Digital Ischemic Necrosis of the Upper Limb Due to Thoracic Outlet Syndrome

Thoracic outlet syndrome encompasses diverse clinical disorders resulting from compression of the neurovascular structures running from the base of the neck to the upper limbs at its thoracic outlet. Arterial involvement in thoracic outlet syndrome is infrequent but serious due to the potential risk of thromboembolic complications that threatens the functionality and viability of the extremity. (1) Digital ischemic necrosis is a form of presentation of this condition.

This is the case of a 32-year-old woman, heavy smoker, with a 4-month history of intermittent left upper limb claudication associated with hand paresthesia. The patient progressed with pain at rest and dry ischemic necrosis in the third finger of the left hand (Figure 1A). Evaluation and treatment were performed by a multidisciplinary team (Traumatology, Internal Medicine, Rheumatology, Emergency); the patient was referred to the Vascular Surgery Division. Physical examination revealed coolness and pallor of the left forearm and hand; absence of axillary, humeral, radial and ulnar pulses with contralateral pulses present; a painful non-pulsatile hard-stony mass in the left supraclavicular fossa, which the chest X-ray revealed to be a supernumerary cervical rib. Doppler ultrasound showed subclavian, humeral, radial and left ulnar arteries with a post-obstructive monophasic flow pattern, with very low velocity (20 cm/sec in the subclavian artery) and reduced resistance. CT angiography revealed an anterior costal fusion of the bilateral cervical rib and the first costal arch only on the left side, associated with thrombosis of the subclavian artery (Figure 2A). Angiography evidenced recanalization of a thrombosed aneurysm of the left subclavian artery distal to the vertebral artery through collaterals in the axillary artery, distal occlusion of the humeral artery, but patency in the deep humeral artery and recanalization through collaterals in the ulnar and radial arteries with incomplete palmar arch (Figure 2B). It was decided to perform surgical resection of the cervical and first thoracic ribs, anterior and middle scalenectomy, exclusion of the subclavian aneurysm and arterial reconstruction with subclavian-axillary bypass with a ringed 6 mm-polytetrafluoroethylene (PTFE) prosthesis.

The patient was referred to a smoking cessation program. After one and a half-year follow-up, the patient is asymptomatic, with healing of the digital trophic lesion (Figure 1B) and patent bypass.

Upper limb ischemia is less common than lower limb ischemia. (2, 3) Depending on the time of evolution, ischemia can be acute or chronic, the latter being less common. Vascular trauma, embolism and arterial thrombosis are causes of acute ischemia, whereas subclavian artery atherosclerosis and thoracic outlet syndrome often present with chronic upper limb ischemia. (2, 3)

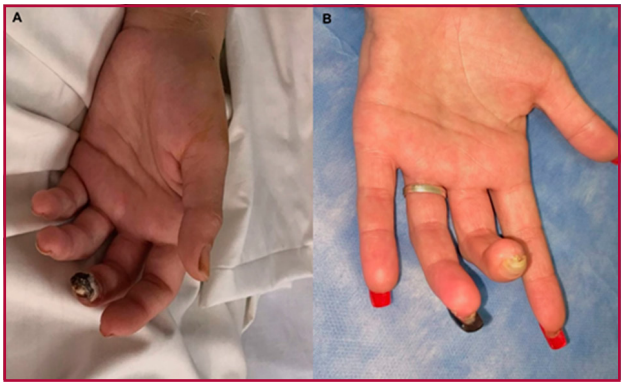

Fig. 1. A. Digital ischemic necrosis of the third finger. **B.** Trophic lesion healing following revascularization

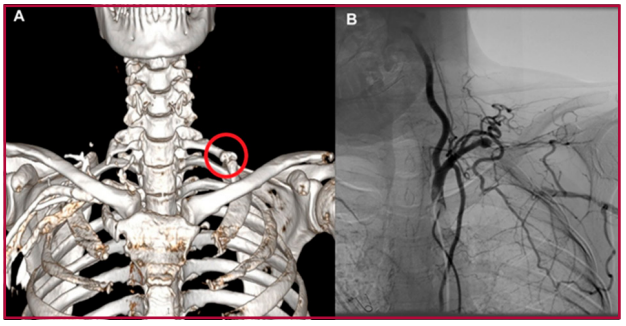

Fig. 2. A. CT Angiography: Anterior costal fusion of the bilateral cervical rib and the first costal arch only on the left side. **B. Angiography:** Recanalization of a thrombosed aneurysm of the left subclavian artery distal to the vertebral artery through collaterals in the axillary artery

Thoracic outlet syndrome is a complex entity characterized by extrinsic compression of the brachial plexus and/or subclavian vessels (vein or artery), by bony abnormalities or by hypertrophy of the scalene and/or subclavian muscles. Depending on which structures are compressed, it can be neurogenic (95%), venous or arterial TOS. Arterial thoracic outlet syndrome is characterized by subclavian artery disease in the setting of anatomic factors that produce compression, most commonly bony abnormalities such as a cervical rib, first hypoplastic rib, clavicle fracture, rib or hypertrophic calluses from healed fractures. (1, 4) It is the least frequent form of thoracic outlet, representing only 1-3% of patients in the largest reported series, and occurring mostly in young women. (1)

Arterial complications at the thoracic outlet represent the end stage of an undiagnosed condition in which the subclavian artery has been chronically compressed and wall changes develop, including intimal injury with or without post-stenotic dilatation or aneurysm formation, and thrombus formation leading

to distal embolization and serious complications, such as upper limb ischemia and posterior circulation ischemic stroke. (1, 4, 5)

