Valve-Sparing Aortic Root Replacement in Marfan Syndrome

Duke E. Cameron and Luca A. Vricella

Marfan syndrome is the most common inherited connective tissue disorder, affecting approximately 1 in 10,000 live births. The cardinal features of Marfan syndrome are the abnormalities of the skeleton (tall stature, arachnodactyly, and joint hyperelasticity), eye (lens subluxation), and aorta (root aneurysm with proclivity toward rupture and dissection). Aortic catastrophe accounts for most of the premature mortality among Marfan patients, a risk that climbs steeply during adolescence and results in death of half of Marfan patients by the age of 40 years. Most of the improvement in life expectancy that has been achieved in Marfan syndrome is attributable to early recognition of aortic root aneurysms and prophylactic replacement with composite grafts (mechanical valve prostheses within Dacron conduits) before rupture or dissection occurs. Despite the excellent early and late results with composite grafts, there has been growing interest in operative procedures that replace the sinuses but preserve the aortic valve leaflets, to avoid anticoagulation and minimize the risk of prosthesis-related endocarditis. These procedures are still in evolution and late results are not yet known, but as with mitral repair in the setting of myxomatous disease, valve-sparing procedures in Marfan syndrome have weathered a storm of initial criticism and skepticism and are steadily gaining acceptance.

Marfan syndrome was originally described as a skeletal malformation in 1897. Nearly 30 years passed before the associated aortic aneurysm was recognized, and it is not widely appreciated that Helen Taussig provided one of the first accounts of death from aortic catastrophe associated with Marfan syndrome. McKusick offered the comprehensive characterization in the mid 1950s and developed the Marfan clinic at Johns Hopkins where a prophylactic aortic replacement program was begun in the 1970s. The first full aortic root replacement is generally credited to Bental and DeBono, who in 1968 fashioned a composite prosthesis using a plastic conduit and mechanical valve at the operating room table to replace an aortic sinus aneurysm and regurgitant valve in a young woman. The success of this procedure dramatically changed the outlook for Marfan syndrome patients with root aneurysms and led to an aggressive program of prophylactic aortic root replacement for patients for whom risk of rupture was estimated to exceed risk of operation.

Low operative and late mortality were achieved by many centers throughout the world over the next 20 years. Further refinement came with introduction of valve-sparing procedures, independently by Tirone David and Sir Magdi Yacoub. In the David I (reimplantation procedure), the aortic sinuses are excised and a Dacron cylinder is slid down and around the aortic valve complex (Fig 1). It provides superior hemostasis and stabilizes the anular diameter but in its original form sacrificed sinuses of the root. Because of concerns that the lack of sinuses would lead to valve leaflet damage, the David II (remodeling) procedure was introduced, though it was performed earlier by Yacoub. In this procedure, the sinuses are excised and a Dacron tube is trimmed to have three tongue-like extensions that are sewn to the annulus of the aortic valve. This technique provided sinuses, but lacked anular stabilization, and in some cases progressive anular dilatation led to late valvar incompetence. Various technical modifications to stabilize the annulus with external prosthetic strips (David III) were tried with mixed results. The David IV and V procedures used oversized grafts that are plicated at the anulus and sinotubular ridge to create a bulging sinus proximally but with anular stabilization. The Valsalva graft (CarboMedics, Austin, TX) is a commercially available graft with prefabricated sinuses.
Because the surgical techniques used for valve-sparing surgery have evolved continuously over the last 12 years, it is difficult to be certain of the results of current operative approaches.\textsuperscript{7} The issues surrounding this surgery have been eloquently summarized recently by Miller.\textsuperscript{8} Because composite grafts have set high standards for surgical outcomes, valve-sparing procedures have been adopted cautiously, and certain key issues, such as the integrity of Marfan aortic valve leaflets and the value of preserving sinuses, remain unaddressed, and will only be defined by long-term clinical outcome studies.

