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CLINICAL CASE

The image is the key: unilateral absence of a branch of the pulmonary artery

La imagen es la clave: ausencia unilateral de una rama de la arteria pulmonar

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What do we know about the subject matter of this study?

Unilateral absence of a branch of the pulmonary artery is an infrequent and underdiagnosed entity, generally described as clinical cases and, when it occurs in isolation, it may not cause symptoms until advanced age. There is no consensus on its treatment, which is generally reserved for complications, making clinical follow-up essential.

What does this study contribute to what is already known?

Through the presentation of two cases of unilateral absence in both branches of the pulmonary artery in isolation, diagnosed very early in life, we demonstrate that routine and noninvasive imaging methods, such as echocardiography, are key in the detection and follow-up of this entity.

Abstract

Unilateral absence of pulmonary artery (UAPA) is a rare and underdiagnosed entity. Due to its varied clinical expression, especially respiratory and most frequently associated with congenital heart disease, it can also present in isolation and remain asymptomatic for a long time. There is no consensus on its treatment, which is generally reserved for the presence of complications, mainly pulmonary hypertension, hemoptysis, or recurrent respiratory infections. **Objective:** To describe two cases of UAPA identified early in life, and the usefulness of routine imaging tests for its diagnosis. **Clinical Cases:** We present two clinical cases of asymptomatic children referred for pediatric cardiological assessment. The first one was 1 month old, with a prenatal diagnosis of right aortic arch, that was referred to rule out associated anomalies. An echocardiographic evaluation confirmed left isolated UAPA, sustained by plain radiography. CT angiography confirmed UAPA and left lung hypoplasia. Only clinical follow-up was determined. The second case is a 2-week-old neonate who was referred due to a systolic murmur. Right pulmonary artery agenesis was diagnosed by echocardiography with no other associated defects and was confirmed by CT angiography. Plain chest radiography showed

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no alterations initially, however, after 1.5 months of follow-up, right lung collapse was detected so clinical follow-up was decided. Both cases were suspected by echocardiography and confirmed by CT angiography. At 1.5 years and 6 months old, respectively, they show a good evolution, with good growth, and close monitoring of their intercurrent respiratory processes. **Conclusions:** UAPA can be asymptomatic when occurs in isolation and go unnoticed. Through simple imaging methods, especially echocardiography, it can be suspected and must be confirmed later by CT angiography or MRI angiography.

Introduction

Unilateral absence of pulmonary artery (UAPA) is a rare clinical entity with an estimated incidence of 1/200000 people and the absence of the right branch reaches up to 60% of the cases[1].

Usually associated with congenital heart disease, it can present clinically in isolation in approximately 30% of cases[2,3]. In the latter case, it can remain asymptomatic or with little respiratory symptomatology for a long time, hence its underdiagnosis[4].

Embryologically, it results from the involution of one of the two pairs of the sixth aortic arch, at the proximal level, which is intended to become a branch of the pulmonary artery[5]. This may go unnoticed in the fetal period due to the relatively normal development of the affected lung, caused by the vascular supplementation of the distal part of this undeveloped arterial branch by the ductus arteriosus. After birth and with ductal closure, progressive pulmonary hypoplasia will occur[6,7]. In this situation, collateral arteries may develop with a high risk of hemoptysis and, often, patients present with a small hyperlucent lung on the affected side on chest x-ray[5].

There are few cases reported in children and there is still no consensus on its treatment, generally reserved to the presence of complications, mainly pulmonary hypertension, hemoptysis, or recurrent respiratory infections[8].

The objective of this study was to report two cases of unilateral absence of pulmonary artery identified at early ages, and the usefulness of routine imaging for its diagnosis.

Clinical Cases

Case 1

A 1-month-old female infant was referred for cardiologic reevaluation due to a prenatal diagnosis of isolated right aortic arch in the fetal stage. No other history of interest; born at term at 39 weeks, weighing 3,220 g, with an uneventful neonatal period, no cardiovascular symptoms, and adequate weight and length growth. Prenatally, a basic genetic study was performed, including 22q11 deletions, with negative results. No significant findings on physical examination, with normal oxygen saturation.

