Infant Repair of Massive Aortic Aneurysm With Prosthetic Valved Conduit

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A 4-month-old child with severe infantile Marfan syndrome underwent successful repair of an extremely dilated aortic root and severe aortic valve insufficiency using a prosthetic valved conduit.

Marfan syndrome (MFS) is a progressive multiorgan connective tissue disease that typically manifests in adolescence or early adulthood. Rarely, MSF may be diagnosed in neonates, and these patients frequently suffer from a more severe form of the disease than those diagnosed later in life [1]. One of the most common cardiovascular manifestations of this disorder is progressive dilation of the aortic root [2], the consequences of which account for as many as 80% of premature deaths in these patients [3]. Prophylactic surgery is, therefore, indicated for patients with aortic root dilation and has been shown to improve the prognosis in young children with MFS [1]. The Bentall procedure [4] is the procedure of choice when the aortic valve cannot be salvaged [5]. Although this operation has been reported in young children [1,6,7], this case represents the youngest patient reported in the literature.

A 4-month-old boy was diagnosed at birth (weight 3.8 kg) with infantile MFS. His initial echocardiogram demonstrated mild mitral regurgitation, mild aortic regurgitation, and significant aortic root dilation, measuring 1.7 cm in diameter (Z-score 5.64).

At 7 weeks of age (weight 4.61 kg), the patient started to show signs of heart failure. A repeat echocardiogram demonstrated an increased aortic root dimension, now measuring 2.4 cm (Z-score 10.03). It also revealed mildly depressed left ventricular function, moderate aortic insufficiency, and moderate mitral regurgitation due to mitral prolapse. Anticongestive medical therapy, beta-blockade, and angiotensin-converting enzyme inhibition were initiated. Over the next several weeks, the patient improved symptomatically. However, by 14 weeks of age (5.32 kg), the aortic root dimension had increased to 3.0 cm in diameter (Z-score 13.83), the aortic insufficiency had progressed, and the left ventricular function appeared more depressed (Fig 1). A subsequent magnetic resonance image confirmed significant dilation of the aortic root (Fig 2), with a Z-score of the sinuses of Valsalva between 8 and 12, with associated severe aortic and mitral insufficiency. In addition, the patient was also incidentally discovered to have a vascular ring, in the form of a right aortic arch and aberrant left subclavian artery. Secondary to the rapid progression of his aortic root dilation, the decision was made to surgically intervene.

At the time of surgery, at 15 weeks of age (weight 5.32 kg), the patient’s aortic root measured in excess of 3.2 cm (Z-score 15.30). Intraoperatively, the aortic valve was trileaflet but was very dysplastic and considered not to be repairable. A composite root replacement using a 19-mm St. Jude prosthesis (St. Jude Medical, St. Paul, MN) was performed. A mitral valve repair was completed using a 28-mm Carpentier-Edwards annuloplasty ring (Edwards Lifesciences, Irvine, CA), and the vascular ring was also repaired. Postoperatively, the patient experienced low cardiac output and underwent elective placement on extracorporeal membrane oxygenation in the intensive care unit. He was weaned off extracorporeal membrane oxygenation uneventfully after 4 days, and discharged home on the 18th postoperative day with no further complications.

Three months after surgery, the patient is thriving (weight 7.06 kg). His electrocardiogram is normal, and his echocardiogram demonstrates improving ventricular function, a well-functioning aortic prosthesis (Fig 3), and trivial mitral insufficiency.

Comment

Aortic aneurysms are rarely encountered in infants and, therefore, not much attention has been paid to the
indications for repair and the outcomes of surgery in this population [8]. Even less literature exists on infants receiving composite valved grafts because the need for long-term anticoagulation therapy after this procedure, and the difficulty in implanting adult-sized prostheses in infants has made valve-sparing techniques preferential [5]. However, some investigators suggest that caution should be used when applying valve-sparing procedures to children with connective tissue disorders [5]. In the only long-term pediatric study on this topic, Cattaneo and colleagues [7] found that composite aortic root replacement in children carries a low surgical risk and yields excellent long-term results; however, the youngest patient in their cohort receiving a composite valved graft

Fig 1. (A) Significant central aortic valve insufficiency on transthoracic echocardiography, with wide vena contracta and regurgitant jet directed into left ventricular apex. (B) Transesophageal echocardiogram demonstrating further significant widening of vena contracta and extension of regurgitation, immediately before repair.

