

Case 6/2012 – Newborn with Pulmonary Atresia, Ventricular Septal Defect and Double Aortic Arch

Edmar Atik

Instituto do Coração do Hospital das Clínicas da Faculdade de Medicina da Universidade de São Paulo, São Paulo, SP - Brazil

Clinical Data

36-week preterm infant born by C-section due to polyhydramnios; birth weight: 2,300 g and birth length: 42 cm, with Apgar scores of 8 and 9. Heart murmur was heard on the fourth day of life and an echocardiogram revealed the diagnosis of pulmonary atresia with ventricular septal defect, patent ductus arteriosus, and hypoplastic pulmonary arteries. Patient received prostaglandin E1 and remained acyanotic with 95% oxygen saturation, HR = 154 bpm and BP = 77/25 mmHg. When transferred to our service (InCor), the picture remained similar, but with PGE1 withdrawal, saturation decreased to 68%, which required the introduction of the intravenous agent.

Physical Examination

Good general status, eupneic, acyanotic, with normal pulses in all four limbs. The aorta was not palpable at the suprasternal notch. There were no deformities or impulsions at the precordium and the apical impulse was not palpated. Heart sounds were normal and there was a mild continuous murmur, ++ intensity, in the pulmonary area radiating along the left sternal border. The liver was not palpable and the lungs were clear.

Complementary Examinations

Electrocardiogram

Shows sinus rhythm and signs of right ventricular overload, with qR complex (5 mm) at V1, RS from V2 to V5 and Rs at V6. AP: +40°, AQRs: +110°, AT: +20°.

Chest X-ray

Shows normal heart size, excavated middle arch and pulmonary vascular network slightly more highlighted on the right (Figure 1).

Keywords

Pulmonary atresia; double aortic arch; VSD.

Echocardiogram

Shows normal cardiac chambers, with pulmonary atresia and ventricular septal defect, aorta originating from the right ventricle and small pulmonary arteries, with the right one being more developed (3.5 mm).

Chest Angiotomography

Shows, in addition to the elements found on the echocardiography, the existence of double aortic arch, with one of them to the right, from which emerged the left carotid and subclavian arteries, and then continuing with the descending aorta to the right. From the other main arc emerge the carotid and the left subclavian arteries, then interrupted and continuing with a large patent ductus arteriosus, which flows into the left pulmonary artery, showing stenosis at this juncture. The pulmonary trunk is of large caliber and the right pulmonary artery is larger than the left (Figure 2).

Clinical Diagnosis

Pulmonary atresia and ventricular septal defect, Double Aortic Arch with Interruption of the Main Arc continuing into large Ductus Arteriosus with stenosis at the junction with the Left Pulmonary Artery, with hypoxemia.

Clinical Rationale

Clinical findings are consistent with the diagnosis of cyanotic congenital heart disease, with hypoxemic effects, considering the saturation < 70% without the use of prostaglandin E1. Cardiac auscultation leads to the diagnosis of pulmonary atresia with patent ductus arteriosus, as observed in some of the anomalies that are similar from the functional point of view. In this context, pulmonary atresia with intact ventricular septum, tricuspid atresia Ia and IIa, and also all anomalies that accompany these two main defects, pulmonary atresia and patent ductus arteriosus, must be mentioned.

On the electrocardiogram, the right ventricular overload indicates the presence of two well-formed ventricles, as in pulmonary atresia with ventricular septal defect; the diagnosis was confirmed by echocardiography. The angiotomography established the accurate diagnosis of vascular anomaly of double aortic arch and the other variants described.

Differential Diagnosis

Other congenital heart defects that are associated with right ventricular overload and continuous murmur of the ductus arteriosus and pulmonary atresia include the double

Mailing Address: Edmar Atik •

InCor - Av. Dr. Enéas Carvalho de Aguiar, 44. Postal Code 05403-000, São Paulo, SP - Brazil

