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A Tetralogy of Fallot Patient Survived Only with a Classical Blalock-Taussig Shunt for more Than 50 Years

Fallot Tetralojisi Hastasının Klasik Blalock-Taussig Şantı ile 50 Yıldan Uzun Süre Sağ Kalımı

CASE IMAGE
OLGU GÖRÜNTÜSÜ

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This case report discusses the case of a patient with tetralogy of Fallot (TOF) who underwent a classic Blalock–Taussig shunt (BTS) at the age of 2 years. Despite having no routine follow-up visits and taking no medications, he had a long asymptomatic life. At the age of 53 years, he started to complain of dizziness and recurrent attacks of syncope for 2 months for which he was admitted and diagnosed with complete heart block. A permanent pacemaker was inserted to regulate heart function. Two-dimensional and three-dimensional echocardiography findings (Figure 1, Video 1*) revealed uncorrected TOF with an overriding aorta, two ventricular septal defects (VSDs)—a large perimembranous VSD and another small muscular one, and marked infundibular stenosis with marked right ventricular hypertrophy. Cardiac computed tomography (Cardiac CT) (Figure 2, Video 2*) confirmed the diagnosis of TOF with infundibular and valvular pulmonary stenosis. In addition, it showed a patent classic BTS between the left subclavian artery (SCA) and the left pulmonary artery (LPA), aneurysmal dilatation of the LPA, and anomalous origin of the left anterior descending (LAD) artery arising from the right coronary cusp (RCC) and passing in front of the right ventricular outflow tract (RVOT) to reach the anterior interventricular groove as well as multiple small aortopulmonary collaterals. Because of his stable condition (well-maintained oxygen saturation and normal 6-minute walk test) and the high mortality associated with any corrective or palliative surgery, he was discharged on close follow-up.

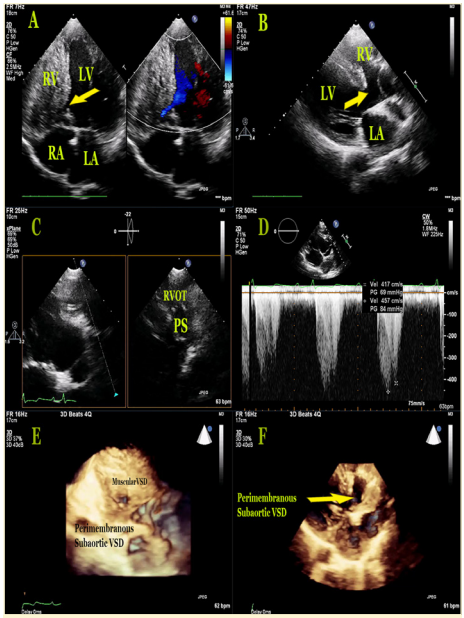

Figure 1. (A-F) 2D and 3D TTE showing uncorrected TOF with an overriding aorta, two VSDs; a large perimembranous VSD and a small muscular one with marked right ventricular hypertrophy (RV free wall=12 mm) with hypoplastic MPA with two levels of stenosis, infundibular pulmonary stenosis and bicuspid pulmonary valve with valvular stenosis (Pvel=4.5 m/s, PG=84 mm Hg).
MPA, main pulmonary artery; RV, right ventricle; TOF, tetralogy of Fallot; TTE, transthoracic echocardiogram; VSD, ventricular septal defect.

The survival of patients with uncorrected TOF is rarely reported, and frequently associated with a well-developed right ventricle, mild pulmonary stenosis, or well-maintained pulmonary blood flow by systemic to pulmonary collaterals or persistent patent ductus arteriosus. The prognosis of TOF treated only with classical BTS remains unclear. In this case, the causes for long-term survival may be the combination of the for-

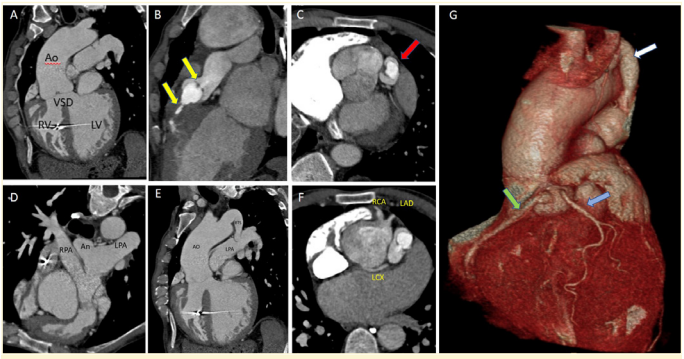

Figure 2. (A-G) Cardiac CT showing (A) Overriding aorta and subaortic perimembranous VSD; (B) Infundibular and valvular stenosis (yellow arrows); (C) Bicuspid thickened pulmonary valve (red arrow); (D) Aneurysmal dilation (An) of the LPA; (E) Classic BTS between the left SCA and LPA; (F) Anomalous origin of the LAD from the RCC; (G) Classic BTS (white arrow), anomalous LAD passes in front of the RVOT (blue arrow) and RCA (green arrow).

BTS, Blalock-Taussig shunt; CT, computed tomography; LAD, left anterior descending; LPA, left pulmonary artery; RCA, right coronary artery; RVOT, right ventricular outflow tract; SCA, subclavian artery; VSD, ventricular septal defect.

ward pulmonary flow (despite severe pulmonary stenosis) and the well-formed BTS shunt (with nonsignificant stenosis). The patient has small aortopulmonary collaterals that originate from the descending aorta, yet there are no major aortopulmonary collateral arteries (MAPCAs). This case shows that classical BTS has a potential of long-term patency with good functional capacity resulting in appropriate pulmonary blood flow by the BTS resulting in long-term survival.

*Supplementary video files associated with this article can be found in the online version of the journal.

Informed Consent: Informed consent was obtained from the patient for the publication of the case image and the accompanying images.