Unilateral hand or digital ischemia is a common clinical presentation in patients with arterial outlet syndrome, which may be accompanied by numbness, tingling, temperature and impaired sensitivity. It is important to point out that these findings can be observed even with palpable radial and ulnar pulses. Loss of ulnar, radial or humeral pulses, as well as decreased blood pressure measurement in the affected limb, may also occur. (4)

Digital ischemia attributable to microembolization is the most common presentation of arterial thoracic outlet syndrome; however, this clinical presentation remains a challenge for physicians due to differential diagnoses, as it can also occur in other conditions such as vasculitis, thromboangitis obliterans, Raynaud's syndrome, ergotamine intoxication, and upper limb atherosclerosis. (4, 6)

The presence of bilateral symptoms suggests a systemic etiology, but the arterial thoracic outlet syndrome should not be ruled out, because cervical ribs tend to occur bilaterally in 50% of cases. (6)

Patients with significant subclavian or axillary artery stenosis, in the absence of acute ischemic complication, may present with typical symptoms of upper extremity claudication. Vemuri et al reported 42% of patients with images of chronic thoracic outlet arterial compression, but with associated symptoms of neurogenic compression, or even asymptomatic, concluding that the incidence of arterial thoracic outlet syndrome is underestimated. (1, 5) Al-Jundi et al suggest that smoking might be a predisposing factor for the symptomatic presentation of arterial thoracic outlet syndrome due to increased risk of thrombosis . (6)

An initial assessment with a chest X-ray including cervical spine views can demonstrate aneurysmal changes or elevated flow velocities correlated with a compressive stenosis of the subclavian artery. (6) Analysis of the causes of embolization should include transthoracic echocardiography, as well as computed angiotomography of the aortic arch and upper limbs, the most readily available modality that provides anatomical details regarding the presence of a bony abnormality and a subclavian artery post-stenotic dilatation. (4) However, for treatment planning purposes, angiography remains more sensitive for assessing the degree of subclavian artery involvement, distal vascular beds and level of embolization. (4, 6) In asymptomatic patients, it also confirms extrinsic compression of the subclavian artery in early stages, when there are no lesions of the arterial wall, performing the procedure during hyperextension maneuvers of the upper limb.

Surgical treatment for arterial thoracic outlet syndrome consisted of supraclavicular thoracic outlet decompression with complete anterior and middle scalenectomy with bone resection (resection of cervical rib and/or first rib or other bony abnormality) plus vascular reconstruction depending on the degree of subclavian artery involvement (stenosis/aneurysm) and the presence or absence of distal embolism. Revascularization of the upper limb is performed using bypass surgery with saphenous vein or ringed PTFE graft, associated with distal embolectomy to treat the embolic complication. (1, 4)

In summary, thoracic outlet syndrome is a rare cause of upper limb arterial ischemia that should be suspected in young patients; early surgical treatment (outlet decompression surgery and arterial reconstruction) is indicated to achieve limb salvage and avoid functional sequelae.

Conflicts of interest

None declared.

(See authors' conflict of interests forms on the web/Additional material.)

Ethical considerations

Not applicable.

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Management of a Premature Infant (1460 g) with Ductal-Dependent Tetralogy of Fallot

Patients with a diagnosis of Tetralogy of Fallot (TOF) requiring early intervention are those who are prostaglandin dependent, or with severe cyanosis due to

decreased pulmonary blood flow resulting from unfavorable right ventricular outflow tract (RVOT) or pulmonary arteries anatomy. Management of these patients with associated risk factors (low weight, prematurity, age < 3 months, unfavorable pulmonary artery anatomy, anomalous coronary distribution and critical preoperative conditions) continues to raise debate. Palliative procedures may include RVOT opening, systemic pulmonary anastomosis, pulmonary valve replacement, or patent ductus arteriosus or RVOT stenting. Early repair in these patients can be performed successfully; however, despite its many advantages, it is associated with a great number of postoperative complications and subsequent reinterventions. (1-4)

The purpose of this study is to describe the management of a TOF patient with multiple risk factors, who underwent successful surgery without complications at the Hospital Interzonal Especializado Materno Infantil in the city of Mar del Plata, Buenos Aires, Argentina.

It was an infant born at 32 weeks of gestation (weight 1460 g), with trisomy 21 on karyotype, delivered by emergency cesarean section due to large anterior ventricular septal defect (VSD), infundibular and valvular pulmonary stenosis (valve Z-score -4), confluent good size pulmonary arteries, overriding aorta, small atrial septal defect (ASD) and patent ductus arteriosus, diagnosed by fetal Doppler echocardiography. Due to the unfavorable cardiac anatomy, continuous prostaglandins infusion was indicated to maintain ductal patency.

At 20 days of life, the patient developed Escherichia Coli bacteremia and sepsis, associated with severe respiratory distress syndrome. After 7 days of sepsis treatment, a 7x7 mm vegetation was found in the right atrium; the condition was assumed to be infective endocarditis due to Escherichia Coli. After 4 more weeks of antibiotic therapy, follow-up echocardiography showed no vegetation. After 58 days on mechanical ventilation and neonatal intensive care unit discharge, the patient required oxygen therapy due to bronchopulmonary dysplasia. Patient comorbidities included chronic diarrhea due to protein-losing enteropathy, for which he was fed with hydrolyzed milk via parenteral nutrition (a total of 68 days). Lab tests showed hypoproteinemia, hypogammaglobulinemia and hypoalbuminemia.

Due to the multiple risk factors described above (ductus-dependent TOF, prematurity, low weight, respiratory distress syndrome, and hypoproteinemia), and once infective endocarditis resolved, RVOT stenting appeared to be the safest option to postpone surgical repair and perform it with more favorable clinical features.

