Valve-sparing replacement of the ascending aorta and aortic arch in a child with Loeys-Dietz syndrome

Emre Özker*, Can Vuran, Bülent Sartaş and Rıza Türköz

Department of Cardiovascular Surgery, Istanbul Medicine and Research Hospital, Baskent University, Istanbul, Turkey

* Corresponding author. Acibadem MH. Ibrahimimag Konutları, A5 D12 Kadıkoy, Istanbul, Turkey; Tel: +90-532-5154225; fax: +90-262-6414569; e-mail: dremreozker@yahoo.com (E. Özker).

Received 27 August 2011; received in revised form 13 October 2011; accepted 18 October 2011

Abstract

We describe a successful surgical treatment in a 2.5-year old boy with Loeys-Dietz syndrome, in whom we performed aortic arch and ascending aorta replacement with a valve-sparing operation (VSO) of the aortic root because of significant aortic insufficiency and dilation of the aortic root. We believe that VSO is ideal for treating young patients with aortic root aneurysm with normal or minimally diseased aortic cusps to avoid the disadvantages of prosthetic valve replacements.

Keywords: Aortic aneurysm • Aortic arch replacement • Valve-sparing surgery

INTRODUCTION

Loeys-Dietz syndrome (LDS) is a newly recognized autosomal-dominant genetic syndrome that demonstrates clinical characteristics similar to Marfan syndrome, including aortic pathologies with more malignant natural history.

We describe a successful surgical treatment in a 2.5-year old boy with LDS, in whom we performed a valve-sparing replacement of the ascending aorta (David 1) (David 1 is the technique to reimplant the aortic valve described initially by Tirone David in 1992. The 3 commissures of the aortic valve are resuspended and secured in the Dacron graft. The remnants of the aortic sinuses are sutured to the Dacron. The coronary arteries are reimplanted and the other end of the graft is anastomosed to the distal ascending aorta. The coronary arteries are reimplanted and the other end of the graft is anastomosed to the distal ascending aorta (David 1) and aortic arch because of significant aortic insufficiency (AI) and aneurysm of the ascending aorta and the arch [2].

CASE REPORT

A 2.5-year old boy was admitted to our clinic with rapid aortic root dilatation, AI, ascending aorta and aortic arch aneurysm diagnosis. In the referring paediatric clinic, the patient has been followed up with echocardiography since the neonatal period. The patient was found to have joint laxity and the genetic scanning revealed Type 2 LDS. The patient was put on metoprolol treatment and followed up with echocardiography. In his routine examinations, progressive aortic root and ascending aorta dilatation with an increase in AI were determined (Fig. 1). The patient was then transferred to our clinic for the operation.

In his physical examination, there was no facial dimorphism. The body weight was 11 300 g. Echocardiography revealed Grades 2–3 AI. In his aorticography, the diameters were measured as 38.8 mm at the aortic bulb, 27.1 mm at the sinotubular junction and 38.9 mm at the ascending aorta. Ascending aorta and aortic arch replacement operation was planned (Supplementary Videos S1 and S2).

In the operation, following median sternotomy, cardiopulmonary bypass was achieved via the right innominate and right atrium cannulation. Annulooaortic ectasia and the ascending aortic aneurysm advancing to the aortic arch were observed. Following dissection of the branches of the aortic arch, an aortic cross-clamp was applied and cardiac arrest was maintained with blood cardioplegia. Aortic valve reimplantation was performed by dissecting the aortic root circumferentially down to a level just below the nadir of the aortic annulus, detaching the coronary arteries from their sinuses, excising the aortic sinuses except for 3 mm, and placing the aortic cusps anulus and subcommisural triangles of the non-coronary cusp inside a 26 mm Hemashield Dacron tube graft (Meadox Medical Inc., Oakland, NJ, USA). The aortic valve competence was checked with saline; there was trivial valve insufficiency. The coronary buttons were reimplanted to the graft and cardioplegia solution was used to control any leakage. Intermittent antegrade hypothermic blood cardioplegia was used throughout the procedure to maintain cardiac arrest. The patient was then continued to cool down to 19°C. Selective cerebral perfusion (SCP) was achieved via the innominate artery. The aortic arch was excised to the origin of the left subclavian artery. One arm of a 10/20 mm Dacron Y-graft was sewn to the innominate artery. A 6 mm-ringed polytetrafluoroethylene which was sewn to the Y graft previously was anastomosed to the left common carotis artery. The other arm of the Y graft was sewn to the distal aortic arch. The SCP was terminated. The free ends of the 20 and 26 mm Dacron grafts were
anastomosed to each other while warming. The cross-clamp and cardiopulmonary bypass times were 174 and 270 min, respectively. The SCP time was 34 min.

The postoperative course was uneventful.

DISCUSSION

LDS is a disorder that is caused by mutations in the genes-encoding transforming growth factor β receptor 1 or 2 and is characterized by vascular and skeletal manifestations. It is classified into two types, the first with craniofacial features such as hypertelorism and bifid uvula, and the second type without these features. Patients falling into the first type tend to demonstrate more severe cardiovascular anomalies than those placed in the second group. Our patient would have been categorized in the second type [3, 4].

LDS is an aggressive aortic aneurysm disease with a propensity towards rupture and dissection at a younger age and smaller aortic diameters than in other connective tissue disorders such as Marfan and Ehlers Danlos syndromes. It is usually recommended that patients with Marfan syndrome undergo aortic root replacement at root diameters >5.0 cm; however, lower thresholds are offered in paediatric LDS patients. For patients with severe craniofacial features aortic root z-score >3.0 and for patients with mild craniofacial features aortic root z-score >4.0 or rapidly expanding (>0.5 cm over 1 year) are offered early prophylactic intervention [5]. The z-score was 9.45 in our patient and there was a rapid increase in both aortic diameter and valve insufficiency.

Although there have been some reports on LDS syndrome recently, there are few paediatric patients who underwent total arch replacement like our case. In Williams et al’s [5] report of 65 LDS patients, nearly two-thirds of the patients had aneurysm disease extending beyond the ascending aorta; however, there was only one patient who had undergone aortic valve-sparing operations with aortic arch replacement.

We believe that the David 1 procedure is an ideal method to treat very young patients with aortic root aneurysm with normal or minimally diseased aortic cusps. These operations help to avoid the disadvantages of prosthetic valve replacements; however, they require prolonged aortic cross-clamp times and therefore should be avoided in patients with diminished ventricular functions and in complex operations with concomitant procedures. Given the widespread involvement of the arterial tree in patients with LDS, periodical scanning from the head to pelvis is needed.

SUPPLEMENTARY MATERIAL

Supplementary material is available at EJCTS online.

CONFLICT OF INTEREST

None declared.

REFERENCES


Figure 1: Preoperative multislice computerized tomography showing the ascending aorta and aortic arch aneurysm.