

## Case 2/2014 – 20-year-old Woman with Corrected Transposition, Pulmonary Atresia and Aortopulmonary Collateral Arteries

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**Clinical data:** In the course of life, the patient developed three types of clinical manifestations. The first one was characterized by dyspnea and tachypnea, from birth to five months of age, increased pulmonary flow and heart failure, with cardiomegaly and hepatomegaly. The second phase was characterized by stability without significant symptoms, which lasted until the age of three, presumptive of balanced aortopulmonary flow. The third phase, with decreased pulmonary flow, had mild cyanosis that worsened two years ago. Exertion fatigue was noted for four years ago. The patient maintained saturation above 85% and up to 75% two years ago, hemoglobin = 17 g/dL and hematocrit = 62%.

**Physical examination:** Eupneic, cyanotic + + +, normal pulses, no jugular venous distention. Weight: 54 kg; height: 160 cm; BP: 115/60 mmHg; HR: 78 bpm; oxygen saturation = 78%. Aorta was palpated + + in the suprasternal notch.

In precordium, apical impulse in the 4<sup>th</sup> right intercostal space and discrete systolic impulses at the right sternal edge. Second hyperphonic heart sound, + + +, at pulmonary area, increases toward the lower left sternal edge. Discrete and mild continuous murmur, + / + +, at pulmonary area, irradiating to the back. Liver was not palpated.

### Complementary tests

**Electrocardiogram** (Figure 1) showed junctional rhythm and signs of left ventricular overload on the right. The P wave was negative in II, III, aVF and precordial areas. The QRS complex morphology was RS in V1, rs in V6 and Rs in V6R. The T wave was negative in I, L and more positive in V6R than in V6. AQRs: +160°, AT: 120°, AP: –80°.

**Chest radiography** shows normal cardiac silhouette (cardiothoracic ratio = 0.45) on the right in situs solitus (gastric bubble on the left) with rounded right ventricular arch. The upper arch on the left was long. Normal pulmonary vasculature and moderate scoliosis (Figure 1).

### Keywords

Pulmonary Atresia / surgery; Heart Defects, Congenital; Corrected Transposition of Great Arteries; Hypertension, Pulmonary.

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**Echocardiography** showed heart in dextrocardia and situs solitus. There were atrioventricular and ventriculoarterial discordances, pulmonary atresia (PA) and large ventricular septal defect (VSD) in the 25 mm inflow. The pulmonary trunk was not viewed, neither were pulmonary arteries. The diameters of the cavities were normal and ventricles were functioning normally.

**Catheterization:** (Figure 2) In the second month of life, the pulmonary arteries were hypoplastic and confluent and irrigated by the ductus arteriosus directed to the right lung and the left upper lobe. The aortopulmonary collateral vessel of the descending aorta was directed to the left lower lobe. A new examination showed left upper vasa as an aortopulmonary collateral vessel instead of the ductus arteriosus. The left pulmonary artery was 8 mm in diameter; the right was 13 mm in diameter with left stenosis (diameter = 5 mm). The pressure along the pulmonary circuit were systemic (100/64 mmHg).

