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Case report

Concomitant valve-sparing aortic root replacement with tetralogy of Fallot repair in a 30-year-old patient: A case report

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ABSTRACT

Introduction and importance: Aortic regurgitation and aortic root dilatation are common features in adults with both repaired and unrepaired tetralogy of Fallot (TOF). Valve-sparing aortic root replacement (VSARR) is an effective repair for aortic regurgitation due to progressive aortic root dilatation with TOF after repair. However, the effectiveness of this technique for unrepaired patients has rarely been reported.

Case presentation: We reported a case of a 30-year-old man with cyanosis and exertional dyspnea. Echocardiography and computed tomography showed unrepaired TOF with significant aortic regurgitation and massively dilated aortic root. Aortic root replacement was mandatory. He underwent successful concomitant VSARR with TOF repair. At 6-month follow-up, he remains stable with trivial aortic regurgitation on echocardiogram.

Clinical discussion: In unrepaired TOF, the absence of sub-annular muscular rim and the unbalance of aortic sinuses make VSARR utmost challenging. If can be done successfully, VSARR preserves the native aortic valve and avoids lifelong anticoagulation therapy.

Conclusion: Concomitant VSARR with TOF repair can be safely and effectively applied for unrepaired patients presenting with progressive aortic root dilatation and significant aortic regurgitation.

1. Introduction

Aortic root dilatation is a common clinical feature of both repaired and unrepaired tetralogy of Fallot (TOF) [1]. In the repaired patients, significant aortic regurgitation (AR) and progressive aortic root and ascending aorta dilatation can be well managed with valve-sparing aortic root replacement (VSARR) [2]. This procedure ensures the durability of the native aortic valve and avoids lifelong anticoagulation therapy for the prosthetic valve. However, the efficacy of VSARR in unrepaired TOF has rarely been reported. This report describes a successful concomitant VSARR in a 30-year-old patient with unoperated TOF and moderate aortic regurgitation with aorto-ventricular dilatation.

The work has been reported in line with the SCARE 2020 criteria [3].

2. Presentation of case

A 30-year-old man was admitted to our hospital with cyanosis and exertional dyspnea. He was diagnosed with TOF and underwent a left Blalock- Taussig (BT) shunt in another hospital in his infancy. Unfortunately, his parents refused further operation for the complete repair and ceased to visit the outpatient clinic. He could not stand his general fatigue and dyspnea at the age of 30. Upon arrival, he presented with heart failure symptoms in New York Heart Association (NYHA) functional class III and cyanosis with peripheral saturation of 86%. Echocardiography and cardiac computed tomography showed unrepaired TOF with a 30 mm large sub-aortic ventricular septal defect (VSD), an overriding aorta with massively dilated sinuses (56 mm) and ascending aorta (43 mm) leading to moderated AR, sub-valvular and valvular pulmonary stenosis (peak gradient of 98 mm Hg), and a left BT shunt (6.5 mm) (Figs. 1 and 2). Left ventricle (LV) was dilated with decreased

