Neoaortic valve and root replacement after Fontan operation

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CASE REPORT

This report describes a 17-year-old woman born with hypoplastic left heart syndrome (mitral stenosis/aortic atresia), who was palliated with a Norwood/right modified Blalock-Taussig shunt, bidirectional cavopulmonary connection, and extracardiac fenestrated Fontan. Although she was clinically stable (New York Heart Association functional class II), she was followed with saturations in the low 90s, an enlarging aortic aneurysm (Figure 1), and progressive neoaortic (native pulmonary valve) insufficiency. Preoperative magnetic resonance imaging revealed a massive ascending aortic aneurysm (maximum dimensions: 77 × 95 mm) involving the arch (Video 1). The native ascending aorta was generous (8.9 × 15 mm proximal to the aortopulmonary anastomosis) and provided retrograde flow to usual coronary arteries. There was extrinsic compression of the left pulmonary artery by the aortic aneurysm, with only 16% of pulmonary blood flow directed to the left lung (Figure 2). Neoaortic insufficiency was severe and associated with mildly reduced right ventricular ejection fraction (45%) and moderate-to-severe right ventricle dilation (156 mL/m²). Preoperative catheterization was favorable with low Fontan pressures (11 mm Hg), low transpulmonary gradient (common atrial pressure, 6 mm Hg), and no evidence of obstruction in systemic or pulmonary venous pathways, or atrioventricular valve insufficiency. Comorbidities included scoliosis (Harrington rods placed) and proteinuria. She was presented at a multidisciplinary surgical conference and accepted for double root replacement and arch reconstruction with an operative risk of 10% to 15%. The institutional review board of Children’s Health Dallas did not require approval for this study because it is a retrospective case report. The subject provided written consent for the publication of this report.

OPERATIVE PROCEDURE

The operation (Video 1) was sequenced as follows to minimize crossclamp duration. The 58-kg patient was taken to the operating room for elective repair. Arterial access was

Aneurysm of the ascending and transverse aorta in a patient with a single ventricle.

CENTRAL MESSAGE

With excellent and reproducible outcomes possible for the repair of ascending aortic aneurysms, surgical repair should be offered earlier in a patient’s course to minimize operative risk.
obtained via the right axillary artery (chimney graft, 8 mm Gore-tex tube graft; W.L. Gore & Associates Inc, Medical Products Division) and direct cannulation of the right common femoral artery (15Fr) and vein (23Fr); the inferior vena cava cannula was positioned in the inferior aspect of the Fontan conduit. Redo sternotomy and mediastinal dissection were performed. Then, on cardiopulmonary bypass, 2/3 20Fr vents were established via the common atrium, and dissection continued at normothermia until the aneurysm and arch vessels were mobilized. Cooling was initiated, and head and neck vessels were debranched (4-Branch Plexus Vascular Graft, 24 mm bore size, arch branch diameters 10 × 8 × 8 mm; Terumo Cardiovascular Group). Debranching began by placing a vessel loop around the distal left subclavian, followed by ligation of the vessel origin at the aneurysm, and then the appropriate limb of the vascular graft was anastomosed to the vessel end to side. This debranching was then performed for the left common carotid artery and, lastly, the innominate artery. At a minimum core temperature of 18 °C, deep hypothermic circulatory arrest was initiated, the native ascending aorta was opened cephalad to the coronary ostia, and antegrade cold del Nido cardioplegia was administered directly.

The aortopulmonary anastomosis was incised. The distal graft-to-descending aorta anastomosis was performed end to end (branched Gelweave graft; Terumo Aortic). The graft was clamped in the ascending aortic segment, and after 29 minutes of deep hypothermic circulatory arrest, full cardiopulmonary bypass resumed. During rewarming, the proximal end-to-end anastomosis was performed between

FIGURE 1. A lateral view, volume-rendered 3-dimensional reconstruction of the preoperative ascending aortic aneurysm, which had maximum dimensions of 77 × 95 mm.

VIDEO 1. Preoperative magnetic resonance imaging (MRI) (0:00:03-0:00:22): A coronal coronary reconstruction from the 3D bSSFP obtained during the preoperative MRI demonstrates the extent of the aneurysm, which extends from the neoaortic root to the proximal descending aorta, including the arch and branch origins. Operative procedure (0:00:23-0:04:19): The operative procedure illustrating the massive aneurysm mobilized enough to allow for debranching and grafting to the head and neck vessels, followed by replacement of the ascending aorta, transverse aorta, and mechanical valved conduit replacement of the incompetent pulmonary valve. Graft diagram (0:04:20-0:04:43): A diagram of the completed repair showing the direction of blood flow through the mechanical valved conduit and the branched graft, followed by a volume-rendered 3-dimensional reconstruction of the thoracic aorta and systemic venous return to the pulmonary arteries. Video available at: https://www.jtcvs.org/article/S2666-2507(23)00208-0/fulltext.

FIGURE 2. An axial view from the 3-dimensional balanced steady-state free precession obtained during the preoperative magnetic resonance imaging scan demonstrates the aortic aneurysm (*) and compressed left pulmonary artery (arrow).
the native ascending aorta and proximal Gelweave graft. The graft was deaired, the crossclamp removed (55-minute crossclamp), and the heart reperfused. With the heart beating, the native pulmonary valve was excised and a 27-mm mechanical valved conduit (St Jude Medical Masters HP Series Valved Graft; Abbot Cardiovascular) was implanted; the distal conduit was anastomosed end-to-side to the neotransverse arch (Gelweave graft). Cardiopulmonary bypass was weaned after 243 minutes. The patient was extubated on postoperative day 1, and discharged on postoperative day 13.

COMMENT

Neoaortic root dilation is frequently encountered among patients who have undergone staged palliation for hypoplastic left heart syndrome and can require intervention if neoaortic insufficiency is severe. The Society of Thoracic Surgeons Congenital Heart Surgery Database reports 479 aortic root replacements over the past 4 years with an operative mortality risk of 2.9% (database accessed April 13, 2023), and valve-sparing neoaortic root replacements have been reported in patients who have previously undergone staged palliation. A valve-sparing procedure was not considered for the morphologic pulmonary valve in this case because of the degree of annular dilation and concern for increasing the duration of an already long cardiopulmonary bypass time. With excellent and reproducible outcomes possible for the repair of thoracic aortic aneurysms, these operations should be offered earlier in a patient’s course, when operative risks are minimized. In the case of a late referral of a patient with a single ventricle with a giant aortic aneurysm, a successful surgical outcome can still be achieved, as evidenced by this complex case.

References