Partial anomalous pulmonary venous connection with interventricular communication: the rare and possible difficulty on the anatomic definition of left atrial isomerism

Conexão anômala parcial de veias pulmonares com comunicação interventricular: a rara e possível dificuldade na definição anatômica de isomerismo atrial esquerdo

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CLINICAL DATA
9-month-old male child, 7 kg, born in Fronteira, MG. Born at full-term with 3.3 kg. On the 10th day of life heart murmur was noted, but referred to the pediatric cardiologist with 5 months due to fatigue while breastfeeding, excessive sweating and inappropriate weight gain. With the clinics indicating congenital heart disease with pulmonary hyperflow and congestive heart failure (CHF), chest X-ray, electrocardiogram and echocardiogram were requested, which were complemented by multi-detector computed tomography (MDCT).

Under use of digital and diuretic, on the physical examination the patient presented in good general condition, afebrile, acyanotic and with tachyypnea. Ictus cordis positioned to the right, regular heart rhythm with two clicks, ejection systolic murmur ++/4+ in right middle sternal border. Pulmonary auscultation with sparse bullous rales. Flaccid abdomen, liver at 3 cm from the left costal margin. Pulses were palpable and symmetrical in the four members.

ELECTROCARDIOGRAM
Sinus rhythm, heart rate 150 beats/min. PR interval of 80ms, QRS 80ms and QT 200ms. Negative P wave in D1, aVL indicating situs inversus totalis. Possible right ventricle overload (Figure 1).

Fig. 1 - Electrocardiogram with negative P wave in D1 and aVL suggesting situs inversus totalis

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RADIOGRAM

Slight increase of the cardiac silhouette with apex to the right, cardiothoracic index of 0.71. Signs of increased pulmonary vascular network with characteristics of hyperflow. There is compression on the right side wall of the trachea distal portion, suggesting aortic arch to the right. Liver in horizontal position (Figure 2A). In profile, it is noted increased of the anterior ventricular cavity (contact greater than 2/3 to the sternum), and in the pulmonary hila, two epibronchial arteries and bronchi with the same length (Figure 2B).

ECHOCARDIOGRAM AND TOMOGRAPHY

Echocardiogram diagnosed *situs inversus totalis* in dextrocardia, interatrial communication (IAC) *ostium secundum* of small size, wide ventricular septal defect (VSD), mild pulmonary valve stenosis by thickening of the valves.

Fig. 2 - Chest radiography: (A) Front in PA with dextrocardia and increased cardiac area. (B) Left profile with two epibronchial arteries and bronchi of the same length

Fig. 3 - Multi-detector computed tomography after 3D reconstruction: (A) Posterior view with descending aorta to the right, symmetrical and epibronchial pulmonary arteries. Bronchi also symmetrical originating two similar lobe bronchi, all with characteristics anatomically left. (B) Rear view with superior vena cava to the left, supra-hepatic veins draining into a common vein that, in its turn flows into the atrium to the left and two venous pulmonary trunks draining into each of the atriums.
and commissural fusion, dilated azygos vein draining into the superior vena cava (SVC) located to the left near the right atrium (RA) also positioned to the left. Presence of vein abnormally draining into the posterior region of the RA positioned to the left, which suggests such veins are left pulmonary veins, showing partial anomalous connection of left pulmonary veins (PAPVC) in RA. At Doppler, there was mean gradient between the right ventricle and pulmonary trunk (PT) of 24 mmHg and no significant gradient across the VSD.

MDCT, in addition to PAPVC, concluded that there was left atrial isomerism, with the pulmonary veins that drained the lung to the left and the SVC flowing into the atrium at left, which showed characteristics of left atrium (LA), and the pulmonary veins that drained the lung to the right flowing into the atrium at right also with characteristics of LA. The inferior vena cava (IVC) continued as azygous vein, which drained into the SVC positioned to the left. There was also a IAC and the aortic arch was to the right (Figures 3A and 3B).

**DIAGNOSIS**

The clinical presentation of CHF in the presence of PAPVC and IAC were dominant and sufficient to indicate the surgical correction, however, conflicting data respect to the situs was evident on the physical examination, electrocardiogram and echocardiogram, suggesting *situs inversus totalis*. A radiography with bronchi of the same length and liver in horizontal position favored the left atrial isomerism, according the findings of MDCT that showed two bronchii with left configuration, symmetrical, sealed; two epibronchial pulmonary arteries and both the bilobulated lungs, in addition to the presence of polysplenia and interruption of the IVC, which is an extremely reliable data of left atrial isomerism. Also, the fact of having two left pulmonary veins draining to the atrium positioned to the left, pointed to the left atrial isomerism, which was confirmed during the operation.

It is important to emphasize that the external aspect of the atrial appendage is not the best model of right or left morphology, and the presence of trabeculations on the atrial inferior wall should be valorized in order to characterize such atrium as right or the absence of them in order to designate an atrium as left [1].

**OPERATION**

With the surgeon positioned to the left of the patient, as previously described for patients with *situs inversus totalis* [2], after transsternal median thoracotomy and opening the pericardial sac, it was possible to note the external characteristics of the heart, with difficult definition respect to whether the atrium positioned to left was anatomically right or left (Figure 4).

Regardless of external aspect, the operation consisted of making circular purse-string sutures on the aorta using 5-0 polypropylene yarn and 6-0 polydioxanone yarn on the IVC and supra-hepatic veins. Heparinization with 4 mg/kg and introduction of cannulas of diameters appropriate for the weight.

Conventional cardiopulmonary bypass was initiated, hypothermia at 25ºC, aortic clamping, opening of the atrium positioned to left. Two pulmonary veins from the left lung were found draining into the aforementioned atrium (one highest pulmonary vein and near the great IVC and the
REFERENCES
