

Case Report

Beating Heart Replacement of the Pulmonary Valve in a Patient with Surgically Corrected Tetralogy of Fallot

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We describe the case of a young man who underwent anatomical correction of Tetralogy of Fallot in childhood. The patient developed severe pulmonary valve regurgitation, moderate tricuspid regurgitation and dilation of the right heart chambers. He underwent a redo sternotomy, replacement of the pulmonary valve and tricuspid annuloplasty with beating heart under extracorporeal circulation. Although this technique is more technically demanding, it avoids the use of cardioplegia and the complications of myocardial ischemia and reperfusion.

Tetralogy of Fallot is one of the most common cyanotic congenital cardiac diseases. The first successful repair was performed in 1954 and long-term results showed a 30-year survival of 91%.¹ However, the progressive right ventricular volume overload from the long-standing regurgitation of the pulmonary valve (PV) results in severe late complications. Exercise limitation, right and left ventricular dysfunction, electrocardiographic abnormalities and, most importantly, the development of life threatening atrial and ventricular arrhythmias, are the commonest complications.² It seems that the main substrate of sudden death presentation in these patients is pulmonary regurgitation and the preservation or restoration of PV function may reduce that risk.²⁻⁵

Case description

This case concerns a 32-year-old man with a medical history of tetralogy of Fallot surgically corrected 19 years before. On routine follow-up, the patient had developed dyspnoea on exertion. Echocardiographic

study revealed severe PV regurgitation (4th grade), a 5.5 cm aneurysm of the main pulmonary artery (PA) extending from the right ventricular outflow tract to just before the PA bifurcation, mild distension of the ascending aorta and significant dilation of the right heart chambers with moderate (2nd grade) tricuspid valve regurgitation and mild right ventricular systolic dysfunction (Figure 1). His ECG showed right bundle branch block and prolongation of the QRS complex (185 ms). His remaining medical history was unremarkable. A redo sternotomy was performed and cardiopulmonary bypass was instituted with bicaval and ascending aorta cannulation. After the opening of the main PA, the pericardial patch that was responsible for the aneurysmatic dilation was excised. A bioprosthetic PV (Mosaic Medtronic, Inc.) No. 29 was inserted and reconstruction of the PA was performed with a Dacron graft. The right atrium was incised and a Kay type annuloplasty of the tricuspid valve was carried out. All the above procedures were performed with the beating heart technique on cardiopulmonary bypass. The patient was tak-

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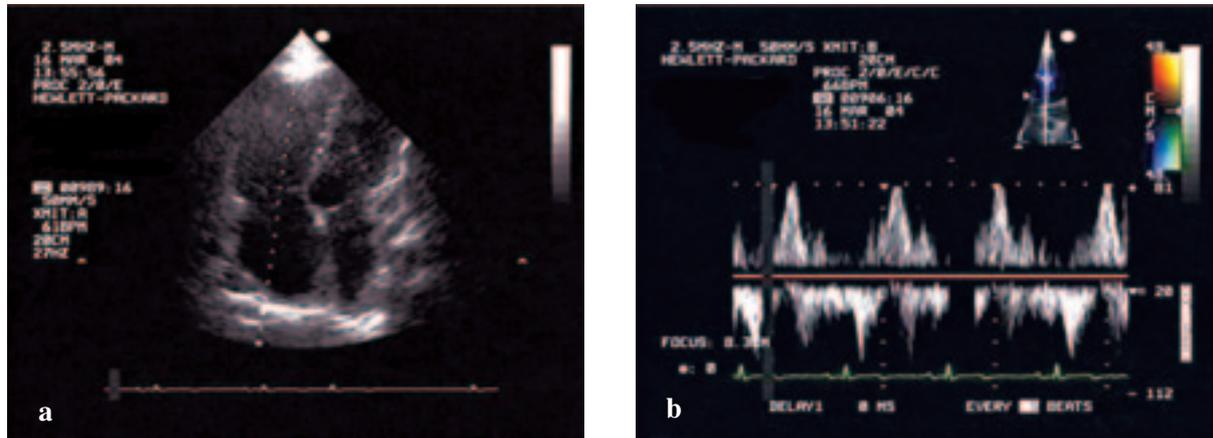


