Echocardiographic Diagnosis of Transposition of the Great Arteries with the Posterior Aorta: Presentation of Two Cases and Literature Review

Transposition of the great arteries (TGA) is the most common cyanotic congenital heart disease in the neonate. It is characterized by ventricular-arterial (V-A) discordance. Great arteries usually run in parallel, with the aorta in the anterior position and to the right of the pulmonary artery (PA). The posterior artery is the PA and there is mitro–pulmonary continuity.

We report two cases of newborns with TGA where the aorta was posterior to the pulmonary artery (P-TGA), and with mitro-aortic continuity as in a normal heart –a very rare anatomical variant–, with diagnostic difficulties and particular clinical and surgical implications.

Case 1

A 10-day old infant, weighing 3,460 g, was referred due to cyanosis since birth. Physical examination revealed mild cyanosis, symmetrical, wide pulses, enlarged S2, soft ejective systolic murmur at the base, and S3 at the apex.

Chest X-ray showed moderate cardiomegaly and pulmonary hyperflow. The electrocardiogram revealed right axis and signs of biventricular enlargement.

Transthoracic echocardiography showed situs solitus, levocardia, and atrioventricular concordance. Apparent V-A concordance was observed at the parasternal short axis, as a heart in normal position. At the parasternal long axis, mitro-aortic continuity through a ventricular septal defect (VSD) was visualized; however, the aorta appeared to be more related with the right ventricle (RV) (Figure 1). The PA seemed to emerge from the left ventricle (LV). Mitro-pulmonary discontinuity was detected. The aortic and pulmonary annuli were similar in size. The pulmonary valve was bicuspid. The right coronary artery (RCA) and the anterior descending artery (ADA) emerged from the right coronary sinus, in "shotgun tube". The circumflex artery (Cx) emerged from the left coronary sinus.

Arterial switch operation with VSD closure was performed at 13 days of life. During coronary reimplantation, both buttons remained in the same sinus (bicuspid valve). The Lecompte maneuver was not necessary. After the switch procedure, the neoaorta had a left anterior location, and the neopulmonary artery had a right posterior location.

The immediate postoperative course was favorable, and the patient was discharged 12 days after surgery. Echocardiography before discharge showed a satisfactory repair.

Case 2

A 31-week gestational age infant, weighing 1,600 g. was referred due to cyanosis 4 days after birth. The physical examination revealed moderate cyanosis and symmetric peripheral pulses. S1 was normal and S2 was enlarged, with ejective systolic murmur at the base.

Echocardiographic findings were similar to those of the first case, with some differences. While the aorta was posterior to the PA, it was rather side by side with it. Mitro-aortic continuity was also detected, as in a heart with normal position of the great vessels. There was a single coronary artery arising from the left posterior sinus. The right coronary artery surrounded the pulmonary annuls and ran in front of the aorta to the right AV sulcus (Figure 2).

Due to low weight at birth and hypoxemia associated to restricted foramen ovale, a balloon atrial septostomy was performed. After reaching a weight of 2,200 g, at 46 days of age, an arterial switch operation with VSD closure and coronary reimplantation was performed, without Lecompte maneuver. Postoperative echocardiography showed no residual defects.

Postoperative course was torpid. The patient developed Acinetobacter sepsis with refractory shock and died on the 21st postoperative day.

In TGA, the vessels usually emerge in parallel with the aorta located anteriorly and to the right of the PA (R-TGA). In these two cases, we had cyanotic patients whose echocardiographies simulated normal position of the great arteries (right posterior aorta), with mitro-aortic continuity. However, when assessing the connection between the ventricles and the arteries, a V-A discordance (TGA with posterior aorta or P-TGA) was confirmed.

In the transposition of the great arteries, the aorta is usually anterior and to the right of the PA; the aorta being anterior and to the left (L-TGA) is less common.

Posterior TGA is the most uncommon anatomical variations of the transposition of the great arteries. Echocardiographic diagnosis is complicated. It is necessary to evaluate various planes and search for V-A discordance even if at first glance there seems to have V-A concordance.

Mitro-aortic continuity was visualized through the parasternal axes, as in a heart in normal position, but the aorta was associated with the RV. This continuity occurs through a subaortic VSD. In the same axis, the connection between the PA and the LV was visualized, showing mitro-pulmonary discontinuity.

We used a view from the right infraclavicular area, performing a basal short axis view that allows the simultaneous visualization of both sigmoidal arteries and both AV valve annuli. In this view we evaluated the relationship between vessels and with the aorta located between both annuli (tricuspid-aortic and mitro-aortic continuity).

