

## Case 4/2011 - 49-year-old Man with Pulmonary Valve Stenosis and Atrial Septal Defect as Residual Lesions after Correction of Tetralogy of Fallot, which Occurred 30 Years Ago

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### Clinical aspects

After correction of tetralogy of Fallot, carried out at age 19, with expansion of the right ventricle outflow tract, pulmonary valvotomy and closure of the ventricular septal defect, pulmonary valve stenosis at a small pulmonary annulus and atrial septal defect deliberately remained as residual defects. Still, a good evolution lasted for 28 years, when the first complication emerged, with paradoxical cerebral embolism with a mild stroke, with no sensory or motor effects. Moreover, a year ago there was arrhythmic complication by atrial fibrillation with fast heart rate, reverted with amiodarone and electrical shock. Recently, new atrial fibrillation, but with low ventricular rate (50 to 60 bpm) and arterial desaturation (70%), after walking a few minutes, have motivated diagnostic investigation and more accurate conduct.

On physical examination, the patient presented good general condition, eupneic, normal color, normal pulses, weighing 114 kg, height of 1.70 cm, BP of 110/80 mmHg and HR of 60 bpm in atrial fibrillation rhythm. The aorta was discreetly palpated at the suprasternal notch. In the precordium, there were no deformities or impulses and apical impulse was not palpated. Heart sounds were faint, and systolic murmur was auscultated, +/+ + of intensity, of ejection, rough, in the third and fourth left intercostal spaces at the sternal border and at the suprasternal notch. Liver could not be felt.

### Complementary tests

Electrocardiogram after surgery showed signs of right bundle branch block with QRS duration of 0.16", with ventricular rate of 60 bpm, in atrial fibrillation rhythm (Figure 1).

Chest radiography shows mild enlargement of the cardiac area at the expenses of the right cavities, globular cardiac form and slightly increased pulmonary vasculature (Figure 1).

### Keywords

Congenital heart disease, tetralogy of Fallot, pulmonary valve stenosis, right ventricular hypertrophy.

Echocardiogram showed moderate obstruction at pulmonary valve with average pressure gradient of 50 mmHg, a slight enlargement of right cavities and atrial septal defect of 14 mm in length, next to the superior vena cava. The measurements corresponded to 37 mm in the aorta, 50 mm in the left atrium, 38 mm in right ventricle, 49 mm in the left ventricle, 12 mm in the ventricular septum and 11 mm in the posterior wall. Left ventricular ejection fraction was 69% and there was a mild right ventricular hypocontractility with unusual movement of the ventricular septum (Figure 1).

Hemodynamic study also showed signs of pulmonary stenosis at valve with pressure gradient of 50 mmHg and shunt in atrial septal defect by contrast injection in the right superior pulmonary vein. The pressures in the pulmonary artery were 25/8 mmHg, and in the right ventricle were 75/15 mmHg. In the atria, the patient had mean pressure of 15 mmHg, in the left ventricle, 135/12 mmHg and 135/85 mmHg in the aorta. The right ventricle was slightly dilated, hypertrophied and hypocontractile. The right ventricle outflow tract was more dilated and with more pronounced hypocontractility. Right ventricular angiography showed marked pulmonary stenosis at the valve level, with passage of contrast through a well-defined central hole (Figure 2).

### Diagnosis

Moderate pulmonary valve stenosis and right ventricular hypertrophy responsible for shunting blood from right to left by atrial septal defect, such as residual defects after correction of tetralogy of Fallot, performed 30 years ago, causing severe arterial unsaturation upon small exertion.

### Clinical reasoning

The residual defects of the surgery performed 30 years ago for correction of tetralogy of Fallot, which seemed innocuous, became fraught with time. The accentuation of pulmonary stenosis caused major hemodynamic disorder with shunt from right to left at atrial level and progressively increased (recently, oxygen saturation decreased from 90 to 70%, even with minimal exertion and severe tiredness), in addition to paroxysmal atrial fibrillation and paradoxical embolism two years ago.

