

Anomalous origin of the right pulmonary artery from the ascending aorta associated with aortopulmonary window

José Miguel Torres-Martel^{1*}, Lydia Rodríguez-Hernández¹ and Joaquín Rodolfo Zepeda-Sanabria²

¹Department of Pediatric Cardiology; ²Department of Cardiovascular Surgery, UMAE, Pediatrics Hospital, Centro Médico Nacional Siglo XXI, IMSS, Mexico City, Mexico

Abstract

Anomalous origin of one pulmonary artery from the aorta is rare. We report a case of a three-month-old infant with aortopulmonary window and anomalous origin of the right pulmonary artery from the ascending aorta. He underwent surgery with anastomosis of the right pulmonary artery, ligation of the aortopulmonary window and the patent duct. He was released under medical treatment and had no signs of pulmonary hypertension or heart failure. (Gac Med Mex. 2016;152:102-5)

Corresponding author: José Miguel Torres Martel, jmiguelmex@hotmail.com

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Introduction

The anomalous origin of one pulmonary artery branch from the ascending aorta is an infrequent anomaly that occurs in 0.05% of patients with congenital heart disease¹. One of the lungs is irrigated by the aorta, whereas the other is perfused by the main pulmonary artery, in the presence of two semilunar valves². Approximately 40% of the cases occur associated with other cardiovascular anomalies such as aortopulmonary window, which increases the risk for complications³. It has to be early diagnosed in order to enable opportune surgical repair, due to the high risk for the development of irreversible pulmonary disease⁴. If not corrected at an early age, survival reported at one year of life can be 30% or lower⁵. In 1982, Berry et al. described the

association between the aortic origin of the right pulmonary artery, distal aortopulmonary septal defect, intact interventricular septum, patent ductus arteriosus and aortic isthmus interruption or coarctation in 5 patients, and suggested this could be a syndrome, rather than a mere coincidence⁶.

The case of a patient with anomalous origin of one pulmonary artery branch from the ascending aorta and aortopulmonary window undergoing successful repair in a single surgical procedure is next described.

Report of the case

This is the case of a 3-month-old male infant with the following history: son of a 28-old mother with 4 gestations and 4 births, maternal history without complications; he was born by eutocic delivery, full-term, with a

Correspondence:

*José Miguel Torres-Martel
Departamento de Cardiología Pediátrica
UMAE Hospital de Pediatría
Centro Médico Nacional Siglo XXI, IMSS
Av. Cuauhtémoc, 330
Col. Doctores
C.P. 06725, Ciudad de México, México
E-mail: jmiguelmex@hotmail.com

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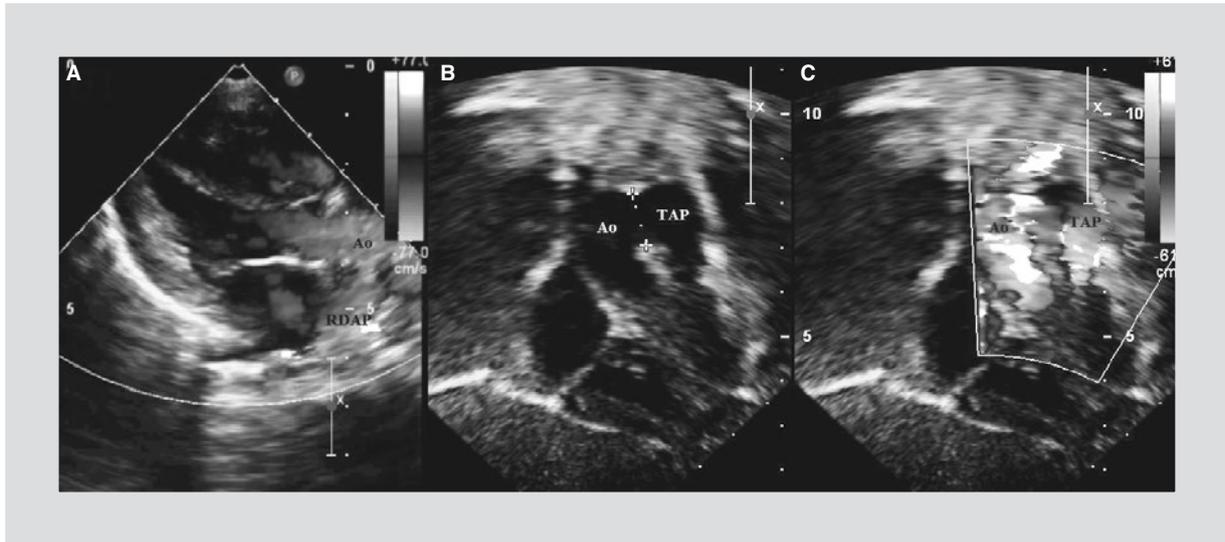


Figure 1. Two-dimensional transthoracic echocardiography of the anomalous origin of the pulmonary artery right branch and aortopulmonary window in an infant. **A:** the right branch of the pulmonary artery emerging from the posterior and proximal wall of the ascending aorta is observed at the long parasternal axis. **B:** the aortopulmonary window between the ascending aorta and the trunk of the pulmonary artery is observed at the subcostal axis. **C:** Color-Doppler showing the aortopulmonary window, the aorta and the trunk of the pulmonary artery. RDAP: right branch of the pulmonary artery; Ao: ascending aorta; TAP: pulmonary artery trunk.