Figure 1. Preoperative cardiac echo. a. Significant dilation of the right heart chambers; b. Severe pulmonary valve regurgitation (4th degree).

en off the cardiopulmonary bypass without any inotropes and the postoperative course was uneventful. Three months after the operation, the echo findings showed good function of the bioprosthetic PV, trivial (1/2+) tricuspid regurgitation and significant decrease of the right chambers (Figure 2).

Discussion

Some degree of PV regurgitation is present in almost every patient after anatomical correction of tetralogy of Fallot. PV regurgitation is tolerated well for many years, but the chronic effects on right ventricular function may be dramatic.⁶ Exercise limitation, secondary development of tricuspid regurgitation, presence of supraventricular or ventricular arrhythmias, and the risk of sud-

den death are the most important complications. Patients often do not notice symptoms until right ventricular dysfunction becomes severe. In addition, for undetermined reasons, a ventricular-ventricular interaction occurs and concomitant left ventricular dysfunction is often observed.⁶

PV regurgitation in these patients is frequently underestimated. The diastolic murmur is soft and short, since there is rapid equalisation of the diastolic pressures in the PA and right ventricle. The regurgitant jet is also often missed on two-dimensional echocardiography, because the jet has a low velocity and the flow is laminar.⁶

PV regurgitation is the most common lesion and replacement of the valve is a frequent indication for reoperation in these patients.⁴ PV replacement is un-

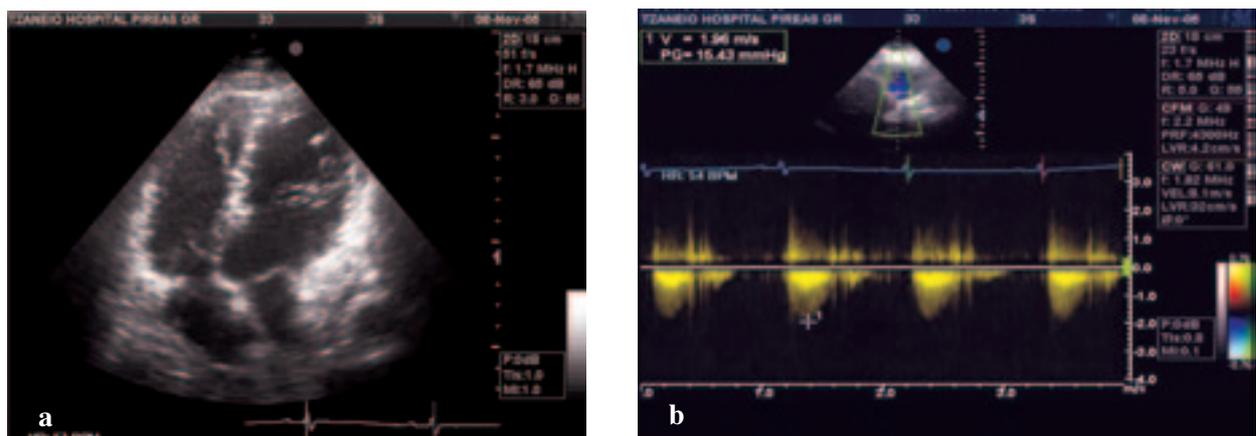


Figure 2. Cardiac echo three months after the operation. a. Significant decrease in the right heart dimensions in comparison with the preoperative findings; b. Normal flow through the bioprosthetic pulmonary valve (no stenosis or regurgitation is detected).

