Unilateral Absence of the Left Pulmonary Artery in a Patient with Acute Pulmonary Embolism

We report the case of a 49-year-old male patient, former smoker and living a sedentary lifestyle, with a history of prolonged travel in the last week and treatment with beta-blockers for an unspecified arrhythmia.

He was admitted to the coronary care unit with a diagnosis of non-ST segment elevation acute coronary syndrome, typical angina and FC III-IV dyspnea (NYHA).

On physical examination, he weighted 76 kg and was 1.72 m tall; blood pressure was 120/70 mm Hg and heart rate was 70 bpm. He had no fever, the respiratory rate was 17 bpm and the arterial oxygen saturation was 94%.

The electrocardiogram showed sinus rhythm, heart rate of 65 bpm, vertical electric axis and abnormal ventricular repolarization in the anteroseptal wall, corresponding to the right ventricle (Figure 1A). The anteroposterior chest X-ray showed absent hiliar shadow and mediastinal shift to the left side with left hemidiaphragm elevation, and hyperinflation of the contralateral lung (Figure 1 B).

The remarkable results of the laboratory were high-sensitivity troponin of 213/176/90 pg/mL (normal value: ≤ 34) D-dimer of 3427 ng/mL (normal value: ≤ 198) and BNP of 73 pg/mL (normal ≤100).

On transthoracic echocardiography, left ventricular function was preserved and wall motion was normal. The right cardiac chambers and the main pulmonary artery diameters were in the upper normal limit.

The patient underwent coronary angiography. There were no signs of obstructive coronary artery disease; pulmonary sequestration of the right coronary artery (RCA). A right aortic arch was present. (Figure 2A)

The computed pulmonary angio tomography showed absence of the main left pulmonary artery with ipsilateral shift of the mediastinum and great vessels, and right aortic arch (Figure 1B). There was

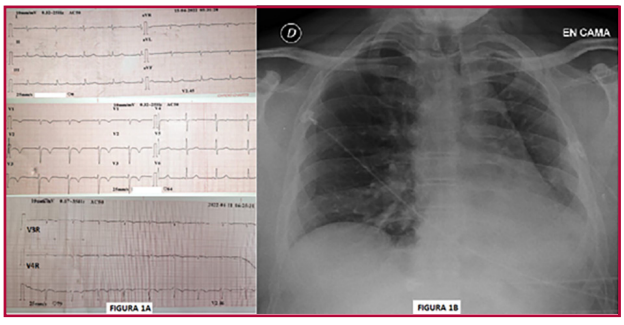

Fig. 1. A. Electrocardiogram with right-sided leads. **B.** Chest X-ray

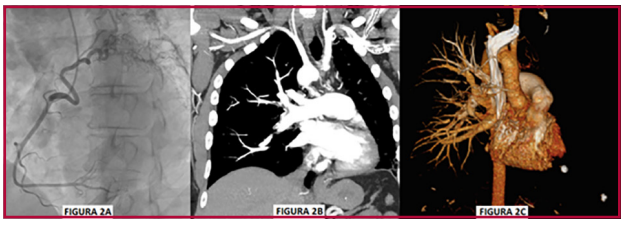

Fig. 1. A. Coronary angiography. **B.** Computed tomography pulmonary angiogram. C. 3D reconstruction

a filling defect in the lobar segmental and subsegmental right pulmonary artery branches consistent with acute pulmonary embolism (PE). The left lung was hypoplastic and the right main pulmonary artery and right cardiac chambers were enlarged. (Figure 2B-2C).

The lower limb ultrasound revealed deep venous thrombosis in the popliteal vein and left tibioperoneal trunk.

The final diagnosis was venous thromboembolism and high-risk pulmonary embolism with right ventricular infarction in the context of absence of the left main pulmonary artery. The patient received medical treatment with anticoagulation and had favorable outcome.