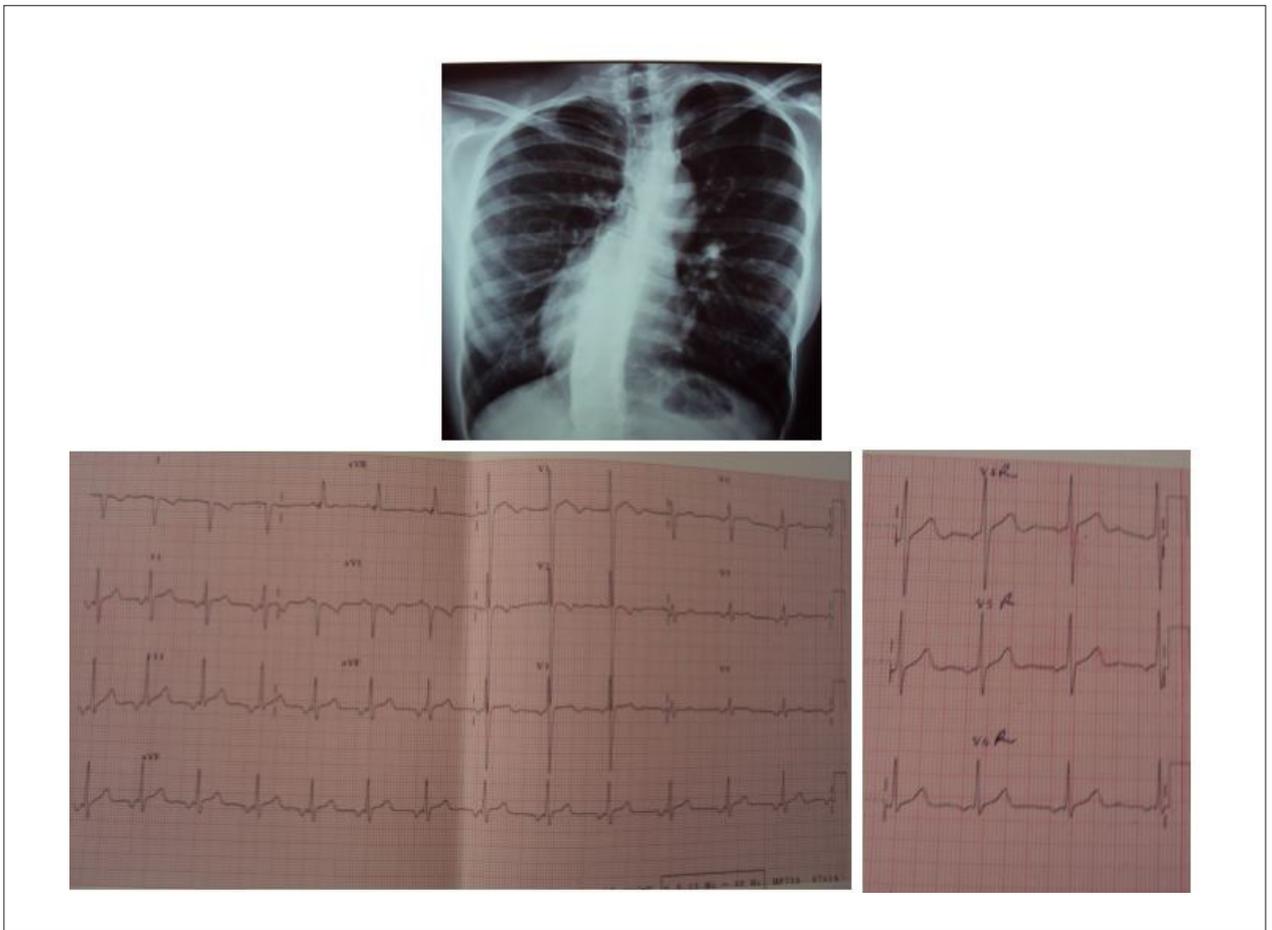
**Clinical diagnosis:** Corrected transposition of the great arteries (CTGA), pulmonary atresia and ventricular septal defect in dextrocardia and situs solitus. Pulmonary circulation is nourished by aortopulmonary collateral arteries with pulmonary arterial hypertension in severe hypoxia and natural evolution.

**Clinical reasoning:** Clinical findings of cyanogenic heart diseases, accompanied by PA + VSD may vary, often with signs of overflow, balancing flows or decreased pulmonary blood flow, as noted in this patient under natural evolution. The initial overflow gave way others due to predominance of decreased pulmonary blood flow. In this patient, the last stage was due to increased pulmonary arterial hypertension, not diagnosed initially. Other clinical signs were indicative of CTGA: second heart sound in the pulmonary area, which increases to the lower right sternal edge, suggesting that the aorta arises from the right ventricle to the left, backed by radiographic imaging, in which the ascending aorta is located on the left. The continuous murmur of collateral arteries expresses the presumed associated pulmonary atresia. The left ventricle is located on the right, directing the T wave to the right and systolic and diastolic overload of this ventricle.

**Differential diagnosis:** Heart diseases of type VSD + PA are externalized in the same manner, and the base diagnosis of other anomalies must be performed based on other clinical guiding elements, as demonstrated in this case.

**Conduct:** In view of the marked reactive pulmonary hypertension, sildenafil was introduced at a dose of 25 mg every 8 hours. There was improvement in physical tolerance and arterial saturation rose to 85%.

**Comments:** This rare association, PA + VSD + CTGA and pulmonary circulation dependent on aortopulmonary collateral arteries was reported only once in the literature<sup>1</sup>.