### Conduct

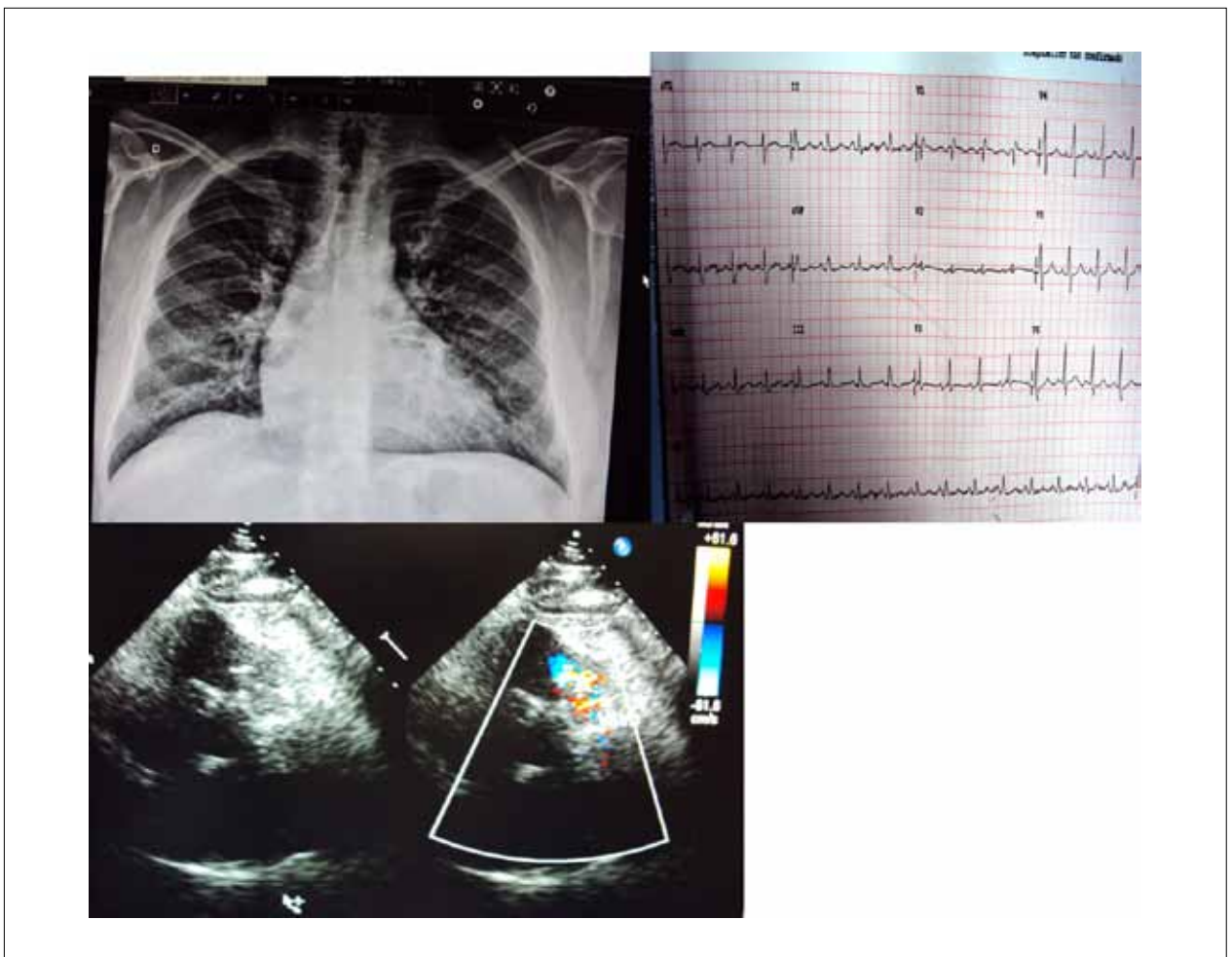
New indication for surgery was incisive due to hemodynamic disorders of high clinical risk. Upon surgery, the pulmonary

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**Figure 1** - Chest X-ray in late postoperative period of tetralogy of Fallot shows moderate increase in right cavities, excavated medial arch and increased pulmonary vasculature. Electrocardiogram reveals the usual findings after previous correction of the defect such as right bundle branch block. Atrial fibrillation with ventricular rate of 51 bpm. On a left parasternal short axis view, there is a clear obstruction of the pulmonary valve in the the echocardiography image.



**Figure 2** - Angiogram shows a normal left ventricle (LV) in connection with the aorta (Ao) in A, slight increase in right ventricle (RV) with myocardial hypertrophy and pulmonary stenosis at valvar level, in B. Abbreviation: PT - pulmonary trunk.

valve was thickened into a small pulmonary annulus. The right ventricle outflow tract was dilated and with calcification of the pericardial patch, and the septal defect increased 20 mm in diameter. Expansion of right ventricle outflow tract and pulmonary annulus with placement of homograft number 24, and atrioseptoplasty with pericardial patch, resulting in good postoperative outcome, given the normal hemodynamics obtained. Sinus rhythm returned after treatment of atrial fibrillation by periostial incision encircling the pulmonary veins. Systolic murmur disappeared and oxygen saturation remained around 95%. In the electrocardiogram, right bundle branch block persisted with greater conduction delay with QRS duration of 0.18". Postoperative echocardiogram showed a slight gradient of 8 mmHg between the right ventricle and the pulmonary trunk. The patient was discharged on 10<sup>th</sup> postoperative day.

## Considerations

Any residual defect except those of small magnitude, after prior cardiac correction causes some hemodynamic disturbance in a variable evolutionary period. Upon early correction of tetralogy of Fallot, it was estimated that a pulmonary obstructive residual lesion could not cause as much comorbidity as that produced by pulmonary valve insufficiency after pulmonary annulus enlargement. As we have found, such an assertion is not true for the pathological consequences of the case presented with serious life risks due to severe arterial unsaturation that came up with little exertion. Today, hemodynamic normalization is preferred even after the opening of the pulmonary annulus, perhaps by placing a biological valve in pulmonary position or a bicuspid valve. Such conduct alleviates the possibility of adverse consequences both for residual lesions of pulmonary insufficiency and pulmonary stenosis.