The echocardiographic evaluation showed normal pulmonary trunk and normal right pulmonary artery in dimensions, flow, and trajectory. In this evaluation, no left pulmonary artery was identified, visualizing a small vessel compatible with small ductus arteriosus at that level. Chest X-ray performed at 2 months of life showed a completely veil-like opacity in the left pulmonary field with ipsilateral tracheal deviation. A CT angiography confirmed the findings of pulmonary aplasia and left lung hypoplasia. Perfusion scintigraphy performed by pediatric pulmonology specialists did not show uptake in the left lung.

The patient has continued only with clinical follow-up with good evolution. At one and a half years of age, she has been under close follow-up of her intercurrent respiratory processes, without usual treatment except for exacerbations of such pathology, with adequate growth and normal cardiac function.

Case 2

A 2-week-old full-term neonate, born by vaginal delivery with a controlled pregnancy and with normal course, weighing 3,380g, was hospitalized in the neonatal ward due to poor oral tolerance and mild asymptomatic hypoglycemia. In addition, he needed mild oxygen therapy to maintain saturation intermittently, with initial chest radiological evaluation without alterations. He was referred to cardiology due to a murmur found in the routine examination.

In the cardiological evaluation, he was asymptomatic, physical examination showed no significant alterations and adequate saturation. Echocardiography showed a main pulmonary artery of normal caliber and flow arising from the right ventricle and confluent left pulmonary artery of normal caliber and flow; no right pulmonary artery or ductus was observed. Normal pulmonary pressure values by indirect estimation. There was a small vessel starting from the descending thoracic aorta to the right with a continuous flow, cal-

iber 2 mm, and it was not identified where it ended. Therefore, a diagnosis of right pulmonary artery aplasia was made. The chest X-ray performed at that visit showed no significant alterations.

In the follow-up of the patient, the X-ray performed at 6 weeks of life showed a collapsed right lung, with volume loss, and dextrocardia. No alterations were observed in the left lung. A heart CT scan performed at 2 months confirmed the findings.

The patient has continued with clinical follow-up; in the last evaluation, at 6 months of age, he presented an uneventful clinical evolution, with no need for oxygen therapy, adequate growth, and without having required hospital admissions.

The comparison of both cases is shown graphically. In Figures 1 and 2 of the X-ray images, the development of pulmonary hypoplasia after ductal closure in the case of agenesis of the right pulmonary artery is noteworthy (figure 2). Figure 3 (A and B) shows a typical ultrasound section of the pulmonary trunk and artery, which are key to clarifying the image of the left (figure 4A) and right (figure 4B) pulmonary artery agenesis. Figure 5 corresponds to the confirmatory CT angiography in both cases.

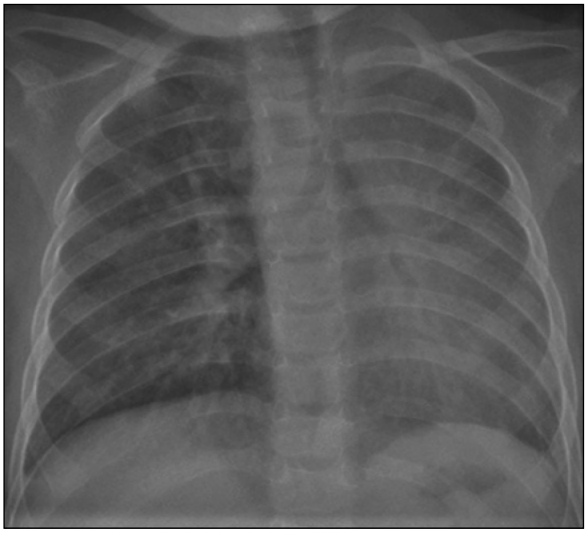

Figure 1. Case 1. X-ray of the case of left pulmonary branch agenesis performed at 2months. Note the completely veiled left lung field with ipsilateral tracheal shift and levocardialization.

