Case reports

Late concomitant repair of tetralogy of Fallot and aortic valve replacement following successful pregnancy

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The importance of patient follow-up after repair of tetralogy of Fallot, with special attention to the development of arrhythmias, has been widely studied. It is only recently that postoperative problems relating to the aortic root of these individuals have been looked into. The present case report refers to a patient with tetralogy of Fallot who underwent complete correction, together with aortic valve replacement at 33 years of age following a successful pregnancy. To our knowledge, this is the third report of primary repair of tetralogy of Fallot with simultaneous aortic valve replacement for severe aortic regurgitation. Our patient is unique in that she had uncorrected and unpalliated tetralogy of Fallot and severe aortic regurgitation and still went through an uneventful pregnancy and delivery. There is only one report of successful pregnancy and delivery in uncorrected tetralogy, but this was not associated with aortic regurgitation.

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Introduction

The importance of patient follow-up after repair of tetralogy of Fallot, with special attention to the development of arrhythmias, has been widely studied. It is only recently that postoperative problems relating to the aortic root of these individuals have been looked into[1]. The present case report refers to a patient with tetralogy of Fallot who underwent complete correction, together with aortic valve replacement at 33 years of age following a successful pregnancy.

To our knowledge, this is the third report of primary repair of tetralogy of Fallot with simultaneous aortic valve replacement for severe aortic regurgitation, and the first report of a successful pregnancy in these circumstances.

Case report

Our patient is the eldest of six siblings born to unrelated parents. A clinical diagnosis of ventricular septal defect was made at 15 months of age. She remained asymptomatic but at 15 years of age, she was admitted to hospital with chest pain, dyspnea and joint pains. At this stage, she was not cyanosed but clinical signs of right ventricular outflow tract obstruction were noted, along with an early diastolic murmur in the left second intercostal space. ECG showed left ventricular hypertrophy and chest X-ray showed an increased cardiothoracic ratio with mild pulmonary plethora. The patient was transferred to a tertiary center for further investigation. Cardiac catheterization showed tetralogy of Fallot along with marked aortic incompetence, and a decision for conservative management was taken. Her clinical conditions remained stable and 15 years later, she had a successful pregnancy with a normal vaginal delivery. Two years after the delivery, she began to experience frequent episodes of chest pain and dyspnea on minimal exertion. Her clinical findings remained unchanged. Echocardiography revealed tetralogy of Fallot with prolapse of an aortic valve cusp into the ventricular septal defect and significant incompetence, a left ventricular end-diastolic diameter of 60 mm and severe right ventricular outflow tract obstruction occurring mostly at the pulmonary infundibular level.

Repeat catheterization revealed a peak subvalvar right ventricular outflow tract gradient of 40-50 mmHg, and predominantly right-to-left shunting across the ven-

Table I. Tetralogy of Fallot (TOF) associated with aortic valve disease.

Author	Case/s
Suzuki et al.[2]	Uncorrected TOF with aortic valve prolapse into VSD (n = 5)
Abraham et al.[3]	Uncorrected TOF with aortic root dilation and regurgitation (n = 10)
Niwa et al.[4]	Corrected TOF with late aortic root dilation and regurgitation (n = 32)
Dodds et al.[5]	TOF with iatrogenic aortic regurgitation complicating surgical repair (n = 1)
Dodds et al.[5]	TOF with aortic valve replacement and recurrent VSD closure (n = 3)
Kiziltan et al.[6], Takeuchi et al.[7]	TOF repair with simultaneous aortic valve repair (n = 1)
Gelfman and Levine[8], Glancy et al.[9]	TOF with aortic valve disease following bacterial endocarditis (n = 3)
Collins et al.[10]	TOF with aortic valve disease following rheumatic fever (n = 1)
Glancy et al.[9]	TOF with bicuspid aortic valve (n = 1)

VSD = ventricular septal defect.

tricular septal defect (left to right at 0.6 l/min; right to left at 1.7 l/min). The aortic saturation was 91%. There was no left ventricular outflow tract obstruction and angiography showed severe (grade 3) aortic regurgitation with left ventricular dilation and dye crossing to the right ventricle demonstrating a subaortic ventricular septal defect. Right ventricular angiography showed severe subvalvar outflow tract obstruction.

Total correction of tetralogy of Fallot was carried out via patch repair of the ventricular septal defect and resection of the right ventricular outflow tract obstruction. The pulmonary valve was normal. Aortic valve replacement with a 25 mm St. Jude valve was carried out concomitantly. She was discharged from hospital 12 days after surgery.

Discussion

Significant aortic regurgitation is an uncommon finding in patients with tetralogy of Fallot, usually due to aortic root dilation and occasionally necessitating surgical intervention to the left outflow tract (Table I)[2-10]. Aortic dilation is worse in those individuals who are not corrected within the first decade of life[5,11]. This is believed to be due to an increased blood flow through the left ventricular outflow tract secondary to right-to-left shunting[12-14]. Biopsies from these dilated aortic roots are histologically characterized by non-inflammatory loss of smooth muscle cells, mucoid degeneration, and fragmentation of the elastic fibers within the media[15]. These changes closely resemble those found in association with bicuspid aortic valves and Marfan's syndrome. Aortic regurgitation due to trauma of the aortic root at total surgical correction has also been implicated[4].

However, only a small proportion of patients will eventually need aortic valve surgery. In 1975, Rieker et al.[16] reported that 15-18% of their patients had mild aortic regurgitation due to aortic root dilation after complete repair of tetralogy of Fallot. More recently, Dodds et al.[5] reported 4 patients with complete Fallot repair who required aortic valve replacement. Another recent study showed that 15% of corrected patients had aortic root dilation with incompetence severe enough to require valve replacement in 2 patients[4]. In addition, Chugh et al.[17] reported 46 out of 62 individuals with aortic regurgitation after Fallot correction, and in 76% of these, the regurgitation was mild.

Niwa et al.[4] have identified the potential risk factors relating to aortic root dilation in tetralogy of Fallot. These were male sex, pulmonary atresia, right aortic arch, and delayed total correction, irrespective of previous palliation.

Our patient is unique in that she had uncorrected and unpalliated tetralogy of Fallot and severe aortic regurgitation and still went through an uneventful pregnancy and delivery. Our patient was fortunate in that the degree of her right ventricular outflow tract obstruction was not severe, as evinced by an outflow gradient of only 40-50 mmHg with bidirectional shunting. Pregnancy requires a significant increase in cardiac output, and the hemodynamic changes worsen both aortic stenosis and right-to-left shunting, if present. Aortic incompetence, on the other hand, is well tolerated because of the decrease in afterload that occurs in pregnancy and is precipitated by an unsupported aortic root. There is only one report of successful pregnancy and delivery in uncorrected tetralogy, but this was not associated with aortic regurgitation[18]. Surgical repair in congenital heart disease, just as so many other things in life, is better late than never.

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