Fig 2. Severe aortic root dilation on magnetic resonance imaging, with comparison to size of main pulmonary artery superiorly, with associated aortic valve regurgitation.

Fig 3. Still frame in diastole on postoperative echocardiogram shows minimal aortic insufficiency with significant improvement in left ventricle dilation.
was 6 years of age. Dervanian and coworkers [6] described a 22-month-old infant as being the youngest patient to successfully undergo a Bentall procedure. Our patient experienced rapid and extreme aortic root dilation, with a growth rate of 15 mm over 15 weeks to a diameter in excess of 32 mm at the time of surgery. At an age of 15 weeks, we believe this is the youngest patient reported to have undergone implantation of a prosthetic aortic valve conduit. Our case demonstrates the utility of this approach in situations where preservation of the aortic valve is not possible.

References


Severe Aortic Valve Regurgitation Due to Takayasu’s Aortoarteritis in a Child

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An 8-year-old Japanese boy with severe aortic valve regurgitation was treated by the Ross procedure with use of the full root technique. Takayasu’s aortoarteritis was diagnosed 2 months after the operation. At 8 months after the operation, follow-up echocardiography revealed an aortic root pseudoaneurysm, which was surgically repaired. At 24 months after operation, the patient continues to receive prednisolone, azathioprine, and cyclophosphamide and is in good health, with good pulmonary autograft function.

Takayasu’s aortoarteritis (TA) is a chronic, nonspecific inflammatory disease affecting the aorta and its major branches and the coronary and pulmonary arteries.Predominantly occurring in young women, TA is rarely documented in childhood [1]. Establishing a definitive diagnosis of TA in childhood, especially in the early phases of the condition, is challenging [2]. Here we report the case of an 8-year-old Japanese boy who underwent the Ross procedure for severe aortic valve regurgitation, which was subsequently diagnosed as TA.

An 8-year-old Japanese boy weighing 27.3 kg was admitted to our hospital with severe aortic valve regurgitation and congestive heart failure. He had a 1-month history of fever, fatigue, and shortness of breath. A grade 3 to-and-fro murmur was heard at the fourth left sternal border. Chest roentgenography revealed cardiomegaly (cardio-thoracic ratio, 56.1%) and pulmonary congestion. Trans-thoracic echocardiography revealed a thickened ascending aorta wall, severe aortic valve regurgitation with right and noncoronary cusp detachment, and noncoronary cusp perforation. The thickened wall of the ascending aorta was enhanced on contrast medium–enhanced multislice computed tomography. There were no stenoocclusive lesions of the aorta and its major branches. Laboratory studies revealed an increased leucocyte count of 10,000/μL (neutrophil count, 62.6%) and an elevated C-reactive protein level of 3.72 mg/dL. Blood culture was negative. These findings suggested infectious endocarditis of the aortic valve, with aortic root abscess or aortic dissection. Aortic root replacement with a composite graft or pulmonary autograft (Ross procedure) was subsequently planned and performed 48 hours after admission on account of deterioration of the boy’s condition.

During the operation, it was observed that the wall of the ascending aorta was entirely edematous (Fig 1A), whereas the aortic arch and pulmonary trunk appeared normal. Cardiopulmonary bypass was initiated with aortic arch arterial and bicaval venous cannulations. The left vent cannula was inserted into the left ventricle through the right upper pulmonary vein. An aortic cross-clamp was placed distally on the ascending aorta. Transverse aortotomy was performed, and cardioplegic solution was directly administered into both coronary orifices. The noncoronary Valsalva sinus was aneurysmally enlarged. The commissure between the right and noncoronary cusps was detached from the Valsalva sinus wall and carried a vegetative mass. Noncoronary cusp perforation was also evident (Fig 1B). An inflammatory lesion that chiefly involved the Valsalva sinus wall and the right and noncoronary cusps was noted. The inflammation was also associated with partial intimal disruption of the right coronary orifice (Fig 1C). The pulmonary autograft was obtained, and the Ross procedure was performed with...