Parkes-Weber Syndrome with Pulmonary Hypertension Due to Multiple Arteriovenous Fistulas

Parkes-Weber syndrome (PWS) is an arteriovenous vascular malformation. It occurs at birth and affects mainly the lower limbs (77%). Detection of arteriovenous fistula associated with the Klippel-Trénaunay syndrome (KTS) triad confirms the diagnosis of this syndrome. (1, 2)

The KTS is characterized by the typical triad of cutaneous vascular port wine stains, soft tissue and/or bone hypertrophy and varicose veins (Figure 1). The difference between PWS and this syndrome is that PWS presents high-flow vascular lesions and arteriovenous fistula, no anomalous lateral veins, lymphatic malformations are rare, and the musculoskeletal involvement and dissymmetry of the affected extremity is lower than in KTS. (3)

The major complication in PWS is the increased cardiac output that can lead to heart failure, pulmonary hypertension, and cutaneous ischemia, caused by high-flow arteriovenous fistulas. (3)

Although its etiology is unknown, PWS has been associated with mutation of the E133K gene in the angiogenic factor VG5Q, which controls the growth of blood vessels for angiogenesis and vasculogenesis during embryonic development, and is transmitted by autosomal dominant inheritance with variable expressivity. PWS is much less common than KTS. (4)

High cardiac output with heart failure and pulmonary hypertension is associated with different conditions, including traumatic arteriovenous fistulas and therapeutic and congenital ductus arteriosus. The primary pathophysiological event is reduced peripheral vascular resistance due to peripheral vasodilation or arteriovenous fistula, and both scenarios can lead to a reduction of systemic blood pressure, neurohormonal activation, ventricular remodeling, and heart failure.

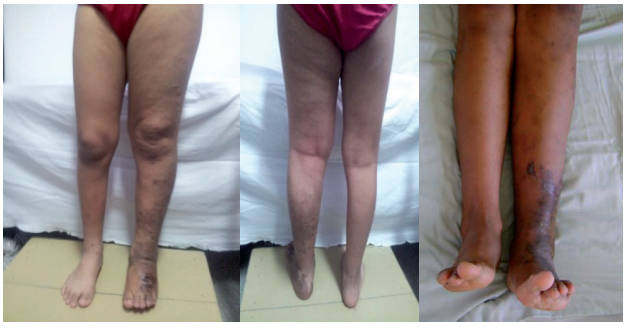

Fig. 1. Echocardiographic image following self-expandable valve implantation.

In these cases, conventional therapy for heart failure, such as angiotensin-converting enzyme inhibitors, beta blockers, and angiotensin receptor blockers, reduces peripheral vascular resistances and may result in the patient's clinical deterioration, although the literature reports this treatment in some cases.

Treatment consists of elastic compression, ligation of arteriovenous fistulas, orthopedic therapy, and eventually percutaneous embolization, in many cases resulting in limb amputation.

We present the case of an 11-year-old male Argentine patient, with no family history of this condition. The patient presented with an 8-year history of the disease, with difficulty in walking during childhood associated with multiple falls, increased volume, hyperpigmentation and indurations in the left lower limb.

He referred FC II dyspnea (New York Heart Association [NYHA] classification) associated with nocturnal dry cough. He also presented with elevated lesions and pruritic honey-colored scabs, moderate pain and heaviness of the left lower limb together with spontaneous hemorrhages described as a continuous stream of blood shooting up to 50 cm, with a frequency of five episodes in the last month.

The patient was oriented and afebrile; BP in right arm: 100/60 mm Hg; BP in left arm: 100/50 mm Hg; HR: 94/min; T: 36.6 °C.

Cardiovascular system: Jugular venous distension at 0 plane level, normal S1; regular S2: (P2 > AO2), no S3, non-irradiated systolic murmur in pulmonary focus 2/6, apex beat in 5th LIS.

Limbs: Left lower limb: increased volume, 11 cm in the thigh and 6.6 cm in the leg, 5 cm in the ankle, and 3 cm high with respect to the contralateral limb; increased temperature, multiple isolated brownish macular lesions of net, irregular borders, indurated hyperpigmented lesions ('port wine' stains), and visible venous pathways; macules, peripheral scaling and multiple blood stretch marks were observed in the dorsum of the ipsilateral foot.

Murmur with thrill in the dorsum of the left foot; pulses, altered ambulation.

Chest X-ray: CTI > 0.50.

