AORTIC ORIGIN OF THE RIGHT PULMONARY ARTERY ASSOCIATED WITH *DUCTUS ARTERIOSUS* IN AN ADULT. A CASE REPORT*

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RESUMEN

ORIGEN DE LA ARTERIA PULMONAR DERECHA EN LA AORTA, ASOCIADO A LA PERSISTENCIA DEL CONDUCTO ARTERIOSO EN UN ADULTO. INFORME DE UN CASO

Los autores presentan el caso de una mujer de 26 años, en clase funcional I (según criterio de la NYHA), con origen de la arteria pulmonar derecha en la aorta, asociado a la persistencia del conducto arterioso y severa hipertensión arterial pulmonar (101/40-70 mm Hg), la cual permaneció elevada (89/40-60 mm Hg) después del suministro de O_2 al 100%. La presión de la arteria pulmonar derecha (125/60-86 mm Hg) resultó más alta que la presión del tronco de la arteria pulmonar y similar a la aórtica. La paciente fue tratada con éxito mediante la división quirúrgica del conducto arterioso y la anastomosis término terminal de la arteria pulmonar con la arteria pulmonar derecha. Seis meses después de la cirugía, la presión sistólica de la arteria pulmonar, determinada por medio de la ecocardiografía Doppler, fue 60 mm Hg. El ecocardiograma bidimensional mostró la correcta anastomosis de la arteria pulmonar derecha con la arteria pulmonar. La gammagrafía pulmonar mostró la perfusión de ambos pulmones por la arteria pulmonar; si bien, la perfusión del pulmón derecho fue menor que la del izquierdo: 30 vs. 70%, respectivamente.

El origen anómalo de una de las ramas de la arteria pulmonar en la aorta se observa mayormente en niños menores de 1 año; la frecuencia de este padecimiento es <1% de todas las cardiopatías congénitas y la sobrevida de los pacientes con esta cardiopatía es muy corta. La originalidad del caso clínico aquí descrito es que se trata de un paciente con una cardiopatía rara en el adulto, tratada quirúrgicamente.

SUMMARY

We describe the case of a 26-year-old female in functional class I (NYHA), with aortic origin of the right pulmonary artery associated with a persistent ductus arteriosus and severe pulmonary artery hypertension (101/40-70 mm Hg), which remained elevated (89/40-60 mm Hg) after the administration of 100% oxygen. Right pulmonary artery pressure (125/60-86 mm Hg) was higher than that of main pulmonary artery and similar to aorta pressure. The patient was successfully treated: surgical closure of the ductus arteriosus and end-to-end anastomosis between the pulmonary artery and right pulmonary artery were carried out. Systolic pulmonary arterial pressure, estimated by echocardiography Doppler, was 60 mm Hg six months after surgery. Cross-sectional echocardiogram showed the anastomosis of the right pulmonary artery with the main pulmonary artery. Pulmonary gammagraphy showed both lungs perfused through the main pulmonary artery; right lung perfusion was lesser than left lung perfusion, 30 vs. 70% respectively.

Aortic origin of a right or left pulmonary artery is a heart disease seen in patients during the course of the first year of life. Its frequency is <1% among all the congenital cardiopathies and the survival rate to adult life is very low. The originality of this paper is the presentation of a rare congenital cardiopathy treated surgically in an adult.

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RESUME

ARTERE PULMONAIRE DROITE NAISSANT DE L'AORTE, ASSOCIEE A LA PERSISTANCE DU CANAL ARTERIEL, CHEZ UNE ADULTE

On présente le cas clinique d'une femme âgée de 26 ans, de classification fonctionnelle I (NYHA), avec l'artère pulmonaire droite naissant de l'aorte et associée à la persistance du canal artériel perméable avec hypertension pulmonaire accentuée (101/40-70 mm Hg). Celle-ci était restée élevée (89/40-60 mm Hg) après inhalation d'oxygène pur. La pression de l'artère pulmonaire droite (125/60-86 mm Hg) était plus élevée que la pression du tronc de l'artère pulmonaire et que celle de l'artère pulmonaire gauche. La malade a été operée avec succès: section-suture du canal artériel, suivie de l'anastomose de l'artère pulmonaire droite avec le tronc de l'artère pulmonaire. Six mois après l'intervention chirurgicale, la pression systolique de l'artère pulmonaire, estimée par échocardiographie Doppler, était de 60 mm Hg. L'anastomose adéquate de l'artère pulmonaire droite a été démontrée par échocardiographie bidimensionnelle. D'après la gammagraphie pulmonaire, les deux poumons étaient irrigués par l'artère pulmonaire, le poumon droit étant moins irrigué que le gauche: 30% contre 70%. L'artère pulmonaire droite naissant de l'aorte est une malformation congénitale observée surtout chez des enfants âgés de moins d'un an. Cette malformation se présente avec une fréquence de moins de 1% de toutes les cardiopathies congénitales et la survie est très courte. L'interêt du cas, qu'on présente ici, consiste en la rareté de cette malformation traitée par chirurgie à l'âge adulte.

