

Case 4/2013 - Woman with Transposition of the Great Arteries and Delayed Corrective Progress

Edmar Atik

Clinica privada do Dr Edmar Atik, São Paulo, SP – Brazil

Clinical Data: After surgical correction of the transposition of the great arteries associated with other defects and heart failure, beginning with termino-terminal coarctectomy and pulmonary artery banding at three months (weight = 3800 g) and use of the Jatene technique with the closing off of interventricular communication, and pulmonary banding at ten months (weight = 5200 g), the patient progressed well, recovering weight and remaining symptom-free, despite the appearance of pulmonary artery stenosis at five years of age, improved after dilation by balloon catheter and resulting in a reduction in the pressure gradient, down from 72 mmHg to 26 mmHg. Since then, she has been taking 40 mg/day of propranolol for slight but persistent stenosis of the right ventricular outflow tract. She currently tolerates routine exercises well and does not mention symptoms.

Physical Examination: Good general condition, eupneic, acyanotic, normal pulse. Weight: 73 kg. Height: 156 cm. Right Arm Blood Pressure (RABP) and Right Leg Blood Pressure (RLBP) = 130/80 mm Hg. Heart rate (HR): 73 bpm. The aorta was not palpated and there is a mild systolic murmur at the suprasternal notch.

In the precordium, the *ictus cordis* was not palpated and there were no systolic impulses. The heart sounds were normophonetic, auscultating a moderate systolic murmur of ++ intensity, harsh, on the upper left sternal edge. The liver was not palpated and the lungs were clear.

Supplementary Examinations

Electrocardiogram: showed the sinus rhythm, with no signs of overload, unaltered since the correction of the defects. Electrical axis of P wave (PA): +50°. Electrical axis of the QRS complex (QRSA): +70°. Electrical axis of the T wave (TA): +70° (Figure 1).

Chest X-ray: showed the cardiac area as normal, with the pulmonary vascular network also normal (figure 1).

Echocardiogram: showed cardiac cavities with normal dimensions with no myocardial hypertrophy (septum –

posterior wall = 10 mm) and a 40 mmHg pressure gradient in the right ventricular outflow tract. A pressure gradient was detected in the descending aorta.

Chest Tomography of the Aorta: showed mild hypertrophy of the right ventricle, slight dilation in the aortic isthmus region at the location of the previous coarctation correction, and normal biventricular function. The dilation of the ascending neoaorta persists, due to the dilated pulmonary artery, subject to hyperflow over a lengthy period of time, pre-operative (Figure 2).

Clinical Diagnosis: Transposition of the great arteries associated with interventricular communication and aortic coarctation corrected in stages, with good late post-operative progress and with mild stenosis in the right ventricular outflow tract.

Clinical Reasoning: The clinical elements of her progress were compatible with the diagnosis of a slight obstruction of the right ventricular outflow tract. Despite the clear systolic murmur and the pressure gradient of 40 mmHg found in the echocardiogram, the absence of right ventricle potentials in the electrocardiogram attests to the slight repercussion of the dynamic obstruction.

Differential Diagnosis: After anatomical correction of the transposition of the great arteries, neopulmonary stenosis was found more frequently among patients undergoing surgery in the early days of this technique, with most of them requiring surgical expansion of the obstruction. Dilation by balloon catheter was rarely effective, as also occurred in this case. The continuation of dynamic subpulmonary stenosis became an element for differential diagnosis with native anomalies in this region that are hard to distinguish.

Conduit: Watchful clinical conduct was adopted, guided by the 40 mmHg pressure gradient and normal electrocardiogram, as expressions of the slight repercussion of the right ventricular outflow tract obstruction.