Electrocardiogram: RS HR: 84/min, axis at 40°, left atrial and ventricular overload.

Color Doppler echocardiography: LVDD: 56 mm, LVSD: 32 mm, IVS: 10 mm, PW: 7 mm, Ao root: 26 mm, LA: 38 mm, RVDD: 18 mm, EF 70%, ShF: 42%. Pulmonary annulus: 26 mm, inferior vena cava: 15 mm, collapse <50%, TAPSE: 23. Left chamber dilation; preserved biventricular function. Moderate pulmonary hypertension, PASP: 50 mm Hg.

Arterial and venous lower limb Doppler ultrasound: Calibers: FV: 10.9 mm, PV: 10.8 mm, PV: 4 mm, CFA: 8.8 mm, PA: 7.6 mm, TPI: 4.6 mm, pedial artery: 3.36 mm. Multiple high-flow arteriovenous fistulas in CFA, proximal and distal popliteal artery, and at the tibial-peroneal trunk. Impaired venous flow, increased density and velocity were found on pulsed Doppler. Right lower limb: Without vascular disorders.

Multislice computed tomography angiography of the lower limbs: Hypertrophy of the left lower limb is observed. During scan acquisition of the arterial phase, opacification of the superficial and deep venous drainage system is seen, suggestive of microfistulas or fistulas (Figure 2).

Impaired bone structure at the calcaneal and mid third of the calf bones is found, as part of the vascular disorder.

Varicose dilatation of the superficial venous drainage system is observed. No signs of superficial and deep vein thrombosis are detected.

Changes are appreciated in the density of subcutaneous tissue of the leg, associated with edema.

Right lower limb: No vascular disorders are detected. Bone structures and soft tissues are normal.

The case reported matches the clinical data and imaging findings characteristic of this syndrome, together with the musculoskeletal hypertrophy of the left lower limb and increased circumferential and longitudinal diameter, cutaneous vascular disorders and high-flow arteriovenous fistulas in popliteal, femoral, and tibial vessels demonstrated with venous Doppler ultrasound and multislice CT angiography (Figure 3). (5)

Over the course of time, high-output arteriovenous fistulas cause dilation of the heart chambers, pulmonary hypertension, and heart failure, (6) which is the most severe complication. The effective fistula treatment is essential to prevent this hemodynamic abnormality.

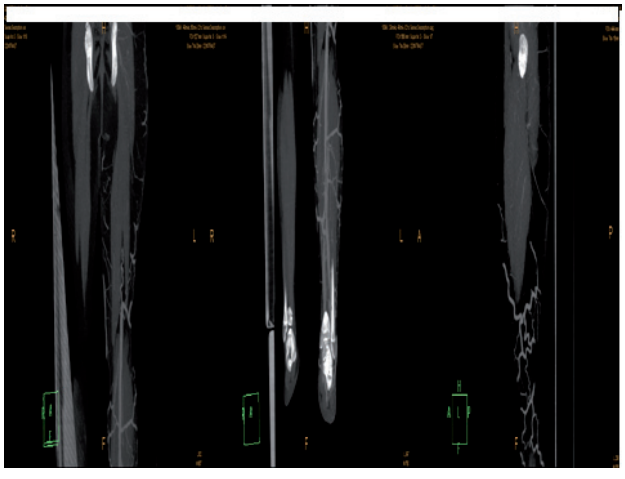

Fig. 2. Echocardiographic image following self-expandable valve implantation.

Fig. 3. Echocardiographic image following self-expandable valve implantation.

Conflicts of interest

None declared.

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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).

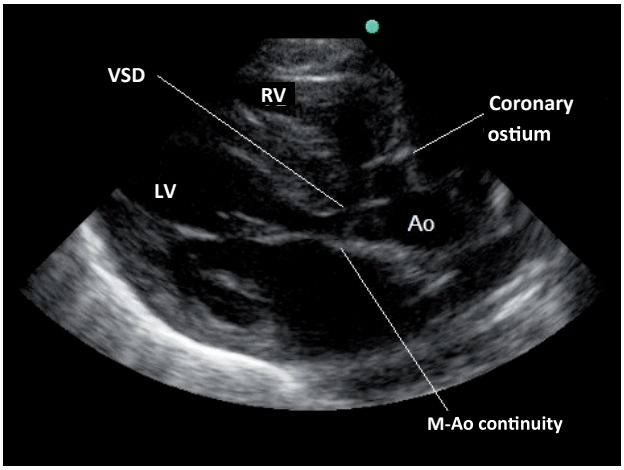

Fig. 1. A & B. Long axis parasternal view. Case 1. Relationship between the ventricles and the great arteries. Aortic valve continuity with the anterior mitral leaflet and ventricular septal defect is observed. VSD: Ventricular septal defect. RV: Right ventricle. LV: Left ventricle. Ao: Aorta. MPA: Main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