Discussion

UAPA is a rare condition that usually occurs in association with complex heart disease with very definite clinical manifestations, such as cyanosis (Fallot, truncus, pulmonary atresia); or in isolation, as in our cases, where it can remain asymptomatic for a long time[6,7,9]. In isolated cases, hemoptysis or frequent respiratory infections may be the initial clinical manifestation. Pulmonary hypertension frequently develops during the disease due to ipsilateral pulmonary hyperflux[5], which may occur in up to 45% of cases and constitutes the most feared complication[7,9].

Imaging is the key to diagnosis. Accurate diagnosis may require several modalities. Chest radiography raises suspicions[6,7,10]. Echocardiography has been defined as a useful screening modality but according to some authors, it is insufficient to make a definitive diagnosis[7,8,11], however, we believe it is a key diagnostic test. Ultrasound evaluation of the pulmonary artery and its branches is routine and measurable in pediatric practice[12]; the demonstration of the blind end of this artery is therefore totally objectifiable as has been demonstrated in the cases described.

In this context, and within the differential diagnosis, we must consider possibilities such as the anomalous origin of the pulmonary artery from the ascending aorta or ductus arteriosus, or a pulmonary artery sling, where the left pulmonary branch leaves the right one, creating a vascular ring with respiratory symptomatology. It is important to evaluate the pulmonary irrigation from collaterals or from the descending aorta, and the ipsilateral pulmonary venous return to the agenesis, which normally will be significantly decreased or absent. Likewise, the frequent association of congenital heart diseases is confirmed or ruled out by echocardiography.

Echocardiography allows the detection and evolutionary estimation of pulmonary arterial pressure. It is risk-free, non-invasive, and fast and cost-effective[11,13]. In addition, it could be considered the main method to detect UAPA, but it still needs to rely on technology and operator experience, as well as an understanding of the disease, so there is still a high rate of misdiagnosis. Therefore, the definitive diagnosis is made by CT angiography or MR angiography, which also accurately defines the different lung fields and their flow sources[12,13].

There is no consensus on treatment, and different alternatives are offered in the presence of clinical manifestations[3,6,7,9]. In general, the approach is conservative, and periodic monitoring is recommended, at least annually, by echocardiography to control the appearance and development of pulmonary hypertension, a fundamental clinical element in its follow-up[14]. When it appears, treatment with specific vasodilators (calcium channel blockers, phosphodiesterase inhibitors, endothelin antagonists) can improve the patient's evolution. In recurrent or severe hemoptysis, embolization, lobectomy, or pneumectomy is indicated; surgical actions are also reserved for