Palabras clave: Origen de la arteria pulmonar derecha en la aorta. Persistencia del conducto arterioso. Ectopia de la arteria pulmonar.

Key words: Right pulmonary origin from the ascending aorta. Patent ductus arteriosus. Unilateral ectopia of the pulmonary artery.

INTRODUCTION

The aortic origin of a pulmonary artery (AO-a-PA) is an infrequent congenital cardiopathy: seven cases among 7329 patients with congenital heart diseases.[1] Fifty percent of patients with aortic origin of the right pulmonary artery (AO-RPA) died before 1 year of age and 75% before 10 years of age. Few patients survive beyond 20 years of age. In 1868, Fraetzel published the first case of AO-RPA associated with an aortopulmonary window in a 25-year-old girl (Op. cit. Fontana).[2] Purcaro informed of the case of a 43-year-old man with isolated aortic origin of the left pulmonary artery (AO-LPA) (Op. cit. Tagliente).[3] Another lengthy survivor was a 21-year-old female with AO-RPA and associated persistence of *ductus arteriosus* (PDA).[4] The purpose of this article is to present the clinical case of a 26-year-old female with AO-RPA associated with a PDA, treated surgically, to discuss the clinical, echocardiographic, and hemodynamic studies, and to review the treatment aspects of the AO-a-PA.

Case report

This is the case of a 26-year-old female, in functional class I (NYHA) without treatment, with a cardiac murmur present at birth. Seven years previously, she presented moderate dyspnea and fatigue provoked by exercise. The clinical examination revealed a blood pressure of 110/60 mm Hg. Jugular vein distension was absent. Cyanosis was apparent only at exercise. A continuous murmur was heard at the left sternal border and the second heart sound was prominent and constantly split. Peripheral pulses were wide. Respiratory rate, body temperature T, N, E, head, abdomen, and extremities examination were normal. The chest roetgenogram showed signs of cardiomegalia (cardiac index 0.52), right ventricle enlargement, concavity of the main pulmonary arterial segment, and compared with the left lung, right lung vascularity was increased *(Figure 1)*. The electrocardiogram at rest (not illustrated) showed sinusal rhythm; ÂQRS at 70º, signs of right incomplete bundle branch block, right ventricle enlargement, and pulmonary artery hypertension.

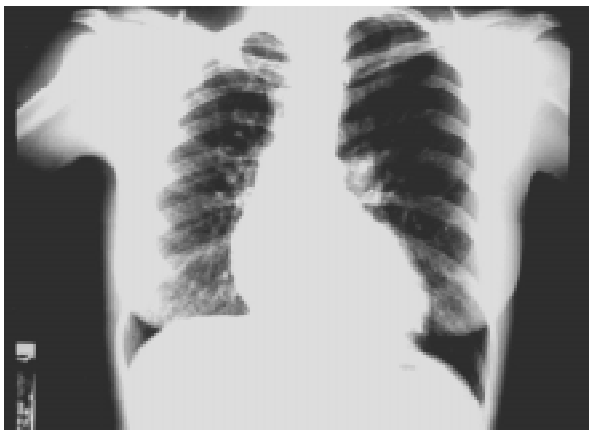

Fig. 1: Chest PA roentgenogram. See text.

The echocardiogram showed signs of a left patent *ductus arteriosus (Figure 2A),* abrupt interruption of the right pulmonary from the pulmonary artery trunk *(Figure 2A),* origin of the right pulmonary artery from the ascending aorta *(Figure 2B)*, and tricuspid insufficiency *(Figure 2C)*.