Unilateral absence of a pulmonary artery (UAPA) is a rare congenital abnormality, with a prevalence of 1 in 200,000. It occurs equally in both sexes. Most of the cases have associated cardiovascular anomalies, as coarctation of the aorta (isolated or associated with interventricular septal defects), subvavular aortic stenosis, transposition of the great vessels, patent ductus arteriosus, tetralogy of Fallot and right aortic arch. (1)

The pulmonary arteries develop in week 16 of gestation from the sixth proximal aortic arch. Absence of pulmonary artery occurs due to failure of the sixth aortic arch to connect with the main pulmonary trunk, hypothetically secondary to low blood flow. (2) Absence of the left or right pulmonary arteries may present with ipsilateral pulmonar aplasia or hypoplasia. The hypoplastic lung is perfused by remnants of the aortic arches persisting as embryonic arteries, collaterals of the bronchial arteries, subclavian arteries, intercostal arteries, diaphragmatic arteries and even by coronary arteries, which could cause steal phenomenon with coronary hypoperfusion and ischemia. (3)

Although few patients with UAPA remain asymptomatic until adulthood (15%), the diagnosis is usually made early due to clinical manifestations such as recurrent pulmonary infections, decreased exercise tolerance and exercise-induced dyspnea (40%). Twenty percent of cases present with hemoptysis due to extensive collateral circulation and 25% develop pulmonary hypertension secondary to increased blood flow to the contralateral lung, with implications for long-term survival. (1, 3, 4).

Other form of presentation is dyspnea in the context of pregnancy or at high altitude, as a manifestation of silent pulmonary hypertension. (5)

The diagnostic workup includes chest X-ray with the findings previously described. Doppler-echocardiography may confirm the diagnosis and suggest the presence of pulmonary arterial hypertension. Computed tomography pulmonary angiogram allows visualization of the entire pulmonary vascular tree, including the distal beds, detection of hilar pulmonary arteries by pulmonary vein wedge angiography and the presence of bronchiectasis. (5- 6)

The association between UAPA and pulmonary embolism has rarely been reported in the literature. The differential diagnosis with chronic PE should be made when amputation of the main branches occurs due to thrombotic obstruction.

A small group of patients with UAPA remain asymptomatic during their lifetime. This condition should be suspected in the presence of recurrent respiratory infections, hemoptysis or pulmonary arterial hypertension. In the presence of angiographic evidence of extensive or enlarged collateral arteries, as a potential cause of pulmonary hypertension or bleeding, embolization may be beneficial. The early diagnosis of this disease and treatment of its complications can reduce the associated morbidity and mortality. The differential diagnosis with chronic pulmonary thromboembolism is important due to the therapeutic implications.

Conflicts of interest

None declared.

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

Ethical considerations

Not applicable.

Paula Boggio[1], Franco Mátar[1], Valentina Rodriguez[1], Andrea Astesiano[1], Nicolás Renna[1], Alfredo Astesiano[1]

[1] Hospital Español de Mendoza. Coronary Care Unit.

Paula Boggio - Hospital Español de Mendoza. Unidad Coronaria - Av. San Martín 965 - 5501 - Mendoza - E-mail: boggiopaula@gmail.com

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Papillary Fibroelastoma: A Benign Tumor with Potentially Devastating Consequences

We report the case of a 69-year-old woman with hypertension, hypothyroidism and rheumatoid arthritis, under treatment with methotrexate.

The patient was transferred from the Emergency Department to the Interventional Cardiology Section for acute myocardial infarction with anterolateral ST-segment elevation (Killip-Kimball IV).

The patient persisted with an oppressive central chest pain radiating to both arms. She was hypotensive (Blood Pressure; 90/60 mmHg), with poor peripheral perfusion despite continuous dobutamine infusion at 5 mcg/kg/min. In addition, arterial oxygen saturation was around 90% despite the use of a reservoir mask at 15 L/min, and pulmonary auscultation revealed crepitations up to mid-zones.