Remarks: Post-operative progress after the correction of the transposition of the great arteries, despite associated defects such as interventricular communication and aortic coarctation, has proven adequate even over the long term, as shown in this case report. Complications in progress after anatomical correction using the Jatene technique, such as pulmonary artery stenosis, coronary obstruction and aortic insufficiency, have become increasingly rare, due to appropriate technical alterations over time. The progress of this patient underscores the normalization of anatomical and functional elements found in the transposition of the great arteries. This is why this congenital defect has become a condition with an anatomical and functional cure, despite belonging to the cyanogenic anomalies group and with marked prior hemodynamic disorders. This case exemplifies adequate progress noted after anatomical correction through the Jatene technique over the long term.

Keywords

Heart Defects, Congenital; Transposition of Great Vessels / surgery; Heart Septal Defects, Ventricular / complications; Aortic Coarctation.

Mailing Address: Edmar Atik •
Rua Dona Adma Jafet 74, cj 73 01308-050
São Paulo, SP - Brazil
E-mail: conatik@incor.usp.br

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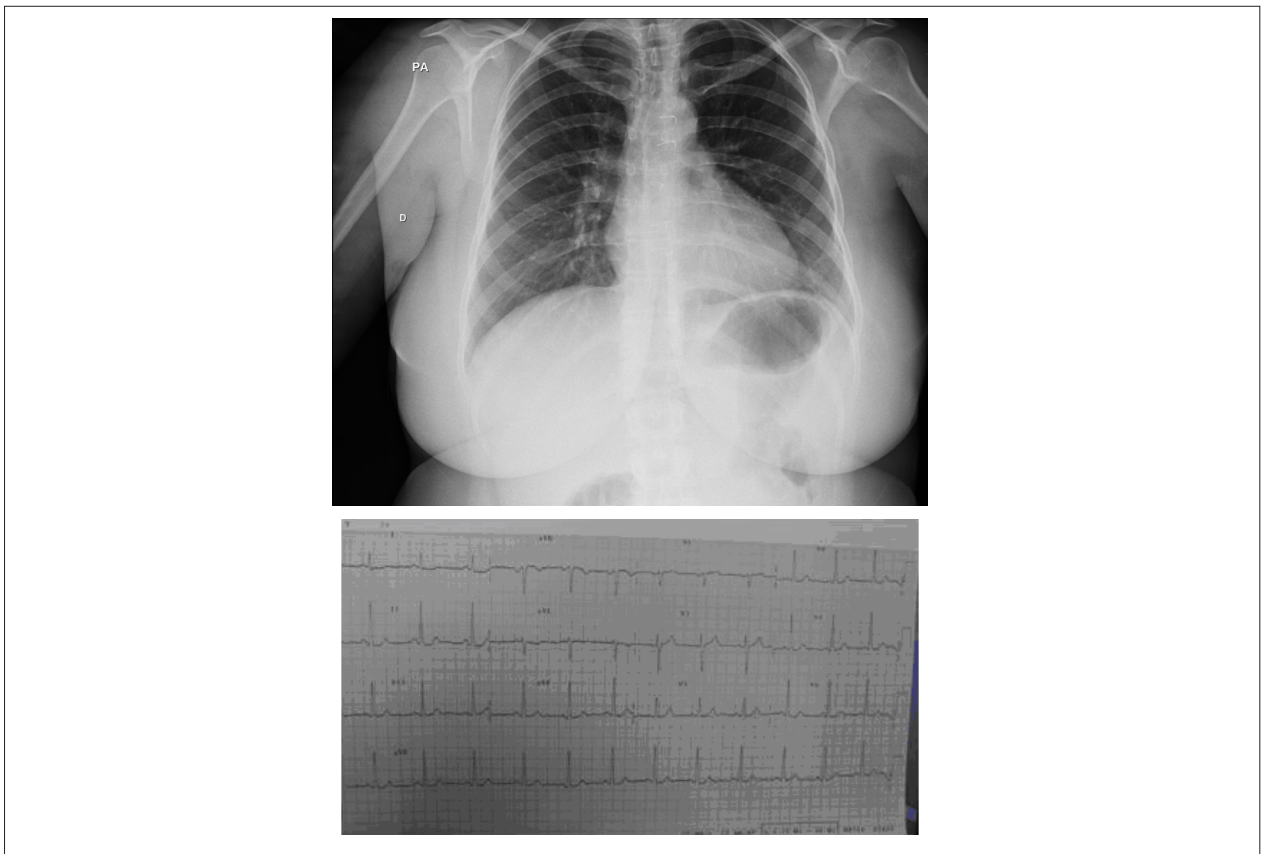