At intravascular catheterization *(Tables I and II)*, the following occurred: a) a *ductus arteriosus* was catheterized from the left branch of the pulmonary artery; b) the blood O_2 analysis, breathing atmospheric O_2, or 100% O_2, suggested the presence of a left-to-right blood flow shunt at the pulmonary artery level, and the presence of a right-to-left blood flow shunt; c) at rest, severe pulmonary artery hypertension was present, remaining elevated after breathing 100% O_2, and e) the pressure and O_2 content of the right pulmonary artery were

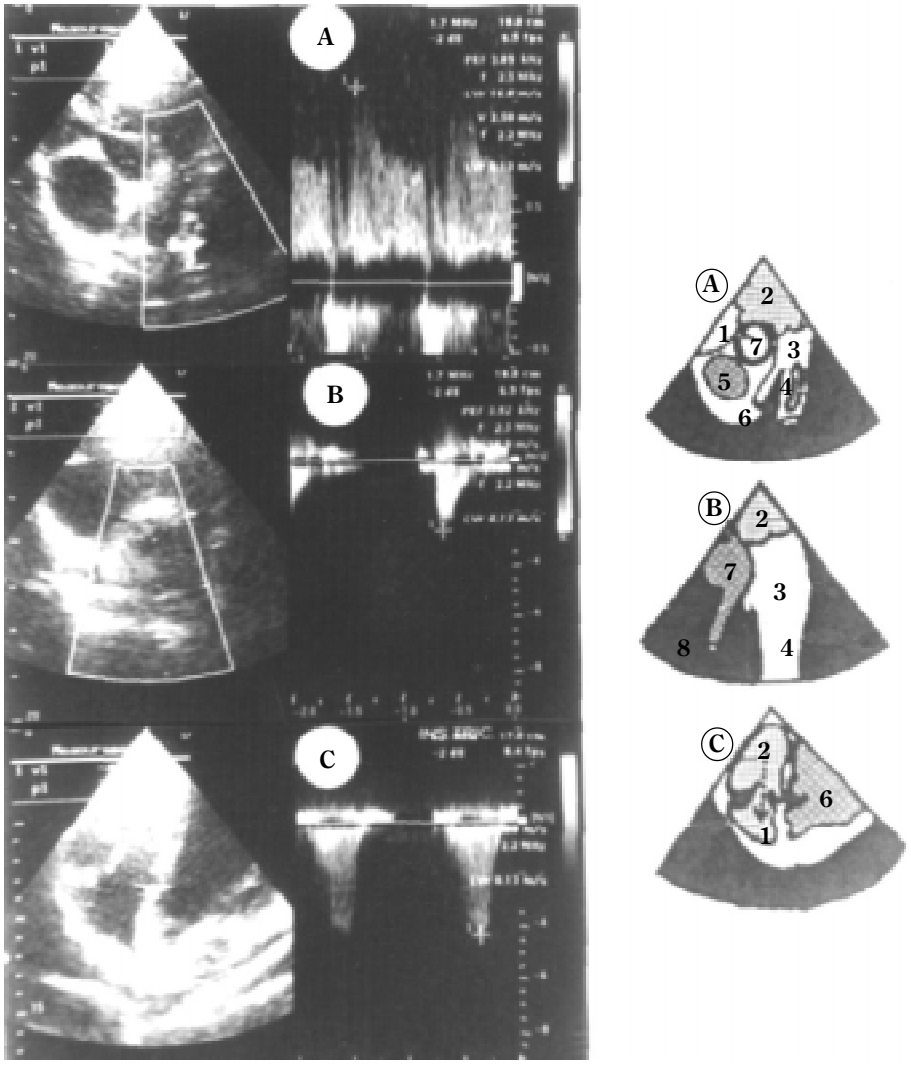

Fig. 2: Doppler echocardiograms before surgical treatment. A. Parasternal short axis view. B. Modified parasternal short axis view, and C. Apical four chambers view. 1= Right *atrium;* 2 = right ventricular outflow chamber; 3= pulmonary artery; 4= left pulmonary artery (In A, continuous Doppler shows the blood flow jet emerging from the *ductus arteriosus*); 5= left *atrium*; 6= left *atrium* and ventricle; 7= Aorta, and right pulmonary artery.