birth weight of 3,700 g, 51-cm length and an 8/9 Apgar. Symptoms started at 26 days of extrauterine life with cyanosis during crying; poor feeding was referred, as well as data consistent with dyspnea, noted by the mother, and therefore he was referred to this hospital for assessment. Physical examination reflected tachypnea, 2/6 holosystolic murmur at the left lower parasternal border, with accentuated pulmonary component of the second heart sound. The liver was palpated 3 cm below the right costal margin. Chest X-ray showed a slightly enlarged cardiac silhouette and increased pulmonary vasculature, predominantly at right hemithorax. Electrocardiogram showed sinus rhythm, QRS axis at $+60^\circ$, with left ventricular forces predominance. Echocardiogram revealed an aortopulmonary window, with a 15 mmHg gradient throughout, pulmonary artery systolic pressure of 75 mmHg; the origin of the pulmonary artery right branch from the ascending aorta was identified (Fig. 1), in addition to patent ductus arteriosus. These anatomical features were confirmed by heart and large vessels angiotomography (Fig. 2).

Surgical repair was managed by a median sternotomy, using hypothermia and cardioplegia; simple ligation of the aortopulmonary window was performed; the anomalous pulmonary left branch was implanted into the pulmonary artery trunk, and arteroplasty was performed with bovine pericardial patch at the aortic defect site left by the vascular button resection. The anterior

portion of the right pulmonary artery was reconstructed with bovine pericardial patch. Ductus arteriosus ligation was simultaneously performed. After reheating, extracorporeal circulation exit was achieved at first attempt. During postoperative evolution, the patient had two infections, one of them caused by *Pseudomonas aeruginosa* and the other by *Staphylococcus hominis*; he received cefepime and vancomycin and the



Figure 2. Axial angiotomography of the heart and large arteries showing the aortopulmonary window, the right branch of pulmonary artery arising from the ascending aorta and the left pulmonary artery arising from the main pulmonary artery. Ao: ascending aorta; TAP: pulmonary artery trunk; RDAP: right branch of the pulmonary artery; RIAP: left branch of the pulmonary artery.

infection remitted. A postoperative echocardiogram was performed, which demonstrated the absence of residual short-circuits at the aortopulmonary window site, and no stenosis was observed at the junction of the right pulmonary branch with the pulmonary trunk; gradient was 4 mmHg, and pulmonary artery systolic pressure, 25 mmHg. The patient was discharged with medical treatment without showing heart failure or pulmonary hypertension signs.

Discussion

In 1868, O. Fraentzel reported the first case of this pathological entity. The anomalous origin of the right pulmonary branch from the ascending aorta is 5-8 times more frequent than that of the left pulmonary artery⁷. Embryologically, it is caused by incomplete migration towards the left of the right sixth aortic arch. According to C. Cucci, it may result from a common arterial trunk septation defect. This septation process starts with the appearance of two edges in the primitive trunk extending cephaladly towards the base of the conus and coinciding with the conotruncal septum. This divides the aorta from the pulmonary trunk of the two outflow tracts. If the right conotruncal edge originates more dorsally than normal from the primitive trunk, the proximal portion of the aortic arch (and hence the pulmonary branch) will arise from the ascending aorta⁸.

There are other cardiovascular anomalies that occur frequently in this condition, such as the following: patent ductus arteriosus (75%), ventricular septal defects, aortopulmonary window, aortic coarctation, aortic arch interruption, atrial septal defect and contralateral pulmonary veins stenosis. Clinical presentation is characterized by early appearance of respiratory distress due to increased pulmonary flow and congestive heart failure and cyanosis when pulmonary pressure and pulmonary vascular resistance are too elevated. The electrocardiogram shows right ventricular hypertrophy, and left ventricular hypertrophy can occur in 25% of patients. Chest X-ray shows cardiomegaly, and increased pulmonary flow, more markedly on the side where the anomalous vessel is found^{9,10}.

Early diagnosis has to be made aided by an echocardiogram: a posterior vessel can be visualized originating from the ascending aorta and perfusing the lung on the parasternal and suprasternal axes. Subcostal views can be useful if the anomalous pulmonary artery arises from the ascending aorta lateral portion but, generally, it has a posterior origin. Cardiac catheterization and angiographies provide additional

information. Prognosis is poor due to a tendency to develop early congestive heart failure, in addition to irreversible pulmonary vascular disease, which can be fatal¹¹.

The most frequently used surgical approach is direct anastomosis of the anomalous pulmonary branch to the main pulmonary artery; however, direct implantation is associated with high residual gradient across the anastomosis site, with high frequency of surgical reintervention. Alternative methods have been proposed when direct implantation is not possible, by performing an end-to-end anastomosis with a synthetic graft, with homograft or autologous pericardial patch interposition, which allows for the anomalous pulmonary branch size to be enlarged, thus avoiding stenosis¹².

Peng et al. have reported a 0% operative mortality; among the long-term complications they mention the presence of anastomotic stenosis (12.5%) and the need to increase the size of the patch (12.5%), without reporting long-term mortality¹³. Prifti et al. report operative mortality of 20%, with 100% survival at long-term follow-up and with no need for re-operations¹⁴.

Conclusions

The case is described of a patient with anomalous origin of the right branch pulmonary artery from the ascending aorta, associated with aortopulmonary window, who underwent an end-to-end anastomosis with placement of a bovine pericardial patch, in addition to ligation of the aortopulmonary window; he did not show residual anastomotic stenosis and had an adequate short-term evolution and marked pulmonary pressure reduction.

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Conflict of interests

There are no conflicts of interests for the development of this publication.

Ethical standards

Informed consent was obtained from the patient's parents for the publication of this case.

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