dertaken with low operative risk (1-2%) in experienced centres. The choice of the appropriate timing for PV replacement is very important and should be considered before the development of irreversible right ventricular dysfunction.^{2,3,5} The precise indication for PV replacement remains uncertain, although evidence suggests that in many centres it is performed too late.⁶ The development of tricuspid regurgitation is an indication for PV replacement⁷ and optimally the procedure should be performed before the onset of significant tricuspid regurgitation in order to preserve right ventricular function.² Even when subjective improvement in clinical symptoms is noted, there is often no improvement in right ventricular function and volumes when chronic myocardial exposure to severe PV regurgitation has resulted in irreversible contractile impairment.⁸ According to Davlouros et al,⁹ we could classify the indications for PV replacement as follows: a) asymptomatic patients with severe PV regurgitation and evidence of progressive right ventricular dilation and dysfunction and/or progressively diminished exercise tolerance; b) symptomatic patients with long-standing severe PV regurgitation and right ventricular dilation with or without right ventricular dysfunction; c) asymptomatic or symptomatic patients with moderate to severe PV regurgitation and haemodynamically significant associated lesions that need surgical intervention; d) patients with serious ventricular arrhythmias when associated with severe PV regurgitation and right ventricular dilation, with or without ventricular dysfunction. In this case combination with cryoablation seems more effective. Therrien et al¹⁰ concluded that PV replacement should be undertaken before the right ventricular end-diastolic volume reaches 170 ml/m² or the right ventricular end-systolic volume reaches 85 ml/m², in order to increase the chances that patients will reach normal right ventricular volume after repair. In a recent study, Dave et al¹¹ showed that timely insertion of a PV in young patients, when right ventricular end-diastolic volume exceeds 150 ml/m², is directly associated with improvement in right ventricular dimensions and functions over a six-month period. Right bundle branch block is expected in almost 95% of patients and is associated with the right ventricular size. The size of the right ventricle is directly proportional to the duration of the QRS complex on the ECG. QRS duration also provides a clear indication for reoperation.^{5,6}

However, early reoperation entails the risk of structural degeneration in the bioprosthetic valve. The placement of a mechanical prosthetic valve in the pulmonary

position is problematic, because of the poor washout of the valve due to its being in a low pressure system, and of course the need for lifelong anticoagulation with its associated risks among young people. On the other hand, a bioprosthesis is superior to a mechanical valve¹² and the durability of the newer (3rd generation) bioprosthetic valves appears very promising, with intermediate results reaching a 10-year survival of 95%, at which time 90% are free from reoperation.¹³ In an effort to prolong the durability of a bioprosthetic valve we have developed the following strategy: 1) use of 3rd generation bioprosthetic valves (AOA [Amino Oleic Acid] anti-mineralization treatment, physiological [zero pressure] fixation) which will prolong the valve duration; 2) oversized bioprosthetic valves, in order to minimise the gradient between the right ventricle and pulmonary artery with resultant valve damage from the high pressure gradient; 3) use of oversized stented bioprosthetic valves that will not be compressed by the sternum after chest closure.¹³ Nevertheless, patients who undergo a pulmonary valve replacement in the third or fourth decade may require re-operation in the future.⁶

During reoperation, apart from the PV replacement, tricuspid valve repair, resection of aneurysmal outflow tract patches, closure of residual septal defects and augmentation of stenotic pulmonary arteries may also be performed.²

In our case, PV replacement, PA reconstruction and tricuspid valve annuloplasty were performed with the beating heart technique under extracorporeal circulation. Although this makes the surgical technique more difficult, it has the significant advantage of avoiding the myocardial ischemia/reperfusion syndrome that occurs during cardiac arrest. The beating heart technique should be decided upon only after intracardiac defects (patent *foramen ovale*, atrial/ventricular septal defect) have been ruled out, because of the risk of air embolism into the brain and other vital organs. For the same reasons this technique should be avoided when intracardiac repair in the left heart is needed.

In conclusion, patients with operated tetralogy of Fallot need periodic cardiac follow up in order to achieve timely PV replacement before the development of complications due to volume overload of the right ventricle. In experienced centres, PV replacement with bioprosthesis is achieved with low morbidity and mortality (especially with the beating heart technique) and should be performed before irreversible dysfunction of the right ventricle has become established.

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