Valve-sparing aortic root surgery in children and adults with congenital heart disease

Edward Buratto, MBBS, PhD,a,b,c and Igor E. Konstantinov, MD, PhD, FRACSa,b,c,d

Aortic root surgery in patients with congenital heart disease was traditionally reserved for children with connective tissue disorders.1-3 In recent years, there has been an increasing number of patients, especially those with repaired conotruncal anomalies, who develop aneurysmal aortic root dilatation with or without concomitant aortic valve insufficiency.4,5 However, rupture and dissection of the aorta are exceedingly rare in childhood,6,7 and as such the precise criteria for aortic root surgery have not been defined.3,4 Although valve-sparing aortic root replacement has been the gold standard for decades,1-3 there is a newer approach combining replacement of the ascending aorta and external root stabilization, as well as a renewed interest in external support of the entire root,8-10 which have evolved from previously described aortic root wrapping.11 Of note, stabilization of the ascending aorta and entire aortic root can be performed without cardiopulmonary bypass,9,10 and if its effectiveness in root stabilization and reduction of aortic insufficiency is demonstrated, could further modify indications for aortic root surgery. The purpose of this focused review is to highlight key references on the current state of aortic root surgery in patients with congenital heart disease.

INDICATIONS FOR SURGERY

Aortic root surgery may be required because of progressive aortic regurgitation (AR) or prophylactically to prevent potential dissection and rupture of the aneurysmal aorta. The indications for aortic root surgery in children are elusive, derived from adult guidelines,12,13 and mainly focused on those with connective tissue disorders.2 The current guidelines are not data-driven or evidence-based, but rather derived from those developed for adults.14 In adults, to account for variation in patient size, aortic cross-sectional area has been indexed to height (Svensson’s index) to predict the risk of aortic dissection.15,16 Subsequently, the simplified aortic diameter to height ratio was found to be predictive of dissection, rupture, and death.17 Although the Svensson’s index has been shown to correlate well with aortic z-scores in children,18 it has not been validated for the prediction of dissection and rupture in children or adults with repaired congenital heart disease. When should we perform aortic root surgery in children without significant aortic insufficiency? Should we replace the aorta in children with an aneurysmal root when an adult-size graft can be placed? Should the aneurysmal aorta be replaced when it compresses adjacent structures even though the aortic size threshold has not been reached? What is the risk of aortic dissection or rupture in patients with repaired congenital heart disease? None of these important questions can be answered by the data derived from the current literature. Yet, with an ever-increasing population of children and adults with repaired congenital heart disease we will inevitably face these questions more often. In children, the largest study published to date described the outcomes of 100 patients, of whom 90% had connective tissue disorders, and provided institutional guidelines for aortic root replacement.2 At the present time, this study provides the best guidance on indications for aortic dissection.

From the aDepartment of Cardiac Surgery, Royal Children’s Hospital, Melbourne, Australia; bDepartment of Paediatrics, University of Melbourne, Melbourne, Australia; cHeart Research Group, Murdoch Children’s Research Institute, Melbourne, Australia; and dMelbourne’s Centre for Cardiovascular Genomics and Regenerative Medicine, Melbourne, Australia.
Received for publication June 25, 2020; revisions received Aug 4, 2020; accepted for publication Aug 11, 2020; available ahead of print Sept 28, 2020.
Address for reprints: Igor E. Konstantinov, MD, PhD, FRACS, Royal Children’s Hospital, Flemington Rd, Parkville 3052, Australia (E-mail: igor.konstantinov@rch.org.au).

J Thorac Cardiovasc Surg 2021;162:955-62
0022-5223/$36.00
Crown Copyright © 2020 Published by Elsevier Inc. on behalf of The American Association for Thoracic Surgery
https://doi.org/10.1016/j.jtcvs.2020.08.116
root surgery in children with connective tissue disorders and nonsyndromic congenital heart disease. Current surgical indications are summarized in Table 1.