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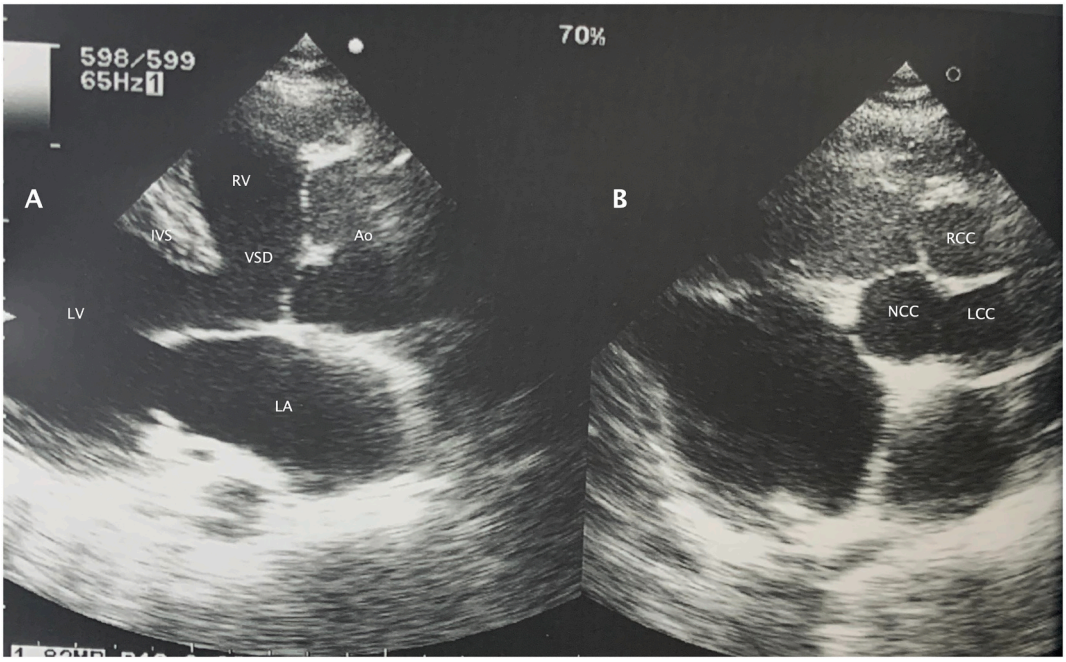

Fig. 1. (A) Parasternal long-axis echocardiographic view, showing large ventricular septal defect and dilated ascending aorta overriding the interventricular septum. (B) Parasternal short-axis view, showing three dilated aortic cups. Ao, aorta; LA, left atrium; LCC, left coronary cusp; LV, left ventricle; NCC, noncoronary cusp; IVS, interventricular septum; RCC, right coronary cusp; RV, right ventricle; VSD, ventricular septal defect.

