Surgical correction of tetralogy of Fallot in a 61-year-old patient

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Abstract  Tetralogy of Fallot (TOF) is a congenital heart disease that is usually diagnosed and treated during infancy. Only 3% of such patients reach the age of 40 without surgery. We describe a 61-year-old woman with uncorrected TOF that was successfully treated by radical surgery. The patient presented with exertional dyspnea, insomnia, and malaise. Echocardiography and cardiac catheterization indicated a dilated, severely hypertrophic right ventricle, ventricular septal defect, an overriding aorta, and infundibular stenosis in the right ventricular outflow tract. All symptoms disappeared after full surgical correction, which remains the preferred treatment for adult TOF because it confers long-term survival and an improved quality of life.

Key words  Tetralogy of Fallot · Adult congenital heart disease

Introduction

Tetralogy of Fallot (TOF) is the most frequent cyanotic type of congenital heart disease, and it is generally treated by total surgical correction during the first few years of life. Without surgery most of these patients would die during childhood. Survival rates are 66%, 40%, 11%, 6%, and 3% at 1, 3, 20, 30, and 40 years of age, respectively.1 Here we describe a woman with TOF who survived until 61 years of age without surgical intervention.

Case report

A 61-year-old woman was admitted to our hospital with slowly increasing symptoms of exertional dyspnea, insomnia, and malaise. She had been troubled with insomnia for more than 30 years and had used sleeping pills regularly. Exertional dyspnea had appeared 6 months ago, and it had been worsening gradually. Her medical history included only a cardiac murmur during childhood, and she had delivered a child.

Blood gas analysis showed pH 7.48, with PaCO2 and PaO2 of 26.9 and 40.5 mmHg, respectively. Routine biochemistry showed normal renal function (creatinine 0.9 mg/dl, blood urea nitrogen 16.7 mg/dl), white blood cell count 6500/mm3, hemoglobin 15.2 g/dl, and C-reactive protein <0.1 mg/dl.

Electrocardiography showed a sinus rhythm with signs of right atrial overload, right axis deviation, and right ventricular hypertrophy with strain. Echocardiography confirmed a dilated, severely hypertrophic right ventricle (RV) and a ventricular septum defect (VSD) in the perimembranous region. An overriding aorta and severe infundibular stenosis in the right ventricular outflow tract (RVOT) were also evident. The maximum gradient in the RVOT was 144 mmHg. The left ventricle (LV) was small, but wall motion was normal [left ventricular end-diastolic volume index (LVEDVI) 51.1 ml/m2 and LV ejection fraction (LVEF) 64%].

Cardiac catheterization demonstrated normal LV function and coronary artery system. A VSD with bidirectional
flow and RVOT stenosis were reconfirmed (Fig. 1). The systolic pressures in the RV and LV were 140 and 150 mmHg, respectively. The peak gradient across the pulmonary valve/RVOT was 120 mmHg. In room air, the arterial, pulmonary artery (PA), and RV saturations were 87.1%, 61.4%, and 52.3%. The branch pulmonary arteries were relatively large, and the Qp/Qs ratio was about 0.9:1.0.

The patient was started on diuretics and β-blocker therapy, which is a typical therapeutic strategy for TOF in children. However, the symptoms worsened, and the PaO₂ dropped to 25.3 mmHg. Stopping drug therapy somewhat relieved the worsening symptoms.

Full surgical correction was planned in the light of the severity of the symptoms, the failure of drug control, preserved RV function, and normal PA pressure. The surgery proceeded under full cardiopulmonary bypass. The hyper-trophic muscle obstructing the RVOT was resected (Fig. 2), and the VSD was closed with a Gore-Tex patch. The pulmonary valve appeared intact, and the diameter was large enough (>20 mm) that we judged that valvotomy was unnecessary. The RVOT was enlarged using an equine pericardium patch that was preserved in glutaraldehyde.

The postoperative period was uneventful, and the patient was discharged 2 weeks later with class I New York Hospital Association (NYHA) status. She continues to do well 4 years after the procedure without the need for care.

Discussion

Tetralogy of Fallot is the most frequent form of cyanotic congenital heart disease worldwide. First described in 1888 by the French physician Etienne-Louis Arthur Fallot, it comprises a ventricular septal defect, an overriding aorta, obstructed right ventricular outflow, and right ventricular hypertrophy.

Most patients with TOF have cyanosis from birth or from the first year of life due to right-to-left shunting. The operative mortality of surgery in children is <3%, and early correction permits normal growth and development. Thus, early primary repair is regarded as the treatment of choice. Without surgery, few patients with TOF reach adulthood, as the average life expectancy is 12 years. Although 10% of TOF patients might survive into their thirties, only 3% reach their forties or beyond. Our patient had no medical history except a cardiac murmur identified during childhood, and she had delivered a baby without any problems. Her symptoms appeared after the age of 60 years.

Several factors might contribute to the increased survival of patients with uncorrected TOF. The gradual development of RVOT obstruction might be associated with increased survival, and the development of LV hypertro-