Coexistence of mitral regurgitation and pulmonary regurgitation in an adult with surgically corrected tetralogy of Fallot

Dimitrios Z Mytas,¹ Meletios A Kanakis,² Lamprini K Kosma,³ Fotios A Mitropoulos²

¹Department of Cardiology, Sismanoglion General Hospital, Athens, Greece

²Department of Paediatric and Congenital Heart Surgery, Onassis Cardiac Surgery Centre, Athens, Piraeus, Greece

³Department of Radiology, A Fleming General Hospital, Athens, Marousi, Greece

Correspondence to

Dr Dimitrios Z Mytas,
dmytas@yahoo.com

SUMMARY

In patients with corrected tetralogy of Fallot (TOF), progressive right ventricular volume overload from longstanding regurgitation of the pulmonary valve, results in severe late complications. The presence of additional major clinical conditions may aggravate the clinical status in these patients. We describe the case of a patient with coexistence of mitral and pulmonary regurgitation in the setting of corrected TOF who underwent successful surgical treatment.

BACKGROUND

Tetralogy of Fallot (TOF) is one of the most common cyanotic congenital cardiac diseases. In 1954, the first successful repair was performed and long-term results showed a 30-year survival of 91%.¹ However, progressive right ventricular volume overload from longstanding regurgitation of the pulmonary valve (PV), results in severe late complications. Exercise limitation, right and left ventricular dysfunction, electrocardiographic abnormalities and most important, 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 However, the presence of additional problems may aggravate the clinical status in these patients as in this case. Mitral regurgitation of degenerative aetiology due to tendinae chordae rupture has never been described in the setting of corrected TOF to the best of our knowledge.

CASE PRESENTATION

A 55-year-old man with medical history of surgically corrected TOF, 40 years ago presented with a 2 week history of progressive dyspnoea and fatigue. He reported exercise limitation in the last 6 months and denied any further follow-up during the last 2 years. Clinical examination revealed that the patient was tachypnoeic without cyanosis, blood pressure of 90/60 mm Hg, oxygen saturation of 94% (on room air), irregular pulse of ± 120 bpm. He also had bilateral pretibial oedema, ascites, hepatomegaly and jugular vein distention. Cardiac auscultation revealed a pansystolic murmur audible throughout the precordium, and pulmonary auscultation showed diminished breath sounds in the lower third of the right hemithorax.

INVESTIGATIONS

The electrocardiogram showed atrial fibrillation (AF) with rapid ventricular response and right bundle branch block. QRS duration was 140 ms. Chest x-ray showed cardiomegaly and right pleural effusion with increased pulmonary vascular markings. Transthoracic echocardiography revealed severe mitral regurgitation with normal left ventricular internal dimensions and a good systolic function. It also showed significant dilation of right heart chambers, severe pulmonary regurgitation with main pulmonary artery dilation, moderate tricuspid regurgitation, while the estimated pulmonary arterial systolic pressure was 70 mm Hg. Transoesophageal echocardiography demonstrated severe mitral regurgitation with a flail leaflet due to tendinae chordae rupture (figure 1). In addition, there was a small ventricular septal defect (VSD) patch detachment with left to right shunt (figure 2). Cardiac MRI confirmed the previous echocardiographic findings and showed a normal left ventricular ejection fraction with a right ventricular ejection fraction of 40% (figure 3). Coronary angiogram was normal. Initial treatment included spironolactone, furosemide, digoxin, dopamine and dobutamine. After substantial clinical improvement and stabilisation ACE inhibitors, β -blocker and amiodarone were also administered, while he was in anticoagulant therapy. The patient was referred for surgery.

TREATMENT

A redo sternotomy was performed. The patient was placed on cardiopulmonary bypass and the heart was arrested. A transeptal approach was used to access the mitral valve. There was myxomatous degeneration of the posterior and the anterior leaflet and the tendinae chordate of the anterior leaflet were ruptured. The valve was not amenable to repair. A mechanical prosthesis was implanted with the technique of subvalvular apparatus preservation. The residual VSD was sutured closed. Then, the right ventricular outflow tract (RVOT) patch was excised and a mechanical prosthesis was implanted and the RVOT was reconstructed with a patch. The tricuspid valve was repaired with a Kay technique. The patient was weaned off cardiopulmonary bypass without any difficulty with mild inotropic support.

OUTCOME AND FOLLOW-UP

His postoperative course was uncomplicated. One month after the operation he presented sustained VT (under amiodarone and β -blocker) and he underwent cardioversion with subsequent implantation of an

To cite: Mytas DZ, Kanakis MA, Kosma LK, et al. *BMJ Case Reports* Published online: 18 December 2012
doi:10.1136/bcr-2012-007535

Figure 1 Transoesophageal echocardiography showing severe mitral regurgitation with a flail leaflet due to tendinae chordae rupture (PML, posterior mitral leaflet; AML, anterior mitral leaflet; LA, left atrium; LV, left ventricle; AO, aorta; RV, right ventricle; MVR, mitral valve regurgitation and RV, right ventricle).

implantable cardioverter defibrillator (ICD). At 6 months follow-up he is symptom free in New York Heart Association class I.