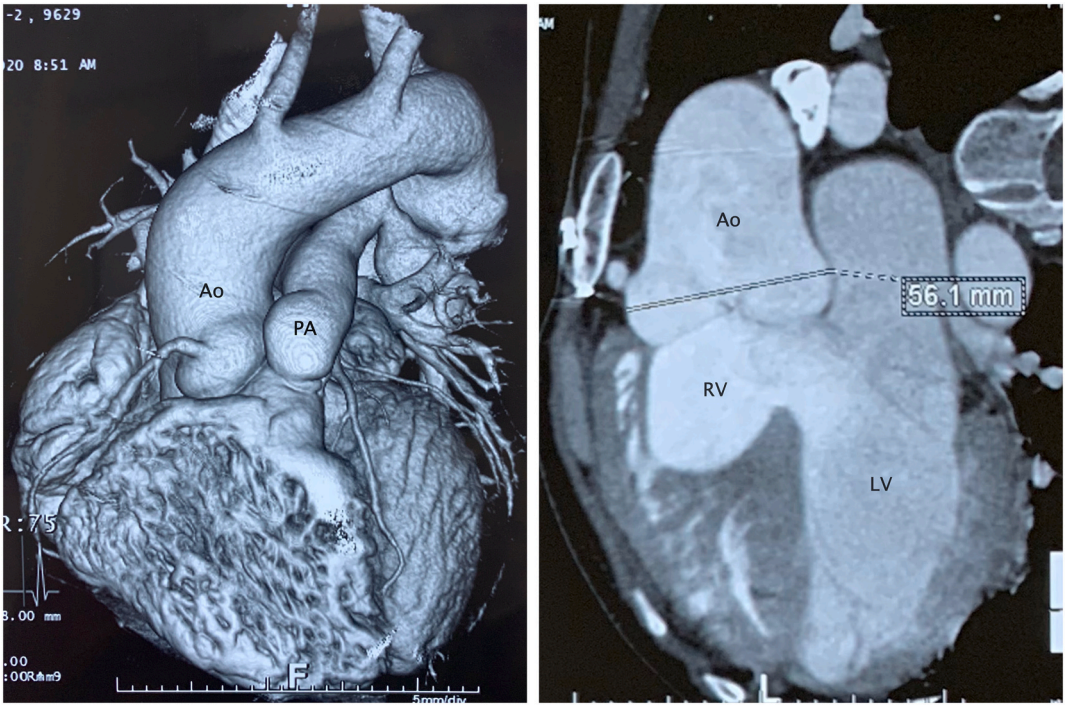

Fig. 2. Computed tomography view, showing massively dilated ascending aorta with 56 mm aortic sinus. Small pulmonary artery was also presented. Ao, aorta; LV, left ventricle; PA, pulmonary artery; RV, right ventricle.

ejection fraction (EF) (LV diastolic diameter = 56 mm, LVEF Simpson = 42%). Surgical procedures including TOF repair and VSARR were proposed.

Under median sternotomy, total cardiopulmonary bypass was established between the distal ascending aorta and both vena cavae. The BT shunt was taken down. Myocardial protection was achieved using moderate systemic hypothermia (28 °C) and antegrade Custodiol cardioplegia. Transatrial-transpulmonary approach was performed showing TOF features with a large sub-arterial VSD extending toward the membranous portion, a bicuspid pulmonary valve (PV) with a quite good PV annulus (approximately 20 mm). Sub-pulmonary portion was resected and VSD was closed by a SURGIFOC Pericardial Patch (FOC Medical, City Bell, Argentina). Pulmonary valvotomy, valve ring widening were performed through pulmonary arteriotomy. The main

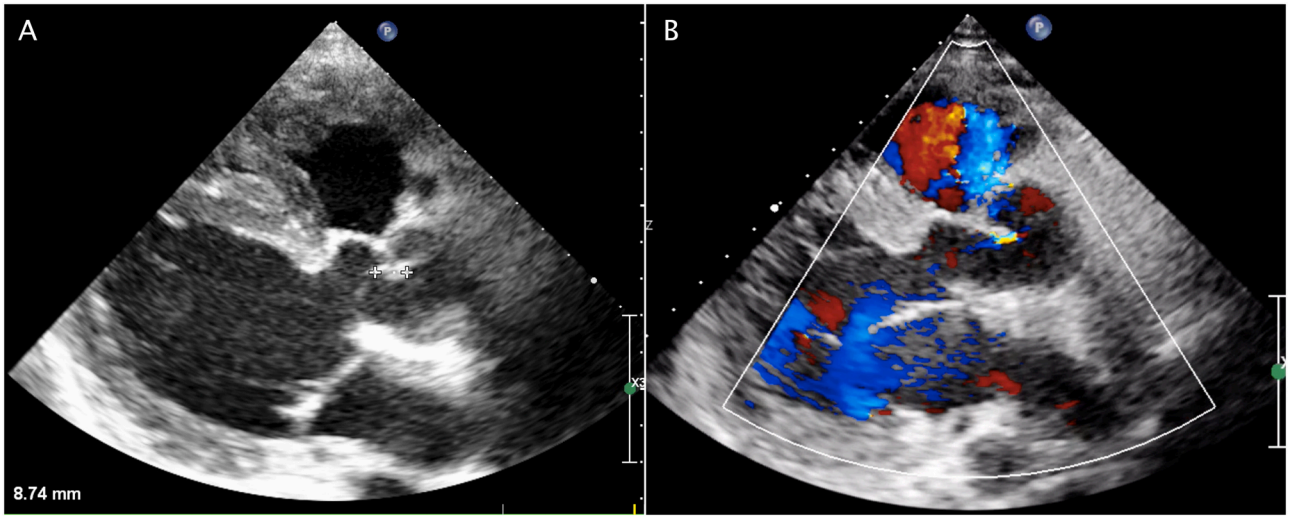

Fig. 3. Postoperative echocardiographic view, showing (A) adequate coaptation height (8.74 mm) with (B) trivial aortic regurgitation.

pulmonary artery augmentation was completed.

