Modified pediatric Bentall procedure: A novel technique in a rare case

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ABSTRACT

Aneurysms of ascending aorta are rarely seen in pediatric age group. Only few cases with Marfans syndrome have been reported in the literature. Preferred treatment for these children has been the standard Bentall procedure (aortic root replacement with composite graft prosthesis). We report a 4-year-old male child with huge aneurysm of ascending aorta and aortic root dilation with severe aortic regurgitation, having phenotypic features of Loeys-Dietz syndrome type I. He underwent Bentall procedure with a novel modification (medial trap-door technique for coronary reimplantation). Short-term result of this procedure is encouraging and he is asymptomatic for the last 14 months of follow-up.

Keywords: Pediatric Bentall, Loeys-Dietz syndrome, medial trap-door technique

INTRODUCTION

Ascending aortic aneurysm is rare in children. It usually occurs in patients with underlying connective tissue disorders such as Marfans syndrome (MFS) and Ehler-Danlos syndrome.[1] The standard treatment is replacement of the aortic root and ascending aorta with a composite valve conduit or a cryopreserved homograft or a xenograft,[2] or an aortic valve-sparing ascending aortic replacement.[3]

We report a child with Loeys-Dietz syndrome (LDS) type I, having dilated aortic root with aneurysm of ascending aorta with severe aortic regurgitation (AR), who underwent a modified technique of Bentall procedure.

CASE REPORT

A 4-year-old male child presented with breathlessness on exertion and easy fatigability (NYHA- New York Heart Association class III). He had hypertelorism, malar hypoplasia, retrognathia, bifid uvula, and developmental delay suggestive of LDS phenotype I. Clinically, child had significant AR. Transthoracic echocardiography (TTE) revealed aortoannular ectasia with dilated aortic root and ascending aorta. The aortic valve was tricommissural with severe AR (Grade IV). Biventricular function was normal.

Contrast-enhanced computerized tomography of aorta showed a fusiform dilatation of ascending aorta involving the aortic root with ballooned sinuses extending until the origin of innominate artery. There was diffuse thickening of aortic leaflets. Aortic annulus measured 4.8 × 3.3 cm and the sinus of valsalva measured 5.9 × 5.3 cm [Figure 1].

The child was taken up for surgery through median sternotomy. Huge aneurysm of the ascending aorta was noted involving the aortic root [Figure 2]. Aorta, pulmonary artery (PA), branch PAs, and all cephalic vessels were dissected and looped. Cardiopulmonary bypass was established with distal aortic, right atrial, and inferior vena caval cannulation. Under moderate hypothermia, with vent into the right

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superior pulmonary vein, aorta was cross-clamped and opened. Direct osteal cardioplegia was administered and subsequently repeated at regular intervals. Thickened, distorted, and noncoapting aortic leaflets were excised. Coronary buttons were harvested. Remaining aneurysmal portion of ascending aorta was excised starting from few millimeters from the annulus proximally to the point next to origin of innominate artery where the aorta was normal, leaving a narrow aortic wall collar at the annulus. Multiple interrupted pledgetted horizontal mattress sutures (2-0 braided polyester) were taken at the annulus and then passed through the sewing ring of a composite valve conduit no. 21 (Dacron composite graft with a mechanical valve; St. Jude Medical Inc., Minnesota, MN, USA), suturing it to the annulus [Figure 3]. Another layer of continuous polypropylene suture was taken between the adjacent aortic wall collar and sewing ring. Distal end of graft was appropriately fashioned and was anastomosed to the distal aorta. Aorta was then deaired by direct injection of saline and cross-clamp was released momentarily to allow the graft to distend. Exact sites of both coronary button implantations were marked and aorta was clamped again. Coronary buttons were then reimplanted on to the graft using ``medial trap-door technique''[4] [Figure 4]. Aortic cross-clamp was removed with standard precautions. Remaining procedure was performed in routine fashion and was uneventful.