DISCUSSION

PV regurgitation is almost present in every patient after anatomical correction of tetralogy of Fallot. It 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, ventricular arrhythmias and the risk of sudden 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 a concomitant left ventricular dysfunction is often observed.⁶

PV regurgitation is the most common lesion and its replacement is the most common indication for re-operation in these patients.⁴ PV replacement is undertaken with low operative risk (1–2%) in experienced centres. The choice of 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 has been performed too late.⁶ The development of significant tricuspid regurgitation is an indication of PV replacement⁷ and this can be optimised before the onset of significant right ventricular

dysfunction.² The indications for PV replacement generally follow these categories: (1) asymptomatic patients with severe PV regurgitation and evidence of progressive right ventricular dilation and dysfunction and/or progressively diminished exercise tolerance, (2) symptomatic patients with long-standing severe PV regurgitation and right ventricular dilation with or without right ventricular dysfunction, (3) asymptomatic or symptomatic patients with moderate to severe PV regurgitation and significant haemodynamically associated lesions that need surgical intervention and (4) 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.⁸ Therrien *et al*⁹ concluded that PV replacement should be undertaken before right ventricular end-diastolic volume reaches 170 ml/m² or the right ventricular end-systolic volume reaches 85 ml/m² to increase the chances that patients will reach normal right ventricular volume after repair. Dave *et al*¹⁰ showed that timely insertion of a PV in young patients, when RV end-diastolic volume exceeds 150 ml/m², is directly associated with improvement in right ventricular dimensions and functions, in 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 in electrocardiogram. QRS duration also consists of a clear indication for re-operation.^{5 6}

Figure 2 Transoesophageal echocardiography showing ventricular septal detachment with left to right shunt (VSD, ventricular septal defect).

Figure 3 Cardiac MRI depicting the presence of severe pulmonary regurgitation (PA, pulmonary artery; PVR, pulmonary valve regurgitation).

Bioprosthetic, oversized stented valves seem to be superior to a mechanical valve in these patients^{11 12} and survival is 95% at 10 years, at which time 86% are free of valve re-operation.³ Degenerative mitral valve disease is a common disorder affecting around 2% of the population.¹³ The most common finding is leaflet prolapsed due to elongation or rupture of the chordal apparatus. Mitral regurgitation in operated patient with TOF due to myxomatous degeneration with tendinae chordae rupture causing a flail leaflet is a very rare entity and it has never been described in the English literature so far, to the best of our knowledge. In contemporary practice, mitral valve replacement for degenerative disease in a primary operation should be a rare surgical option and if it is necessary, subvalvular apparatus preservation technique should be performed.

However, in this rare case, the valve was not amenable to repair and because of his young age since the prosthesis choice for the mitral valve was mechanical we also elected to implant a mechanical valve as well in the pulmonary position.

During re-operation besides PV replacement, tricuspid valve repair, resection of aneurysmal outflow tract patches, closure of residual septal defects and augmentation of stenotic pulmonary arteries may be also performed as in our case. In the absence of a VSD, PV replacement, PA reconstruction and tricuspid valve annuloplasty could be performed with safety by the beating heart technique and under extracorporeal circulation. This technique has the significant advantage of avoiding myocardial ischaemia/reperfusion syndrome which occurs during cardiac arrest.¹⁴ The acceptance of beating heart technique should be performed after exclusion of intracardiac defect (patent foramen ovale, atrial/ventricular septal defect) due to 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.

Arrhythmia is an important cause of morbidity and mortality in patients with repaired TOF. Sudden death occurs in up to 6% of patients late after repair.¹⁵ Several risk factors have been identified, but sustained ventricular tachycardia and impaired residual haemodynamics are now widely accepted to be the most significant. Pulmonary regurgitation was the main underlying haemodynamic lesion for sudden cardiac death and sustained ventricular tachycardia. Sustained ventricular tachycardia developed 1 month after the operation in our patient and finally, the patient underwent ICD implantation. Preoperative VT was not documented in this patient, although there was existence of significant right ventricle dilation and severe pulmonary regurgitation preoperatively.

AF is often related to residual VSD and left sided overload from long standing systemic to pulmonary artery shunts in patients with operated TOF.⁸ However, the risk of atrial arrhythmias may also correlate with longer duration of follow-up from repair.¹⁶ In addition, in this case mitral regurgitation was an additional significant factor for the development of AF.

