Butchart EG, Li HH, Payne N, Buchan K, Grunkemeier GL. Twenty years' experience with the Medtronic Hall valve. *J Thorac Cardiovasc Surg.* 2001;121;1090-100.
22. Lim KH, Caputo M, Ascione R. Prospective randomized comparison of Carbomedics and St Jude Medical bileaflet mechanical heart valve prostheses: an interim report. *J Thorac Cardiovasc.* Surg 2002;123: 21-32.
23. Birks EJ, Webb C, Child A, Radley-Smith R, Yacoub MH. Early and long-term results of a valve-sparing operation for Marfan syndrome. *Circulation* 1999;100(19 suppl):II29-35.
24. De Paulis R, De Matteis GM, Nardi P, Scaffa R, Colella DF, Bassano C, et al. One-year appraisal of a new aortic root conduit with sinuses of Valsalva. *J Thorac Cardiovasc Surg.* 2002;123:33-9.

Discussion

Sir Magdi Yacoub *(London, United Kingdom).* I congratulate de Oliveira and colleagues from Toronto for an excellent presentation and thank them for sending me the text for review. This report concerns a single-center retrospective study of patients undergoing either repair or replacement of the aortic root. The authors reported what they described as excellent survival for both the repair and the replacement groups, with 96% and 87% estimated survivals at 10 years, respectively. They also confirmed that patients undergoing replacement did have complications inherent to the use of artificial valves, even though the incidence was slightly lower than in isolated valve replacement, and they speculated that this might be due to the absence of knots inside the aorta. They then compared the two types of valve-sparing operations and found that there were no differences in survival or valve function, at least in the text that I saw, but there was a difference in terms of a tendency toward dilatation of the sinuses, but not so much the annulus, after the remodeling operation. I have several questions and one comment.

The first question relates to the pattern of survival, with 10-year survivals of 96% and 87%. Although statistically there was not a significant difference, the lack of significance was probably, as indicated in the text, due to the small number of repair patients, with only 2 patients followed up for 10 years. If this trend were to continue, however, it might become significant, and we do look forward to further reports about this. The second question related

to the pattern of survival is whether de Oliveira and colleagues have considered, in view of the very young age of these patients, comparing this pattern of survival with age-matched control subjects from the general population?

The next question relates to the indication for operation. What were the incidences of acute and chronic dissection in this series? This could have an important bearing on both the early and long-term results. In a previously reported series of patients with Marfan syndrome undergoing repair at our center, we observed an incidence of 35% of dissection, which did indeed influence both the early and long-term results. So I would like to know whether de Oliveira and colleagues looked at that. Equally, did they consider prophylactic repair, particularly in view of the very good results, for high-risk patients with aneurysms of the aortic root before there is significant regurgitation? Prophylaxis might be particularly relevant for patients known to be at high risk, as indicated by several prognostic indicators. This could avoid dissection, which is catastrophic, both in the early and longer term. It could also, importantly, halt the progressive changes in the cusps, because aortic regurgitation has been shown to increase the amount of matrix metalloproteinases, which cause the cusps to shrink. What was the cutoff point in terms of size, if prophylactic repair was indeed used?

Another point has to do with endocarditis. The authors stated that there was no incidence of endocarditis and that there were no reoperations in the repair group, but then somewhere else in the article they did mention that a patient with repair required replacement of the root. How do they reconcile those two statements? Also, what was the indication for the use of a prosthetic valve, a homograft, or an unstented xenograft? Was there a rationale behind the choice?

In a larger series from our center of remodeling operations, we have confirmed the results of this study. In particular, we have seen that the results depend critically on timing of the operation but equally, as de Oliveira and colleagues stress in the article, on technical details. For example, during the last 4 years or so we have introduced several technical refinements in the remodeling operation. The first is undersizing of the graft, which we think is really important; the second is insertion of the top of the commissures within the Dacron polyester fabric graft. The third, which is probably the most important, is in the length and shape of the tongues of the Dacron polyester fabric tube, which we think should be as long as possible and tapering at its end. This has the effect of remodeling of the sinuses. By the way, all the diseased aortic wall is excised in the remodeling operation. This is essential, and the stitches have to be in the annulus. But a thin, long tongue would reshape the sinuses and form a vortex, which in effect pushes the bottom of the sinuses inward.

Dr de Oliveira. Starting with the first question, yes, we believe that with a larger sample size the difference in survival between root replacement and valve sparing operations will become significant. The lower risk of infective endocarditis and the absence of anticoagulation therapy after preservation of the native valve are important factors associated with lower morbidity and mortality after valve-sparing operations.

We did not attempt to match the patients in this study with the general population to compare the long-term survival because of the relatively small sample size.

As far as aortic dissection is concerned**,** there were 15 cases of acute aortic dissection; 9 of these patients had undergone valve-sparing operations with reimplantation of the aortic valve and 6 had undergone root replacement. We believe that reimplantation of the aortic valve gives a more stable repair and is associated with a lower risk of postoperative bleeding. There were 4 chronic dissections, and these patients all had undergone root replacement.

Regarding prophylactic surgery in high-risk patients, such as female patients during child-bearing age and patients with a family history of aortic dissection, we tend to be more aggressive and to recommend repair when the root reaches 45 mm, whereas we use 50 mm to recommend surgery in other patients. We should try to avoid dissection and aortic complications in this very young group of patients because of the negative effect on operative mortality and long-term survival. In most series, including ours, an important cause of late death has been rupture of the false lumen. In our series 4 patients required reoperation on the residual aorta because of expansion or rupture of the false lumen. One patient died, and 1 had paraplegia develop.

With respect to the issue of infective endocarditis, there were 5 episodes of endocarditis: 4 after root replacement and 1 after remodeling. The latter occurred 11 years after the operation. That is why the freedom from endocarditis was 100% at 10 years.

Finally, I can only speculate on the indications for using homograft or bioprosthetic valves. They probably were used because the patients did not wish to take anticoagulant. I thank you again for your comments and questions.

Dr Hartzell V. Schaff *(Rochester, Minn).* Technique is terribly important, but did you look at the outcome related to the degree of preoperative aortic valve regurgitation? What degree of preoperative aortic regurgitation would make you hesitant to try to preserve the valve?

Dr de Oliveira. There was a correlation between the size of the aortic root and the probability of preserving the valve. If the root was less than 5 cm in diameter, most cusps were normal. If the root was between 5 and 6 cm, about 50% of them were normal. If the aneurysm was more than 6 cm, then most cusps were abnormal, and those patients were more likely to undergo aortic root replacement. Is that the answer to your question?

Dr Schaff. Not directly. The question is the degree of aortic valve regurgitation, understanding that the larger annuli will have more leakage. Is there a degree of aortic valve regurgitation about which you would worry?

Dr de Oliveira. We did not look at that specifically, but I believe that the more severe the degree of AI, the less the chance to repair the valves. The cusps will more likely become thinner and fenestrated, making the repair more difficult and risky. However, 6 of our patients had prolapse of one cusp and yet were amenable to repair with good results.