**Patients With Connective Tissue Disorders**

Currently, surgery is recommended for asymptomatic children with connective tissue disorders (Figure 1, A) and a maximum aortic diameter greater than 5.0 cm in children with Marfan syndrome or greater than 4.5 cm in those with additional risk factors (family history dissection or rupture) or significant aortic valve insufficiency, whereas in children with Loey-Dietz syndrome (LDS), the threshold of aortic root diameter is decreased to greater than 3.5 cm or an aortic Z-score greater than 3. Rapid progression of the aortic root dilatation of greater than 0.5 cm per year appears to indicate aortic root surgery in any patient with connective tissue disorder.1,3

**Patients With Repaired Congenital Heart Defects**

There is now an ever-growing population of children and adults with repaired congenital heart disease who have an enlarged aortic root. It is not unusual to see patients after repair of conotruncal anomalies, particularly truncus arteriosus, tetralogy of Fallot, transposition of the great arteries (Figure 1, B and C), with asymptomatic aortic root aneurysm.4 In patients without significant aortic valve insufficiency, indication for surgery is based on adult criteria, namely, maximal aortic root diameter greater than 5.5 cm in both bicuspid aortic valve and nonsyndromic aortic aneurysms.2

Neoaoortic dilatation may also occur in patients who have undergone the Ross procedure (Figure 1, D). Autograft dilatation is more common in growing children. In older children and adults, the autograft root may be stabilized, thus reducing the rate of late dilatation.21,22 Furthermore, in the reoperative setting, the rate of autograft failure appears to be decreased, possibly due to postsurgical adhesions present prior to sternotomy that may provide an additional extrinsic support for the autograft.23 Attempts were made to predict autograft dilatation using proteomic technology.24 Although such prediction appears elusive at the present time, it may become useful with further technologic refinement in the future.25 Contemporary studies demonstrate that when the Ross operation is performed in childhood, the freedom from autograft reoperation is approximately 80% at 15 years,21 whereas when the Ross operation is performed in adults, the freedom from autograft failure is 96% at 15 years.22 Although described in a single case report, aortic dissection after the Ross procedure is exceptionally rare.26 Thus, autograft reoperation is mostly performed for aortic insufficiency. Rarely, aortic root replacement for autograft dilatation has been performed without aortic insufficiency for neoaoortic root dilatation alone, based on autograft diameter greater than 5 cm or Svensson’s index greater than 10 in adults.27,28 Does neoaoortic root dilatation alone in children without significant aortic valve insufficiency after, for example, the Ross procedure or arterial switch operation warrant aortic root surgery? If so, then at what size? These questions are yet to be answered.

As the survival of children with univentricular physiology is improving, there is also a growing population with enlarged neoaoortic roots (Figure 1, E) after univentricular palliation. Neoaoortic root dilatation and insufficiency appear to progress over time.29 The exact indication based on the aortic root diameter in these patients is even more obscure. Thus, aortic root surgery in patients with repaired congenital heart disease is typically performed for significant aortic, or neoaoortic, valve insufficiency or compression of the adjacent structures by the aneurysm30 and the exceptionally rare event of dissection.7

**Surgical Techniques**

Valve-sparing surgical techniques that have been applied to aortic root dilatation are wrapping of the ascending aorta (Figure 2, A), replacement of aortic root using reimplantation (Figure 2, B), which appears to be the gold standard, or remodeling (Figure 2, C), replacement of the ascending aorta with root stabilization (Figure 2, D), or personalized external root support (PEARS) (Figure 2, E).