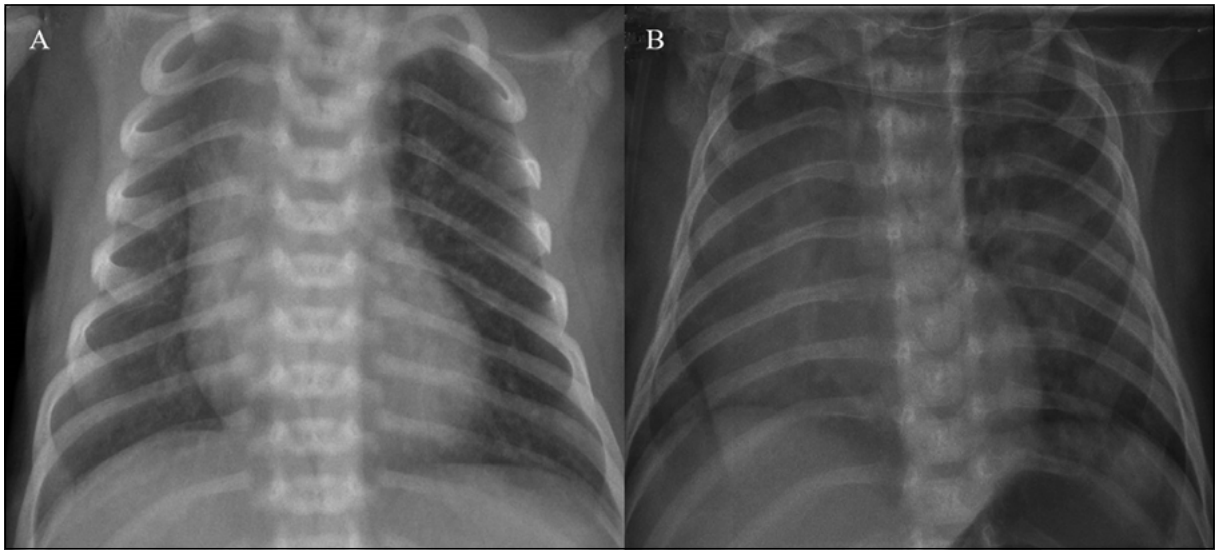

Figure 2. Case 2. X-ray of the case of right pulmonary branch agenesis. (A) At 2 weeks of life. No alterations are evident. (B) At 1.5 months. Here we can see the dextrocardialization and collapse of the pulmonary field itself.

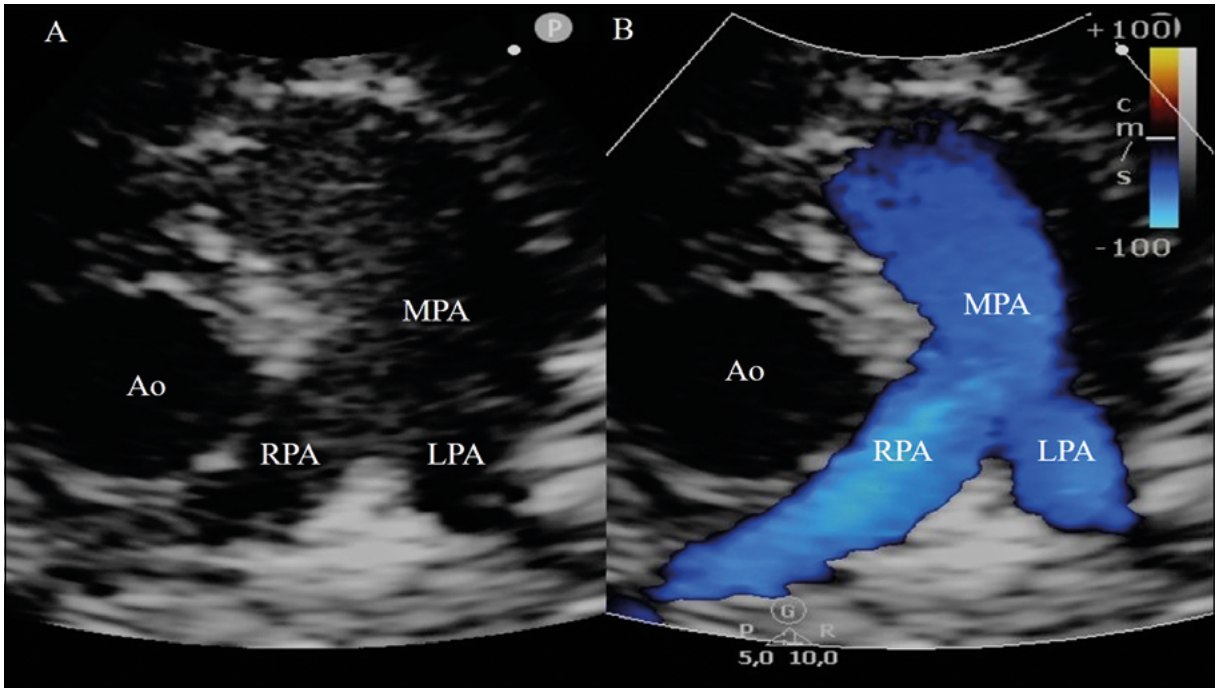

Figure 3. Echocardiographic projections to assess the pulmonary trunk and branches in 2 dimensions (A) and with color Doppler (B). Ao: Aorta; MPA: Pulmonary trunk or main pulmonary artery; LPA: Left pulmonary branch; RPA: Right pulmonary branch.

presentations with repeated pulmonary infections, and with significant impact on the patient's health. Alternatively, revascularization procedures could be attempted in selected cases detected early in life, with minimal pulmonary artery branch remnant, using grafts (autologous pericardium, allografts, or prosthetic material), with successful results reported on an individual basis[15,16].