E-mail: conatik@incor.usp.br

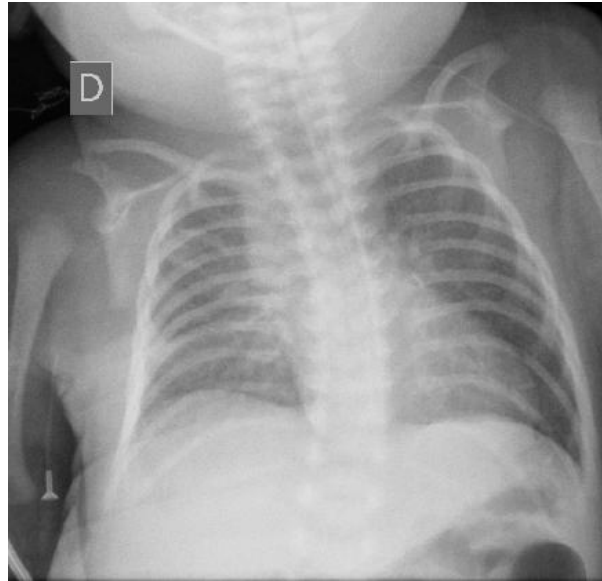


Figure 1 – Chest X-ray showing slightly enlarged cardiac area due to prominent left ventricular arch, excavated middle arch and somewhat increased pulmonary vascular network to the right.

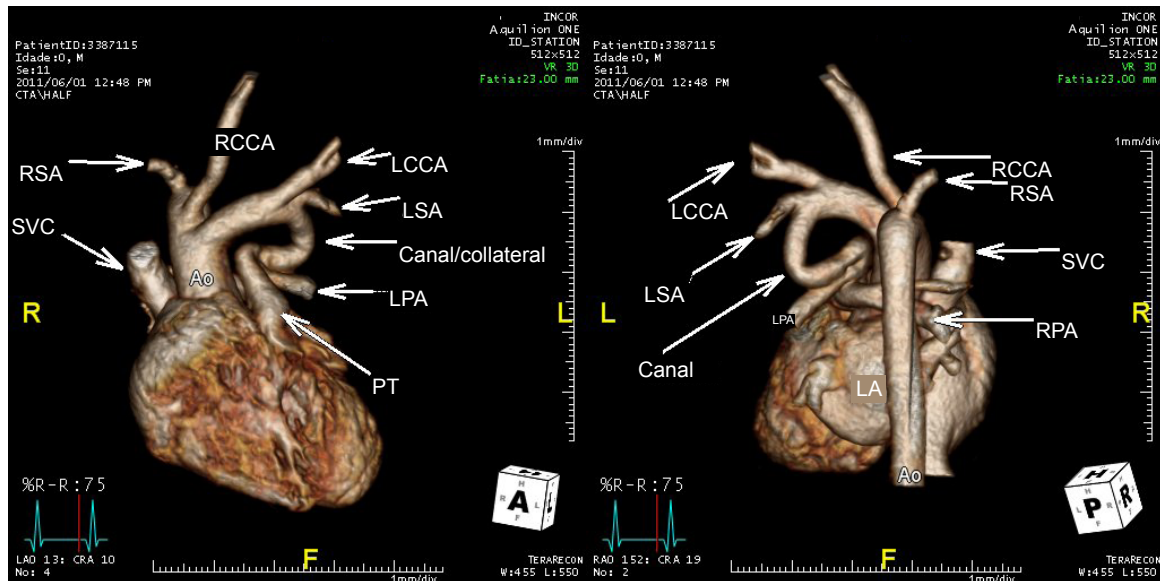


Figure 2 – Angiotomography in the anterior (A) and posterior (B) views shows a double aortic arch; one of them continues to the descending aorta to the right and the other is interrupted, but continuing with large ductus arteriosus and joining with the left pulmonary artery.

RCCA: right common carotid artery; LCCA: left common carotid artery; RPA: right pulmonary artery; LA: left atrium; LPA: left pulmonary artery; RSA: right subclavian artery; LSA: left subclavian artery; PT: pulmonary trunk; SVC: superior vena cava; Ao: aorta.

Clinicoradiological Session

inlet single right ventricle, corrected transposition of great arteries, double outlet right ventricle, among other more complex anomalies.

Conduct

We considered the need for surgical intervention, given the severe hypoxemia after PGE1 withdrawal. Considering the presence of a pulmonary trunk with adequate size, it was decided to open the right ventricle outlet and enlarge the pulmonary arteries. In fact, through a median thoracotomy, we performed the opening of the right ventricle outlet and even the closure of the large ventricular septal defect by the persistence of high pulmonary artery pressure of 70/20

mmHg. However, the patient died after three hours due to pulmonary hemorrhage, probably caused by an inadequate pulmonary arterial tree.

Comments

The diagnosis of cardiac anomalies is easily established by clinical elements confirmed by echocardiography. Anatomical alterations such as double aortic arch with all the characteristics of arterial vessel emergence from the two arcs are established by angiographic examinations or by cardiac catheterization or by angiotomography and magnetic resonance imaging. Thus, it was possible to plan the entire strategy for this patient, with these peculiar anomalies. In retrospect, a systemic-pulmonary anastomosis might have been the best choice.