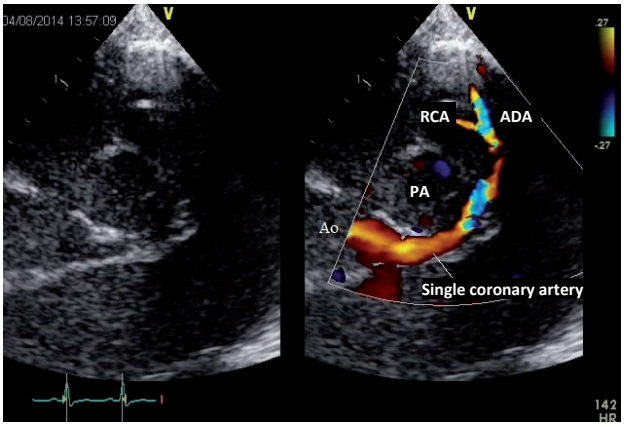

Fig. 2. Case 2 coronary pattern. Single coronary artery arising from the left sinus.

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.
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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.

Diagnostic arteriography showed an ulcerated plaque causing severe stenosis at the level of the distal abdominal aorta, involving the origin of the inferior mesenteric artery (Figure 1 B), severe stenosis in the origin of the right primitive iliac artery, two severe stenoses in the origin and distal portion of the left primitive iliac artery (Figure 1 C), and opacification of the posterior tibial artery in the right foot; the anterior tibial and peroneal arteries were not visualized (Figure 1 D).

The condition was interpreted as BTS due to spontaneous atheroembolism as a result of ulcerated atherosclerotic plaque in the distal abdominal aorta, worsened by ischemia of the lower limbs secondary to severe stenoses described in both primitive iliac arteries.

It was decided to continue with endovascular treatment of the aorto-iliac axis disease.

Our case showed a large, ulcerated plaque in the distal abdominal artery as source of embolism. One of the options for endovascular resolution was covered stent placement to prevent embolization during the procedure. Covered stent implantation in the aorta was ruled out because the inferior mesenteric artery originated in the described plaque and would be occluded during stent placement. Bare self-expanding nitinol stents were chosen instead.

Access was performed with an 8F introducer in the right femoral artery and a 6F introducer in the left femoral artery, and the described stenoses were managed with bilateral 0.035" hydrophilic guidewire. A 14 mm wide by 60 mm long stent in the distal aorta, a 9 mm wide by 60 mm long stent in the right primitive iliac artery, and two stents, one 9 mm wide by 40 mm long and the other 8 mm wide by 40 mm long, in the left primitive iliac artery, were implanted. The aortic stent was dilated with a 10 mm balloon and the procedure was finished with a kissing balloon at the level of the aorto-iliac bifurcation with two 8 mm balloons, with positive angiographic outcome and without involving the origin of the inferior mesenteric artery (Figure 2).

The patient evolved with no toe pain, persistence of cyanosis, recovery of pedal pulse, and marked improvement of posterior tibial pulse. The patient was discharged 72 hours after the procedure with dual antiplatelet therapy, statins, vasodilators, and hypoglycemic agents.