Catheterization revealed good-sized left main coronary artery with a convex thrombus occluding the distal portion (Figure 1). After bypassing the occlusion

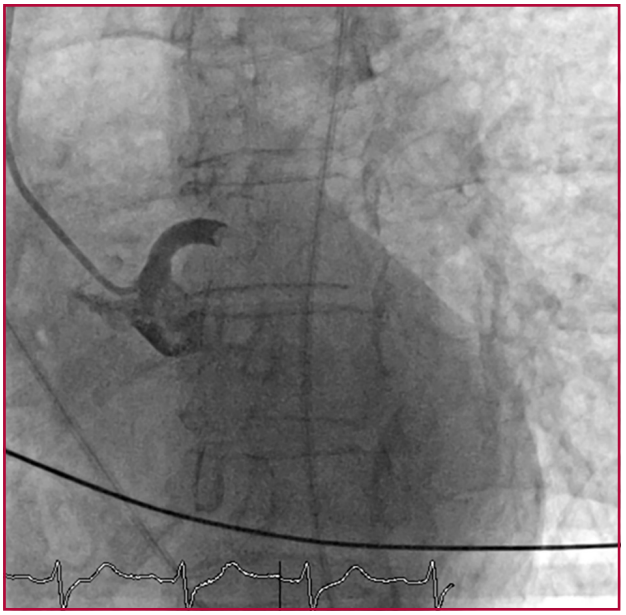

Fig. 1. Cardiac catheterization, left anterior oblique projection (28°). Image showing a good-sized left main coronary artery with a convex thrombus occluding the distal portion; no visualization of the anterior descending artery or the circumflex artery.

with a guidewire and locating it in the left anterior descending artery (LAD), the vessel did not recover flow and manual thromboaspiration was performed, resulting in LAD flow recovery and macroscopic material collection.

Due to thrombus migration to the first obtuse marginal artery, a bolus was started, followed by continuous tirofiban infusion, together with aspirin and prasugrel loading. However, final TIMI 1 flow and abundant thrombotic burden remained.

Since thrombotic etiology was initially suspected, intracoronary imaging with optical coherence tomography (OCT) was performed, showing no endothelial lesion at any level.

The patient was transferred to the Coronary Care Unit, hemodynamically unstable but with improved peripheral perfusion, no chest pain and good respiratory progression.

Laboratory findings included glomerular filtration rate of 57 ml/min/1.73 m^2, normal electrolyte panel, blood count and coagulation tests, and ultrasensitive troponin T that initially was 23 ng/L, reaching a peak of 28041 ng/L. Further evaluation with a transthoracic echocardiography (TTE) revealed severely depressed left ventricular function due to apical akinesia extending to the mid-septal segments and almost all the anterior and lateral walls.

Despite the absence of significant valve diseases, a 9 x 6 mm sessile nodular image was targeted on the ventricular side of the non-coronary cusp of the aortic valve (Figure 2).

Agitated saline was also administered, confirming the absence of right-to left bubble passage.

Given the poor hemodynamic response —requiring higher doses of vasoactive drugs and balloon counterpulsation—, the transplant referral center in our area was contacted to evaluate the case.

Coronary embolism is considered a rare phenomenon, likely under-diagnosed, with an incidence to be around 0.06%. (1) The most frequently documented etiology is infective endocarditis (IE), followed by atrial fibrillation and prosthetic heart valve thrombosis.

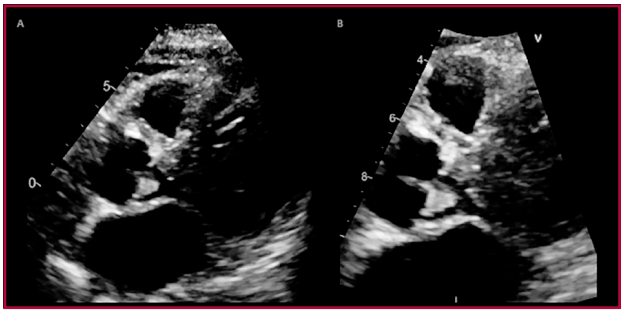

Fig. 2. Transthoracic echocardiography. **A.** Long axis parasternal view showing a 9 x 6 mm sessile nodular image on the ventricular side of the non-coronary cusp of the aortic valve. **B.** Zoom-in on the nodular image

Other etiologies include iatrogenic embolism, paradoxical embolism, atrial myxoma, and papillary fibroelastoma. (1) IE seemed unlikely in our patient, who was afebrile, with no stigmata of endocarditis on physical examination, and infectious parameters were not significantly elevated on admission. The patient had no history of trepidation, did not have a prosthetic valve, or had not undergone a recent medical intervention that could account for iatrogenic embolism.

