Case 4/2010 - Seven-month-old Female Infant with Transposition of the Great Arteries and Subpulmonary Ventricular Septal Defect (Taussig-Bing Anomaly)

Edmar Atik, Lucilia S. Faria, Marcelo Jatene
Hospital Sírio Libanês, São Paulo, SP - Brazil

Clinical Data

The patient presented fatigue since birth, which had become more evident in the last two months, in addition to failure to thrive and presence of persistent coughing since the infant was one month old. The patient had received specific medication (digoxin, furosemide and captopril) since she was three months old, which did not alter symptom progression.

At physical examination, the patient presented with malaise, dyspnea+++, acyanotic, very irritated and had normal pulses. Arterial saturation was 60% in the awake state and 95% after sedation. The aorta was not palpable at the suprasternal notch.

The precordium showed marked impulses and bulging in the left sternal border and at the right 4th and 5th intercostal spaces and there was deviation of the ictus cordis. Heart sounds were muffled and a holosystolic murmur++ was heard in both hemithoraces, accompanied by thrill in the same sites. The liver was palpable at 5 cm from the right costal border and the xiphoid process. There were no adventitious lung sounds.

The electrocardiogram showed sinus rhythm and signs of left atrial and biventricular overload. AQRS was +110º, AP: +50º, AT: +60º (Figure 1).

Radiographic Image

The x-ray shows an enlarged cardiac silhouette, with markedly displaced left ventricular arch, bulging mid-arch+++ and increased pulmonary vascularity, mainly in the pulmonary hila (Figure 1).

Diagnostic Impression

The image is compatible with the diagnosis of cardiopathy

Key words

Infant; heart defects, congenital; transposition of great vessels; heart septal defects, ventricular; double outlet right ventricle.

Mailing address: Edmar Atik •
Rua Dona Adma Jafet, 74 conj.73 - Bela Vista - 01308-050 - São Paulo, SP - Brazil
E-mail: conatik@incor.usp.br, eatik@cardiol.br
Manuscript received July 30, 2009; revised manuscript received November 11, 2009; accepted November 12, 2009.

accompanied by marked left-to-right blood shunt, such as ventricular septal defect and/or ductus arteriosus.

Differential Diagnosis

All other cardiopathies, both acyanotic (atrial septal defect, double outlet right ventricle (DORV) + subaortic ventricular septal defect (VSD)) and cyanotic ones (DORV + subpulmonary VSD, atrioventricular valve atresia without pulmonary stenosis, transposition of the great arteries with VSD and TAPVC) must also be recalled.

Diagnostic Confirmation

The clinical elements were decisive for the diagnosis of cyanotic cardiopathy, considering the marked arterial unsaturation, even though it was reverted during sedation, caused by the marked pulmonary venocapillary congestion, as in cases with transposition of the great arteries with VSD. The echocardiogram confirmed the diagnosis, disclosing arterial vessels that were side-by-side, with the aorta on the right and a large subpulmonary VSD. The left cavities were very enlarged, as well as the pulmonary trunk and arteries.

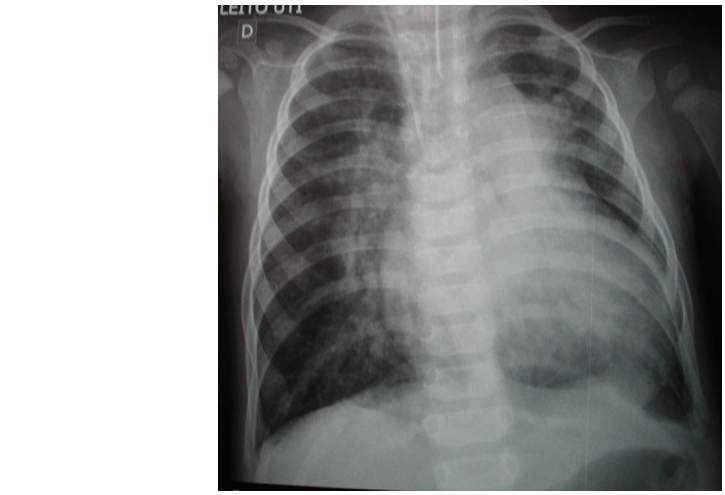
Management

For two days, before the surgical correction was carried out, the patient was maintained under mechanical ventilation with oxygen at 50%, with dobutamine and furosemide, in order to achieve a better compensation of the clinical status, which presented marked congestion and hypoxemia. Subsequently, the surgical procedure was considered inevitable, in spite of the possibility of associated pulmonary hypertension, obligatory in a seven-month-old infant with TGA+VSD.

The congestive heart failure caused by an excessive pulmonary arterial flow supported the surgical management and the diagnostic hemodynamic confirmation was then considered dispensable.

On-pump surgery with 180' of ECC was carried out through Jatene's technique, directing the LV to the PT through the subpulmonary VSD. Intense therapy with dobutamine, milrinone, NO, sodium nitroprusside, adrenaline, sildenafil, furosemide, digoxin and peritoneal dialysis with Tenckhoff catheter kept the pulmonary pressure within normal range (Chart 1).

