

# Alternative to an extracardiac Fontan-type operation: direct anastomosis between pulmonary trunk and inferior vena cava

*Alternativa para operação tipo Fontan extracardiaco: anastomose direta entre o tronco pulmonar e a veia cava inferior*

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## CLINICAL DATA

We report a case of a 9-year-old boy, weighing 20.8 kg, born in Salvador, BA. Cyanosis had been reported since birth by his mother, and two years ago she noticed a worsening in his physical condition when he started presenting episodes of dyspnea and fatigue with mild exertion. He was not taking any medication and had no previous surgical interventions.

On physical examination, he was in a good health condition, red-faced, hydrated, cyanosis ++++/4+, and digital hipocratism. Systolic fremitus ++/4+ on the left upper sternal border was perceptible on the precordial region, cardiac auscultation with two clicks with a hyperphonic second heart sound ( $S_2$ ), and systolic murmur ++++/6+ on upper sternal border irradiating to the right. Abdomen was normal. Peripheral pulses were present and symmetrical, and peripheral saturation was 73% on ambient air.

## ELECTROCARDIOGRAM

Electrocardiogram showed sinus rhythm and a heartbeat of 100 bpm;  $S\hat{A}P +30^\circ$ ,  $S\hat{A}QRS +240^\circ$ , PR interval of 0.12 seconds, and QTC of 0.43 seconds. Right ventricular overload evidenced by Rs complex in Vs and rS complex in lead V6.

## RADIOGRAM

On radiogram visceral situs solitus was present at levocardia. The cardiac area was within normal limits presenting a cardiothoracic index of 0.41. We could not see

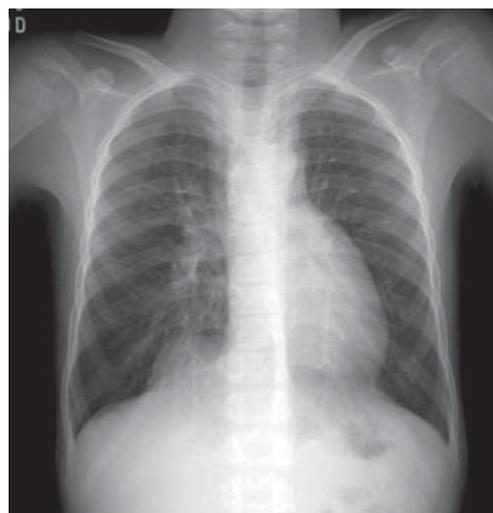


Fig. 1 – Chest radiography showing the positioning of the heart with absence of a suggestive image of atrium to the right

atrium image on the right, which attracted our attention in addition to the lobulation of the medial portion of right hemidiaphragm suggesting Morgagni hernia or fat tissue hernia. The transparency of the pleuropulmonary fields was normal (Figure 1).

## ECHOCARDIOGRAM

Situs solitus was present at levocardia. Venoatrial and

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atrioventricular connections were normal. Ventriculoarterial connection was abnormal with two arteries originating from the right ventricle. On Doppler, a turbulent and accelerated flow in the double outlet of the right ventricle and in the pulmonary trunk was seen, what was compatible with infundibular stenosis; maximum peak gradient was 66 mmHg. The following measurements were obtained: interventricular communication (IVC) = 18 mm, aortic valve ring = 20 mm, pulmonary valve ring = 6.5 mm, pulmonary trunk = 7.5 mm, right pulmonary artery (RPA) = 11 mm, and left pulmonary artery (LPA) = 6 mm. Thus, the echocardiogram displayed double outlet of the right ventricle with unrelated interventricular communication, anterior aorta, posterior pulmonary trunk, significant infundibular pulmonary stenosis, and hypoplasia of left pulmonary artery.

#### DIAGNOSIS

Taking into consideration the patient's extremely acute clinical condition, due to his anatomy, age, degree of cyanosis, and pulmonary impairment, it was chosen to perform a repair with univentricular physiology. The disproportion between pulmonary artery diameters (Figure 2) was well documented by the hemodynamic study, which also evidenced the double outlet of the right ventricle with anterior aorta and posterior pulmonary trunk, ostium secundum interatrial communication of moderate size, significant supralvalvar pulmonary stenosis and provided a pulmonary artery mean pressure of 25 mmHg.



Fig. 2 – Hemodynamic study showing the positioning and disproportion between the pulmonary arteries