TTE showed no right-to-left bubble passage, and given the mass characteristics and location, papillary fibroelastoma (PF) seemed to be the most likely etiology of coronary embolism.

Primary cardiac tumors are very rare, with a documented prevalence at autopsy of 0.01%. (2)

Myxoma is the most frequent primary cardiac tumor, but PF is the most common valvular tumor of the heart, accounting for 15% of all primary cardiac tumors. (2) In most series, it occurs predominantly in adult men (mean age of detection 60 years). While tumors may occur on any cardiac structure, around 77% are located on the valvular surface, the aortic valve being the most commonly involved valve. (3) Most of PFs are discovered incidentally, but may present with a broad range of symptoms, depending on their location, size, rate of tumor growth and possible embolization of the tumor. Cerebral embolism is the most common presentation; however, myocardial infarction and sudden death often occur when PF is located in the aortic valve. (3) These major events are mainly attributed to two mechanisms: occlusion of the coronary ostium during valve movement or embolization of the tumor fragments to the coronary arteries. (4)

TTE is a useful tool in a first evaluation, but it may fail to detect up to one third of the PFs, which are evident in the transesophageal echocardiography (TEE); (5) therefore, TEE should be performed in cases of high diagnostic suspicion, as it can also locate the anchor site to design the surgical treatment. (2) The typical echocardiographic image appears as a pedunculated, mobile, homogeneous, small mass (typically < 20 mm), with a characteristic stippling along its edges. (2)

The decision on management of PF is difficult as there have been no randomized controlled trials. (2) Literature published to date suggests that surgical excision in symptomatic patients is curative with an excellent long-term prognosis and no documented postoperative recurrences. (3, 5) Surgery is also recommended if the tumor is mobile, since tumor mobility is considered a predictor of mortality and risk of embolism. (2) However, in asymptomatic patients with nonmobile tumors, close follow-up is recommended. (3) Although papillary fibroelastoma is a benign tumor, given its potential to cause both cardiac and noncardiac fatal complications and the curative treatment available, PF is a differential diagnosis that should be considered in myocardial infarctions with normal coronary arteries.

Conflicts of interest

None declared.

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

Ethical considerations

Not applicable.

Uxue Idiazabal Rodríguez[1], Iñigo Pereiro Lili[1], Ane Elorriaga Madariaga[1], Rafael Martínez de Bourio Uriarte[1], Paula María Mendoza Cuartero[1], Jesús Roberto Sáez Moreno[1]

[1] Hospital Universitario Basurto. Department of Cardiology. Coronary Care Unit. Osakidetza

Uxue Idiazabal Rodríguez – Hospital Universitario Basurto. Department of Cardiology. Coronary Care Unit. Osakidetza - Av. Montevideo 18 – 48013, Bilbao, España. E-mail: Uxue_278@hotmail.com

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Coronary Subclavian Steal Syndrome: A Not-So-Rare Cause of Acute Coronary Syndrome

We report the case of a 65-year-old male patient, ex-smoker, with a history of high blood pressure and dyslipidemia under medical treatment.

In June 2019, he was admitted to our center with non-ST-segment elevation acute coronary syndrome (Killip-Kimball I). Catheterization showed severe coronary artery disease involving the left main coronary artery and 3 vessels; triple coronary artery bypass (left mammary artery to middle third of anterior descending artery, right Y-graft using the mammary artery to first obtuse marginal branch, and saphenous vein to posterior descending artery) was performed. Mild left ventricular dysfunction due to mid-basal and inferoposterior akinesia persisted at discharge.

