Turk Gogus Kalp Dama 2016;24(2):360-362
doi: 10.5606/tgkdc.dergisi.2016.11996

Aortopathy following the correction of tetralogy of Fallot

Fallot tetralojisi düzeltme ameliyatından sonra gelişen aortopati

Şafak Alpat, Mustafa Yılmaz (Uzm. Dr.), Recep Oktay Peker, Mustafa Yılmaz (Prof. Dr.)

Department of Cardiovascular Surgery, Medical Faculty of Hacettepe University, Ankara, Turkey

ABSTRACT

Tetralogy of Fallot is one of the most common forms of cyanotic congenital heart disease. Thanks to the evolution of surgical concepts and improved intra- and postoperative management strategies, its 30-year survival rate is 90%. However, once the number of these patients has grown, clinicians can face with new challenges. Pulmonary valve regurgitation is the most common long-term complication which eventually requires surgery. Although aortic valve and ascending aorta is mostly overlooked, aortic root dilatation and associated aortic valve regurgitation are rare, but major long-term complications. There are no guidelines or clear evidence-based approaches; only sporadic case series exist in the literature. Herein, we report a 31-year-old male case who was incidentally diagnosed with aortic root dilatation 23 years after total surgical repair of tetralogy of Fallot.

Keywords: Aortic regurgitation; aortic root dilatation; aortic valve; tetralogy of Fallot.

ÖZ

Fallot tetralojisi, siyanotik doğuştan kalp hastalığının en sık formlarından biridir. Cerrahi tekniklerin gelişmesi ve cerrahi sırası ve sonrası bakım stratejilerinin iyileşmesi sayesinde, 30 yıllık sağkalım oranı %90'dır. Ancak hasta sayısı arttıkça, hekimler yeni sorunlarla karşılaşabilmektedir. Uzun dönemde en sık görülen komplikasyon, nihayetinde cerrahi gerektiren pulmoner kapak yetmezliğidir. Aort kapağı ve çıkan aort genellikle gözden kaçsa da, aort kök genişlemesi ve bununla ilişkili aort kapak yetmezliği nadir fakat önemli uzun dönem komplikasyonlardır. Herhangi bir kılavuz veya aşikar kanıta dayalı yaklaşımlar mevcut değildir; literatürde yalnızca nadiren olgu serileri mevcuttur. Bu yazıda, Fallot tetralojisi total cerrahi tamirinden 23 yıl sonra tesadüfen aort kök genişlemesi tanısı konan 31 yaşında bir erkek olgu sunuldu.

Anahtar sözcükler: Aort yetmezliği; aort kök genişlemesi; aort kapağı; Fallot tetralojisi.

Surgical treatment of tetralogy of Fallot (TOF) has been evolved, since the first total correction was performed by Walton C. Lillehei in 1954. Since then, operative mortality has decreased dramatically and correction can be currently performed in infancy or early childhood.[1] Therefore, the adverse long-term effects of cyanosis and chronic hypoxia can be avoided. Although it is lower than that of the overall population, its long-term survival rates have been reported to be excellent. Pulmonary regurgitation and right ventricular outflow tract complications are the two most common indications for re-do surgeries. However, aortic root dilatation and aortic regurgitation are less common etiologies for secondary interventions.[2] In this report, we present a male case who was incidentally diagnosed with aortic root dilatation 23 years after total surgical repair of TOF.

CASE REPORT

A 31-year-old asymptomatic male patient was referred to our institution for routine follow-up after TOF repair and pulmonary valve replacement procedures. His medical history revealed total surgical repair of TOF and pulmonary valve replacement, when he was eight and 25 years old, respectively. He had no complaint and he was in the New York Heart Association class I. Physical examination findings were completely normal, except grade IV diastolic murmur over the

Available online at
www.tgkdc.dergisi.org
doi: 10.5606/tgkdc.dergisi.2016.11996
QR (Quick Response) Code

Received: May 24, 2015 *Accepted*: September 28, 2015

Correspondence: Şafak Alpat, MD. Hacettepe Üniversitesi Tıp Fakültesi Kalp ve Damar Cerrahisi Anabilim Dalı, 06100 Sıhhiye, Ankara, Turkey.
Tel: +90 312 - 305 17 73 e-mail: safakalpat@gmail.com

aortic area. Electrocardiogram showed right bundle branch block. Chest X-ray findings were normal. Transthoracic echocardiography showed moderate to severe aortic valve regurgitation with ascending aortic dilatation (48 mm), a left ventricle ejection fraction of 66%. The left ventricle end-diastolic and end-systolic diameters were 56 and 35 mm, respectively. Cardiac magnetic resonance imaging was scheduled to elucidate the ascending aortic aneurysm, right ventricular function, and any accompanying lesions regarding TOF repair. Aortic root was 52 mm and ascending aorta was in normal size interval around the origin of brachiocephalic trunk (Figure 1). Pulmonary valve was normally functioning. No accompanying lesion existed. Based on the retrospective examination of previous echocardiographic results, all aortic sizes were found to be normal. According to those findings, surgical repair was planned and a written informed consent was obtained from the patient.

Mid-line re-do sternotomy was performed. After removal of surgical adhesions, aortic root aneurysm was seen. It was substantially aneurysmatic through the right and non-coronary cusps. The aortic wall was very thin over these areas. Following establishment of cardiopulmonary bypass with left femoral artery and right atrial cannulation, the Bentall procedure with composite mechanical aortic valve conduit (27 mm) was performed under mild hypothermia. Postoperative course was uneventful; the patient was taken to the ward in the second postoperative day and discharged in the seventh postoperative day. Repeated echocardiography demonstrated normally functioning aortic valve. Left ventricle ejection fraction was 62% and left ventricle end-diastolic and systolic diameters were 50 and 33 mm, respectively. Histopathological examination of the aortic wall revealed cystic medial degeneration with disrupted elastic fibers and medial fibrosis.

