

Successful Surgical Repair of an Elderly Patient with Tetralogy of Fallot: A Case Report

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We report on a rare case of a 61-year-old female patient with tetralogy of Fallot who had a late radical repair. The patient had no previous surgical interventions, and her postoperative course was uneventful. One year after the repair, the patient is doing well and has no symptoms of cardiac failure. Surgical treatment greatly benefited our patient by improving her functional status and extending her life expectancy. (Ann Thorac Cardiovasc Surg 2007; 13: 125–7)

Key words: tetralogy of Fallot, adult patient, late surgical repair

Introduction

Although surgical repair of untreated adult patients with tetralogy of Fallot (TOF) remains controversial, we believe that late surgical repair in selected patients who have appropriate indications for radical surgery is effective. We report a rare case of an elderly patient with TOF in whom a successful radical operation was done. We also review the specific issues that should be considered prior to surgery in this type of patient.

Case

A 61-year-old female patient was referred to our department for surgical treatment of TOF. Her chief complaint was dyspnea on exertion, and she was categorized as New York Heart Association (NYHA) functional class III. At 35 years of age, the patient was initially diagnosed as having TOF; surgical correction was suggested, but the patient refused treatment. At 61 years, the patient's symptoms worsened, and she requested surgical intervention. The preoperative electrocardiogram (ECG) showed sinus rhythm, complete right bundle branch block, and occa-

sional monofocal premature ventricular contractions. Laboratory data included a hemoglobin level of 21.1 g/dL, a hematocrit of 64.2%, and a mean systemic arterial saturation of 80%. Other laboratory data were normal. The echocardiogram showed a large subaortic type of ventricular septal defect (VSD), stenosis between the right ventricular outflow tract (RVOT) and the main pulmonary artery trunk, and a left ventricular ejection fraction of 73% with normal wall motion. The results of the cardiac catheterization are shown in Table 1. The peak pressure gradient across the RVOT was 96 mmHg. Angiography revealed stenosis between the RVOT and the main pulmonary artery with a hypoplastic left pulmonary artery and dilatation of the right pulmonary artery (Fig. 1, A and B). A ventilation and perfusion scintiscan was done to evaluate left pulmonary function. The ventilation capacity was normal, but the vascular capacity was reduced due to left pulmonary artery hypoplasia. Based on these preoperative findings, the patient had normal left and right ventricular function. Hypoxia and not cardiac failure was considered to be the primary problem; therefore, a radical operation was indicated. During the operation, the RVOT was opened across the pulmonary valve, and the incision was extended to the bifurcation and into both the right and left pulmonary arteries to release the stenosis. All leaflets of the pulmonary valve were removed, since they were severely thickened with calcification. Surgical repair included patch augmentation from the RVOT to the main pulmonary artery across the pulmonary valvular annulus and patch closure of the VSD and the atrial septal defect. After declamping

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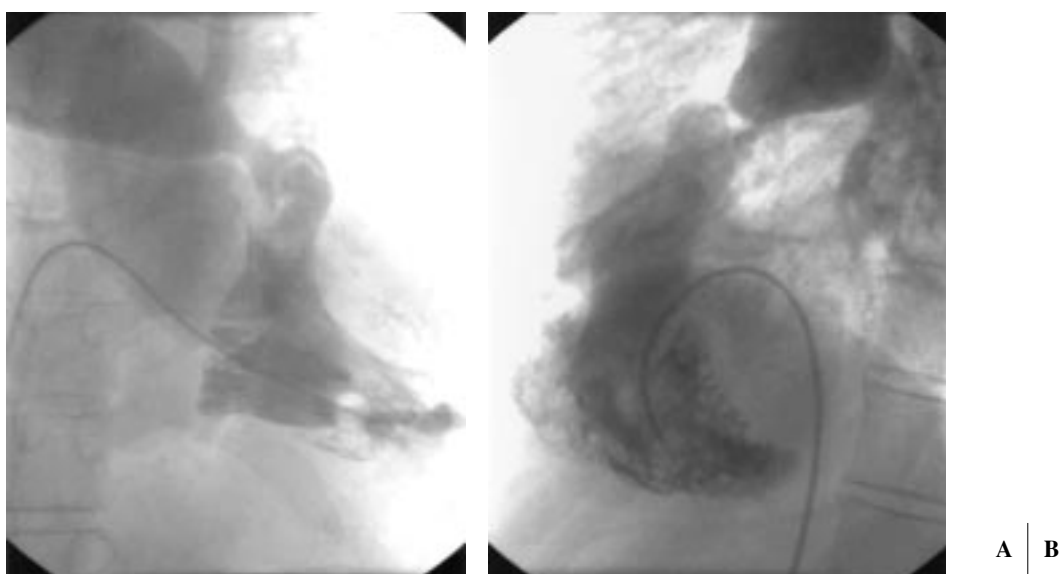
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Table 1. Preoperative cardiac catheterization results

Pressure study (mmHg)
RA 8, RV 121/10, PA 25/9 (17), PCWP 12/7 (8), LV 118/5, Ao 122/54
Coronary artery angiography
1) Normal structure without significant stenoses
2) Collateral flow from the left coronary artery to the left pulmonary artery
Right ventricular and aortic angiography
1) RVOT obstruction and a large subaortic type of VSD
2) Slight dilatation of the ascending aorta and closure of the ductus arteriosus
Calculated pulmonary-systemic flow ratio
Qp/Qs 0.79

RA, right atrium; RV, right ventricle; PA, pulmonary artery; PCWP, pulmonary capillary wedge pressure; LV, left ventricle; Ao, aorta; RVOT, right ventricular outflow tract; VSD, ventricular septal defect; Qp/Qs, pulmonary-systemic flow ratio.