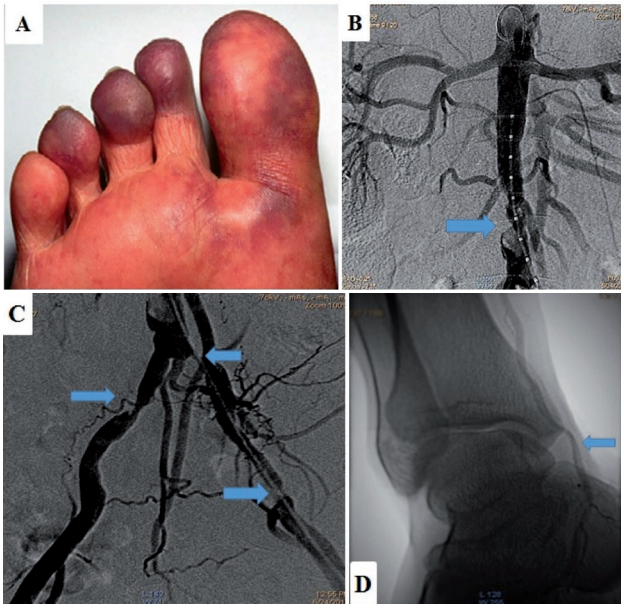

Fig. 1. A. Right foot toes showing cyanosis. **B.** Ulcerated atherosclerotic plaque in distal abdominal aorta involving the origin of the inferior mesenteric artery (*arrow*). **C.** Severe stenosis in both primitive iliac arteries (*arrows*). **D.** Distal circulation. Only the right posterior tibial artery shows opacification (*arrow*).

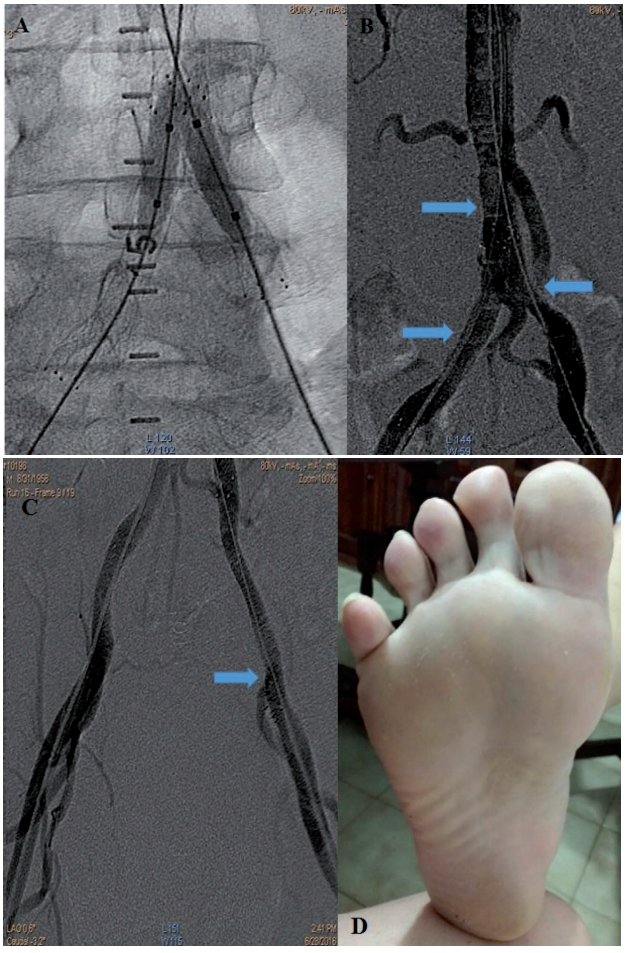

Fig. 2. A. Kissing balloon placement is observed. **B & C.** Angiography shows implanted stents and absence of inferior mesenteric artery involvement (*arrows*). **D.** Complete resolution of cyanosis.

At 10-month follow-up, the patient has made good progress with no recurrent embolic phenomena, complete resolution of cyanosis in right toes (Figure 2 D), preserved distal pulses, and arterial Doppler ultrasound showing triphasic flow in the right lower limb.

There is no specific therapy for BTS. Supportive measures to prevent the progression of atherosclerotic disease include modifications of risk factors, use of statins and antiplatelet agents, and avoidance of anticoagulation. (3)

Traditionally, the treatment options for BTS include endarterectomy or bypass to exclude the embolic source. Due to the bad prognosis of patients with BTS, there is still much controversy about the best treatment for these patients. For this reason, various studies focusing on the resolution of the entity have been published. Stents proved to be effective in the treatment of this arterial condition, providing a platform to prevent future embolisms and promote atherosclerotic plaque remodeling. Although there is concern for increasing distal embolization during stent placement, there is no clinical evidence to demonstrate it in the period immediately after placement of the device. (4-6)

Blue toe syndrome is a relatively rare condition, but it is a manifestation of atherosclerotic disease that can be devastating due to its morbidity and mortality. Cardiologists should bear this entity in mind when encountering a patient with the clinical manifestations described above, since early revascularization can prevent limb amputation or patient death.

Conflicts of interest
None declared.
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