Stent implantation

The right femoral vein was punctured, and a 6 French introducer was deployed. A 15 mm-long outflow tract was observed, with a marked dynamic systolic narrowing at 4 mm from the pulmonary valve plane. It was decided to place a 4x16 cobalt chromium coronary stent. The stent was progressed taking the valvular plane and the RVOT. The balloon was inflated and a stable infundibular diameter of 4 mm was achieved. Subsequent angiography showed good flow through the stent and severe pulmonary valve regurgitation due to collapse of the valve by the stent.

After the procedure, the patient was transferred to the neonatal ward; arterial oxygen saturation was between 93-95%; prostaglandin was discontinued, and closure of the arterial ductus was confirmed 48 h post-procedure, with saturation dropping to 90%. The patient remained in the pediatric ward for nutritional recovery.

At the age of 7 months, arterial oxygen saturation was 85%, requiring 0.5 L oxygen through a nasal cannula. Physical examination showed chronic malnutrition (weight 3.4 kg), active precordium, presence of Dressler's syndrome, and systolic murmur in pulmonary focus 3-4/6. Echocardiography revealed a large subaortic VSD, RVOT stent gradient of 55 mmHg, right ventricular hypertrophy, confluent good size pulmonary arteries, overriding aorta, and small ASD. A surgical repair was decided.

Surgical procedure

It was performed by median sternotomy. A patch was reserved for pulmonary artery repair, and the patient was heparinized. Cannulation of the aorta and the superior and inferior vena cava was performed, temperature was lowered to 28°C, the aorta was clamped, cardioplegia was infused antegrade, and a left heart vent cannula was placed. The main pulmonary artery was opened at the level of the annulus, extending to the bifurcation of the pulmonary branches (Figure 1). The stent was attached to the posterior wall, at the level of the RVOT and pulmonary valve; it was completely resected without complications (Figure 2). Infundibular resection was performed due to right ventricular hypertrophy, and the large VSD was closed with a 0.6 mm polytetrafluoroethylene patch. A monocusp autologous pericardial valve was sutured to the pulmonary annulus and transannular patch, from the ventriculotomy to the bifurcation of the pulmonary branches. Air was purged from the left heart and the aorta was declamped after 61min. The patient recovered sinus rhythm. When the patient reached 36.5ºC, he was weaned from cardiopulmonary bypass after 100 min, and was decannulated.

Immediate postoperative course required low doses of inotropes. The patient was extubated 48 hours after surgery, and was on nasal cannula oxygen due to his bronchopulmonary dysplasia. On the 5th postoperative day, he was transferred to a less complex area for nutritional recovery. Echocardiography 1 month after surgery showed closed VSD, no residual VSD, closed ASD, mild pulmonary stenosis, mild to moder-

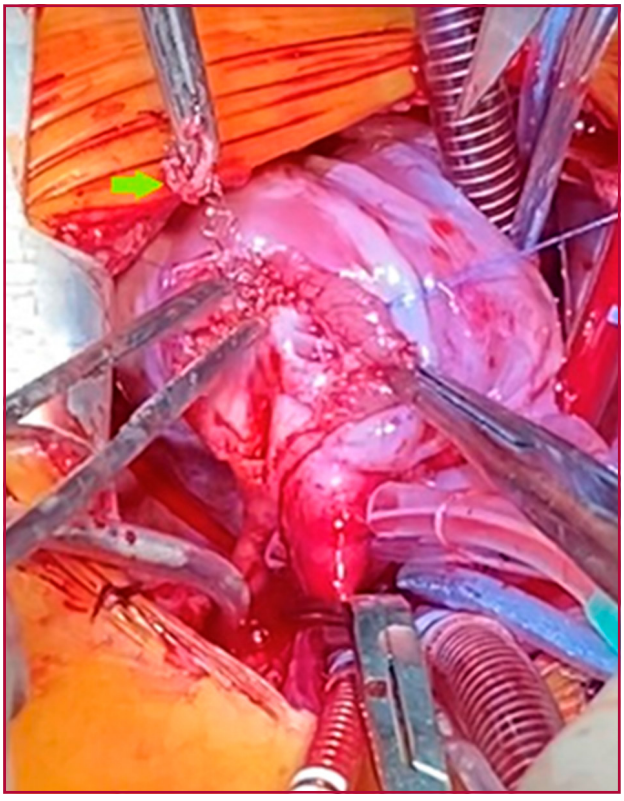

Fig. 1. The stent is removed (green arrow) through ventriculotomy; it is attached to the posterior wall of the right ventricular outflow tract

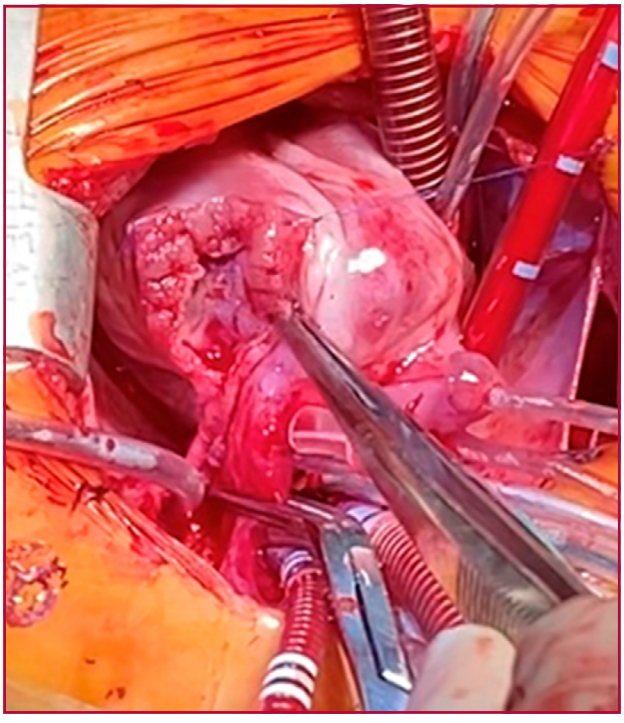

Fig. 2. Complete stent removal, without complications

ate pulmonary insufficiency, and good-caliber confluent pulmonary branches.