**Fig. 1.**

- A:** Preoperative right ventricular cineangiography in the anteroposterior (AP) view shows stenosis around the pulmonary valve. Supravalvular narrowing extends to the origin of the main pulmonary artery. Poststenotic dilatation is present from the main pulmonary artery to the right pulmonary artery; hypoplasia of the left pulmonary artery is also present.
- B:** Left anterior oblique (LAO) view also shows stenosis in the right ventricular outflow tract (RVOT).

the aorta, the patient was easily weaned from cardiopulmonary bypass. Although a left pleural effusion required drainage, the patient's postoperative course was otherwise uneventful. A postoperative ECG showed sinus rhythm without premature ventricular contractions. She improved to NYHA functional class I and remains in good health 1 year following the radical operation.

Discussion

The natural history of patients with TOF and pulmonary

artery stenosis is determined by the severity of their right ventricular and pulmonary arterial outflow tract obstruction. Most surgically untreated patients with TOF and pulmonary stenosis die within the first 10 years of life, and the remaining patients rarely survive beyond 40 years of age.¹⁾ Nevertheless, there are occasionally patients who remain undiagnosed or untreated until adulthood. These patients have a mild form of RVOT obstruction and pulmonary artery stenosis, as well as a low right-to-left shunt ratio. However, over time, due to pressure overload, even these patients develop right ventricular hypertrophy and

RVOT obstruction, including valvular and infundibular obstruction. Furthermore, these patients have particular problems that are related to the effects of long-standing hypoxia. In particular, in untreated adult patients with TOF, such problems include: (i) impairment of ventricular function due to fibrosis; (ii) progressive arterial desaturation and polycythemia that is perpetuated not only by an increase in pulmonary stenosis but also by a tendency to thrombosis of the pulmonary arteries; (iii) valvular impairment, including aortic regurgitation, due to dilatation of the aortic root, as well as tricuspid valve regurgitation due to right ventricular dilatation; and (iv) frequent ventricular arrhythmias, which are thought to cause sudden death. Therefore, surgical correction can benefit even untreated adult patients with TOF by alleviating their long-standing hypoxia and ventricular overload. Our present patient had in fact developed hypoxia. Despite progressive oxygen desaturation, polycythemia, slight dilation of the ascending aorta (maximum diameter, 45 mm) without aortic regurgitation, and monofocal ventricular premature contractions, the patient's left and right ventricular function remained within the normal range, and the pulmonary vascular bed was sufficient. We decided that the patient's preoperative condition was adequate for surgical intervention and that an improvement in the patient's functional status could be expected postoperatively. Therefore, a radical operation for TOF without any additional procedures was planned. We did a transannular patch to release the RVOT obstruction caused by severe thickness with calcification of the pulmonary valve and annulus. Based on a review of pertinent reports, transannular patch repair of RVOT obstruction in previously untreated adult patients with TOF physiology is not a common procedure. Up to 21.2% of these patients who required a patch repair of their RVOT obstruction required a transannular patch.²⁻⁵ Moreover, it was reported that reconstruction of the RVOT with a patch was significantly related to perioperative mortality.²⁻⁵ The reason for the higher mortality in this group was suggested to be due to chronic pulmonary regurgitation, which has a detrimental effect on right ventricular function. It is possible that, in this selected group of patients, postoperative pulmonary regurgitation is not tolerated, and, together with impaired right ventricular function, it may lead to irreversible right ventricular decompensation. Previously published reports have also mentioned exclusion criteria, such as severe cyanosis, right and left ventricular dilatation, elevated end-diastolic pressure, and tricuspid and aortic regurgitation. These should be carefully and individually

evaluated when deciding whether corrective or palliative surgery should be recommended for a particular patient.²⁻⁵ According to recently published data, the operative mortality rate in adolescent and adult patients with previously untreated TOF ranged between 2% and 16%.²⁻⁵ Unexpectedly, advanced age does not appear to carry a higher operative risk. Thus, a radical repair can be recommended for adult patients, since the operative procedure has an acceptable morbidity and mortality rate, though it is higher than in pediatric series.²⁻⁵ After surgical repair, a marked improvement can be expected in the patient's functional class and quality of life; the improvement is superior to that seen in patients given medical therapy alone. Furthermore, the literatures show that, following the operation, these patients can have a normal life expectancy.²⁻⁵ However, careful follow-up is required to monitor whether there are residual hemodynamic abnormalities and whether significant rhythm disturbances develop.

Conclusion

We safely performed a radical repair on a previously untreated adult patient with TOF and pulmonary artery stenosis. The patient's postoperative functional capacity improved to NYHA functional class I and the patient remains in good health. Thus, surgical repair is an effective option for this specific group of patients.

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