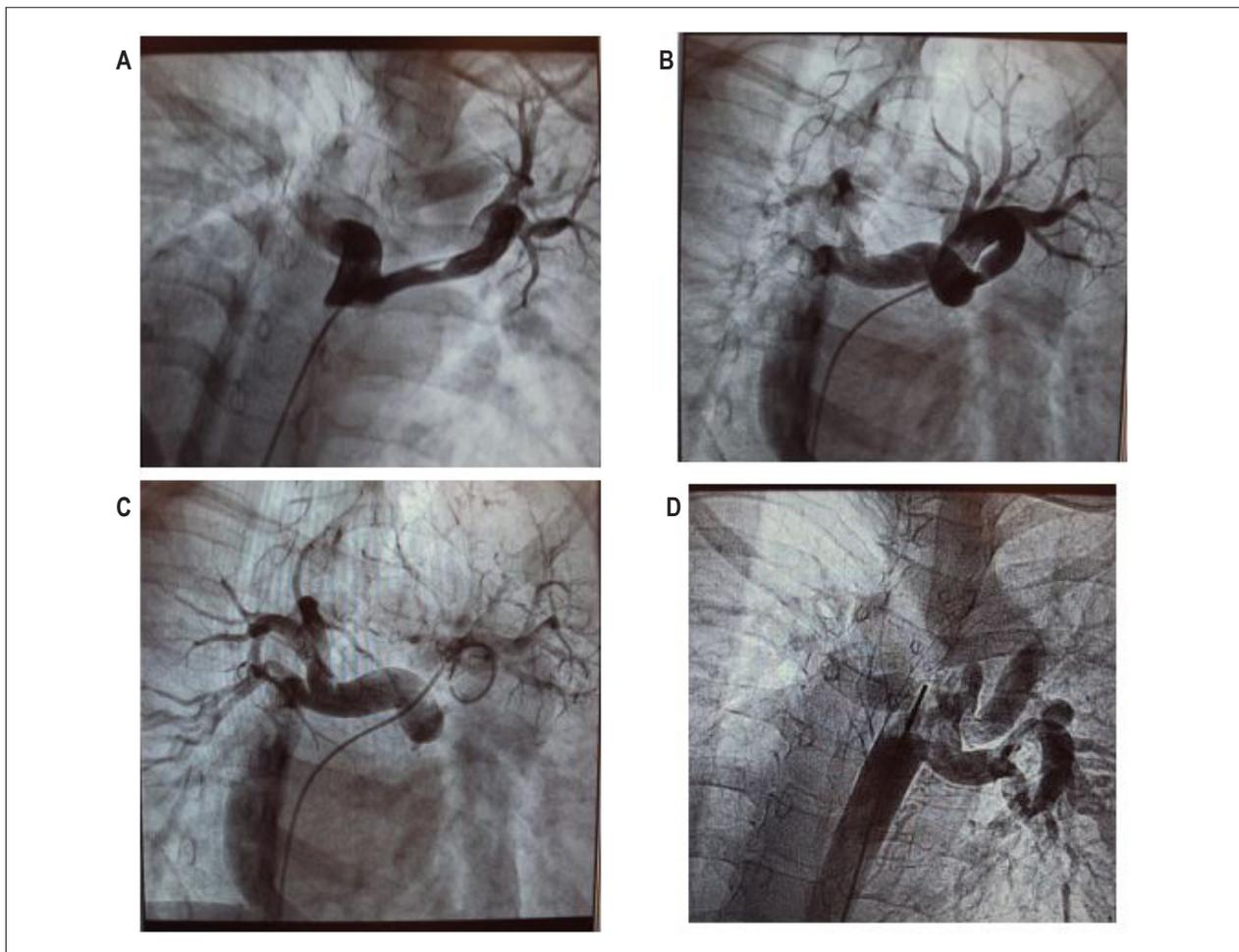


**Figure 1** – Chest radiography shows the heart on the right in situs solitus, with characteristics, together with those of the electrocardiogram, corrected transposition of the great arteries, with the left ventricle on the right and the aorta on the left.

In general, the clinical picture depends on two defects (VSD + PA) guiding the pathophysiology of this set. When the collateral circulation is augmented with increased pulmonary flow, it generally undergoes stabilization and subsequent decrease, characterizing the evolutionary stages. Hypoxemia arises from decreased pulmonary flow through stenosis of

aortopulmonary circulation or the development of pulmonary hypertension. In both situations, this picture naturally evolves to the second or third decade of life. Because of this, indication for surgery in this patient and other similar patients should always be questioned, counterbalancing clinical evolutionary aspects, post-operative conditions and surgical risk.

## Clinicoradiological Session



**Figure 2** - Angiography of aortopulmonary circulation emphasizes the anatomic aspects described. The pulmonary arteries are opacified by left upper aortopulmonary vessel (A and B) and another aortopulmonary collateral vessel to the left lower lobe (D). The left pulmonary artery shows clear stenosis (A and B) and is smaller than the right, which is directed to the entire right lung (C).

## References

1. Ando M, Duncan BW, Mee RB. Anatomic correction for corrected transposition after pulmonary unifocalization. *Ann Thorac Surg.*2003;75(3):1012-4.