TOF is characterized by large subaortic VSD, pulmonary stenosis, right ventricular hypertrophy, and overriding aorta. Surgery in these patients with confluent, and of good caliber, pulmonary branches, usually allows primary repair without complications, with excellent results. Those prostaglandin-dependent patients with severe cyanosis due to severely decreased antegrade pulmonary blood flow require early intervention. In patients with coexisting comorbidities, primary repair can be performed successfully with low mortality rate, but it is associated with more postoperative complications and reinterventions. (1-4) Systemic pulmonary anastomosis is the most common palliative procedure for premature infants with low weight or pulmonary branches hypoplasia; however, it is associated with a higher rate of complications, such as pulmonary artery stenosis. (5) In these patients, RVOT stenting is the bridging option to repair surgery. (6) Surgical repair with RVOT stenting in TOF patients can be performed without complications. Complete intraoperative stent removal can be achieved in up to 95% of the patients, requiring longer cardiopulmonary bypass. (6)

In premature, low-weight patients with coexisting comorbidities, RVOT stenting improves cyanosis, does not alter the anatomy of the pulmonary branches, and allows to solve non-cardiac comorbidities in order to then perform the uncomplicated reconstructive surgery.

Conflicts of interest

None declared.

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Ethical considerations

Not applicable.

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Dasatinib Induced Pulmonary Arterial Hypertension in a Patient from the Peruvian Andes at 3660 m

Pulmonary arterial hypertension (PAH) incidence ranges from 1.5 to 32 patients per million, with poor prognosis, (standardized U.S. mortality rate between 4.5 and 12.3 per 100 00 inhabitants, and a five-year survival rate of 56%). (1) PAH is defined when pulmonary systolic pressure is > 30 mmHg, or mean pulmonary artery pressure (mPAP) is ≥ 25 mmHg; its etiology is varied and includes PAH secondary to drugs (group 1 of the 6th World Symposium on PAH Clinical Classification: drug/toxin induced).

Dasatinib is a second-generation tyrosine kinase inhibitor (TKI), approved by the FDA in 2010 as first-line treatment of chronic myeloid leukemia (CML) and Ph+ acute lymphoblastic leukemia (ALL). Several clinical trials have shown that dasatinib is more effective than imatinib in treating CML as it can produce fast and sustained clinical, hematological and molecular remissions. However, cardiovascular adverse events with dasatinib have been reported, including QTc prolongation, pleural and pericardial effusion, pulmonary parenchymal infiltrates, and PAH.

Considering that there are no literature reports on PAH and dasatinib in subjects inhabiting high altitude settings, our purpose is to describe the case of a patient born in a city at 3660 m high, who was diagnosed with CML and treated with dasatinib, and who developed PAH, in order to determine whether altitude contributed to such development.

This was a 42-year-old male patient from Huancavelica, Peru, diagnosed with CML by flow cytometry in 2009. Imatinib 400 mg/day was prescribed in March 2011. Since the patient was unresponsive to imatinib, in September 2014 he was switched to dasatinib 100 mg/day for 21 months, followed by 140 mg every other day for 25 months. In June 2019, the patient reported dyspnea, fatigue, headache, and weight loss (5 kg), and was admitted to the emergency room for acute respiratory distress and fever. Chest computed tomography (CT) revealed mild pericardial effusion and bilateral pleural effusion requiring drainage, with clinical improvement. ECG showed right bundle branch conduction disturbances (Figure 1). Echocardiography revealed severe right ventricular enlargement, normal contractility, and 3D ejection fraction 42%; marked right atrium enlargement and moderate tricuspid regurgitation. Left ventricular function was normal, with ventricular septal displacement associated with right ventricular overload. Severe pulmonary hypertension, systolic PAP 100 mmHg, and mild pericardial effusion were also detected (Figure 2). Pulmonary CT angiography showed bilateral pleural effusion without pulmonary thromboembolism.

A medical peer review decided to discontinue dasatinib and switch to nilotinib. Medical check-ups were performed every 3 months; no progression of PAH was observed. Symptoms completely resolved within 9 months after dasatinib discontinuation, ECG was normal; systolic PAP on echocardiography reverted to baseline and right chambers returned to normal.

Our patient had baseline systolic PAP of 40 mmHg, typical of an Andean native resident. Peru is a country traversed by the Andes mountains; 8,726,000 people (28.1% of the Peruvian population) live in this region, and are adapted to altitudes above 3500 m. At this altitude, low barometric pressure results in low partial pressure of inspired oxygen compared to coastal residents; hypoxia causes physiological changes in the pulmonary circulation —first described by the Peruvian researcher Dante Peñaloza 62 years ago—, (2) with increased smooth muscle cells (SMC) in the small pulmonary arteries and muscularization of the arterioles, increased pulmonary vascular resistance, and development of PAH. Mean PAP describes a parabolic curve: at 2000 m the value is 15 mmHg and at 4500 m it is 30 mmHg, and can reach up to 40 mmHg, as was the case in our patient. (3) The prevalence of altitude-induced PAH is 5-18% in South America.

