Bigger is not always better

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Disclosures: Author has nothing to disclose with regard to commercial support.

Received for publication Feb 21, 2017; accepted for publication March 6, 2017; available ahead of print April 12, 2017.

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J Thorac Cardiovasc Surg 2017;153:1488-9

0022-5223/\$36.00

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<http://dx.doi.org/10.1016/j.jtcvs.2017.03.023>

Outcomes of the Norwood operation for palliation of hypoplastic left heart syndrome and associated single ventricle anomalies have improved because of several advances in preoperative management, surgical and perfusion techniques, and postoperative care.¹ Shunt-related complications were associated with the highest postoperative morbidity and mortality, and therefore it was natural that variations in the source of pulmonary blood flow have dominated surgical modifications and clinical investigations.² Nonetheless, aortic arch reconstruction remains the most important part of the Norwood operation, and recurrent arch obstruction has been linked to deterioration in systolic and diastolic ventricular function, worsening atrioventricular valve regurgitation, failure to progress to subsequent palliation stages, and increased interstage mortality.²⁻⁵ The reported incidence of recurrent coarctation after the Norwood operation ranges between 2% and 40%, and a recent report from the Single Ventricle Reconstruction (SVR) trial showed that arch reintervention was necessary in 18% of patients.^{4,5} Several surgical modifications have been adopted to address the recoarctation risk and included complete resection of all ductal/coarctation tissue, the extent of arch augmentation, and the use of all native tissue for aortic reconstruction in addition to variations in aortic patch shape and material.^{1,5,6}

In this issue of the *Journal*, Haller and colleagues⁷ from the Hospital for Sick Children in Toronto present an elegant study in which they studied the growth of the reconstructed aorta after the Norwood operation in 73 patients. They have used mainly the interdigitating technique that included complete coarctectomy with extended anastomosis of the distal aortic arch with the posterolateral wall of the descending aorta, anterolateral counter incision on the descending aorta, and wide aortic arch augmentation using a pulmonary homograft patch. The purpose of this interdigitating technique is to increase the area of native aortic tissue used for distal aortic reconstruction to allow for better aortic

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Central Message

Reconstructed arch geometry at the time of Norwood operation is abnormal and remains unchanged. Reinterventions for obstruction or excessive patching are equally associated with morbidity and mortality.

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growth. The group in Toronto has previously reported on this technique and showed a remarkable improvement in recurrent aortic coarctation rates, leading other centers to adopt similar approach for aortic arch reconstruction.⁶ In their current follow-up study, they studied the growth of various segments of the reconstructed aorta on cardiac catheterizations performed before subsequent palliation procedures: the bidirectional cavopulmonary connection and the Fontan operation.⁷ Several of their findings are of interest.

The first predictable finding is that the reconstructed aortic arch geometry is not normal with the ascending aorta and proximal arch being much larger than normal, whereas the distal arch anastomosis area and descending aorta are closer to normal on initial cardiac catheterization performed before the bidirectional cavopulmonary connection. This is naturally due to the use of a large pulmonary artery for the reconstruction of the ascending aorta and the 3-dimensional shape of the patch as it tapers from a wide proximal end to a narrow distal end. Therefore, the Z scores of the ascending aorta and proximal arch were 5.3 and 4.5, respectively, compared with Z scores of the distal arch anastomosis and descending aorta that were -0.5 and -0.7, respectively. The important finding is that on subsequent cardiac catheterization performed before the Fontan operation, those Z scores remained almost unchanged, with Z scores of the

ascending aorta and proximal arch of 6.0 and 4.8, respectively, compared with Z scores of the distal arch anastomosis and descending aorta of 0.2 and 0.1, respectively. That indicates that growth of all segments of the reconstructed aorta is almost uniform and that the geometry of the reconstructed arch remains nearly the same. This growth was not different for patients with a diminutive native ascending aorta or arch or those with a larger native ascending aorta or arch, indicating that compensatory growth of native aortic tissue occurs once unobstructed antegrade aortic flow is established, further giving support to the better use of native tissue for aortic reconstruction, such as in their interdigitating technique.

The echocardiographic findings in this study from Toronto⁷ were naturally associated with a low reintervention rate of 4% for recurrent aortic coarctation, which is a remarkable achievement given the significant morbidity and mortality associated with recurrent coarctation.²⁻⁵ This rate is comparable to their initial report describing zero reintervention need after adaptation of the interdigitating technique on short follow-up and seems favorable compared with the recent multi-institutional SVR trial report, all suggesting the long-term benefit of this technique in decreasing the recurrent aortic coarctation risk.^{4,6} Of note, their aortic reintervention rate for excessive patching causing vascular distortion or compression effect on adjacent pulmonary arteries or bronchi was slightly higher at 6% and might be higher than that usually reported in other series. Most important, although the SVR trial showed that aortic reintervention was not associated with decreased transplant-free survival at 1 year, this was not the case in the current series, and aortic reinterventions for recurrent coarctation or excessive patching were equally associated with mortality.^{4,7} This suggests that there is a fine line between adequate arch enlargement and excessive patching, and choosing the perfect patch that eliminates the risk of recurrent coarctation without the compression or distortion effects might be a challenging task.

In the past 2 decades, surgeons have come a long way and have made important accomplishments in the treatment of hypoplastic left heart syndrome. Although morbidity and mortality have improved remarkably after the Norwood operation, it continues to be one of the highest risk congenital cardiac surgeries, and outcomes of those patients continue to trail behind those of other congenital anomalies, including those with other single ventricle anomalies. The quest of surgeons is to continue to explore further refinements in aortic reconstruction and shunt techniques with the hope to further decrease technical complications associated with morbidity and mortality risks. Although this is a difficult mission given the great variations in the size and orientation of the great vessels and adjacent structures, new exciting technologies in 3 dimensional imaging and printing and flow dynamics studies might prove valuable in further refining aortic reconstruction techniques that might need to be tailored to individual patient anatomy in the near future.⁸

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