Two years later, the patient was admitted to our Coronary Care Unit with a new episode of angina at rest, changes in ECG and raised myocardial injury markers (ultrasensitive troponin T 1148ng/mL). Echocardiogram showed increased ventricular dysfunction, left ventricular ejection fraction (LVEF) 30-35%, at the expense of new-onset akinesia of the anterolateral wall. The condition was interpreted as high-risk non-ST-elevation myocardial infarction. Given the hemodynamic and clinical instability, urgent coronary catheterization was performed at 24 h (Figure 1); all the grafts were patent, without new lesions in the distal beds; however, there was occlusion of the origin of the left subclavian artery.

The computed axial tomography (CAT) (Figure 2) and Doppler ultrasound confirmed complete occlusion of the origin of the left subclavian artery, with reversal of blood flow through the left vertebral artery, suggestive of subclavian-vertebral steal syndrome, in addition to the subclavian-coronary steal causing myocardial ischemia in our patient.

The clinical benefits of using the left internal mammary to bypass the left anterior descending artery are well established, but coronary subclavian steal syndrome is an underestimated complication in these patients. (1)

The prevalence of subclavian artery stenosis is 2% in the general population, and 7% in patients with peripheral arterial disease, (2) figures that increase to 2.5 - 4.5%, (3) and 12% respectively (4) in patients with previous aortocoronary surgery.

The strongest predictors of subclavian artery stenosis include peripheral artery disease, smoking, higher levels of systolic blood pressure, or low levels of HDL cholesterol. Atherosclerosis is responsible for more than 90% of subclavian artery stenosis; less common etiologies include arteritis, inflammation, ra-

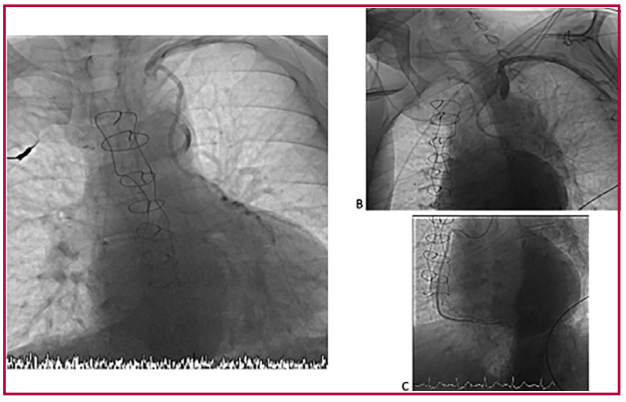

Fig. 1. Urgent coronary catheterization: Occlusion of the origin of the left subclavian artery (A & B); after the administration via left radial access, abrupt cessation of the passage of contrast from the subclavian artery to the aorta is observed. Patent bypass grafts of left internal mammary artery to the left anterior descending artery, right internal mammary artery to obtuse marginal artery (A & B), and saphenous vein to posterior interventricular artery (C) are observed. No lesions in the distal beds.

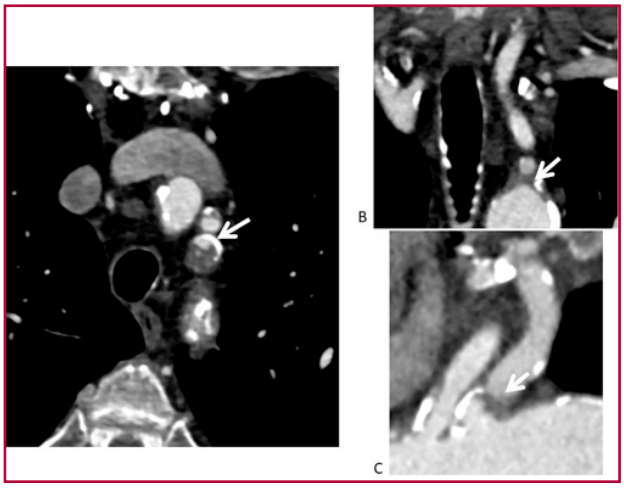

Fig. 2. CAT of supra-aortic trunks and circle of Willis: axial (**A**), coronal (**B**) and sagittal (**C**) images, showing calcified atheromatosis with occlusion of the origin of the left subclavian artery. The left internal mammary artery is patent. No occlusions in the structures of the circle of Willis or in the vertebrobasilar or carotid systems are observed.

diation exposure, compression syndromes, and fibromuscular dysplasia, with the proximal portion of the left subclavian artery being the most common location (> 75%).