Postoperative TTE showed normally functioning prosthetic aortic valve with normal-sized aortic root. The child was discharged on postoperative day 10 on adequate oral anticoagulation therapy. Over the last 14 months, the child clinically remains asymptomatic. Follow-up echocardiography revealed normal study. We aim to keep yearly regular surveillance for other manifestations of the syndrome- cardiac and noncardiac such as dilatation of the aorta, widespread vascular aneurysms and dissections, and generalized arterial tortuosity.[5]

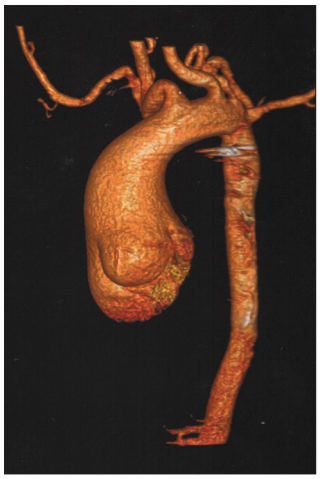

Figure 1: Three-dimensional reconstruction of contrast-enhanced computerized tomography angiogram of ascending aorta, arch of aorta, and descending thoracic aorta

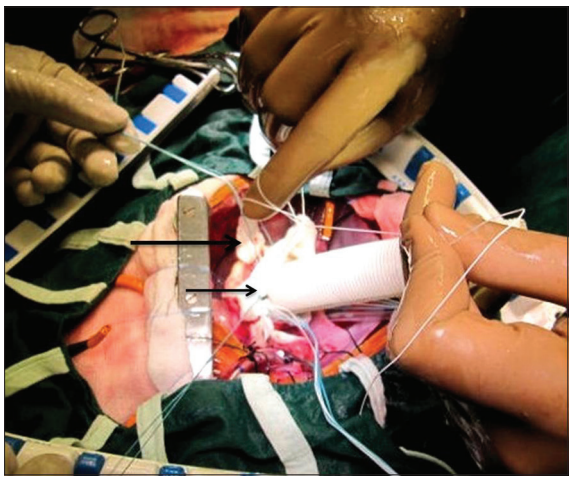

Figure 3: Sutures taken through the aortic annulus and then through the sewing ring (arrow) of composite valve conduit. Coronary button indicated by bold arrow

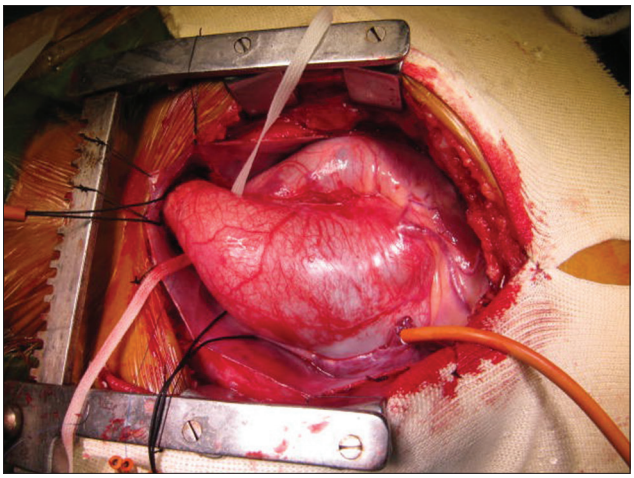

Figure 2: Aneurysm of ascending aorta, involving the aortic root and sinotubular junction, measuring around 5.5 cm in diameter, extending up to the origin of innominate artery