A second mechanism of altitude-induced PAH would be vasoconstriction of the pulmonary precapillary vessels, accounting for 80% of increased pulmonary vascular resistance, and of the small pulmonary veins (< 900μm), which determine the remaining 20%. This vasoconstrictor response to hypobaric hypoxia has an initial stage within seconds of exposure to alveolar oxygen deprivation that reaches its maximum value at 15 minutes, followed by a second stage with a more gradual increase in pulmonary arterial pressure that reaches a plateau at 2 hours and is maintained for 8 hours. The first stage would be a consequence of reactive oxygen species (ROS) generated by mitochondria of SMCs in the middle layer of the arterioles; this excess of ROS induces alterations in the potassium channels, intracellular calcium increase and subsequent vasoconstriction. The second stage would be regulated by alterations in the endothelial function: enhanced endothelin-1 production and thromboxane A2, with reduced prostacyclin and nitric oxide synthesis. (3)

Imatinib, a first-generation TKI, is the treatment for CML. However, second-generation TKIs such as

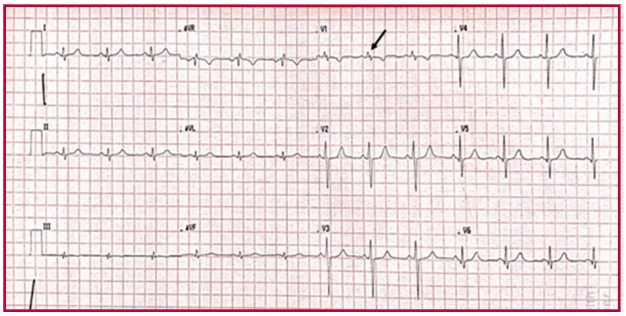

Fig. 1. ECG. Findings on admission: arrow showing right bundle branch conduction disturbances, with QRS complex with rSr' morphology in V1.

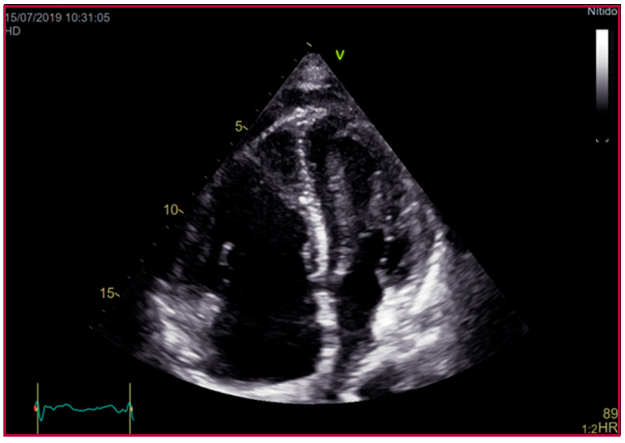

Fig. 2. Echocardiography. Transthoracic echocardiography showing enlargement of the right ventricle and right atrium, ventricular septal displacement to the left, compression of the left ventricle, and mild pericardial effusion.

dasatinib have been approved as first-line treatment for CML since November 2007 in USA, because they produce improved molecular and clinical response due to their potency on non-mutated BCR-ABL1, 325-fold greater than imatinib, with early disease control. (4)

Both imatinib and dasatinib are TKIs that act on BCR-ABL kinase; however, their effects are opposite. Imatinib improves PAH, while dasatinib causes PAH (incidence 0.2-5%). (5) This is a late complication, occurring at around 34 months (range 8-48 months). In our patient, this complication occurred at 46 months of treatment; the fact that he was an Andean resident did not determine earlier presentation or more severe symptoms. The mechanism responsible for PAH in dasatinib-treated patients is still unclear. Dasatinib differs from other TKIs in that it strongly inhibits the Src family kinases, which degrade activated platelet-derived growth factor receptors (PDGFR) and other growth factors, encouraging the proliferation of SMCs in pulmonary artery branches. It has also been demonstrated that increased oxidative stress is a determinant of pulmonary endothelial dysfunction and vascular injury. (5)

Our patient developed pleural effusion, one of the most common adverse reactions caused by dasatinib (incidence 14-60%); 90% of cases occur within one year; the reaction disappears after dasatinib withdrawal. The mechanism may be autoimmune or by inhibition of PDGFR-beta, which would cause fluid retention, and alteration of pulmonary endothelial permeability. Several studies report that PAH is the major comorbidity in patients with dasatinib-induced pleural effusion. Patients with pleural effusion may also develop pericardial effusion (29%). (6)

PAH symptoms are nonspecific, including shortness of breath, dyspnea, chest pain, cough, and hemoptysis. Physical examination is useful but nonspecific. ECG is poorly sensitive in the detection of right ventricular enlargement (20%). Echocardiography features high sensitivity and specificity. Hemodynamic manifestations improve when the drug is discontinued; however, some patients will require PAH treatment. Our patient showed clinical improvement after drug discontinuation; ECG and echocardiography were normal without treatment.

In conclusion, living at high altitude was not associated with changes in the progression of dasatinib-induced PAH.

Conflicts of interest

None declared.

(See authors conflicts of interest forms in the website/ Supplementary material).

Ethical considerations

Not applicable.

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Surgical results in the rehabilitation of Continuous hypoplastic pulmonary branches in patients younger than 6 months. Experience of a center

Severe hypoplasia of the pulmonary branches is life-threatening, unless pulmonary flow through the ductus arteriosus or aortopulmonary collaterals is maintained.