While most subclavian stenosis patients remain asymptomatic, a small percentage may debut with clinical manifestations, such as stable angina, acute coronary syndrome -as was the case in our patient-, heart failure or malignant ventricular arrhythmias. (1)

However, current guidelines for coronary artery bypass grafting (5) lack recommendations for screening of subclavian artery stenosis before or after coronary surgery. Some authors recommend blood pressure measurements in both arms in the preoperative screening of these patients. (1) In patients with a differential systolic blood pressure (SBP) ≥ 15 mmHg, or in those with known or suspected peripheral artery disease (arm claudication, digital wounds, neurological symptoms), specific imaging tests are recommended to rule out subclavian stenosis. (1) Doppler ultrasound and CAT offer good sensitivity (73% and 91% respectively) and specificity (91% and 96% respectively) and are useful tools especially for the diagnosis of subclavian stenosis. However, magnetic resonance imaging (MRI) offers better results for coronary steal detection (sensitivity 90%, specificity 95%): in addition to anatomic information, it shows the directionality of retrograde flow within the mammary graft. Nevertheless, subclavian artery angiography remains the gold standard. (1)

As differential diagnosis for our patient, we could consider bypass obstruction or new lesions in the distal coronary beds, which are ruled out in the catheterization. If the patient had presented with other associated symptoms, such as ipsilateral upper extremity ischemia, aortic disease affecting the left subclavian and brachial arteries should be ruled out.

After confirming the diagnosis, our patient was evaluated by Vascular Surgery. It was decided to perform endovascular intervention, with predilatation and stent placement in the ostium of the left subclavian artery, with good angiographic outcomes.

Based on current evidence, revascularization is indicated in symptomatic (recommendation IIa C) and asymptomatic patients with bilateral subclavian stenosis (recommendation IIb C).

No clinical trials have compared the results with endovascular or surgical treatment, and the risk of major complications —including vertebrobasilar stroke— is low in both procedures (2.6% for endovascular treatment, and 0.9-2.4% for open surgery); therefore, current guidelines recommend both approaches with the same level of recommendation (IIa C) to be individualized depending on the case, (6) even though the endovascular approach is often the default strategy, as in our patient. It generally consists of stent placement, with a higher patency rate at 5 years (up to 85%). The success rate is 100% in stenosis and 80-95% in occlusions, as was the case with our patient. (6)

Open surgery, on the other hand, is limited to selected low-surgical risk patients, generally after failure of endovascular treatment. It offers good outcomes, with 1-year patency 95%, 3-year patency 86%, and 5-year patency 73-96%, (6) being the extra-anatomic bypass procedures (axillo-axillary, carotid-axillary or carotid-carotid bypass) the most used. (5, 6)

Postoperative clinical course of our patient was excellent and uneventful, and with pending follow-up in our Cardiology Department to assess the recovery of ventricular function.

In conclusion, we highlight our case because, despite left internal mammary graft is the most used in coronary revascularization surgery, subclavian artery obstruction is an underestimated entity in these patients. This entity may lead to angina recurrence or even acute coronary syndrome; therefore, we should keep it in mind in the differential diagnosis of previously operated patients debuting with angina, and include screening in the preoperative assessment of patients referred to aortocoronary surgery.

Conflicts of interest

None declared.

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Ethical considerations

Not applicable.

Ana Ruiz, Ane Elorriaga, Amaia Arregi, Paula Mendoza, Abel Andrés, Roberto Sáez
Department of Cardiology.
Hospital Universitario de Basurto (Bilbao, España).
Ana Ruiz Rodríguez - Email: anarrmo@gmail.com
Phone: 619 127 425

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Repair of Coarctation of the Aorta in Preterm Infant Weighting 1180 g