The earliest valve-sparing approach to the aortic root appears to be the wrapping technique described by Robicsek and colleagues,31 in which an aortoplasty is performed with elliptical excision and reapproachimation of the ascending aortic wall. The ascending aorta is then wrapped with a vascular prosthesis, with small semicircles excised above the takeoff of the coronary arteries to avoid encroachment of the coronary orifices.32

**TABLE 1. Indications for aortic root replacement**

<table>
<thead>
<tr>
<th>Condition</th>
<th>Reported indications for surgery</th>
</tr>
</thead>
<tbody>
<tr>
<td>Marfan syndrome</td>
<td>Diameter &gt;5.0 cm</td>
</tr>
<tr>
<td></td>
<td>Growth &gt;0.5 cm/y</td>
</tr>
<tr>
<td></td>
<td>Svensson’s index &gt;10</td>
</tr>
<tr>
<td></td>
<td>Diameter &gt;4.5 cm if family history of AR</td>
</tr>
<tr>
<td></td>
<td>Diameter &gt;4.0 cm if concomitant surgery</td>
</tr>
<tr>
<td></td>
<td>Z score &gt;3-4</td>
</tr>
<tr>
<td>Loey-Dietz syndrome</td>
<td>Growth &gt;0.5 cm/y</td>
</tr>
<tr>
<td>All patients</td>
<td>Svensson’s index &gt;10</td>
</tr>
<tr>
<td>Type I and II</td>
<td>Diameter &gt;3.5 cm or Z score 3</td>
</tr>
<tr>
<td>Type III</td>
<td>Diameter &gt;4 cm or Z score &gt;4</td>
</tr>
<tr>
<td>Type IV</td>
<td>Diameter &gt;4.5 cm or Z score &gt;4</td>
</tr>
<tr>
<td>Bicuspid valve</td>
<td>Diameter &gt;5.5 cm</td>
</tr>
<tr>
<td></td>
<td>Diameter &gt;4.5 cm if concomitant surgery</td>
</tr>
<tr>
<td></td>
<td>Growth &gt;0.5 cm/y</td>
</tr>
<tr>
<td></td>
<td>Svensson’s index &gt;10</td>
</tr>
<tr>
<td>Ross procedure</td>
<td>Diameter &gt;5.0 cm</td>
</tr>
<tr>
<td></td>
<td>Svensson’s index &gt;10</td>
</tr>
<tr>
<td>Nonsyndromic thoracic aortic aneurysm</td>
<td>Diameter &gt;5.5 cm</td>
</tr>
</tbody>
</table>

AR, Aortic regurgitation.
In the current era, aortic root replacement has become the gold standard approach to valve-sparing aortic root surgery in both children and adults. There are 2 widely used techniques for valve-sparing root replacement: aortic root reimplantation (David procedure) and aortic root remodeling (Yacoub procedure). In the reimplantation technique, the aortic valve is sutured within an appropriately sized vascular graft, which supports the entire valve apparatus to a level below the nadir of each cusp. This technique can be used in patients with repaired congenital heart disease and has been reported in patients with univentricular palliation, prior arterial switch operation, and repaired tetralogy of Fallot in the setting of acute dissection. In the remodeling technique, a scalloped vascular graft is sutured to the rim of retained aortic wall on the aortic valve. This technique can be used in patients with repaired congenital heart disease and has been reported in patients with univentricular palliation, prior arterial switch operation, and repaired tetralogy of Fallot in the setting of acute dissection. In the remodeling technique, a scalloped vascular graft is sutured to the rim of retained aortic wall on the aortic valve. In this technique, the aortic annulus is not supported within the graft. To avoid the risk of annular dilatation, an annuloplasty technique has been described using polytetrafluoroethylene suture (Gore-Tex CV-0; W.L. Gore and Associates, Newark, Del) or an aortic annuloplasty ring placed externally (Extra-Aortic; CORONEO Inc, Montreal, QC, Canada) or internally (BioStable Science and Engineering, Inc, Austin, Tex). Alternatively, the similar stabilization of the annulus can be achieved by circular strip of any synthetic material placed externally around the aortic annulus at the time of root remodeling.