It is vital to reestablish pulmonary flow in the first year of life to prevent final lung development from being permanently affected. To this end, a number of surgical techniques that promote the development of pulmonary arteries have been proposed.

This paper describes our experience in rehabilitating continuous hypoplastic pulmonary branches in infants < 6 months of age.

In a retrospective cohort study, we reviewed electronic medical records of patients < 6 months of age with hypoplastic pulmonary branches undergoing surgery to ensure adequate pulmonary flow between January 2015 and December 2020 at the Children's Hospital Dr. Roberto Gilbert Elizalde.

Patients with previous procedures or discontinuous pulmonary arteries were excluded. Pulmonary branches with an echocardiographic Z-score < -2 were defined as hypoplastic. Parameter Z (Children's Hospital of Michigan) was used to calculate the Z-score.

Operative mortality was defined as deaths occurring within 30 days after surgery —or after 30 days if the patient remained in hospital—, and late mortality as those occurring beyond 30 days post-surgery if the patient was discharged.

All therapeutic catheterization or surgical procedures performed after surgery were considered reinterventions.

Ten patients met the inclusion criteria; 70% were male infants. Mean age was 1.87 ± 1.7 months; mean weight 4.04 ± 1.28 kg; mean height, 52.95 ± 5.48 cm; and body surface area, 0.23 ± 0.04 m2. Pulmonary atresia was the most common condition (30% of cases); mean Z-score of the right and left pulmonary branches prior to the procedure was -3.99 ± 1.98 and -2.36 ± 1.67 respectively.

The modified Blalock-Taussig shunt (subclavian pulmonary anastomosis using a 3-4 mm-polytetrafluoroethylene [PTFE] tube) was the preferred procedure (7 cases); 2 central shunts were used in 2 cases, and a 12 mm-valved biological conduit from the right ventricle to the pulmonary artery, with pulmonary branch plasty, was required in the remaining patient.

Mean Z-score of the right and left pulmonary branches after the procedure was -0.6 ± 2.37 and 0.09 ± 1.55 respectively (difference from preoperative values: p = 0.006 and p = 0.007 respectively).

Five patients required reintervention: a surgical reintervention with pulmonary branch plasty and a new central shunt, and four therapeutic catheterizations.

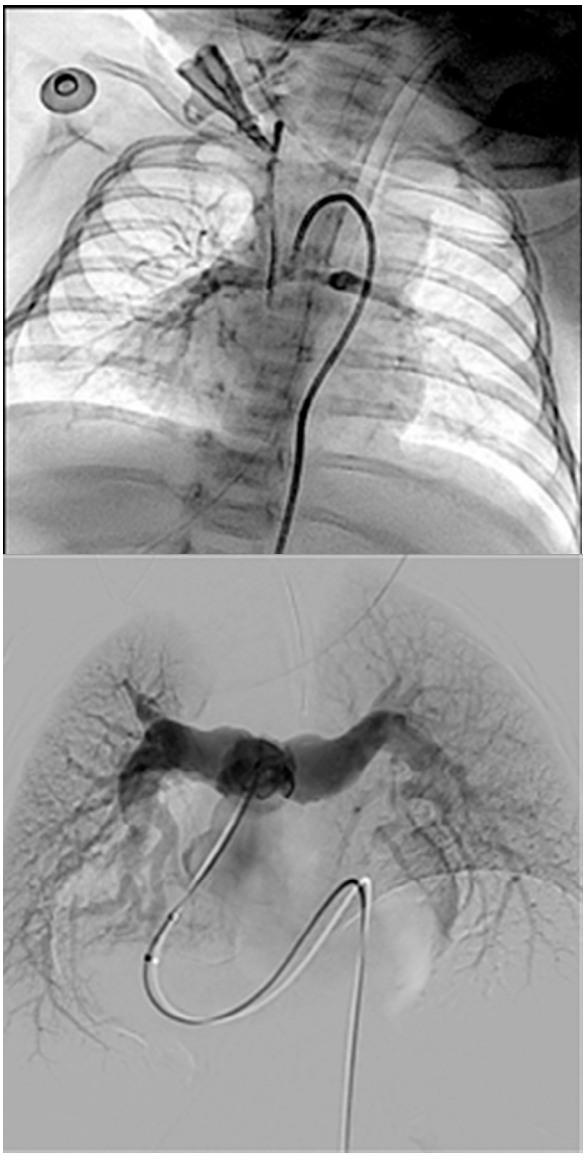
Table 1. Surgical procedures and outcomes.

n	Diagnosis	Z-score Preoperative		Surgery	Catheterization	Postoperative Z-score		Survival
		RPB	LPB			RPB	LPB	
1	Ia TA	-2.66	0.47	BT (3mm)	Yes	.	.	-
2	IIa TA VSD	-2.99	-2.47	BT (3.5 mm)	No	-1.77	0.29	+
3	PA VSD	-2.8	-3.8	RV tube PA + branch plasty	Yes	0.41	1.86	+
4	TOF	-2.73	-2.25	BT (3.5 mm)	No	-2.45	.	+
5	DORV TOF	-2.3	0.37	Central shunt	Yes	0.7	1.3	+
6	UAVC PS	-3.68	-3.4	BT (4 mm)	No	4.14	0.28	+
7	AVC TOF	-4.6	-1.78	BT (3 mm)	No	-0.92	0.32	+
8	PA IS	-7.8	-4.38	BT 4 mm + branch plasty	No	.	.	+
9	PA VSD	-7.3	-3.8	Central shunt 4 mm	Yes	-3.52	2.93	+
10	TOF	-3.12	-2.58	BT (3mm)	No	-1.39	0.73	+

