Yacoub valve-sparing aortic root replacement with a concomitant repair of Tetralogy of Fallot: a case report

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Abstract

Enlargement of the aorta is a severe sequela of tetralogy of Fallot (TOF) that can cause fatal complications including aortic regurgitation, dissection, aneurysm or even rupture. Valve sparing aortic root replacement (VSARR), also known as the Yacoub technique, is a surgical maneuver that corrects regurgitation and avoids requirement for long-term anticoagulation treatment at the same time. Several studies have demonstrated the effectiveness of VSARR in patients with aortic regurgitation complicated by aortic root dilation following a repair of congenital heart defects. To date, little data exists on VSARR in TOF patients. Herein we report a case of a previously unoperated 35-year-old male TOF patient. He was admitted due to dyspnea, initial physical examination demonstrated cyanosis and heart murmurs. Echocardiography suggested secundum atrial septal defect, perimembranous ventricular septal defect and mild-severe aortic regurgitation. Computed tomography angiography indicated aortic root dilation and suspicious right coronary artery aneurysm. Concurrent repair of TOF and VSARR was performed successfully and the patient had a satisfactory recovery.

KEYWORDS: Aortic root replacement; Congenital heart disease; Tetralogy of Fallot

1 — INTRODUCTION

Enlargement of the aorta is a severe sequela of tetralogy of Fallot (TOF), which can lead to fatal outcomes like aortic regurgitation (AR), dissection, aneurysm, or even rupture with fatal consequences, even in those patients who have had a repair of TOF.[1] Valve sparing aortic root replacement (VSARR), also known as the Yacoub technique, is a surgical procedure that has the advantages of correcting AR, preserving native aortic valves, avoiding prosthetic valves and subsequent requirement for long-term anticoagulation treatment.[2] Several studies have demonstrated the effectiveness of VSARR in patients with AR complicated by aortic root dilation following a repair of congenital heart defects.[2, 3] To date, VSARR has not been performed on TOF patients undergone no intervention. Here we described a case of a previously unoperated TOF patient undergoing VSARR and concurrent repair of TOF successfully.

2— CASE PRESENTATION

A 35-year-old male previously diagnosed to have TOF at the age of 5 but did not undergo any intervention for an unknown reason, presented with dyspnea after physical exercises and occasional cyanosis of the lips. He reported no symptoms of chest pain, palpitation, amaurosis, syncope, swelling of limbs, or squatting position. His other past medical history included pulmonary tuberculosis that was fully treated at the age of 25, and an abdominal angioma diagnosed approximately 4 weeks prior to his hospitalization in our institution.

The physical examination revealed evidence of cardiac arrhythmia, tremors, grade 3/6 systolic ejection murmur at the second intercostal space and the left margin of the sternum, grade 4/6 systolic blowing murmur at the fourth intercostal space and the left margin of the sternum. Echocardiography was performed which showed unoperated TOF. A dilated right atrium measuring 52 mm in diameter was evident, while the right ventricle appeared hypertrophic with a thickness of 14 mm. The infundibulum of the right ventricular outflow tract appeared severely narrowed with a diameter of 8 mm. The pulmonary artery measured 18 mm in diameter and the pulmonary artery valves were slightly restrictive. The left ventricle appeared normal in size and the ejection fraction was 73%. The aorta seemed to have moved forward and 50% of its area covered the interventricular septum. Furthermore, there was dilation of the aortic root (55.4 mm) **(Fig.1)** and a mild central AR (VC = 5.7 mm) **(Fig.2)** . The right coronary artery showed evidence of an anomaly with a diameter of 7.5mm. Moreover, there was a secundum atrial septal defect showing bidirectional low-speed shunt and a 15 mm perimembranous ventricular septal defect (VSD). Given the above findings, surgery was recommended to correct the above abnormalities.

