

## Case 3/2007 – A Three-Month-Old Male Infant with Criss-Cross Heart, Atrioventricular Discordance, and Double-Outlet Right Ventricle, without Pulmonary Stenosis

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### Clinical data

A three-month-old white male infant with history of fatigue while breast-feeding since birth, which had gotten worse over the last 60 days; restlessness; difficulty in gaining weight (he had put on only 240 g in the previous month); and, sometimes, dry cough. On physical examination he had dyspnea ++, normal pulses, and weighed 4440 g. No cyanosis was observed. The aorta was not palpable. Chest examination showed mild impulses along the left sternal border and a valve and muscular apical impulse +/++ located at the 4<sup>th</sup> left intercostal space limited by the breadth of two fingers. Heart sounds were loud and most intense in the tricuspid area. The third sound was heard in the mitral area. A harsh systolic ejection murmur+ was heard in the 3<sup>rd</sup>, and 4<sup>th</sup> left intercostal spaces, irradiating to the right sternal border. The liver was palpable 4 cm from the right costal margin and xiphoid process.

The ECG showed sinus rhythm and signs of ventricular overload that could not be determined due to the predominance of S-waves from V3. Left anterior fascicular block may be present, with the QRS axis deviated superiorly and to the right at - 120°. P-axis:+40°, QRS-axis:-120°, T-axis:+80°.

### Radiographic examination

Radiographic findings included an enlarged cardiac silhouette due to the long ventricular arch and right atrium, assuming an oval-shaped configuration (egg-on-side); a straight mid-arch; and increased pulmonary vascularity (Figure 1).

### Diagnostic impression

This oval-shaped configuration is consistent with TGA-type congenital heart disease with increased pulmonary blood flow.

### Key words

Heart defects, congenital; infant; double outlet right ventricle; crisscross heart.

### Differential diagnosis

Other heart diseases associated with increased pulmonary blood flow and exhibiting this morphology, as well as right atrium and left ventricular enlargement, must be considered, such as double-outlet right ventricle, tricuspid atresia without pulmonary stenosis, mitral atresia without pulmonary stenosis, common arterial trunk, and single ventricle.

### Diagnostic confirmation

Clinical findings point to the diagnosis of heart disease with increased pulmonary blood flow and associated ventricular septal defect, given the low-systolic murmur audible along the left sternal border. The QRS axis rightward and superior shift (suggesting left anterior fascicular block) and the undetermined ventricular overload raise doubts and add further suspicion of a more complex heart disease. The echocardiogram (Figure 2) showed a criss-cross heart with atrioventricular discordance and double-outlet right ventricle, arising from the right aorta and pulmonary artery to the left with double infundibulum, with no pulmonary stenosis. It was a restrictive ventricular septal defect. The inlet ventricular septal defect was not artery-related and thus allowed the tricuspid valve to relate to the left ventricle and mitral valve to the right ventricle. The right ventricle, with normal cavity size and lying in a superior and rightward position, was found to be hypertrophic. The left ventricle was dilated and lying in a inferior, leftward position. Marked pulmonary hypertension (mean PAP 68 mm Hg) was present.

### Management

Considering the heart failure and the patient's difficulty in gaining weight, in the setting of a challenging heart condition to be treated surgically, a successful pulmonary artery banding was performed. After some months, the heart failure was controlled. Only when he was ten years old did he become mildly cyanotic, and a systolic murmur ++ was heard in the 1<sup>st</sup> and 2<sup>nd</sup> left intercostal spaces and in the suprasternal notch. During this period, the electrocardiogram findings remained unchanged, and the echocardiogram showed pulmonary banding with a 94-mm Hg pressure gradient. A follow-up chest x-ray revealed a leftward bowing ventricular arch with an elevated cardiac apex; pulmonary vascularity remained slightly increased. As the patient was progressing favorably, even 10 years after pulmonary artery banding, a Fontan-type operation was not considered.