**Indications for Surgery**

The current indications for aortic root replacement in children at our institution include aneurysm diameter $>5$ cm, aneurysm diameter increasing $>1$ cm/year, and progressive aortic valve insufficiency.\textsuperscript{9} In children less than 12 years of age, we have not used Z-scores per se as a threshold for surgery, because we believe rupture and dissection are rare in this age group. Surgical indications for young children thus typically are “giant” aneurysms (those that satisfy adult criteria for intervention) and rapid enlargement over time with progressive valvar insufficiency.

**Operative Techniques**

Our initial experience used the remodeling (Yacoub/David II) procedure because we believed the preservation of sinuses would be important to long-term valve function. Several cases of late aortic valve insufficiency necessitating aortic

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**Figure 1** Reimplantation and remodeling procedures. (Reprinted with permission, top,\textsuperscript{4} bottom,\textsuperscript{5})
Figure 2  Operative view of aortic sinus aneurysm typical of Marfan syndrome, with rapid tapering of ascending aorta to normal diameter.

Figure 3  After institution of cardiopulmonary bypass, aortic cross clamping, and cardioplegic arrest, the aortic sinuses are excised and the coronary arteries widely mobilized. Dissection outside the aorta must be to a level just below the annulus. Stay sutures are invaluable for optimal exposure.
Figure 4  Three non-pledgetted horizontal mattress sutures of 2-0 braided polyester are passed from inside outward, below the annulus at the nadir of each of the three leaflets. These sutures will secure the lower end of aortic root graft at the bottom of the aortic root complex.

Figure 5  The Depaulis “Valsalva” graft is trimmed; usually 2 to 3 horizontal rings are preserved at the bottom end, and 4 to 5 at the top end.
valve replacement led us to reconsider the reimplantation (David I) procedure at the same time as the Valsalva graft became commercially available. This graft has preconstructed sinuses, and when used in the reimplantation procedure, seemed to provide the advantages of anular stabilization as well as sinus preservation. This has been our preferred prosthesis and procedure since May 2002.

Full median sternotomy, in our opinion, is still the optimal surgical approach for safe conduct of the operation. Intraoperative transesophageal echocardiography, blood cardioplegia, and aprotinin are used routinely. Bicaval venous cannulation, with the superior vena cava cannula directed up via the right atrial appendage improves exposure of the root by angling all venous cannulae away from the surgical field and retracting the right atrium. We also routinely inspect the atrial septum for patent foramen ovale and close it when present, which it is in approximately one third of cases.

Because most Marfan aneurysms still have competent aortic valves, even when the root assumes giant dimensions, cardioplegia is administered directly into the aortic root and redosed directly into the coronary ostia every 20 to 30 minutes using soft silastic tipped cardioplegia cannulae. If there is moderate or severe regurgitation, our preference is to cool the patient on cardiopulmonary bypass until ventricular fibrillation, and then crossclamp and open the aorta and deliver cardioplegia directly into the coronary ostia. In our practice, continuous topical cold saline is run over the heart for additional myocardial protection.

After cardioplegic arrest, the aorta is divided just above the sinotubular ridge (Fig 2). Usually a short length of mid ascending aorta is also resected. We do not routinely replace the distal ascending aorta or arch. If the aortic valve is competent, we strive to preserve or slightly reduce the size of the sinotubular diameter. The diameter is measured using valve

Figure 6 The three subanular sutures are passed through the bottom of the graft, 120° apart from one another, and the three stay sutures at the top of the commissures are pulled up within the graft.
sizers, and a Valsalva graft is chosen that is 2 to 3 mm larger, because it will fit outside the reconstructed valve complex. In adolescents and adults, 28 and 30 mm are the most common size grafts used. Measurement of the aortic annulus diameter is performed and noted, but the graft is usually several millimeters bigger than the annulus, and is plicated at the base during the course of the replacement.