In surgery, a median sternotomy was performed and a total cardiopulmonary bypass was established between the distal ascending aorta and the vena cavae. Myocardial protection strategies including systemic hypothermia at 32, antegrade cold cardioplegia, and consistent retrograde cardioplegia through coronary sinus were applied. The right ventricular outflow tract was incised longitudinally and the excessive infundibular muscle was resected. The VSD extending towards the infundibular and perimembranous portion of the right ventricle was clearly visualized. An autologous pericardium was harvested for the closure of the VSD and secured with continuous sutures. The aortic root was then transected to explore the aortic annulus and aortic valves. There was no thickening or calcification found, and all the leaflets appeared equally. Following that, both the coronary arteries were freed, and all the aortic sinuses with the entire ascending aorta were resected. A 3 mm high aortic valvuloplasty ring was made out of a 26 mm artificial vessel. Then, the valvuloplasty ring, aortic root, and the proximal end of the artificial vessel were assembled by continuous suturing to reconstruct the Valsalva sinus. An end-to-end anastomosis was performed between the ascending aorta and the distal end of the artificial vessel with running sutures. Openings of 8 mm in size were made on the artificial vessel and both the coronary arteries were then anastomosed to the openings with continuous suturing using 5-0 Prolene. An autologous pericardium was utilized as a patch to enlarge the pulmonary artery trunk. Partial closure of the atrial septal defect was performed using running sutures, leaving a small remaining 3 mm opening. Upon completion of the surgical procedures, patient rewarming, lung recruitment, discontinuation of cardiopulmonary bypass, and cardiac resuscitation were uneventful. An intraoperative

transesophageal echocardiogram was performed, which revealed trivial central AR and an unobstructed left ventricular outflow tract. Postoperatively, the right ventricular systolic pressure measured 45 mmHg, the pulmonary arterial pressure was 18 mmHg, and the peripheral blood pressure was 88/45 mmHg.

3—DISCUSSION

Aortic dilation is a common sequela among patients with an unrepaired TOF. Studies have suggested that aortic dilation may be attributed to longstanding volume overload of the ascending aorta as a result of right ventricular outflow tract obstruction and right-to-left shunting across the ventricular septal defect.[4],[5] VSARR is a well-established procedure that corrects AR as a result of aortic dilation, with recent studies suggesting that the modified David V technique that preserves sinus-like geometry may improve annular stabilization and hemostasis in the long term compared with those of remodeling techniques (Yacoub/David II).[2] Also, VSARR has also been successfully performed on patients with congenital heart defects.[2, 3] Adachi and colleagues have reported a case of TOF repair with VSARR on a patient who has previously undergone a left original Blalock-Taussig shunt during infancy.[6] Our report represents the first to describe a successfully performed VSARR procedure on a previously unoperated adult TOF patient. In our patient, the long-standing VSD fortunately has not caused a significant impairment to the aortic cusps or annulus. With our previous experience, extreme care was taken to avoid disrupting the balanced aortic root or distorting the aortic annulus during VSD repair. To better preserve the sinuses and considering the hemodynamics of this patient, we chose the Yacoub technique to reconstruct the aorta.

4—CONCLUSION

In conclusion, valve-sparing aortic root replacement using the Yacoub technique for aortic root dilation and severe AR, with a concomitant repair of tetralogy of Fallot in previously unoperated adult patients is feasible with excellent surgical outcomes. Studies examining long-term outcomes are warranted to validate the effectiveness of the VSARR technique further while exploring other innovative surgical approaches in the management of TOF patients with aortic dilation.

AUTHOR CONTRIBUTIONS

All authors were involved in the conception and design, critical revision, manuscript writing, final approval, and agreed to be accountable for all aspects of the work.

FUNDING INFORMATION

No funding was required in the preparation of this case report.

DATA AVAILABILITY STATEMENT

All data generated or analyzed during this study are included in this published article.

ETHICS STATEMENT

This study is in compliance with the declaration of Helsinki.

CONSENT

Written informed consent was obtained from the patient for publication of this case report in accordance with the journal's patient consent policy.

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Figure Legends:

Figure 1 – Preoperative CTA showed the dilated aortic root with a diameter of 55.4 mm in the TOF patient.

Figure 2 – Preoperative TEE indicated existence of dilated aortic root and aortic regurgitation in the TOF patient.