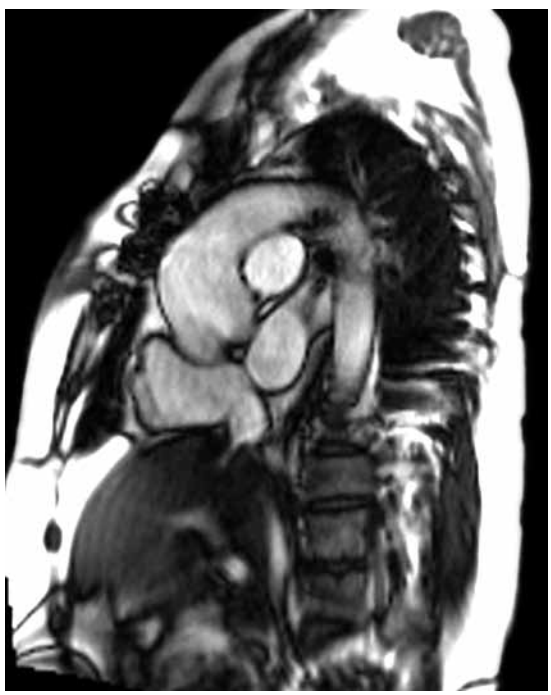

Figure 1. A sagittal section of reconstructed image of cardiac magnetic resonance imaging showing an aneurysmatic aortic root and normal aortic arch.

DISCUSSION

Aortic valve and root pathologies are known complications of unrepaired TOF.[3] However, recent studies have shown that progressive aortic root dilatation and aortic valve regurgitation can be seen even in repaired TOF patients.[3] Its prevalence is 28.9%, when a threshold of 40 mm is used to define an aortic root dilatation.[3] Moreover, 12.5% of patients with corrected TOF and dilated aortic roots have moderate to severe aortic regurgitation.[3]

There are several pathophysiological explanations. According to the developmental (embryological) theory, the failed migration of the neural crest cells to the aorticopulmonary septum and subsequent unequal conotruncal septation may lead to larger aortic component with smaller pulmonary trunk.[4] Also, volume loading of aorta as a consequence of the right-to-left shunt and subaortic jet flow due to subaortic ventricular septal defect may worsen pre-existing aortic pathology.[4] Even in some reports, it was demonstrated that TOF patients had intrinsic aortopathy characterized by fibrosis, 'cystic' medial necrosis, elastic fragmentation, and disruption of elastic lamellae similar to those with Marfan syndrome and bicuspid aortopathy.[5] Irrespective of the pathophysiological mechanism, aortic regurgitation and aortic aneurysm or dissection are encountered in natural history and need surgical intervention, eventually.[6]

Furthermore, several risk factors have been previously defined for aortic root dilatation following the TOF repair including male sex, prolonged time from palliation to repair, presence of pulmonary atresia, right aortic arch, and the older age at repair. However, in a recent multi-center, cross-sectional study, multivariate analysis showed that male sex was the only independent risk factor, although other variables were found to be statistically significant in the univariate analysis.[3] Among these risk factors, our case was male and underwent corrective surgery, when he was eight years old. Although the age at

repair was not found to be statistically significant, we consider that increased transaortic flow might result from the presence of a right-to-left shunt, as the major hemodynamic component in aortopathy.

Despite growing population of these patients, there is still no evidence-based cut-off level of aortic root for surgical intervention. Nonetheless, it has been reported that 40% of patients who presented with dissections had aortic diameters <50 mm. Therefore, in TOF patients with severe aortic regurgitation which requires aortic valve replacement, careful assessment of aortic root and ascending aorta is crucial. In our case, cardiac magnetic resonance imaging helped us to evaluate aortic root and plan procedure.[6]

In conclusion, it is important to assess aortic valve, aortic root, and ascending aorta during routine follow-up of previously operated tetralogy of Fallot patients. Early diagnosis and management would result in improved better functional status and higher survival rates in this patient population.

Declaration of conflicting interests

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

Funding

The authors received no financial support for the research and/or authorship of this article.

REFERENCES

1. Pigula FA, Khalil PN, Mayer JE, del Nido PJ, Jonas RA. Repair of tetralogy of Fallot in neonates and young infants. Circulation 1999;100(19 Suppl):II157-61.
2. Oechslin EN, Harrison DA, Harris L, Downar E, Webb GD, Siu SS, et al. Reoperation in adults with repair of tetralogy of fallot: indications and outcomes. J Thorac Cardiovasc Surg 1999;118:245-51.
3. Mongeon FP, Gurvitz MZ, Broberg CS, Aboulhosn J, Opotowsky AR, Kay JD, et al. Aortic root dilatation in adults with surgically repaired tetralogy of fallot: a multicenter cross-sectional study. Circulation 2013;127:172-9.
4. Tan JL, Gatzoulis MA, Ho SY. Aortic root disease in tetralogy of Fallot. Curr Opin Cardiol 2006;21:569-72.
5. Niwa K, Perloff JK, Bhuta SM, Laks H, Drinkwater DC, Child JS, et al. Structural abnormalities of great arterial walls in congenital heart disease: light and electron microscopic analyses. Circulation 2001;103:393-400.
6. Ishizaka T, Ichikawa H, Sawa Y, Fukushima N, Kagisaki K, Kondo H, et al. Prevalence and optimal management strategy for aortic regurgitation in tetralogy of Fallot. Eur J Cardiothorac Surg 2004;26:1080-6.