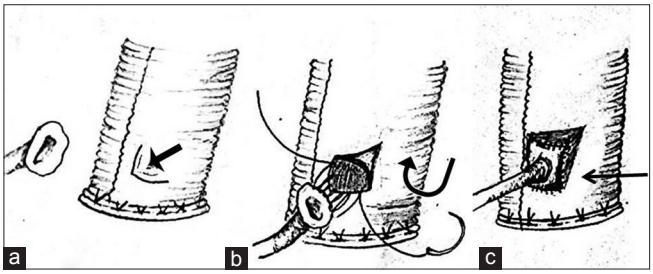

Figure 4: Sketch diagram of medial trap-door technique: (a) "L"-shaped incision (arrow) over the graft with angle of the incision pointing toward the base of coronary button. (b) Coronary button reimplantation using continuous fine polypropylene suture with the graft flap open (curved arrow). (c) Completed anastomosis with the formation of "hood" (arrow)

DISCUSSION

The original technique of Bentall procedure has been modified and used for a large spectrum of various pathologic conditions involving the aortic valve, aortic root, and ascending aorta.[2] Children requiring Bentall procedure is unusual. Only few cases with genetic syndromes have been reported. The most frequent cause of aortic root dilatation in children less than 5 years has been MFS.[1,6]

Our child presented with characteristic phenotypic features of LDS type I. It is a Marfan-like condition associated with severe craniofacial features (craniosynostosis, malar hypoplasia, retrognathia, cleft palate, or bifid uvula and hypertelorism), aortic root aneurysms, generalized arterial tortuosity with widespread vascular aneurysm and dissection, arachnodactyly, pectus deformity, scoliosis, joint laxity, and developmental delay.[7,8] Peripheral Doppler study of the child revealed rest of the vascular tree to be apparently normal.

LDS is associated with mutations of transforming growth factor beta receptor genes 1 and 2 that are inherited in an autosomal dominant manner. There is a variable clinical expression of the disorder, and thus clinical diagnostic criteria do not exist.[8] Mutations of these two genes are also described with MFS or with familial/thoracic aneurysm and dissection. So their detection is not specific.[9]

In one of the largest series of ascending aortic replacement in children (most of them being Marfans), the indications for replacement were either major dilatation (average of 46.3 mm) or rapid rate of progression of dilatation in the preceding 1 year (average of 7.5 mm).[6] It is reasonable to undergo aortic repair in patients with LDS when the aortic diameter exceeds 4.0 cm.[9] Diameter of aortic root and ascending aorta in excess of 5.5 cm with severe AR in the child was an unambiguous indication for an early surgical intervention. Benefits of prophylactic proximal arch replacement at the time of root replacement are not known.[5] So we did not prefer doing it in a pediatric patient. Root replacement was the surgery of choice as aortic valve morphology did not favor valve-sparing procedure.

One of the most important steps of Bentall procedure is coronary implantation technique. Three different anastomotic techniques have been developed – the inclusion, open button, and Cabrol techniques.[2]

In our case, we used the "medial trap-door technique" for coronary implantation. Advantages of this technique over the conventional techniques are the following: (a) accurate location of coronary reimplantation avoids tension or kink; (b) more physiologic "hood" to the coronary anastomosis; and (c) additional length of few millimeters as the trap door opens toward the base of the button.

This technique is particularly more favorable in children because slightest coronary stretch or kink can cause significant change in the flow. The technique suggested by Cabrol *et al.*[10] is undesirable in children because of lack of growth potential. In addition, we are comfortable with the medial trap-door technique in arterial switch operations. Hence, it was easy to reproduce that result in this subset of patients. During the follow-up period of 14 months, the child has done well and remains asymptomatic.

In conclusion, our patient having LDS was probably one of the youngest ever to undergo Bentall procedure for aortic aneurysm with severe AR. We suggest keeping a lower threshold for replacement for patients with LDS because they are known to have rapid progression of dilatation and early dissection as compared with MFS. Our technique of coronary reimplantation that we routinely employ in arterial switch operation ensures tissue-to-tissue, tension-free, and accurate anastomosis. From our experience, we believe that it is a simple, safe, effective, and easily reproducible modification of pediatric Bentall procedure. Short-term results have been encouraging.

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Conflicts of interest

There are no conflicts of interest.

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