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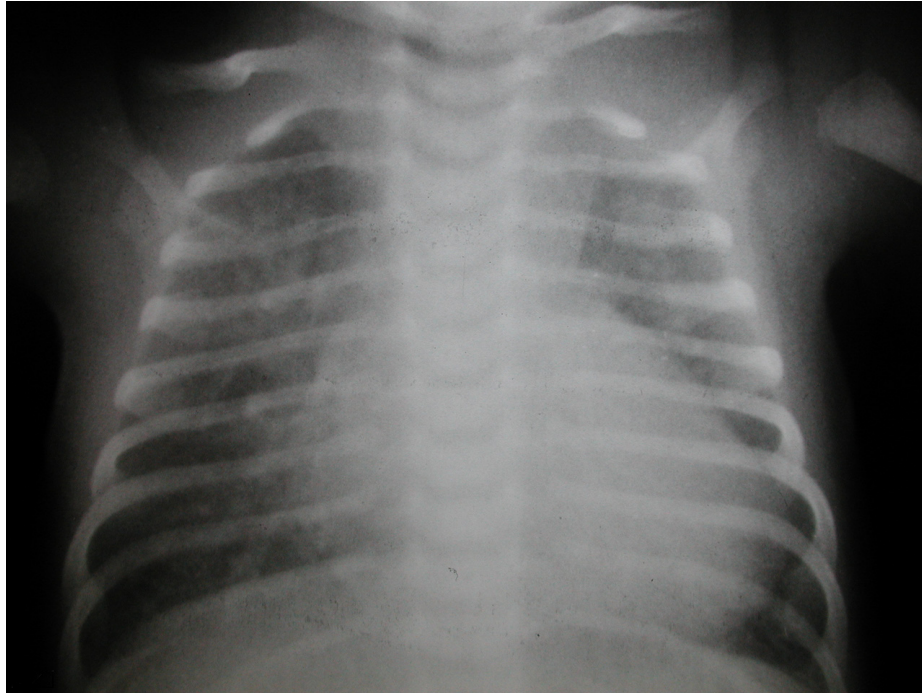


Fig. 1 - Chest x-ray showing the usual signs found in transposition of the great arteries, such as oval-shaped cardiac silhouette and increased pulmonary vasculature.

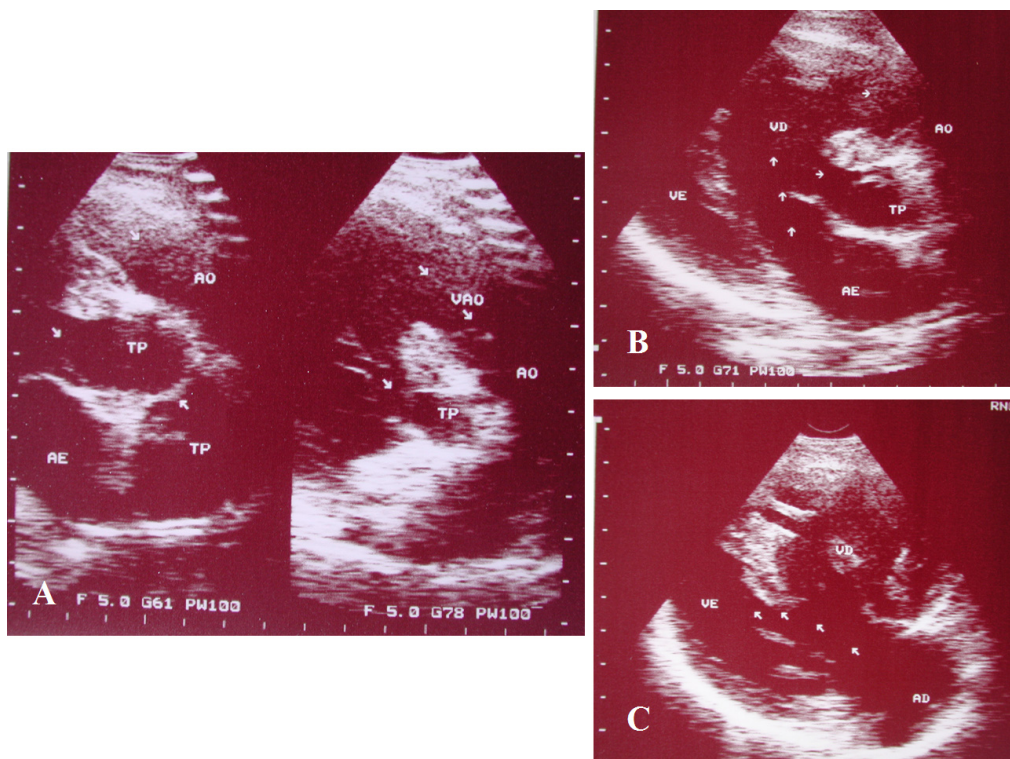


Fig. 2 - Echocardiographic images showing the classic signs of criss-cross heart with the right atrium (RA) communicating with the left ventricle (LV - arrows) in C and the left atrium (LA) communicating with the right ventricle (RV - arrows) in B, in longitudinal sections also characterizing the atrioventricular discordance, with LV to the left and RV to the right. The double-inlet RV with double infundibulum is shown in B and A, and the aorta (Ao) is to the right. The inlet-type ventricular septal defect (VSD) is seen in C, and the pulmonary banding is clearly demonstrated in A (arrow). LV is positioned inferiorly, and RV is positioned superiorly (B,C). PT - pulmonary trunk, AoV - aortic valve.