Extensive dissection at the base of the aorta is carried out, optimally to a level below the annulus (Fig 3). This is straightforward in the non-coronary and left coronary sinuses, but the right coronary sinus dissection is limited by the fibrous continuing between the aorta and pulmonary artery at the left-right commissure, and the thin membranous septum below the right noncoronary commissure. In each of these ar-

Figure 7 Three 4-0 polypropylene pledgetted horizontal mattress sutures are used to fix the top of each commissure post to the sinotubular ridge of the graft. Additional subanular sutures can also be placed at this time, working within the graft.

Figure 8 Continuous 4-0 polypropylene sutures are used to fix the annulus and sinus remnant within the graft. It is easiest to start at the bottom of each sinus and sew upward to the commissure posts.
eas, aggressive dissection may lead to pulmonary artery injury or entrance into the right ventricle. The dissection can be limited in both regions, and small vertical incisions can be made in the base of the Valsalva graft to facilitate seating of the graft low in the root.

The sinus tissue is excised next, leaving 4 to 5 mm of sinus remnant attached to the annulus. The coronary arteries are harvested with several millimeter of collar, and then mobilized widely. Stay sutures are placed at the tops of the commissure posts and the coronary buttons to facilitate exposure of the root and maintain proper orientation of the coronaries.

Three 2-0 braided polyester horizontal mattress sutures are passed under the annulus at the nadir of each of the three leaflets and brought outside the base of the aorta (Fig 4). At this point, a Valsalva graft is prepared by trimming the bottom collar of the graft to about 2 to 3 rings. The distal end of the graft is also shortened to about 4 to 5 rings (Fig 5). If a more extensive replacement of the ascending aorta or arch is required, the additional distal end of the graft is used, but it is still necessary to transect the graft at this point, because suturing of the valve to the inside of the graft is greatly facilitated by working with a short (shallow) graft. The Valsalva graft has three black markings that divide the circumference into three sinuses 120° apart. It is useful to align these three black lines with the commissures and further subdivide each sinus in half with a purple surgical marking pen to assist coronary location and assignment of the nadir of each sinus. We have also found it helpful to notch the bottom of the graft for 5 mm at the locations corresponding to the left-right and right-noncoronary commissures; this will help the graft slide down low enough over the aortic valve complex.

The three subanular sutures are now passed through the bottom of the graft, inside out, and the commissural stay sutures drawn up through the graft, which is lowered down around the valve (Fig 6). The three subanular sutures are now tied. Three 4-0 polypropylene pledgetted mattress sutures fix the commissural posts at the sinotubular junction of the graft (Fig 7). The skirt of the graft has longitudinal pleats and can adapt to a wide range of leaflet lengths and therefore sinus heights. At this point, with three subanular sutures placed and three commissural post sutures placed (“three below and three above”), the valve is oriented properly within the graft. Additional subanular sutures can now be placed, following the curvilinear course of the annulus, avoiding the membranous septum anteriorly. This technique makes it easier to place the sutures along the true course of the annulus rather than in one horizontal plane corresponding to the sinus nadirs, and eliminates guesswork as to where the sutures should exit the graft when all subanular sutures are placed before passing them through the graft. However, because these sutures are not the hemostatic ones, we recently simplified the technique to include only these three subanular sutures, adding others only if there is substantial oversizing of the graft beyond the anular diameter, in which case the additional sutures will plicate the graft down to the anular size.

Working within the graft, we run continuous 4-0 polypropylene sutures to approximate the sinus remnant and annulus to the Dacron (Fig 8). Because this is the hemostatic layer, it is important to complete this suture line accurately and precisely. It is convenient to start at the bottom of each sinus and sew up to the commissure post and tie the suture to the adjacent sinus suture outside the graft. When this step is completed, assessment of valve competence is now possible. Occasionally it has been necessary to perform leaflet repairs, but this has been rare in our experience. We believe that a
prolapsed leaflet is best treated by leaflet plication at the free edge of the mid-leaflet using a fine braided polyester suture and small pericardial pledget. Commissural resuspensions ("Trusler valvuloplasty") lead to leaflet asymmetry and involve suturing what is often the weakest area of the valve given the frequency of "stress fenestrations" seen at the commissure in Marfan syndrome. We do not regard these fenestrations as absolute contraindication to valve-sparing operations, but should probably be considered a risk factor for late valve failure.