Repair of congenital heart diseases in low-weight infants has proved successful. (1) Critical coarctation of the aorta is a life-threatening lesion in newborns; two different approaches in low-weight preterm infants are adopted worldwide. Whereas some advocate delaying the timing of surgery until the baby grows up and gains weight, others favor early intervention. (2) Many centers have reported low mortality rates for coarctation in these patients. (3) However, most debate continues to focus on the question of optimal timing for surgery and the incidence of recoarctation of the aorta. Results of balloon dilation of native aortic coarctation in low-weight patients are disappointing, with recoarctation rates of 83% in neonates. (4)

We report the case of a preterm male infant with surgical repair of critical aortic coarctation, weighing 1180 grams, at the *Hospital Interzonal Especializado Materno Infantil de Mar del Plata*, with no postoperative complications or signs of recoarctation at one-year postoperative follow-up.

It is a male twin born from a controlled pregnancy, with a maternal history of insulin-requiring gestational diabetes and positive serologic test for syphilis, with inadequate treatment. After 32-week gestation, the patient was delivered by C-section due to pathological fetal Doppler and selective intrauterine growth restriction, weighting 1170 g, with an APGAR score of 9/10, not requiring resuscitation at birth. On admission to the Neonatal Intensive Care Unit, the patient presented with respiratory distress, grunting, recession and decreased saturation, requiring Mechanical Ventilatory Support (MVS) and lung surfactant 100mg/kg. Routine echocardiography revealed a 3 mm patent ductus arteriosus, with left-to-right shunt and enlargement of the left chambers, with no other structural heart disease. Physical examination showed symmetrical, peripheral pulses and signs of heart failure. Treatment with indomethacin for patent ductus arteriosus closure was started; 24 hours later, the patient decompensated hemodynamically with oligoanuria, abdominal bloating, poor peripheral perfusion and nonpalpable femoral pulses, requiring inotropic drugs. Follow-up echocardiography showed closed ductus arteriosus, coarctation of the aorta with a gradient of 27 m/sec and diastolic ramp, preserved left ventricular systolic function. Prostaglandin infusion at 0.01 micrograms/kg/min was used to open the ductus arteriosus. Ventilatory measures were taken to decrease pulmonary hyperflow. The patient showed positive diuresis and palpable pulses in lower limbs.

A multidisciplinary panel decided to perform surgical repair due to worsening of renal function, symptoms of heart failure as a result of ductus arteriosus shunt unresponsive to medical measures, increased MVS, high inspired oxygen fraction, dilatation of the left chambers, and also due to the fact that patent ductus arteriosus opening in preterm infants worsens or increases the incidence of intraventricular hemorrhage. Routine preoperative transfontanellar ultrasound revealed a grade 1 intraventricular hemorrhage.

With the patient (18 days of age, 1180 grams) in lateral decubitus position, a left posterolateral thoracotomy through the fourth intercostal space was performed; the ascending aorta, transverse aortic arch and branches, isthmus, large ductus arteriosus, and descending aorta were dissected. The image shows aortic coarctation at the level of the isthmus (Figure 1). The patent ductus arteriosus was ligated and the left carotid and left subclavian arteries were clamped at the level of the ascending aorta, allowing blood flow to the brachiocephalic trunk for cerebral perfusion; the descending aorta was also clamped. The ductus arteriosus was sectioned and the area of coarctation was resected; the incision was extended along the inferior transverse arch and over the left carotid artery. Finally, an extended end-to-end anastomosis was performed, with a clamping time of 16 min and no surgical complications (Figure 2). With good descending aortic pulse and no

palpable anastomotic thrill, the patient was transferred to the cardiac recovery room with low-dose adrenaline (stopped 12 hours post-procedure) and milrinone 0.75 micrograms/kg/min. Postoperative echocardiography reported a gradient in the area of the aortic isthmus of 4 mmHg, apical intraventricular septum of 1.7 mm without hemodynamic repercussion, left ventricular diastolic and systolic diameters of 18 mm and 11 mm respectively, and patent foramen ovale with left to right shunt. Postoperative course was uneventful (no bleeding, chylothorax, diaphragmatic paralysis or arterial hypertension); transfontanellar ultrasound showed no changes compared to the previous ultrasound. The patient was transferred to the Neonatal Intensive Care Unit, remaining in MVS due to sepsis associated with a central venous catheter, and requiring low-dose inotropic drugs. Elective extubation was performed (total MVS time, 40 days) with nasal cannula support for 48 hours, without recurrent nerve palsy. The patient remained in the pediatric ward for nutritional recovery.

