

Case 5/2011 - Infant of 17 Months of Age with Tetralogy of Fallot in Open Heart Failure Due to Hemodynamic Dominance of the Ventricular Septal Defect

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Clinical Aspects:

Since birth, the patient has had persistent dyspnea, psychomotor agitation and poor weight gain. Over this period, no cyanosis has appeared, even with the crying. On physical examination, there were characteristic signs of Down syndrome and moderate tachydyspnea; the patient has been acyanotic and with normal pulse. The patient had a weight of 8,200 g. and height of 74 cm. The precordium showed pronounced bulging with moderate systolic impulses. Apical impulse was diffuse in the fourth and fifth intercostal spaces. There was systolic murmur in the third and fourth intercostal spaces at the left sternal edge irradiating to the upper sternal edge. The second sound was accentuated and split. The liver was palpable two centimeters at the right coastal edge.

Complementary tests:

Electrocardiogram revealed signs of severe biventricular overload with no left anterior hemiblock. The QRS axis was deviated to the right at $+110^\circ$, and the RS waves were large from V2 to V5 and polyphasic in V1 (Figure 1).

Chest radiography revealed marked increase in heart size and pulmonary vasculature. The left ventricular arch was rounded and long, continuing with the medial arch (Figure 2).

Echocardiogram showed 12 mm perimembranous ventricular septal defect (VSD), aortic dextroposition of 50.0%, anterior deviation of the infundibular septum with mild stenosis with 30 mmHg gradient. The shunt was bidirectional for ventricular septal defect.

Diagnosis

Tetralogy of Fallot in open heart failure due to hemodynamic dominance of the ventricular septal defect.

Keywords

Heart defects congenital; tetralogy of fallot; heart failure; heart septal defects, ventricular/complications.

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Conduct: In surgery, extracorporeal circulation of 85 minutes, through the right atrium, a large subaortic ventricular septal defect was found, which was closed with pericardium patch. The opening of the pulmonary trunk identified a bivalve pulmonary valve, with fine texture and good opening in a small pulmonary annulus of 12 mm, which was preserved.

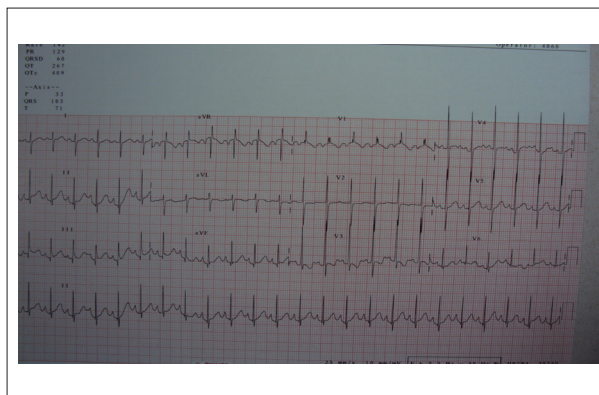


Figure 1 - Electrocardiogram reveals through biventricular overload diagnostic guidance for cardiac interventricular septal defects.



Figure 2 - Chest X-ray shows cardiomegaly and increased pulmonary vasculature consistent with heart disease with shunting of blood from left to right, suggesting interventricular septal defects.

After opening the right ventricle outflow tract, infundibular myectomy was performed in addition to enlarging the region with bovine pericardium and the pulmonary region.

The postoperative course was complicated by spontaneous pneumothorax, consequent low cardiac output, bronchial aspiration and pneumonia, which required prolonged treatment for 14 days until discharge.

Considerations: In the first months of life, tetralogy of Fallot is externalized by heart murmur that resembles that of the ventricular septal defect, low in the left sternal border, resulting in blood flow from left to right and therefore no cyanosis is found in this period. With a larger anterior infundibular septum, and the consequent worsening of obstruction of right ventricular outflow tract, blood flows from right to left with the appearance of cyanosis and hypoxemia crises,

which usually appear from three to six months of age. In rare cases, ventricular septal defect may predominate over the infundibular stenosis, which leads to increased pulmonary flow, volume overload of the heart chambers and consequent cardiac failure, as was the case presented.

Thus, the clinical manifestation of pulmonary venous congestion in heart defects that behave as large ventricular septal defect with heart failure may also be present in tetralogy of Fallot in the presence of a clear prevalence of ventricular septal defects on pulmonary infundibular stenosis, yet despite the presence of dextroposition of aorta and right ventricular hypertrophy. It is noteworthy that in the classic case of tetralogy of Fallot, the dominant clinical manifestation undoubtedly stems from the prevalence of infundibular pulmonary stenosis.