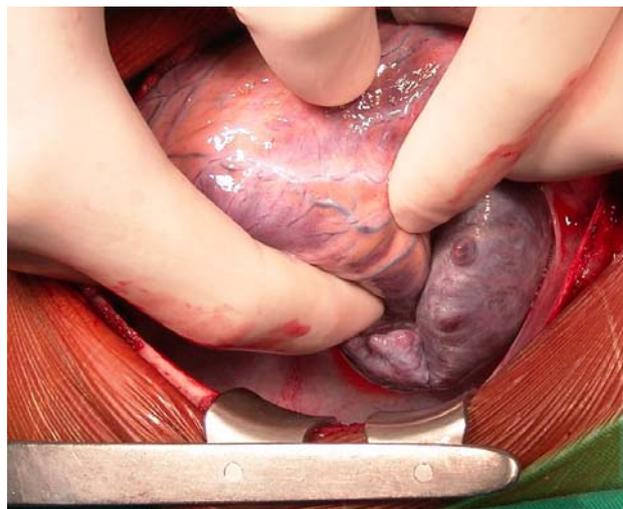


Fig. 3 – Juxtapposition of atrial appendices to the left

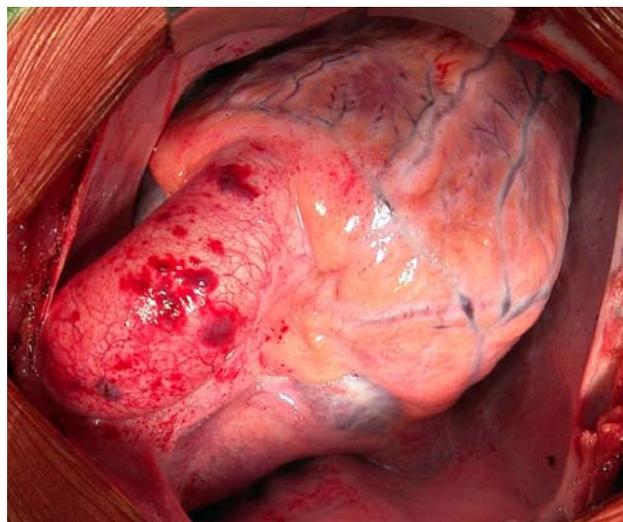


Fig.4 – Image of the anterior aorta and the posterior pulmonary trunk. A huge superior vena cava grafted directly into the atrium with absence of right atrial appendix can be observed

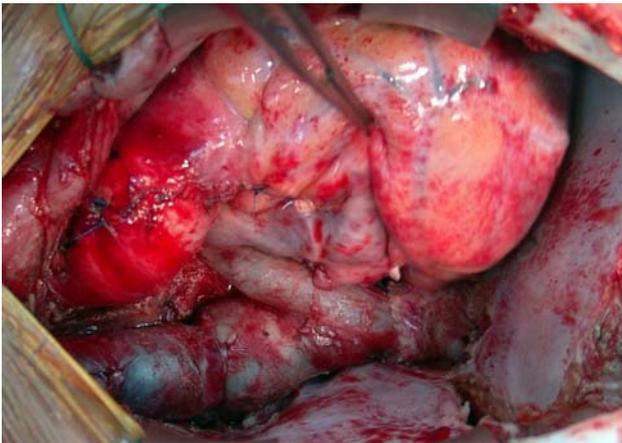
#### OPERATION

A median transsternal thoracotomy was performed; the pericardium was opened and an analysis of external cardiac structures and a wide dissection of superior vena cava (SVC), inferior vena cava (IVC), pulmonary trunk, left and right pulmonary arteries was made. Some aspects attracted attention such as the length of right pulmonary artery was 3 times that of the left pulmonary artery, the atrial appendices were juxtaposed to the left (Figure 3) and the pulmonary trunk, besides being posterior, was discreetly positioned to the right; also, there was a supralvalvar pulmonary stenosis (Figure 4). These features made us think about

the possibility of a direct connection between the pulmonary trunk and the inferior vena cava.

Patient was heparinized, conventional cardiopulmonary bypass (CPB) support was established with aorta cannulation, superior vena cava was close to the brachiocephalic vein (innominate vein), and the inferior vena cava was close to the suprahepatic veins. Cardiopulmonary bypass was started with hypothermia at 26°C, intermittent antegrade blood cardioplegia at every 20 minutes and hypothermic blood cardioplegia at 4°C.

The pulmonary trunk was sectioned close to the base of heart, resection of pulmonary valve and suture of the proximal stump with 5-0 and 6-0 polypropylene threads in order to avoid external bleeding and preventing thrombus to buildup internally to the heart. The pulmonary trunk was widely mobilized with dissection of pulmonary arteries up to the pulmonary hila, and the pulmonary trunk ostium was discreetly augmented using a longitudinal incision at the local where there was a supravalvar pulmonary stenosis. After, PT was positioned to the right of the heart following the inferior vena cava and parallel to the atrioventricular sulcus.



*Fig. 5 – Final aspect of the operation with the pulmonary trunk directly anastomosed to the inferior vena cava and the superior vena cava anastomosed to the right pulmonary artery. A significant disproportion between the diameter of the pulmonary arteries can be noticed*

Inferior vena cava was sectioned from the atrium to a much great extent as possible, in such a way to allow a direct connection between the vein and the pulmonary trunk (Figure 5); the anastomosis was performed with a 5-0

polydioxanone thread (extracardiac Fontan-type operation). With such a spatial positioning, the azygos vein was grafted and sectioned previously to the superior vena cava also being cut off from the atrium and anastomosed in the best anatomic possible way directly into the right pulmonary artery and sutured with a 6-0 polydioxanone thread (bidirectional Glenn operation).

The atrial stumps were sutured using 5-0 polypropylene thread and the interatrial septum was completely cut off. Perfusion time was 95 minutes and myocardial ischemia time was 45 minutes [1,2].

At the Intensive Care Unit, the patient coursed with pleural effusion to the right during 14 days, which has subsided with a low-fat diet. The patient was discharged home on day 21 postoperatively. Immediate postoperative echocardiogram showed, on Doppler, absence of flow between the right ventricle and the pulmonary trunk, biphasic laminar flow with maximum velocity in the anastomosis between the inferior vena cava and the pulmonary trunk, and in the anastomosis between the superior vena cava and the right pulmonary artery of 0.59 m/s and 0.68 m/s, respectively, suggesting normal drainage. The size of the interatrial communication after septostomy was enlarged to 12 mm, inferior vena cava to 14.5 mm, right pulmonary artery to 14 mm, and left pulmonary artery to 7 mm; ventricular contractile function was normal.

Four months after the operation, the patient is taking only acetylsalicylic acid, 5 mg/kg, once there is no prosthetic tissue and, thence, there is no need of using dicumarol as it should be usually used in total cavopulmonary operation or Fontan type operation using extracardiac catheter.

#### REFERENCES

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