More recently, newer techniques of external aortic root support have evolved from the concept originally described by Robicsek. The Florida sleeve technique involves placement of a vascular graft around the aortic root, with fenestrations, referred to as “keyholes,” for the coronary arteries. The graft is secured at the annular level, externally supporting the aortic root, and the supracoronary aorta is replaced with a graft. The PEARS technique involves wrapping the aorta with a customized graft, which is secured, at the annular level, and is designed to be incorporated into the aortic wall. This procedure is unique in that it is an entirely extravascular prosthesis, which can be placed on the beating heart, and in most cases without the use of cardiopulmonary bypass.

RESULTS

Connective Tissue Disorders

Fraser and colleagues reported the results of 100 patients, predominantly with Marfan syndrome (51%) or LDS (39%), who underwent valve-sparing root replacement from 1997 to 2017 at a median age of 13.6 years. The mean preoperative aortic sinus diameter was 4.4 cm, and the mean z-score was 7.3. Most patients underwent reimplantation (84%), with the remainder receiving remodeling (13%) or the Florida sleeve (3%). Early mortality was 2%, and 10-year survival was 88%.

FIGURE 1. Aortic root aneurysms in a spectrum of congenital heart disease.

FIGURE 2. Types of valve-sparing aortic root surgeries in congenital heart disease.
Freedom from reintervention was 70% at 10 years, and the risk of reoperation for aortic valve replacement was significantly higher with remodeling than reimplantation (31% vs 5%, \( P = .001 \)). Furthermore, they abandoned the use of the Florida sleeve procedure, because all 3 patients who underwent this technique required reoperation with the reimplantation technique.

Patel and colleagues\(^{17} \) reported 31 patients with LDS who underwent valve-sparing root replacement between 1992 and 2015 at a mean age of 10 years. In most cases, they used the reimplantation technique (27/31, 87.1%) followed by the Florida sleeve (3/31, 9.7%) and the remodeling technique (1/31, 3.2%). The mean preoperative aortic sinus diameter was 4.0 cm, and the mean z-score was 7.8. There were no early deaths, and survival at 10 years was 95%. Freedom from reintervention on the aorta was 54% at 10 years.

Kluin and colleagues\(^{18} \) reported 19 patients, predominantly with Marfan syndrome, who underwent valve-sparing root replacement between 2003 and 2015 at a median age of 13.2 years. The mean preoperative aortic sinus diameter was 4.4 cm, and the mean z-score was 4.9. There were no early deaths and a single late death (5.3%) with a mean follow-up of 4.4 years. Freedom from greater than mild AR was 85% at 10 years.

Roubertie and colleagues\(^{19} \) reported 11 patients, predominantly with Marfan syndrome, who underwent valve-sparing root replacement with the remodeling technique between 1986 and 2007 at a mean age of 10 years. The mean aortic sinus diameter was 4.7 cm, with a mean z-score not reported. There was 1 early death (9.1%), and freedom from reoperation at 5 years was 46%. They cautioned against the use of the remodeling technique in children because of the high reoperation rate.

Rakhra and colleagues\(^{20} \) reported 10 patients who underwent valve-sparing aortic root replacement from 2002 to 2011 at a median age of 15 years. The mean aortic sinus diameter was 4.8 cm, with a mean z-score of 5.9. There were no operative or late deaths at a median follow-up of 4.1 years. Redo aortic root surgery was required in 1 patient for AR and 1 patient for infective endocarditis.

Aalaei-Andabili and colleagues\(^{21} \) reported 37 patients with Marfan syndrome who underwent the Florida sleeve procedure between 2002 and 2014 at a mean age of 37 years. The mean aortic root diameter was 4.9 cm. There were no operative deaths. Survival and freedom from reoperation at 8 years were 95% and 100%, respectively.

