Valve-Sparing Replacement of the Aortic Root after Repair of Tetralogy of Fallot

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Progressive aortic root dilatation is a common feature after surgical repair of tetralogy of Fallot. This report describes a successful valve-sparing replacement of the aortic root in a patient with significant dilated aortic root and aortic regurgitation after repair of tetralogy of Fallot.

Keywords: aortic operation, aortic root, aortic valve repair, congenital heart disease (CHD), tetralogy of Fallot

Introduction

Progressive aortic root dilatation is a common feature after surgical repair of tetralogy of Fallot (TOF).1) Although mild aortic regurgitation (AR) is seen after repair of TOF, significant AR is not common. Little information is available, regarding the management of aortic root dilatation and AR after repair of TOF.

Case Report

A 16-year-old male presented with moderate AR and a dilated aortic root (40 mm). He had a history of TOF with pulmonary atresia for which he received a right Blalock-Taussig shunt as a newborn followed by a left Blalock-Taussig shunt at one year of age. He underwent complete repair (closure of ventricular septal defect and right ventricular outflow tract (RVOT) patch) at four years of age. He had limited exercise tolerance and occasional chest pain. Echocardiogram revealed that the aortic valve was tricuspid without stenosis with moderate central AR (Fig. 1A). There was retrograde diastolic flow in the descending aorta. The ascending aorta was mildly dilated (40 mm, Z-score 7.4). There was stenosis at the right ventricular outflow tract. Aortogram revealed dilatation of the aortic annulus and ascending aorta (Fig. 2). Cardiac magnetic resonance imaging (MRI) revealed moderate pulmonic regurgitation and moderate AR with regurgitant fraction of 34%. Ejection fraction of the left ventricle (LV) decreased to 33%. The coronary artery anatomy was normal. At 17 years of age, the patient underwent valve-sparing root replacement as well as placement of a pulmonary homograft in the RVOT. Total cardiopulmonary bypass was established between the distal ascending aorta and both vena cavae. Myocardial protection was achieved using moderate systemic hypothermia (32°C) and antegrade cold blood cardioplegia followed by retrograde cardioplegia, repeated every 20 minutes. Although the aortic annulus was dilated, valve tissue was intact. Valve-sparing aortic root replacement with aortic valve re-implantation was performed. A 30 mm Gelweave Valsalva prosthesis (Terumo Cardiovascular Systems Corp., Ann Arbor, Michigan) was used for the reconstruction. The right ventricular outflow tract was reconstructed with a 26 mm allograft. The aortic cross clamp was taken off, and the cardiopulmonary bypass was safely discontinued. Intraoperative echocardiogram after termination
of the cardiopulmonary bypass showed trivial AR from the central area. The postoperative clinical course was uneventful, and the patient was discharged home on the fourth postoperative day. His exercise tolerance improved after the surgery. The follow-up echocardiogram, 1 month after surgery, showed mild AR from the central area (Fig. 1B).

Discussion

Preoperative increased aortic flow from right-to-left shunting has been thought to be one of the causes of progressive aortic root dilatation.1) However, recent studies showed that aortic root dilatation in TOF patients is related to the histological abnormalities of this disease.2,3) AR has also been observed in patients after repair of TOF,1,4) and progressive aortic root dilatation is the major cause of AR after repair of TOF.1,5) Ishizaka and associates reported four aortic valve surgeries (valve replacement with mechanical valve in three patients and valve repair in one patient) after the repair of TOF out of 427 patients (0.9%).6) Risk of thrombo-embolic events, endocarditis, and long-term anticoagulation are the limitations of aortic valve replacement with mechanical valve.

Fig. 1 A. Preoperative echocardiogram shows moderate aortic regurgitation, as well as a dilated ascending aorta. B. Follow-up echocardiogram shows mild aortic regurgitation and reconstructed aortic sinuses (arrow). LA: left atrium; LV: left ventricle; Ao: aorta

Fig. 2 A. Anteroposterior view. B. Lateral view. Aortogram shows a dilated annulus of the aortic valve, as well as the ascending aorta.
Ono and associates reported valve-sparing operations for 2 patients with aortic root dilatation and AR after the repair of TOF.5) We performed valve-sparing aortic root replacement with aortic valve re-implantation using a Valsalva graft for patients after the repair of TOF, and valve function is being well maintained in our follow-up. We chose a 30-mm Valsalva graft, according to the height of the native aortic valve as de Kerchove and associates recently reported.6) Since Windkessel function is well maintained in a Valsalva graft compared with a tube graft,7) we believe that a Valsalva graft may be the preferred prosthesis for these procedures. To our knowledge, there is no follow-up study for TOF patients with valve-sparing aortic root replacement with aortic valve re-implantation. Continued follow-up will be needed to assess the long-term ramifications of this approach for children with aortic root dilatation and AR after following TOF repair.

Surgical treatment for dilated aortic root after the repair of TOF is controversial. Current consensus recommendations for adult patients with congenital heart disease are to repair the ascending aorta when it is 55 mm or greater to prevent aortic rupture or dissection.8) Less information is available for Windkessel function as well as LV function in patients with congenital heart disease. Loss of Windkessel function (increased aortic stiffness) increases systolic blood pressure, leading to increased LV afterload and LV hypertrophy.9) Further in-vitro or animal studies, as well as an observational, clinical study, will be needed to clarify the Windkessel function and LV function, in order to determine the optimal indication for surgery in patients with ascending aortic aneurysm.

Disclosure Statement

We have no financial or other interest in the manufacture or distribution of the device.

References