Figure 1 - Chest X-ray and electrocardiogram show elements of normality.

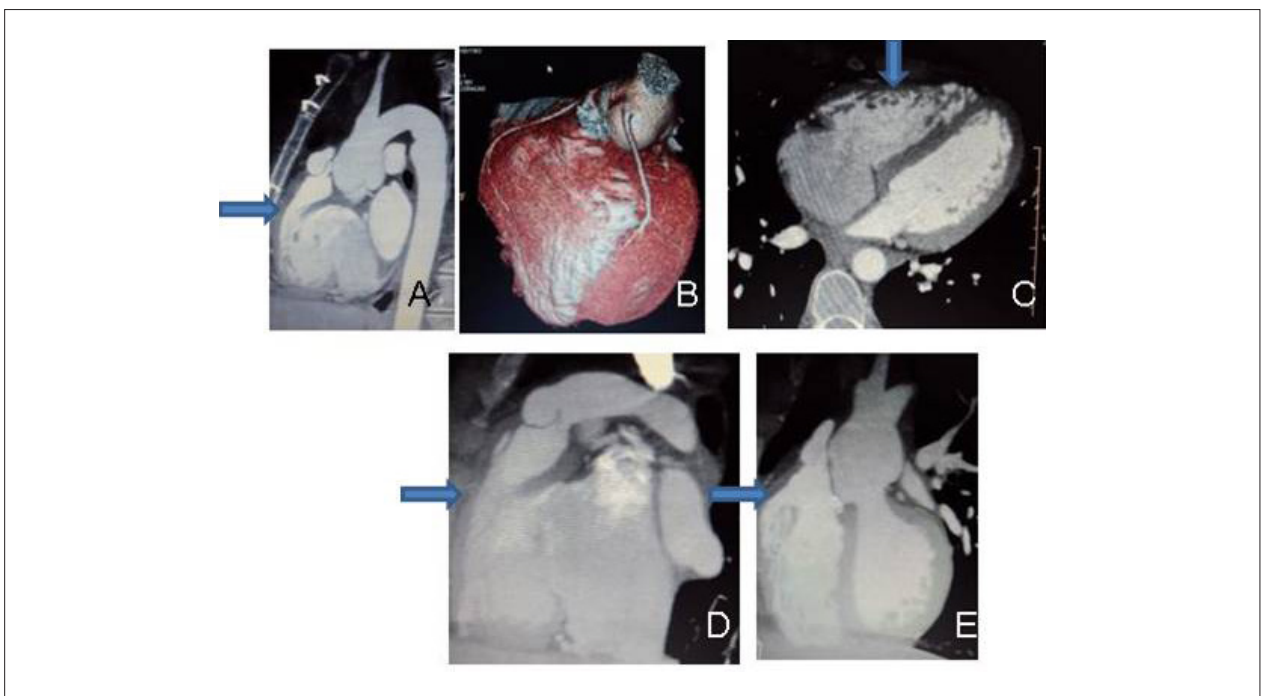


Figure 2 - Chest tomography clearly shows the dilation of the ascending aorta (neoaorta) and the aortic isthmus region (remains of the coarctation correction), as well as the slight subpulmonary stenosis in the right ventricle (arrow) at A and the two transposed coronary arteries in the ascending neoaorta at B, where both ventricles are similar in size and with the right ventricle being slightly hypertrophied at C, D and E (arrow).