TA: tricuspid atresia; Ia TA: tricuspid atresia with pulmonary atresia; IIa TA VSD: tricuspid atresia with transposition of the great arteries plus pulmonary atresia and ventricular septal defect; PA: pulmonary atresia; IS: intact septum; BT: Blalock Taussig; AVC: atrioventricular canal; UAVC: unbalanced atrioventricular canal; VSD: ventricular septal defect; PS: pulmonary stenosis; TOF: tetralogy of Fallot; RV: right ventricle. RPB: right pulmonary branch; LPB: left pulmonary branch; DORV

During mean follow-up of 7.66 ± 5.63 months, there was one operative death 21 days after surgery (Blalock-Taussig shunt with a 4-mm PTFE tube), and one late death 3 months after the procedure (Blalock-Taussig shunt with a 3-mm PTFE tube) in a patient who was readmitted with severe dehydration due to diarrhea. Both of them were cases of pulmonary atresia with intact septum.

Pulmonary artery rehabilitation was achieved in 60% of the patients, but was unsuccessful in two patients, whose pulmonary branches were disconnected.



Diagnostic catheterization: Angiography of pulmonary branches in anteroposterior view.

Fig. 1. A. 4-month-old patient with dextrocardia, double-outlet right ventricle (RV), severe pulmonary stenosis. Hypoplasia of the pulmonary branches. **B.** Post-surgery with right ventricle to pulmonary artery conduit, pulmonary branch plasty, and subsequent pulmonary branch angioplasty

Table 1 includes a summary of the surgical procedures and outcomes.

The modified Blalock-Taussig shunt was the preferred technique in our series. This procedure is the most widely used in the world, since it ensures controlled pulmonary flow with limited coronary diastolic steal, provided the tube size is chosen correctly; it is also easy to be controlled in subsequent surgeries. (1)

The central shunt (aortopulmonary septal defect through a PTFE tube or with a direct window) would offer some advantages: less chances of obstruction, more symmetrical flow, less distortion, and better postoperative management. Both methods may result in pulmonary congestion —due to improper shunt size— and in coronary steal, particularly with the central shunt. (1)

Some studies compared systemic to pulmonary shunts with the right ventricle to pulmonary artery connection, finding no differences in the development of the pulmonary tree. (2, 3)

Depending on the different series, overall mortality with Blalock-Taussig shunt varies between 2.3% and 16%, and between 9% and 11% in neonates. (4) The only operative death in our series was a patient with Blalock-Taussig shunt, who died 21 days after surgery.

Two central shunts were performed. This procedure is an effective option for diminutive pulmonary arteries, offering a pulsatile, symmetrical flow that encourages pulmonary arteries growth. (1, 5) However, one of these patients required therapeutic catheterization, reoperation with a new central shunt and pulmonary branch plasty, but the branches were disconnected during follow-up.

The right ventricle to pulmonary artery conduit is very popular in patients with pulmonary atresia, ventricular septal defect and hypoplastic pulmonary branches with aortopulmonary collaterals, and despite the advantage of enhancing pulmonary artery development has not been demonstrated in all studies, it does ensure greater stability, improved saturation, and decreased time between palliative surgery and the final procedure, with less obstruction, no coronary diastolic steal and less distortion of the pulmonary branches. (3, 6-8) This technique has the disadvantage of requiring cardiopulmonary bypass during surgery; in rare cases, aneurysm due to right ventricular outflow tract patch may occur. (3) It was used in a case of pulmonary atresia and ventricular septal defect, with positive outcomes.

Pulmonary artery rehabilitation was achieved in 60% of our patients. Various studies report ranges of success between 37% and 78% for pulmonary branch repair. (2, 8)

We had a significant number of reinterventions (50%): one surgical reintervention and four therapeutic catheterizations, which highlights the importance of catheterization (Figure 1).

All the procedures performed had been shown to

be useful in encouraging pulmonary arteries growth in all the publications reviewed.

Conflicts of interest

None declared.

(See authors' conflicts of interest forms on the website/ Supplementary material).

Ethical considerations

Not applicable

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Aneurysm of the Brachiocephalic Trunk. A Rare Entity

Injuries of the innominate artery, or brachiocephalic trunk (BCT), are exceedingly rare, and are associated with high blood pressure, smoking, syphilis, and

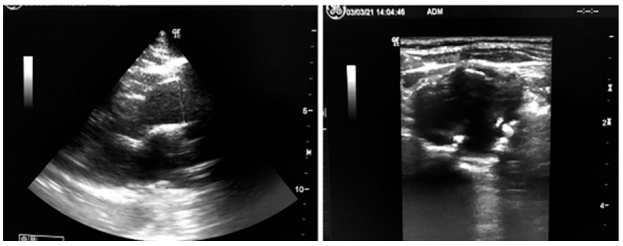

Fig. 1. Echocardiography. **A (left)**. Left parasternal long axis view showing mild elongation of the supravalvular aorta. **B (right)**. Fibro-lipid plaques at the aneurysmal level.