Both patients had small subaortic VSD. In the first case, the pulmonary valve was bicuspid (future neoaorta). Both patients had coronary anomaly; in the first case, the anterior descending artery (ADA) originated from the RCA, and the second case presented single left coronary artery and RCA with an anterior course to both semilunar valves.

Double outlet LV was proposed as differential diagnosis. There is a variant of this anomaly, in which the aorta is posterior and to the right of the PA, with subaortic VSD. In this condition, the aorta emerges mainly from the LV and goes over more than 50% of the VSD. In our two cases, the aorta fully emerged from the right ventricle.

The transposition of the great arteries with posterior aorta was first described by Van Praagh et al. in 1971, after reviewing 4 cases of pathological specimens. (1) In that publication, Van Praagh mentions a case shared by Dr. Luis Becú (Hospital de Niños, Buenos Aires). He reported the case of a patient with TGA who died after mistaken cerclage of the posterior artery (personal communication, Dr. Horacio Capelli).

The term 'posterior transposition' (p-transposition) refers to the aorta originating from the right ventricle but retaining fibrous mitro-aortic continuity, and the pulmonary artery originating from the LV with bilateral conus. (2-5)

The main morphologic characteristics, as described by Anderson (2, 3) in 1975, are the following:

1. Posterior origin of the aorta from the RV.
2. Presence of complete subpulmonary conus.
3. Fibrous mitro-aortic continuity through a ventricular septal defect. (1, 2) Normally, the subaortic conus is absorbed in its central portion forming a fibrous area (mitro-aortic continuity). (3) In the P-TGA, absorption is only partial, and the fibrous part lies posterior and to the left; hence, the aortic root is in fibrous continuity with the mitral valve via the central fibrous body. (2)
4. Wrong orientation of the conal septum with the interventricular septum. (2)

Only a few cases have been reported, most of them discovered when reviewing pathological samples. The in vivo cases reported were diagnosed with catheterization (6); only three had an echocardiographic diagnosis (Béland-Paquet 1988, Sayuri 1993). (7, 8) All of them presented with mitro-pulmonary discontinuty and maintained mitro-aortic continuity through subaortic VSD. Pulmonary stenosis was the most common associated anomaly. Only one case presented with subaortic stenosis. The coronary pattern described for P-TGA is a mirror image of the usual pattern of TGA with anterior aorta. It is associated with coronary anomaly (ADA originating from the RCA).

It is important to be aware of this rare anatomical variant of TGA, since in a neonate with cyanosis and alleged V-A concordance, a pulmonary condition may be assumed as the cause of the cyanosis, which would delay the surgical repair, increasing morbidity and mortality. In addition, it is of great relevance for the surgeon to know the anatomy beforehand to plan the surgical strategy. While these patients are treated with the arterial switch operation, the Lecompte maneuver –bringing the PA to anterior position– is not required (9-11) (in this variant, the PA is the anterior vessel).

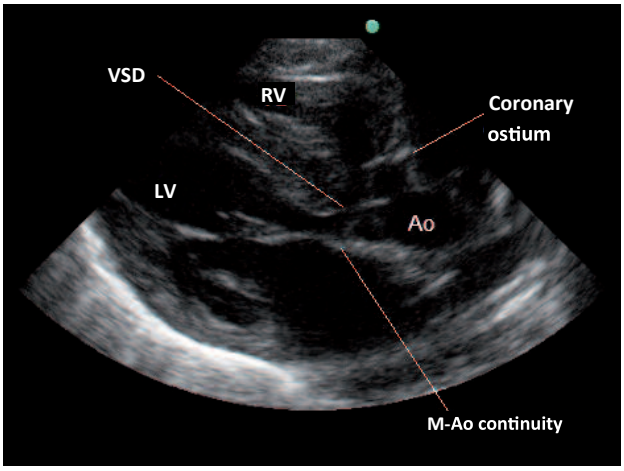

Fig. 1. A & B. Long axis parasternal view. Case 1. Relationship between the ventricles and the great arteries. Aortic valve continuity with the anterior mitral leaflet and ventricular septal defect is observed. VSD: Ventricular septal defect. RV: Right ventricle. LV: Left ventricle. Ao: Aorta. MPA: Main pulmonary artery. M-Ao: Mitro-aortic. M-P: Mitro-pulmonary.