Conclusions

UAPA is a heterogeneous entity in the age and form of presentation and can be asymptomatic for a long time when presented in isolation and goes unnoticed. Through simple and non-invasive imaging methods, among which echocardiography stands out, it can be suspected, but it should be confirmed later by

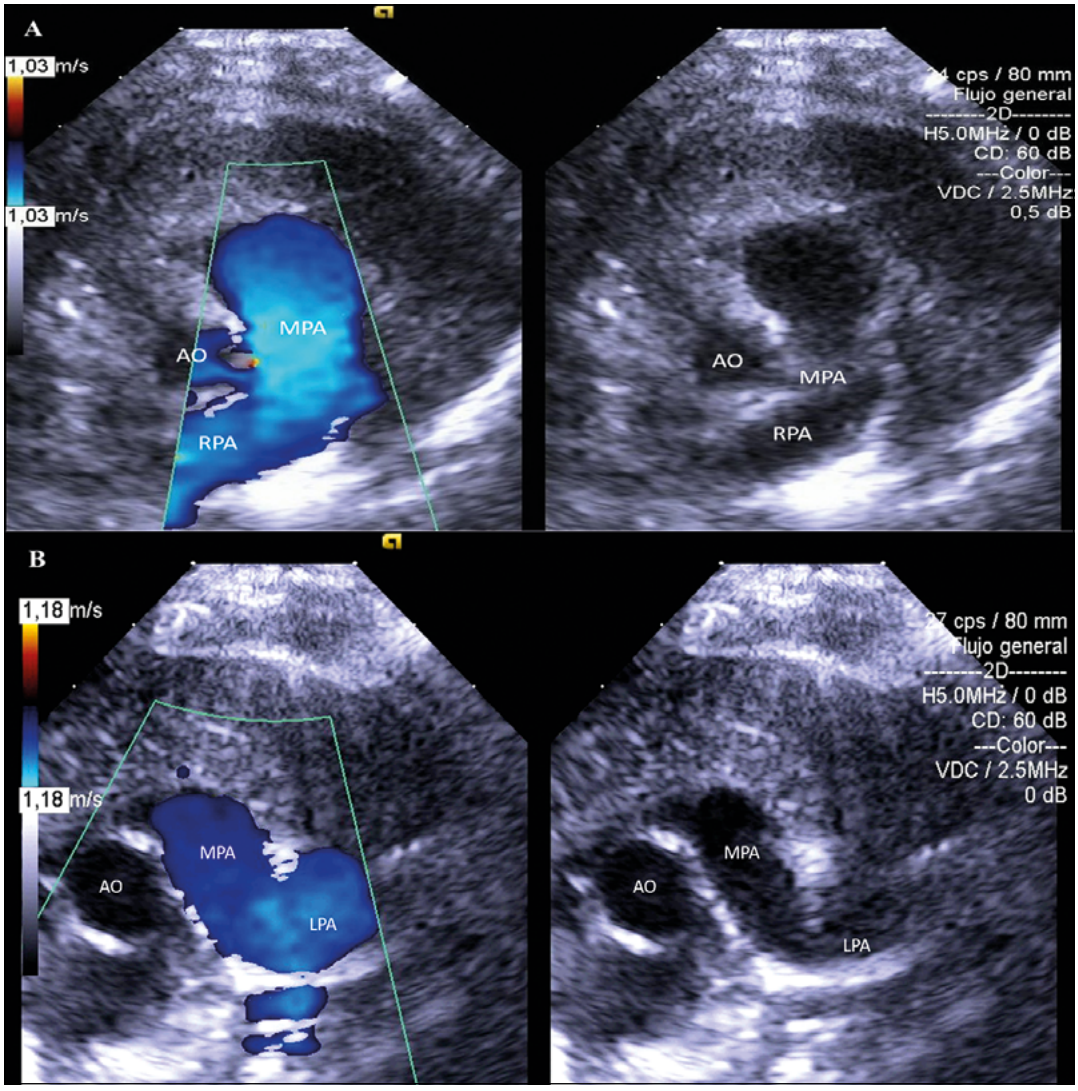

Figura 4. Ecocardiografía siguiendo la proyección base de los casos expuestos. (A) Ausencia de rama pulmonar izquierda (caso 1). (B) Derecha (caso 2). Ao: Aorta; MPA: Tronco pulmonar o arteria pulmonar principal.; LPA: Rama pulmonar izquierda; RPA: Rama pulmonar derecha.

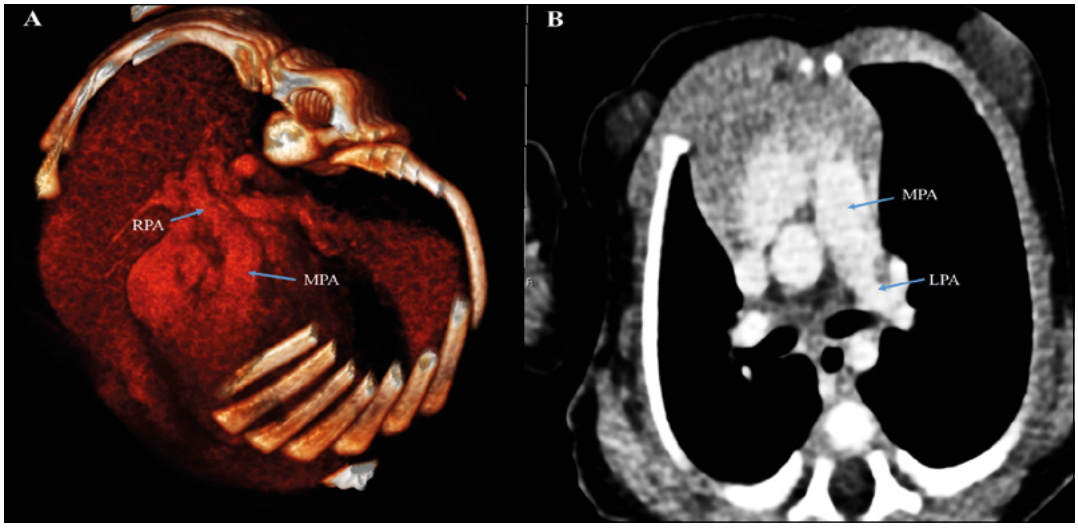

Figura 5. Angiografía por tomografía computarizada. (A) Caso 1. Ausencia de rama pulmonar izquierda con reconstrucción de imagen 3D. (B) Caso 2. Ausencia de rama pulmonar derecha. MPA: Tronco pulmonar o arteria pulmonar principal; LPA: Rama pulmonar izquierda; RPA: Rama pulmonar derecha.

other more accurate procedures such as CT angiography or MR angiography. Echocardiography, in addition to being an excellent screening tool in this case, is the key tool in its follow-up.

Ethical Responsibilities

Human Beings and animals protection: Disclosure the authors state that the procedures were followed according to the Declaration of Helsinki and the World Medical Association regarding human experimentation developed for the medical community.

Data confidentiality: The authors state that they have followed the protocols of their Center and Local regulations on the publication of patient data.

Rights to privacy and informed consent: The authors have obtained the informed consent of the patients and/or subjects referred to in the article. This document is in the possession of the correspondence author.

Conflicts of Interest

Authors declare no conflict of interest regarding the present study.

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