The ascending aorta was transected and the aortic valve was explored. It showed massively dilated, yet unbalanced aortic sinuses with predominance of the right one. Coronary ostia were excised as buttons. The ascending aorta and sinuses were removed with preservation of the aortic annulus, commissures, and valve tissue. Several subannular stitches under the right coronary and noncoronary sinuses were placed at the anterosuperior edge of the VSD patch and passed outside of the aortic annulus. A 30-mm Uni-Graft K DV (B. Braun, Aesculap AG, Germany) was used for the aortic root reconstruction. Subannular stitches of each sinus were distributed to the corresponding sinus of the graft. Care was taken to ensure that aortic commissures were in appropriate position with competent aortic valve coaptation area. The valve is then assessed for competency, symmetry, and height of leaflet coaptation. The coronary arteries were reattached in the graft, followed by distal aortic anastomosis. Cardiopulmonary bypass was safely weaned off. The bypass and aortic cross-clamp times were 394 and 336 minutes, respectively. Intraoperative echocardiogram after termination of the bypass showed trivial AR from the central area without aortic stenosis.

The postoperative clinical course was uneventful. Echocardiogram before discharge showed trivial aortic insufficiency with slightly improved LVEF (49%). At 6-month follow-up, his exercise tolerance gradually improved with NYHA functional class II with increased LVEF (55%) and trivial AR from the central area with adequate aortic valve coaptation height (8.74 mm) on echocardiogram (Fig. 3).

3. Discussion

A dilated aortic root and AR is not an infrequent finding in a growing population of adults with both repaired and unrepaired TOF. Aortic root dilatation is thought to be a combination of hemodynamic stress from volume overloading of the aorta and intrinsic aortopathy [1,6]. In our patient with massively dilated aortic root and significant AR, concomitant aortic root replacement with TOF repair is mandatory. For many years, aortic root replacement with composite mechanical valve graft (Bentall procedure) has been a standard of choice. However, patients with Bentall procedure are at risk of thrombo-embolic events, endocarditis, and long-term anticoagulation. VSARR (David procedure) not only avoids these risks but ensures the durability of the native aortic valve. VSARR has been reported as a feasible and effective procedure for progressive aortic dilatation and AR after TOF repair [4,5], but, to the best of our knowledge, VSARR in unoperated TOF has rarely been reported.

In TOF, a large VSD is located just under the aortic annulus, leaving a sub-annular gap instead of a muscular rim. Therefore, sub-annular stitches must be placed at the edge of the VSD patch which makes the procedure more challenging. The VSD patch can also distort the aortic annulus and the right cusp. Besides, the long-standing existence of VSD may lead to the prolapse of the right coronary or noncoronary cusp, or unbalance dilated sinuses. Hence, cusp plication must be performed to ensure that the cusps are meeting appropriately, and there must be some sutures performed directly on them at the same time. Moreover, the aortic root may have a very horizontal position in TOF in contrast to its normal position of 35–45° to the horizontal plane. This can make visualization of the aortic valve more difficult and compromise the ability to maintain proper geometry of the aortic valve during the procedure [2]. Special attention should be paid to handle these challenges.

4. Conclusion

In conclusion, our result shows that concomitant VSARR can be safely and effectively performed in adults with unrepaired TOF presenting with progressive aortic root dilatation and significant aortic regurgitation.

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Ethical approval

The study was exempt from ethical approval in my hospital.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

CRediT autorship contribution statement

Dr. Nguyen Sinh Hien, Dr. Nguyen Dang Hung, Dr. Pham Tan Thanh: Surgeons.

Dr. Tong Duy Phuc: Study concept, Data collection, Original manuscripts writing.

Dr. Ngo Van Thanh: Data collection, ICU caring.

Research registration

Not a clinical trial.

Guarantor

Dr. Nguyen Dang Hung, M.D.
Dr.Tong Duy Phuc, M.D.

Provenance and peer review

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Declaration of competing interest

None.

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