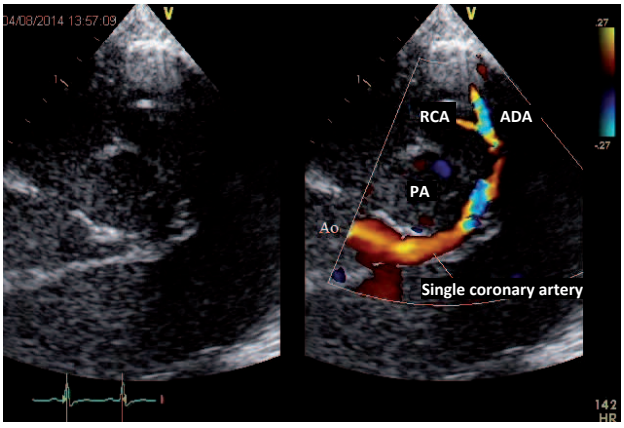

Fig. 2. Case 2 coronary pattern. Single coronary artery arising from the left sinus.

Some cases of transposition require cerclage of the pulmonary artery before the switch operation (premature newborns, multiple VSDs, preparation of the LV, centers with little experience). Therefore, it is important that the surgeon performs cerclage of the left anterior vessel in this variant.

Transposition of the great arteries with posterior aorta is a very rare anatomical variant. Echocardiographic diagnosis is complicated. It is important to assess V-A discordance in different echocardiographic planes to make the diagnosis and decide on the best surgical approach (arterial switch). The left ventricular double outlet, which is also a rare condition but may lead to misdiagnosis due to the anatomical characteristics of P-TGA should be ruled out as a differential diagnosis.

Conflicts of interest
None declared.
(See authors' conflicts of interest forms on the website/Supplementary material).

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Blue Toe Syndrome as Expression of Severe Atherosclerosis

Aortic atherosclerotic plaques are a source of embolic phenomena that can occur with cerebral, splanchnic, or peripheral manifestations.

Cholesterol embolization syndrome (CES) or atheroembolism is the less common manifestation of embolism of an atherosclerotic plaque, and its prevalence is underestimated. It may occur spontaneously or after an arterial endovascular procedure. Cholesterol embolization syndrome is classified as definite and possible. Definite CES presents with cutaneous signs such as livedo reticularis, blue toe syndrome (BTS), and digital gangrene with or without renal involvement. Possible CES only shows renal involvement, that is, serum creatinine >1.3 mg/dl, two weeks after catheterization with normal renal function before the procedure, without cutaneous lesions. Atheroembolism, usually of the abdominal aorta, includes fragments of atherosclerotic plaque that contain cholesterol crystals and fibrin and platelet thrombi leading to disseminated microembolism. Microembolism causes inflammation associated to mechanical occlusion, and both cause ischemia and necrosis. (1)

Blue toe syndrome is a dermatological manifestation of CES with a frequency between 35% and 96%, characterized by tissue ischemia secondary to atheroembolism causing occlusion of the small vessels in the extremities. It presents with focal areas of painful cyanosis in the extremities, surrounded by normal tissue perfusion and preservation of distal pulses. Embolism typically originates from an ulcerated atherosclerotic plaque or from aneurysms located in the aortoiliacfemoral system. It can occur spontaneously or due to several causes (endovascular procedures, vascular surgery, anticoagulation, fibrinolysis). It is important to make differential diagnosis with Raynaud's syndrome, lesions due to hypothermia and idiopathic digital arterial thrombosis, as well as establishing the presence of atheroembolism in BTS, because it is a recurrent phenomenon and can lead to limb amputation or death if the embolism is very extensive. (2)

Diagnosis is predominantly clinical, but definitive diagnosis is made with biopsy of muscle or skin showing cholesterol crystals. Laboratory findings are nonspecific, but reveal a strong correlation with eosinophilia. (1)

Regarding diagnostic imaging, various methods can be used. Doppler ultrasound detects aneurysms or plaques proximal to the affected vascular bed, determining the embolic source. Computed tomography angiography and magnetic resonance angiography reveal the cause and severity of the underlying lesion. Diagnostic arteriography can determine the cause of thrombosis, provide information about the proximal circulation, and accurate details of the extension of collateral circulation and distal flow to the areas occluded by the embolism. (2)

We report a case of definite CES with BTS as dermatological manifestation, in a 57-year-old male patient, with cardiovascular risk factors –hypertension, non-insulin-dependent diabetes mellitus, dyslipidemia and smoking–, and unremarkable past medical history, who was admitted to our center due to 12-hour history of painful cyanosis in the right foot toes (Figure 1 A). The physical examination confirmed lack of pedal pulse and reduced posterior tibial pulse in the right lower limb. Lab tests revealed leukocytosis with eosinophilia and 0.75 mg/dl creatinine. The electrocardiogram was normal.