The patient was removed from mechanical ventilation after 6 days, undergoing treatment of marked pulmonary



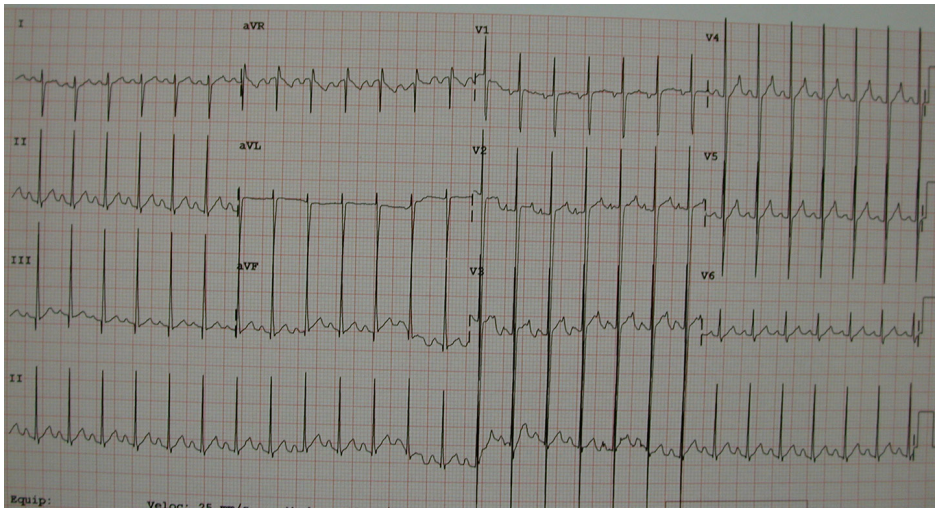


Figure 1 - Chest X-ray showing enlarged cardiac silhouette with marked pulmonary vascularity and bulging mid-arch. The electrocardiogram shows signs of biventricular overload.

hypersecretion. She was released after 15 days in good clinical condition.

Comments

Cyanotic cardiopathies with increased pulmonary arterial flow can be easily mistaken with acyanotic ones, such as VSD and/or PDA, due to the slight arterial unsaturation. However, a greater decrease in oxygen saturation can occur in situations of pulmonary congestion worsening.

In all these cyanotic anomalies and even in those with a more prolonged evolution, such as the TGA, the presence of signs of congestive heart failure (dyspnea, cardiomegaly, tachycardia, hepatomegaly, as well as marked heart murmur) can indicate the surgical approach, even in the supposed presence of pulmonary hypertension.

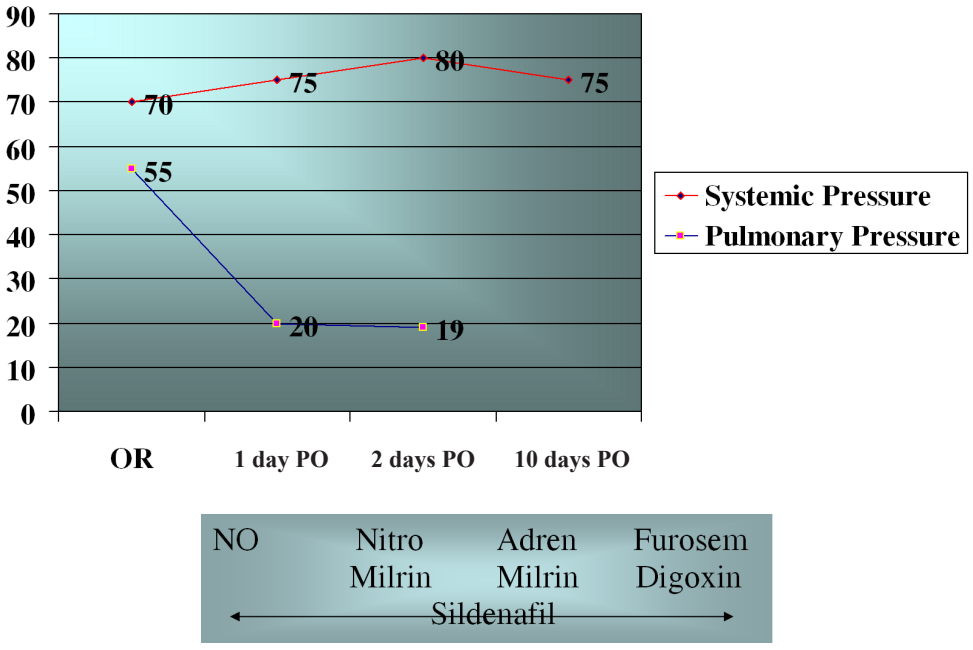


Chart 1 - Chart showing the postoperative evolution of arterial pressures, aorta and pulmonary artery, in relation to the medication prescribed. Abbreviations: Adren - adrenaline, Furosem - furosemide, Milrin - milrinone, Nitro - sodium nitroprusside, NO - nitric oxide, PO - postoperative, OR - operating room.