similar to aortic pressure and O_2 content. Pulmonary artery angiography showed the opacification of the pulmonary artery trunk and subsequent opacification of the left pulmonary artery *(Figure 3A)*. The right pulmonary artery was catheterized from the ascending aorta *(Figure 3B)*. The aortogram (left aortic arch) revealed the abnormal origin of the right pulmonary artery from the posterior aspect of the ascending aorta *(Figure 3C)*. Because the vascular resistance ratio decreased from 0.81 to 0.68 after inhalation of O_2 100% *(Table II)*, surgical treatment was recommended. Surgery was carried out with good results. Through a left posterolateral thoracotomy, section of the *ductus arteriosus* was performed, the pericardium exposed and a Dacron graft sutured to the main pulmonary artery and clamped at the site of the anastomosis. Once left thoracotomy closed, by means of a posterolateral right thoracotomy the right pulmonary artery was clamped and transected from the aorta, and an end-to-end anastomosis between the ectopic right pulmonary artery and the graft at the main pulmonary artery was performed. Six months after surgery, the patient was clinically stable. A new roentgenogram (not illustrated) showed no difference in the vascularity of both lungs. At the Doppler echocardiogram, estimated pulmonary artery pressure was 60 mm Hg. In both pulmonary arteries, velocity of blood flow was normal and turbulence was absent, pulmonary valve closure was normal, and anomalies of the right ventricle were unnoticed. The right pulmonary artery was correctly implanted to the main pulmonary artery *(Figure 4)*. According the gammagraphic study, both lungs were perfused by the main pulmonary artery, but right lung perfusion was lesser than left lung perfusion *(Figure 5)*.

DISCUSSION

In our patient, the right pulmonary artery rose at about the midposition of the back aspect of the ascending aorta *(Figure 3C)*. In approximately 85 to 88% of cases, the AO-RPA is much more common than the AO-LPA.[2,4] As occurs in three-quarters of cases described in the medical literature, our patient presented associated a PDA.[2] The difference in the vascular marking in both lungs, noted at the chest roentgenogram, suggested that the origin of blood flow into the two lungs had different sources

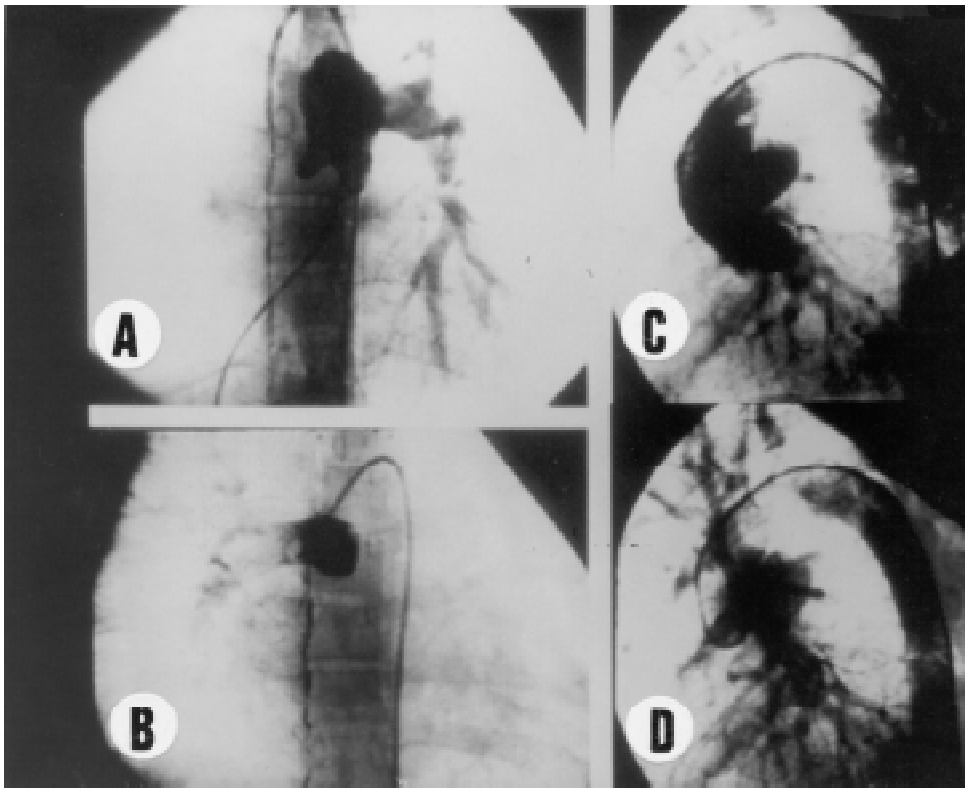

Fig. 3: Pulmonary artery angiography. A. Catheterization of the pulmonary artery from the right heart. The pulmonary artery trunk is observed, with subsequent opacification of the left pulmonary artery, B. Angiography of the right pulmonary artery, catheterized from the ascending aorta, and C. Ascending aortography. Opacification of the ascending aorta, and of the right pulmonary artery originated from the posterior aspect of the aorta. D. Subsequent opacification of the distal branches of the right pulmonary artery.

Table II.
Hemodynamic variables.

Hemodynamic variable	FIO_2 0.20	FIO_2 1.0
Left ventricle PO_2 (mm Hg)	55	246.0
Descending aorta PO_2 (mm Hg)*	55	223.0
Right pulmonary artery PO_2 (mm Hg)	50	321
Left pulmonary artery PO_2 (mm Hg)	36	50
Aortic output (1 min^{-1})	4.1	4.0
Aortic index (1 min^{-1} m^{-2})	2.6	2.5
Pulmonary output (1 min^{-1})	4.5	4.4
Pulmonary index (1 min^{-1} m^{-2})	2.8	2.8
Pulmonary *vs* aortic output	1.1	1.1
Right pulmonary artery resistance (din s cm^{-5}	1528	1600
Pulmonary artery resistance (din s cm^{-5})	1244	1090
Peripheral artery resistance (din s cm^{-5})	1521	1580
Pulmonary *vs* peripheral artery resistance ratio	0.81	0.68

* Normal PaO_2 = 65 ± 5 mmHg at 2240 m altitude at the Mexican plateau.

(Figure 1). Our patient developed pulmonary arterial hypertension, especially in the "systemic" lung. The arteriovenous blood flow shunt through the PDA and the systemic arterial resistance overloading the vasculature of the right lung contributed to the severe increase of pulmonary vascular resistance in our patient. Although no difference in either lung vascularity was observed at the postoperatory chest roentgenogram, pulmonary perfusion scintiscan showed less perfusion of the right lung *(Figure 5)*. This means that right pulmonary obstructive vascular disease was present, in spite of the postoperatory decrease in pulmonary artery pressure.

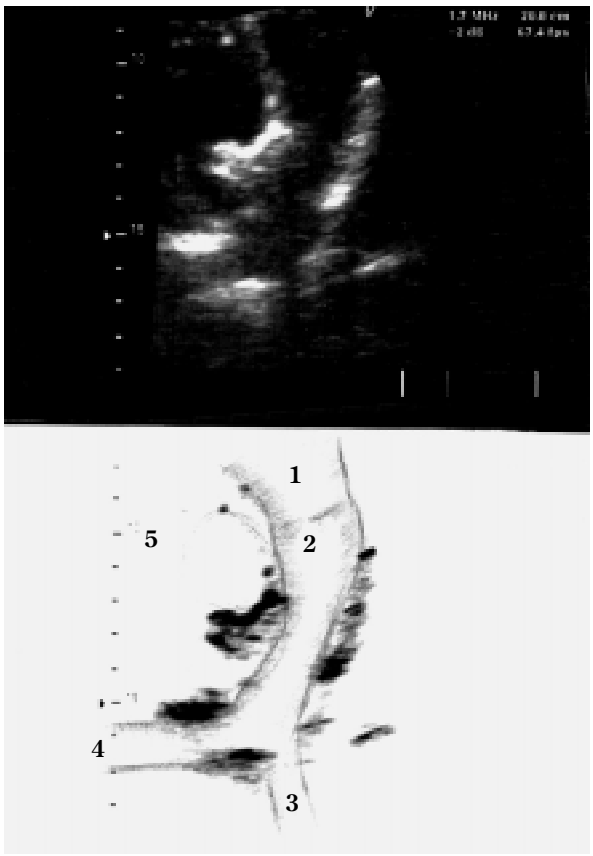

Fig. 4: Subcostal-view echocardiogram after surgical correction. Right pulmonary artery anastomosed (4) to the main pulmonary artery (2) is observed. Higher wave density at the anastomosis site is due to Dacron graft. 1= Right ventricle, 3= Left pulmonary artery and 5= Aorta.

