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The uncommon association between the aortopulmonary window and the aortic coarctation

A rara associação de janela aortopulmonar com coarctação de aorta

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CLINICAL DATA
A 2-month-old female infant, with a birth weight of 3,390 g, prematurely born with a gestational age of 35 weeks and 2,200 g, recovered uneventfully until the first month, when she begun to demonstrate breastfeeding fatigue and cyanotic episodes. She had gastroesophageal reflux. She was clinically treated. Because there was no clinical picture improvement, the doctor ordered an echocardiogram. Upon a diagnosis of heart disease, furosemide and captopril were administered. She was referred to surgical treatment. She was in good general health, active, reactive, red-faced, hydrated, tachypneic, acyanotic, afebrile, and anicteric. The heart rhythm was regular with two normal clicks, with the hypophonetic second heart sound (S2), ejection systolic murmur ++/+4+ in the left upper sternal border. Pulmonary auscultation was normal. There is no abdominal tenderness on palpation, with the liver 3 cm from the right costal margin. Because the increased amplitude in upper limbs and difficult to detect by palpation in lower limbs the pulses draw the attention.

ELECTROCARDIOGRAM
On electrocardiogram sinus tachycardia, heart rate of 166 bpm, AP + 60°, AQRS + 90°, PR interval = 0.08 seconds, QTc = 0.35 seconds, and signs of left atrium overload were all observed.

RADIOGRAM
On radiogram visceral situs solitus, cardiothoracic index of 0.61, discrete rectification of the costal arches and pulmonary vascular prominence with signs of pulmonary hyperflow, augmentation of the heart chambers, mainly the left atrium highlighted on lateral radiograph were all observed.

ECHOCARDIOGRAM
Situs Solitus was present at levocardia. Presence of a 12-mm aortopulmonary window (Figure 1); an 8-mm ascending aorta; a 5-mm aortic arch; and a 3-mm descending aorta in preductal region with an accelerated and turbulent flow in...
the descending aorta; and instantaneous gradient of 50
mmHg characterizing the aortic coarctation (Figure 2) were
observed. Left atrium presented increased diameters, left
coronary artery dilation with a 3-mm trunk and the right
coronary artery with a 1-mm trunk.

DIAGNOSIS
The aortopulmonary window, when associated to other
heart lesions, can be seen with an aortic arch interruption
or an interventricular communication [1]. However, its
association with the aortic coarctation is very uncommon.
Color Doppler Echocardiography has clearly shown such
diagnosis and the indication to surgical repair in a single
phase was chosen. On the differential diagnosis, it must be
kept on mind the following: patent ductus arteriosus,
truncus arteriosus, anomalous origin right pulmonary artery
from ascending aorta, and the tetralogy of Fallot with
pulmonary valve agenesis.

SURGERY
A longitudinal median sternotomy approach was performed,
which facilitated the examination of the anatomical
structures, giving an easy identification of the
aortopulmonary window. (Figure 3) Cardiopulmonary
bypass with cannulas in bicaual and aortic positions was
established. A Profound hypothermia was initiated after
the ligation of the pulmonary arteries. During the cooling-
off period, an extensive dissection of the following
structures: great vessels, aortic arch, and descending aorta.
When the temperature reached 18ºC, hypoflow of 40 mL/
kg, distal clamping of both aortic arch and descending aorta
was performed. The aortic wall was largely resected in order
to remove ductus arteriosus tissue, which had been
previously resected. The coarctation was segmental;
however, it was possible to perform a terminoterminal (end-
to-end) anastomosis with an absorbable surgical suture 6-
0. A normal flow was established, the descending aorta
was clamped, antegrade cardioplegia was performed, with
hypothermia 4ºC, and the repair of the aortopulmonary
window was initiated with removal of pulmonary trunk flap
to allow a greater amount of tissue to the aortic surface and
the direct closing of the aortic wall where it was also
employed the absorbable surgical suture. The pulmonary
wall was repaired with a fresh autologous pericardium (Figure
4). The perfusion time was 93 minutes and myocardial
ischemia was 25 minutes with a 12-minute hypoflow at 18ºC.
The patient is recovering well in the immediate postoperative
period using drugs for pulmonary hypertensión. The
postoperative echocardiogram showed an excellent surgical
outcome with a discrete turbulence on Doppler at the aortic
isthmus (gradient = 22 mmHg) without a clinical
repercussion. The patient was discharged on the 14th day
using captopril and diuretic.

REFERENCE
1. Konstantinov IE, Karamlou T, Williams WG, Quaegebeur JM,
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arch: a Congenital Heart Surgeons Society study. J Thorac