

A Long-term unrepaired Fallot tetralogy survivor treated only with a Classical Blalock-Taussig Shunt

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April 28, 2020

Abstract

Tetralogy of Fallot (TOF) is the most common etiology of congenital cyanotic heart disease, and Blalock-Taussig shunt (BTS) operation is considered the first-step management to maintain pulmonary blood flow in TOF patients. Complete repair of TOF is the standard surgical treatment that should be performed in infancy or early childhood for improved long-term survival. However, the prognosis of TOF patients treated by only palliative operation remains uncertain. We report a man with TOF underwent classic BTS operation at 2 years of age. Despite no medication, he had a long asymptomatic life. At 53 years of age, he started to complain of dizziness and recurrent attacks of syncope due to complete heart block (CHB) and inserted a permanent pacemaker. 2D and 3D Echocardiography showed uncorrected TOF with an overriding aorta, two ventricular septal defects (VSDs) were seen; a large inlet type VSD and another small muscular one with marked right ventricular hypertrophy with marked infundibular stenosis. Cardiac Computed Tomography (Cardiac CT) showed TOF with noted severely stenotic right ventricular outflow tract and pulmonary flow was maintained through a patent classical BTS between left subclavian artery (SCA) and the left pulmonary artery (LPA). Due to his stable condition, he was discharged on close up follow-up visits.

Case Presentation

A Tetralogy Fallot (TOF) patient underwent a classic Blalock-Taussig Shunt (BTS) at 2 years of age. Despite no medication, he had a long asymptomatic life. At 53 years of age, he started to complain of dizziness and recurrent attacks of syncope due to complete heart block (CHB) and inserted a permanent pacemaker. 2D and 3D Echocardiography (Figure 1, Video 1) showed uncorrected TOF with an overriding aorta, two ventricular septal defects (VSDs) were seen; a large inlet type VSD and another small muscular one with marked right ventricular hypertrophy with marked infundibular stenosis. Cardiac Computed Tomography (Cardiac CT) (Figure 2, Video 2) showed DORV-TOF type with noted severely stenotic right ventricular outflow tract and pulmonary flow was maintained through a patent classical BTS between left subclavian artery (SCA) and the left pulmonary artery (LPA) and a noted accessory LAD arises from RCC and passing in front of the RVOT. Due to stable condition of the patient, he was discharged on close up follow-up. Survival of uncorrected TOF patients is rarely reported and frequently associated with a well-developed left 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 our case may the causes for the long-term survival although severe pulmonary stenosis was the well-formed BTS shunt with non-significant stenosis and the aorto-pulmonary collaterals. This case

shows that classical BTS has a potential of long-term patency with good functional capacity resulting in appropriate pulmonary blood flow by BTS resulting in long-term survival.

Disclosure : Authors declare no conflict of interest