The notches created in the graft are next reapproximated with pledgetted 4-0 polypropylene to prevent late graft splaying.

Holes are cut in the graft opposite the coronary arteries and each coronary button encircled with a Teflon felt "lifesaver" ring pledget for hemostasis and prevention of late anastomotic pseudoaneurysm (Fig 9). The left coronary anastomosis is completed first with 4-0 polypropylene. Many surgeons prefer to perform the right coronary anastomosis after the aortic anastomosis is completed and the crossclamp briefly released to find the optimal location of the anastomosis. We prefer to perform the right coronary anastomosis with the aorta still open to minimize risk of injury to the aortic valve. If the coronary artery is widely mobilized and the site of anastomosis is as far anterior as possible (limited only by the anterior commissure) and just proximal to the sinorubular ridge of the graft, anastomotic tension and torsion are avoided.

Finally, the distal graft-to-aorta anastomosis is completed with continuous 4-0 polypropylene (Fig 10). External felt strip reinforcement is used routinely. A left ventricular vent is inserted through the right superior pulmonary vein unless there is complete confidence in valve confidence.

The aortic crossclamp is removed and the heart resuscitated and desired. Residual aortic incompetence is assessed by intraoperative transesophageal echocardiography and left ventricular vent flow. Residual incompetence of 0 to 1+ is satisfactory but 2+ or more is not, especially if it is caused by leaflet prolapse because it is a progressive lesion. In the event of unacceptable valve incompetence, aortic re-crossclamping is necessary to assess whether leaflet repair can salvage the procedure. If the valve is not repairable and replacement is necessary, the native valve can be excised and a prosthesis sewn within the Valvula graft to minimize additional cross clamping. Alternatively, full root replacement with a composite graft, homograft, or porcine bioprosthetic root can be carried out. Often this decision rests on whether there is substantial root bleeding, in which case re-do root replacement is preferred.

Postoperatively, patients are maintained on aspirin for 1 month and Beta-blocker medications indefinitely. Follow-up consists of annual echocardiograms and annual or biennial

![Figure 10](image_url) The distal graft-to-mid ascending aorta anastomosis is completed with 4-0 polypropylene and external felt strip reinforcement.
computed tomography or magnetic resonance imaging of the remainder of the aorta for late dilatation or dissection.

Results

Operative mortality for valve-sparing procedures has been 0% to 5% at several centers throughout the world, and there appears to be no significant difference between Marfan and non-Marfan patients.10-14 Most series include Marfan patients, but these patients constitute a minority of the cohort. Because these patients are among the most challenging cases, with respect to reoperation rates for late valve failure, the proportion of Marfan patients should always be considered when comparing results.15-17

Rates of thromboembolism and endocarditis in general have been lower than with composite grafts, but reoperation rates at 8 to 10 years are approximately 5% to 10%. Many centers have reported substantially lower late failures with reimplantation rather than remodeling procedures, probably because of the anular stabilization provided by the former. There are no clinical studies yet that show clinical superiority of sinus preservation. In general, infants have had higher reoperation rates, as well as concomitant mitral disease, and their long-term prognosis is guarded.

Conclusion

Valve sparing aortic root replacement can be applied to children and adults with the Marfan aortic root aneurysms with low operative risk similar to root replacement with conventional prostheses. The incidence of late endocarditis and thromboembolism are low in both valve-sparing and replacement strategies, and complications with long-term anticoagulation in Marfan patients have been rare. Valve-sparing operations suffer from at least a 10% late valve failure rate, but this rate may now be lower with current operations that stabilize the annulus diameter and preserve anatomic sinuses. Marfan infants with aortic and cardiac disease continue to be a problematic group because the tissue defect appears more severe and reoperation rates are high.

References