This patient was not in a scheduled follow-up by a specialist. Only when his symptoms became severe did he agreed to visit a cardiologist. Undoubtedly, this delay had led to severe right ventricular dilation in the setting of severe pulmonary regurgitation. In parallel, the coexistence of mitral regurgitation and pulmonary hypertension aggravated the clinical status. Patients with operated TOF need periodic echocardiographic examination and a regular follow-up in order to prevent the development of complications that could be detrimental.

Learning points

In patients with corrected tetralogy of Fallot the following facts could be detrimental:

- ▶ The presence of severe pulmonary valve regurgitation.
- ▶ The presence of secondary tricuspid valve regurgitation.
- ▶ Ventricular septal defect patch detachment.
- ▶ Development of arrhythmias.
- ▶ The coexistence of severe mitral valve regurgitation of degenerative aetiology due to tendinae chordae rupture and severe pulmonary regurgitation in a patient with corrected tetralogy of Fallot is a quite rare but very severe clinical entity and a high index of suspicion is needed as prompt management is mandatory.
- ▶ Concomitant surgical treatment of mitral, tricuspid and pulmonary regurgitation with closure of residual ventricular septal defect could be performed with excellent surgical result.
- ▶ These patients need periodic echocardiographic examination and a regular follow-up in order to prevent the development of complications that could be detrimental.

Competing interests None.

Patient consent Obtained.

REFERENCES

- 1 Lillehei CW, Varco RL, Cohen M, *et al.* The first open heart corrections of tetralogy of Fallot. A 26 to 31 year follow-up of 106 patients. *Ann Surg* 1986;204:490–502.
- 2 Warner KG, O'Brien PKH, Rhodes J, *et al.* Expanding the indications for pulmonary valve replacement after repair of tetralogy of Fallot. *Ann Thorac Surg* 2003;76:1066–72.
- 3 Yemets IM, Williams WG, Webb GD, *et al.* Pulmonary valve replacement late after repair of tetralogy of Fallot. *Ann Thorac Surg* 1997;64:526–30.
- 4 Oechslin EN, Harrison DA, Harris L, *et al.* Reoperation in adults with repair of tetralogy of Fallot: indications and outcomes. *J Thorac Cardiovasc Surg* 1999;118:245–51.
- 5 Gatzoulis MA, Balaji S, Webber SA, *et al.* Risk factors for arrhythmia and sudden cardiac death late after repair of tetralogy of Fallot: a multicentre study. *Lancet* 2000;356:975–81.
- 6 Warnes CA. The adult with congenital heart disease. Born to be bad? *J Am Coll Cardiol* 2005;46:1–8.
- 7 Discigil B, Dearani JA, Puga FJ, *et al.* Late pulmonary valve replacement after repair of tetralogy of Fallot. *J Thorac Cardiovasc Surg* 2001;121:344–51.
- 8 Davlourous PA, Karatzas AA, Gatzoulis MA, *et al.* Timing and type of surgery for severe pulmonary regurgitation after repair of tetralogy of Fallot. *Int J Cardiol* 2004;97:91–101.

- 9 Therrien J, Provost Y, Merchant N, *et al.* Optimal timing for pulmonary valve replacement in adults after tetralogy of Fallot repair. *Am J Cardiol* 2005;95:779–82.
- 10 Dave HH, Buechel ERV, Khatami AD, *et al.* Early insertion of a pulmonary valve for chronic regurgitation helps restoration of ventricular dimensions. *Ann Thorac Surg* 2005;80:1615–21.
- 11 Kawachi Y, Masuda M, Tomonaga R, *et al.* Comparative study between St Jude Medical and bioprosthetic valves in the right side of the heart. *Jpn Circ J* 1991;55:553–62.
- 12 Mitropoulos F, Laks H, Kapadia N, *et al.* Bioprosthetic pulmonary valve replacement. Presented at the 42nd Annual Meeting of the Society of Thoracic Surgeons; January 30–February 1 2006, Chicago.
- 13 Enriquez-Sarano M, Akins CW, Vahanian A. Mitral regurgitation. *Lancet* 2009;373:1382–94.
- 14 Mitropoulos F, Kanakis M, Davlouros P, *et al.* Beating heart replacement of the pulmonary valve in a patient with surgically corrected tetralogy of Fallot. *Hellenic J Cardiol* 2006;47:180–3.
- 15 Murphy JG, Gersh BJ, Mair DD, *et al.* Long-term outcome in patients undergoing surgical repair of tetralogy of Fallot. *N Engl J Med* 1993;329:593–9.
- 16 Roos-Hesselink J, Perloth MG, McGhie J, *et al.* Atrial arrhythmias in adults after repair of tetralogy of Fallot. Correlations with clinical, exercise, and echocardiographic findings. *Circulation* 1995;91:2214–19.

Copyright 2012 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- Submit as many cases as you like
- Enjoy fast sympathetic peer review and rapid publication of accepted articles
- Access all the published articles
- Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow