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Case Report

Anomalous origin of pulmonary branches from the ascending aorta. A report of five cases and review of the literature

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ABSTRACT

The anomalous origin of pulmonary branches is a rare entity where, either the right or the left pulmonary arteries arise from the ascending aorta and where the aortic and pulmonary valves are separated. Clinical manifestations usually appear in the infant or, more rarely, in the newborn and include respiratory distress or congestive heart failure due to increased pulmonary resistance.

The survival rate in an Indian series was 94% with the death of one patient from Fallot tetralogy. We have treated five patients with this congenital heart disease in 20 years; the survival rate was of 80% and in all survival cases, the systolic pressure was significantly decreased. The current treatment of choice, consisting of the anatomical correction with translocation of the right pulmonary artery to the pulmonary trunk, was first performed by Kirkpatrick in 1961.

The aim of this paper is to show the Mexican experience in the diagnosis and treatment of the anomalous origin of pulmonary branches from the ascending aorta.

Conclusion: Early surgery with timely correction of this congenital heart disease improves the prognosis and survival rate of patients, with a reduction in pulmonary hypertension.

<**Learning objective:** This article reports a series of cases of anomalous origin of the pulmonary branches from the ascending aorta. This is a rare congenital anomaly of difficult clinical diagnosis that is often accompanied by other associated anomalies. The diagnosis is based on image studies such as computed tomography and echocardiography and the hemodynamic evaluation is essential to determine a surgical procedure. The surgical treatment consists of the anatomists of the pulmonary branch to the pulmonary trunk.>

Introduction

The anomalous origin of the pulmonary branches is a rare entity where, either the right or the left pulmonary arteries arise from the ascending aorta and where the aortic and pulmonary valves are separated [1]. In 1868, Fraentzel [2] reported the first case of this pathology and thereafter several case series have been described. A timely diagnosis leads to better prognosis for these patients [1,3].

The anomalous origin of the pulmonary artery from the ascending aorta (AOPA) is commonly referred to as "hemitruncus" and accounts for 0.1% of all congenital heart diseases. Kutsche and Van Mierop [4] proposed that abnormal migration of pluripotent cells is important in its development.

The anomalous origin of the right pulmonary artery from the ascending aorta (AORPA) is 5–6 times more frequent than that of

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Table 1 Series of cases of anomalous origin of the pulmonary artery in Mexico.

Age	Diagnosis	Complicated anomalies	Hemodynamics	Physical signs	Surgery technique	Outcome of surgery
6 years	Origin of the right pulmonary branch from the ascending aorta.	Subvalvular stenosis of the aorta due to a fibrous ring.	Initial systolic pulmonary pressure of 60 mmHg.	Systolic murmur at the left parasternal border, an intense S2 was found. Pulse oximetry of 90%. Ross functional class II.	Reconstruction of the pulmonary artery with a bovine pericardial patch, ascending aorta transection with terminal-terminal anastomosis and resection of the fibrous ring.	A favorable evolution, with pulmonary systolic pressure measurement of 35 mmHg.
4 months	Anomalous origin of the right pulmonary branch from the ascending aorta.	Patent ductus arteriosus and interatrial communication.	Initial systolic pulmonary pressure of 74 mmHg.	Diaphoresis and polypnea. A continuous murmur at the left infraclavicular region. Oxygen saturation of 85%. Finally, the patient showed hemodynamic deterioration and was proposed for an emergency cardiac surgery.	Reconstruction of the pulmonary artery with a bovine pericardial patch, ascending aorta transection with terminal-terminal anastomosis, section and suture of the patent ductus arteriosus.	During the postoperative period, the pulmonary systolic pressure was 80 mmHg. In spite of the treatment, the patient's response was not satisfactory and finally he died.
10 years	Anomalous origin of the right pulmonary branch from the ascending aorta.	Patent ductus arteriosus.	Initial systolic pulmonary pressure in the left pulmonary artery was 80 mmHg and in the right one was 60 mmHg.	History of cyanosis since birth. At physical examination an expulsive pulmonary murmur, fixed split S2 with an intense pulmonary component, hepatomegaly.	Reconstruction of the pulmonary artery with a bovine pericardial patch, ascending aorta transection with terminal-terminal anastomosis, section and suture of the patent ductus arteriosus.	The postoperative evolution was satisfactory, with pulmonary systolic pressure measured of 40 mmHg.
1 year	Anomalous origin of the left pulmonary branch from the ascending aorta.	Critical stenosis (gradient of 68 mmHg) of the origin of the left pulmonary artery.	Initial systolic pulmonary pressure in the right pulmonary artery was 50 mmHg.	An intense pulmonary component of the S2.	Reconstruction of the left pulmonary artery with a bovine pericardial patch, ascending aorta transection with terminal-lateral anastomosis. Technique without extracorporeal circulation.	The postoperative evolution was satisfactory, without complications. The pulmonary systolic pressure was of 30 mmHg.
7 years	Anomalous origin of the left pulmonary branch from the ascending aorta.		Initial systolic pulmonary pressure was 90 mmHg.	Unspecific chest pain and effort dyspnea. Systolic murmur in the middle region of the chest with an intense pulmonary component of S2.	Reconstruction of the left pulmonary artery with a bovine pericardial patch, ascending aorta transection with terminal-terminal anastomosis. Technique without extracorporeal circulation.	The postoperative evolution was satisfactory, without complications. The pulmonary systolic pressure was 35 mmHg.

the left pulmonary artery [5]. The anomalous pulmonary artery originates from the wall of the ascending aorta, 5–30 mm above the ventricle arterial junction [6].

The lung connected to the normally arising PA receives the entire cardiac output from the right ventricle, while the other lung is exposed to both pressure and volume overload due to unrestricted shunting from the aorta [3]. The vasoconstrictive reaction due to the abnormal state of the vascular bed on the abnormally connected lung is responsible for the development of pulmonary hypertension.

The first successful surgical correction was performed by Armer et al. [7] in 1961 using a Dacron graft placed between the right pulmonary artery and the pulmonary trunk. Kirkpatrick et al. [8] performed the first anatomical correction with translocation of the right pulmonary artery to the pulmonary trunk which is currently considered the treatment of choice.

In a 20-year period (1992–2012) we have treated five patients with this diagnosis. The median age of the patients was 6 years with a minimum of 4 months and a maximum of 10 years (Table 1).

Case 1

We treated an asymptomatic 6-year-old male patient, referred with a diagnosis of heart murmur. During the physical examination, a systolic murmur at the left parasternal border and an intense S2 were found. Oxygen saturation of 90% in the right arm, 87% in the right leg, 92% in the left arm, and 88% in the left leg were reported. The electrocardiogram (ECG) showed sinus rhythm, QRS left-axis deviation, hybrid pattern from V1 to V5. Chest X-ray showed moderate cardiomegaly, right pulmonary branch and pulmonary cone dilation, and increased pulmonary flow. The echocardiogram showed AORPA and a subvalvular stenosis of the aorta due to a fibrous ring. By catheterization, a systolic pulmonary pressure of 60 mmHg was reported (Fig. 1a–c). Cardiothoracic surgery was performed in which the reconstruction of the pulmonary artery with a bovine pericardial patch, an ascending aorta transection with terminal–terminal anastomosis and resection of the fibrous ring were performed (Fig. 1d). There was a satisfactory postoperative evolution. During the follow-up of the patient, the pulmonary systolic pressure measured by echocardiography was lowered to 35 mmHg. The patient remains asymptomatic.

Case 2

A 4-month-old male patient with diaphoresis and polypnea was treated. During the physical examination the oxygen saturation was 85%, and a continuous murmur at the left infraclavicular region was heard. The ECG showed a sinus rhythm, QRS right-axis extreme deviation (210°), an electric *situs solitus*, a prominent R in V1, and an important enlargement of the right chambers. The chest X-ray showed cardiomegaly and increased pulmonary flow. The diagnosis of AORPA, an interauricular communication and *patent ductus arteriosus* was made by scan tomography. The echocardiogram showed a systolic pulmonary pressure of 74 mmHg. The