Neo-aortic Valve and Root Replacement after Fontan Operation

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Glossary of abbreviations:

AA- aortic atresia
bSSFP- balanced steady-state free precession
BT- Blalock-Taussig
CPB- cardiopulmonary bypass
CTA- computed tomography angiography
DHCA- deep hypothermic circulatory arrest
Fr- French
HLHS- hypoplastic left heart syndrome
IVC- inferior vena cava
LCCA- left common carotid artery
LPA- left pulmonary artery
MRI- magnetic resonance imaging
MS- mitral stenosis
NYHA FC- New York Heart Association functional class
OR- operating room
RV- right ventricle
RVEF- right ventricular ejection fraction
STS- Society of Thoracic Surgeons
Central Picture:

Abbreviated Legend for Central Picture: Aneurysm of the ascending and transverse aorta in a single ventricle patient

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.
Case Report:

This report describes a 17-year-old female born with hypoplastic left heart syndrome (MS/AA), who was palliated with a Norwood/right modified BT shunt, bidirectional cavopulmonary connection, and extracardiac fenestrated Fontan. Even though she was clinically stable (NYHA FC II), she was followed with saturations in the low 90’s, an enlarging aortic aneurysm (Figure 1), and progressive neo-aortic (native pulmonary valve) insufficiency.

Preoperative MRI revealed a massive ascending aortic aneurysm (maximum dimensions: 77 x 95 mm) involving the arch (Video). The native ascending aorta was generous (8.9 x 15 mm proximal to the aortopulmonary anastomosis) and provided retrograde flow to usual coronary arteries. There was extrinsic compression of the LPA by the aortic aneurysm, with only 16% of pulmonary blood flow directed to the left lung (Figure 2). Neo-aortic insufficiency was severe and associated with mildly reduced RVEF (45%) and moderate-to-severe RV dilation (156 ml/m2). Preoperative catheterization was favorable with low Fontan pressures (11 mmHg), low transpulmonary gradient (common atrial pressure: 6 mmHg), 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 10-15%. 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) was sequenced as follows to minimize cross-clamp duration. The 58 kg patient was taken to the OR for elective repair. Arterial access was obtained via the right axillary artery (chimney graft, 8mm Gore-tex tube graft, W.L. Gore & Associates Inc, Medical Products Division, Flagstaff, AZ, USA) and direct cannulation of the right common femoral artery (15Fr) and vein (23Fr); the IVC cannula was positioned in the inferior aspect of the Fontan conduit. Redo sternotomy and mediastinal dissection were performed. Then, on cardiopulmonary bypass (CPB), two 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, 24mm bore size, arch branch diameters 10 by 8 by 8 mm; Terumo Cardiovascular Group, Ann Arbor, MI, USA). 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 LCCA and, lastly, the innominate artery. At a minimum core temperature of 18°C, deep hypothermic circulatory arrest (DHCA) 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). The graft was clamped in the ascending aortic segment, and after 29 minutes of DHCA, full CPB resumed. During rewarming, the proximal end-to-end anastomosis was performed between the native ascending aorta and proximal Gelweave graft. The graft was deaired, the cross-clamp removed (55-minute cross-clamp), and the heart reperfused. With the heart beating, the native pulmonary valve was excised and a 27mm mechanical valved conduit (St Jude Medical Masters HP Series
Valved Graft, Abbot Cardiovascular, Saint Paul, MN, USA) was implanted; the distal conduit was anastomosed end-to-side to the neo-transverse arch (Gelweave graft). CPB was weaned after 243 minutes. The patient was extubated post-op day one, and discharged on post-op day 13.

Comment:

Neo-aortic root dilation is frequently encountered among patients who have undergone staged palliation for HLHS\(^1\) and can require intervention if neo-aortic insufficiency is severe. The STS Congenital Heart Surgery Database reports 479 aortic root replacements over the past 4 years with an operative mortality risk of 2.9% (database accessed 4/13/23), and valve-sparing neo-aortic root replacements have been reported in patients who have previously undergone staged palliation\(^2\)\(^-\)\(^5\). 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 single ventricle patient with a giant aortic aneurysm, a successful surgical outcome can still be achieved, as evidenced by this complex case.
References:


Central Picture: Aneurysm of the ascending and transverse aorta in a single ventricle patient

Figure 1: A lateral view, volume-rendered 3D reconstruction of the pre-operative ascending aortic aneurysm, which had maximum dimensions of 77 mm x 95 mm.

Video:

Preoperative MRI (0:00:03-0:00:22): A coronal coronary reconstruction from the 3D bSSFP obtained during the pre-operative 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 3D reconstruction of the thoracic aorta and systemic venous return to the pulmonary arteries.

Figure 2: An axial view from the 3D bSSFP obtained during the pre-operative MRI demonstrates the aortic aneurysm (*) and compressed left pulmonary artery (arrow).