At one-year postoperative follow-up, the patient has symmetrical, peripheral pulses; echocardiography shows a slight narrowing of the isthmus (gradient 15 mmHg) without diastolic runoff; preserved left ventricular systolic function, left ventricular diastolic and systolic diameters of 28 mm and 18 mm respectively, and abdominal aorta with pulsatile flow.

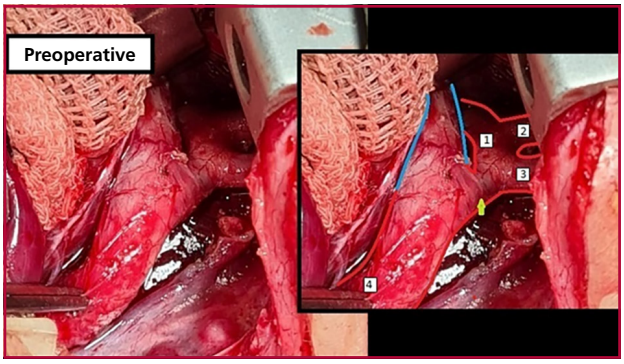

Fig. 1. CImage showing transverse arch (1), left carotid artery (2), left subclavian artery (3), descending aorta (4), patent ductus arteriosus (light-blue lines), and aortic isthmus and coarctation area (green arrow). This image does not show the brachiocephalic trunk.

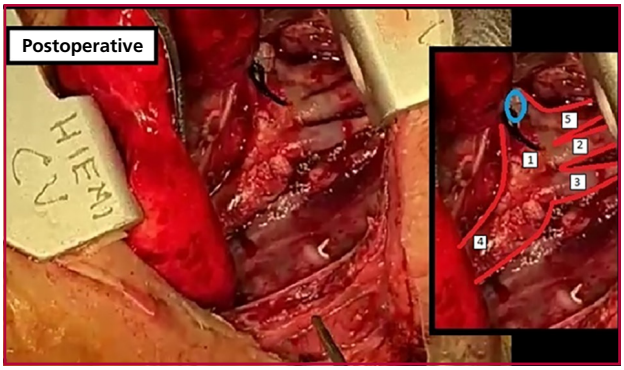

Fig. 2. Postoperative image showing ligated and sectioned ductus arteriosus (light-blue circle), resected aortic isthmus and coarctation area, and extended end-to-end anastomosis; brachiocephalic arterial trunk (5).

Repair of cardiac defects in low-birth and very low-birth weight neonates is increasingly performed in a reparative rather than palliative manner. Only a few risk factors for death after coarctation of the aorta repair in small infants have been recognized so far; however, critical preoperative clinical status and associated complex cardiac lesions have been documented to be the most common and important risk factors to influence mortality after surgery. Bacha et al. found that low-weight infants (< 1.5 kg) have a significantly higher chance of developing a recoarctation. However, Mc Elhinney et al. and Sudarshan et al. concluded that low weight at the time of repair is not a risk factor for recurrent coarctation. (2, 5)

Surgical repair in premature infants with very low birth weight (< 1500 g) and a diagnosis of coarctation of the aorta can be successfully performed. Given that surgical and interventional experience is still limited, the therapeutic decision must be individualized for each patient, and taken together with neonatologists, cardiologists and pediatric cardiac surgeons.

Conflicts of interest

None declared.

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

Ethical considerations

Not applicable.

Ramiro M. Pellicciari[1], Carlos J. Vázquez[1],

[1] Department of Pediatric Cardiovascular Surgery, Hospital Interzonal Especializado Materno Infantil de Mar del Plata. Ramiro Marcos Pellicciari - Avenida Juan José Paso 867, Mar del Plata, Buenos Aires. Argentina

E-mail: ramiropellicciari@gmail.com - Phone: 11-59732992.

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