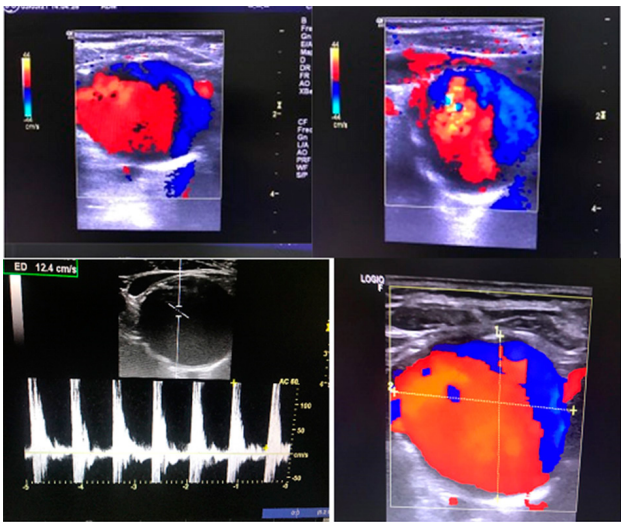

Fig. 2. Doppler ultrasound of the neck vessels. **A** (upper left). Two-dimensional ultrasound confirming BCT aneurysm. **B** (upper right). Color Doppler: bidirectional blue and red flow in Yin Yang. **C** (inferior left). Doppler flow with abnormal, monophasic waveform at the aneurysmal level. **D** (inferior right). Two-dimensional ultrasound showing fibro-lipid plaques and aneurysm size

probably other cardiovascular risk factors. Semiologically, they are detected as asymptomatic pulsatile masses. Clinically, BCT aneurysms are evidenced by their mass effect, compressing neighboring structures (dyspnea, dysphonia, stridor, dysphagia, superior vena cava syndrome). BCT aneurysms may present as arteriovenous fistulas, pseudoaneurysms, dissections or true aneurysms, and the rupture, with or without fistula formation, to the airway or gastrointestinal tract —particularly the esophagus— is the most dreaded complication. They can potentially result in systemic, central nervous system or right upper limb embolisms. Diagnosis is based on clinical suspicion and diagnostic imaging, mainly computed angiotomography, and thoracic and supra-aortic trunk angiography.

We describe the case of a 50-year-old female patient, 1.50 m tall and 64 kg, with a history of controlled hypertension under treatment. The patient started coughing, and a chest X-ray and computed tomography (CT) scan were requested as per protocol

to rule out COVID-19. Chest X-ray profile revealed a superior mediastinal mass compressing and displacing the trachea. CT scan reported BCT aneurysm; the patient was referred to our vascular ultrasound service to decide the course of action to follow.

Physical examination showed a right-suprasternal pulsatile mass and a systolic-diastolic murmur. Vascular ultrasound revealed a 35 x 28 mm BCT aneurysm and hyperechoic fibro-lipid plaques, with no significant obstructions. No aneurysmal dilatations were found in the remaining neck vessels. Echocardiography showed mild supravalvular elongation (33 mm) but no aortic dilatation (Figure 1). Color Doppler showed the Yin-Yang effect filling the aneurysm (Figure 2). No aneurysmal dilations were found in the upper and lower limb arteries. In view of this data, routine laboratory tests, thyroid hormones, glycemia, Hb A1C, and VDRL were requested. Antiplatelet drugs and rosuvastatin 20 mg/day were prescribed. The patient was referred to vascular surgery for either surgical or endovascular approach, but was admitted to hospital due to community-acquired pneumopathy, with good response to antibiotic treatment. After discharge, her case was discussed in a grand round, and it was decided to perform aneurysmal surgery. The patient is currently on the surgery waiting list.

BCT aneurysms are rare entities that may cause life-threatening complications, including rupture, fistulas or thromboembolism that may cause strokes; hence the importance of diagnosis and adequate treatment to prevent these complications. (1) Degenerative aneurysms are the most common type and are associated with thoracic and abdominal aortic aneurysms, with the BTC involvement in cases of type-A aneurysmal dissection. Jiménez et al reported a case of a 52 x 55 x 48 mm BCT aneurysm with intramural thrombus and mild tracheal compression, and visualization of about 6 x 6 cm right-suprasternal, soft, pulsating, non-tender mass in the carotid triangle, with thrill and murmur radiating to the neck. (1) It was associated with a second aneurysm > 5 cm diameter in the descending aorta. Both aneurysms were successfully treated by conventional surgery in two stages.

In a 40-year retrospective study, Bauer et al detected only six true BCT aneurysms from 73 surgeries of supra-aortic trunks and neck vessels. (2)

Pseudoaneurysms are more common due to motorbike or car accidents, and open trauma of the great intrathoracic vessels. Rupture is mainly associated with the presence of posttraumatic pseudoaneurysms. (3)

Autoimmune diseases, such as Takayasu arteritis, Kawasaki disease, giant cell arteritis, Marfan syndrome, or Behcet's disease, can also involve the innominate artery. (4) Surgical or endovascular treatment may be followed, depending on the case. (5) Kieffer et al (6) consider that the approach depends on the extent of the aneurysm. BCT aneurysms are therefore classified into three types: A: aneurysms not involving the origin of the BCT; although they are the most uncommon, they would be the easiest to treat using a termino-terminal bypass or an endovascular prosthesis. B: aneurysms involving the origin of the BCT; they are the most common ones. An aorto-BCT or aorto-carotid subclavian bypass originating from the ascending aorta is performed, and the origin of the BCT is sutured with a patch. C: aneurysms involving the entire BCT and the ascending aorta, requiring ascending aorta replacement, with or without aortic valve replacement.

While case reports are rare, BCT aneurysm is currently uncommon; we must be trained to determine its etiology and surgical timing. Surgical treatment is suggested for all symptomatic or ruptured aneurysms and for those > 3 cm. Surgery in these patients should be performed immediately, due to the risk of rupture and of secondary compression, which can be very serious.

Conflicts of interest

None declared.

(See authors' conflicts of interest forms on the website/ Supplementary material).

Ethical considerations

Not applicable

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