Treasure and colleagues\(^{22} \) reported the results of 30 children and young adults who underwent PEARs between 2004 and 2011 at a median age of 28 years. The mean aortic diameter was 4.6 cm preoperatively. There were no early or late deaths and no reoperations at a mean follow-up of 4.4 years. Izgi and colleagues\(^{23} \) reported 24 patients preoperative and postoperative magnetic resonance imaging who underwent PEARs for aortic root dilatation with a diagnosis of Marfan syndrome between 2004 and 2012 at a mean age of 33 years. The mean preoperative diameter of the sinus of Valsalva was 4.5 cm. They demonstrated that at a mean follow-up of 6.3 years, the PEARs graft could stabilize the aortic root and ascending aorta, with no change in diameter over that period. However, it should be noted that as yet only small series of PEARs root stabilizations have been published, and results have not been reported in children.

Patients With Repaired Congenital Heart Defects

Myers and colleagues\(^{5} \) reported 29 patients who underwent valve-sparing root surgery between 2000 and 2012 at a mean age of 15 years with 62% (18/29) having congenital heart disease. They used aortic root remodeling in 55% (16/29) and reimplantation in 45% (13/29) of patients. The mean aortic root diameter and z-score were 4.1 cm and 6.5 for reimplantation, and 3.7 cm and 5.9 for remodeling, respectively. There were no operative deaths. Freedom from structural valve degeneration was 70% at 1 year. They found that the reimplantation technique and the use of a smaller graft to native annulus ratio were associated with increased risk of valve failure. They suggested that undersizing of the graft in the reimplantation technique could have resulted in earlier structural valve deterioration.

Stulak and colleagues\(^{4} \) reported 81 patients with a dilated ascending aorta after prior repair of conotruncal anomalies who underwent reoperation on the aortic root (9%, 7/81), aortic valve replacement 76% (62/81), aortic valve repair (12%, 10/81), or isolated ascending aorta replacement (2.5%, 2/81) between 1973 and 2008. The mean aortic diameter was 45 mm, but only 9% (7/81) underwent aortic root replacement. Although the aortic root was not replaced in most patients, the 10-year freedom from subsequent reoperation on the aorta was 100%. They concluded that moderate aortic dilatation in patients with conotruncal anomalies is common, but because dissection is rare, the moderately dilated aorta in this setting may be observed.

CONCLUSIONS

Aortic root stabilization in children and adults with congenital heart disease may be achieved by replacing or externally supporting the aortic root. Aortic root replacement with the reimplantation technique appears to provide better freedom from reoperation compared with the remodeling technique. Techniques of external support demonstrate similar results to root replacement in adults, but there has been limited experience in children so far. The indications for both aortic root surgery per se and the type of aortic root surgery in children and adults with congenital heart disease are yet to be refined.

Conflict of Interest Statement

The authors reported no conflicts of interest.
The Journal policy requires editors and reviewers to disclose conflicts of interest and to decline handling or reviewing manuscripts for which they may have a conflict of interest. The editors and reviewers of this article have no conflicts of interest.

References

Key Words: aortic root aneurysm, autograft, connective tissue disorders, conotruncal anomalies, neoaortic root, Ross operation, valve-sparing aortic root replacement
Valve-sparing aortic root surgery in children: Recent Articles from AATS Journals


**Commentary:** Everybody is different: A plea for individualizing treatment of aortopathy. Sievers HH. J Thorac Cardiovasc Surg. 2018;156(2):481-482.


**Commentary:** Height supersedes weight: Height-diameter indexing keeps you ahead of the game. Czerny M. J Thorac Cardiovasc Surg. 2018;155(5):1925.


**Commentary:** Expanding the results of the Ross operation. Mastrobboni S, El Khoury G. J Thorac Cardiovasc Surg. 2018;155(6):2398-2399.


**Commentary:** To pleat or not to pleat…is that the question? Gleason TG. J Thorac Cardiovasc Surg. 2017;153(2):239-240.


**OP TECHS:** Aortic root remodelling. Schneider U, Schäfers HJ. Oper Tech Thorac Cardiovasc Surg. 2018;23(3):102-120.