Table I.
Intravascular catheterization data.

	FIO_2 0.20					FIO_2 1.0				
	Oxygen		Pressure			Oxygen		Pressure		
	CaO_2 (vol %)	SaO_2 (%)	S	D	M	CaO_2 (vol%)	SaO_2 (%)	S	D	M
Right atrium	12.9	65.0	•	•	4	13.7	69.1	•	•	12
Right ventricle	12.9	64.9	106	11		14.3	72.1	86	17	
Pulmonary artery	15.7	78.7	101 (60)	46	70	17.2	86.6	89	40	60
Left pulmonary artery	13.8	69.5	100	40	72	17.0	83.0	89	50	70
Right pulmonary artery	17.0	85.7	125	60	86	19.7	99.0	126	62	88
Left ventricle	17.8	89.5	135	17	•	19.7	99.0	130	18	•
Ascending aorta	16.7	88.7	•	•	•	•	•	•	•	•
Descending aorta	17.7	88.9	133	52	78	19.7	99.0	115	52	79

In parenthesis, systolic pressure 30 days after surgery determined by echocardiography. S, D and M= systolic, diastolic and mean pressures in mm Hg.

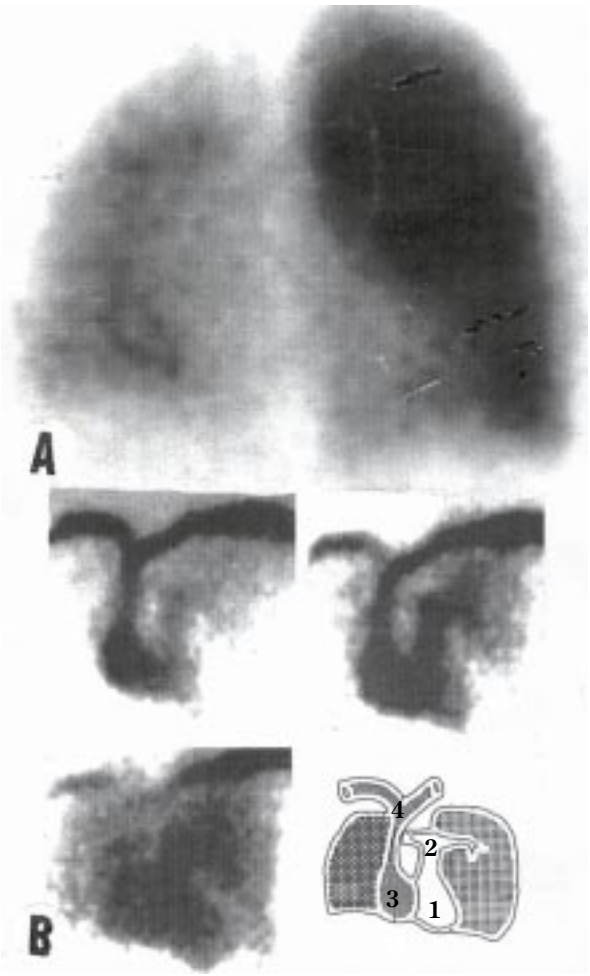

Fig. 5: A. Macroaggregated albumin labeled with I^{131} pulmonary perfusion scintiscan. Heterogeneous distribution of the tracer is observed: 30% for the right lung, and 70% for the left lung. B. First-transit radionucleide (Tc^{99m} dietilentriaminepentacetic acid) angiogram. Radioactive bolus enters the right side of the heart, main pulmonary artery, both pulmonary arteries and lungs. 1= Right ventricle, 2= Main pulmonary artery showing left and right pulmonary arteries, 3= Right *atrium* and 4= Superior vena cava and subclavian veins.

Seven cases of anomalous origin of a pulmonary artery from the ascending aorta were found among 7329 patients with congenital cardiopathies (five of the right pulmonary artery and two of the left).[1] Only three lengthy survivors have been reported in 65 cases of children with aortic origin of the right pulmonary artery.[2] In 1998, Kuinose described the case of a 16-year-old patient, without previous treatment, with AO-RPA, bidirectional shunting across a PDA, and severe pulmonary vascular restriction. In this case, total correction of the malformation was indicated according to preoperative lung biopsy results.[5]

Ninety-five percent of patients with unilateral origin of pulmonary artery in the aorta presented, during their first years of life, discrete signs of congestive heart failure. To the contrary, our patient presented discrete signs of heart failure until she was 14-year-old. Another lengthy survivor was a 21-year-old woman, with associated patent *ductus arteriosus*, generalized cyanosis, exercise intolerance, and episodes of epistaxis.[4] There has been described the case of a 43-year-old woman with differential cyanosis, right pulmonary artery arising from the aorta, pulmonary artery hypertension associated with the persistence of *ductus arteriosus*.[6] In 1978, Purcaro reported the case of a 43-year-old male with isolated anomalous origin of left pulmonary artery from the ascending aorta (Op. Cit. Tagliente).[3] The case of a 23-years-old man with isolated AO-LPA and moderate pulmonary artery hypertension has been described. In this patient post-surgical pulmonary artery and left pulmonary artery pressures decreased to normal.[7] It seems that once patients survive beyond 20 years of age, pulmonary artery pressure stabilizes, leaving behind the period of life in which the probability of death secondary to pulmonary artery hypertension is higher.[8]

The unilateral origin of a pulmonary artery from the aorta is usually fatal without surgical correction. Thirty-five percent of patients with this malformation die in the first month of life, and 70% die by the age of 1 year. A total of 17% survive beyond 10 years of age, and <10% to 20 years. When this anomaly is surgically treated, survival of patients increases from 30 to 84% at 1 year of age.[2] Contrary to the odds, the first case of anomalous origin of a pulmonary artery in the aorta described in the medical literature was not the case of a newborn: in 1868, Fraentzel published the case of a 25-year-old female who died of congestive heart failure secondary to an aortopulmonary window and AO-RPA (Op. cit. Fontana).[2] When no treatment is given, most patients with anomalous origin of a pulmonary artery from the ascending aorta die before 6 months of age due to congestive heart failure. In 94% of the cases, the anomalous origin of a pulmonary artery from the ascending aorta is associated with congenital anomalies derived from the misde-

velopment of the aortic arches: patent *ductus arteriosus* (68%), aortopulmonary window (15%), and hypoplastic or interrupted aortic arch (11%). These findings could suggest that these malformations are embryologically related, although the interruption of the aortic arch and aortic coarctation associated with the origin of a pulmonary artery from the aorta could be secondary to hemodynamically induced anomalies.[1]

The absence of the proximal portion of the left or right pulmonary artery, whose distal portion is supplied via a patent *ductus* or via aortopulmonary collaterals other than the *ductus arteriosus* are two conditions which differ, either in embryologic development and in physiologic behavior, from the AO-a-PA. In the first two conditions, pulmonary artery pressure is often elevated but less than the systemic pressure, whereas in the AO-a-PA the pressure is at the systemic level. In our patient, the difference in the vascular marking in both lungs noted at the chest roentgenogram *(Figure 1)* suggested that the origin of blood flow into the two lungs was from a different source. Our patient developed pulmonary arterial hypertension, especially in the "systemic" lung. Both the arteriovenous blood flow shunt through the patent *ductus arteriosus* and the systemic arterial resistance overloading the vasculature of the right lung contributed to the severe increase of pulmonary vascular resistance in our patient.

Many surgical procedures, as follows, have been proposed to palliate or correct the AO-a-AP and its associated cardiac defects: banding of the anomalous vessel; ligation of the pulmonary ectopic artery and of the patent *ductus arteriosus* and direct anastomosis or interposition of an artificial graft (end-to-end or side-to-side) between the ectopic artery and the main pulmonary artery.[2,9,10] Surgery has been prescribed in cases of AO-a-PA with severe pulmonary artery hypertension, and patent *ductus arteriosus*.[4] Because pulmonary artery pressure and resistance were severely elevated in our patient, prescription of surgical treatment was not obvious. Confronted by this fact, it could be wise to indicate surgical banding of the pulmonary artery, ligation of the ectopic pulmonary artery and *ductus arteriosus*, while pulmonary artery pressure was survey. Complete surgical correction was performed due to our patient being asymptomatic, in functional class I, and because of favorable hemodynamic response to 100% O_2: pulmonary artery pressure and resistance decreased, while PaO_2 increased significantly.

CONCLUSIONS

We presented the clinical case of a heart congenital cardiopathy rare in adults, which was correctly and successfully treated.

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