

Aorto-pulmonary Window, a Rare Congenital Heart Disease: Case Report

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

Article Information

DOI: 10.9734/AJPR/2021/v7i1130206

Editor(s):

(1) Dr. Thomaz Rafael Gollop, Universidade de Sao Paulo, Brazil.

Reviewers:

(1) Monali Patole Mukherjee, University Of Health Sciences, India.

(2) Najia Hassan, Sharda University, India.

(3) Vivek Parameswara Sarma, India.

Complete Peer review History: <https://www.sdiarticle4.com/review-history/74146>

Case Report

Received 08 July 2021
Accepted 18 September 2021
Published 23 September 2021

ABSTRACT

Aortopulmonary window (APW) is an uncommon congenital cardiac malformation, accounting for 0.1% of all congenital cardiac diseases. It is a defect between the ascending aorta and the trunk of the pulmonary artery. Such abnormality may occur as an isolated lesion or it can be associated with other cardiac abnormalities in one third to one half of cases. Clinical and hemodynamic presentation of this condition depends on the size of the defect and on the associated lesions. It is usually fatal in infancy or childhood if untreated with the development of irreversible pulmonary hypertension. Once the diagnosis is made, surgery must be performed quickly to avoid irreversible pulmonary vascular disease. We report the case of a 5-month-old infant with APW who was referred to our center by respiratory symptoms and heart murmurs.

Keywords: *Aortopulmonary window; infants; congenital heart defects; pulmonary arterial hypertension.*

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1. INTRODUCTION

Aortopulmonary window (APW) is a rare congenital heart disease resulting from abnormal communication between the proximal aorta and the main pulmonary artery with two separate arterial valves arising from separate subarterial ventricular outflow tracts. This lesion is representing approximately 0.2% to 0.6% of all congenital cardiac diseases [1-3]. Very few cases have been reported in Africa [4-6]. APW may be an isolated anomaly or may be associated with other cardiac malformations in about one half of cases. The clinical presentation depends on the size of the defect itself as well as on any concomitant anomalies [7-9]. Once the diagnosis is made, surgery must be performed quickly to avoid progression to congestive heart failure or pulmonary hypertension [8]. Herein we report the case of a 5-month-old infant who was diagnosed with a type I aorto-pulmonary window. He was able to benefit from an open-heart surgery with excellent outcomes.

2. CASE PRESENTATION

A 5-month-old boy weighing 3 kg was referred to our hospital with a severe dyspnea and cyanosis. He was born to a 32-year-old mother following a full-term and uneventful pregnancy. The initial Apgar rating was 9 with a normal birth weight (3,5kg) and no abnormalities were noted upon

routine pediatric examination. Physical examination on admission revealed an axial hypotonia, a faltering growth and dysmorphic features: scaphocephaly, exorbitism, pretragial diverticula, and polydactyly. Percutaneous arterial oxygen saturation was 90% on room air with signs of respiratory distress, heart rate at 130 beats per minute and blood pressure at 95/55 mmHg. Cardiac auscultation revealed a holosystolic murmur along the left sternal border; however, lung auscultation was normal. Cardiomegaly and pulmonary congestion were seen in chest X-ray. The echocardiograph revealed a type I aorto-pulmonary window (APW), mild dilated right atrium and ventricle, and normal left ventricular function with ejection fraction of 72% (Fig. 1) which was confirmed on a CT angiography of the chest (CTA chest) (Fig. 2). Angiography was not needed because of the high quality of CT imaging.

The patient was operated using a median sternotomy and cardiopulmonary bypass (Fig. 3). He had a good recovery from the operation except pulmonary arterial hypertension managed by nitric oxide (NO) and vasoactive drugs. The echocardiographic check-up showed normal left ventricular function, and no residual aortopulmonary defect. The clinical outcome was favorable with Improvement of respiratory symptoms and a weight gain of 1 kg during the 2nd month's check-up.

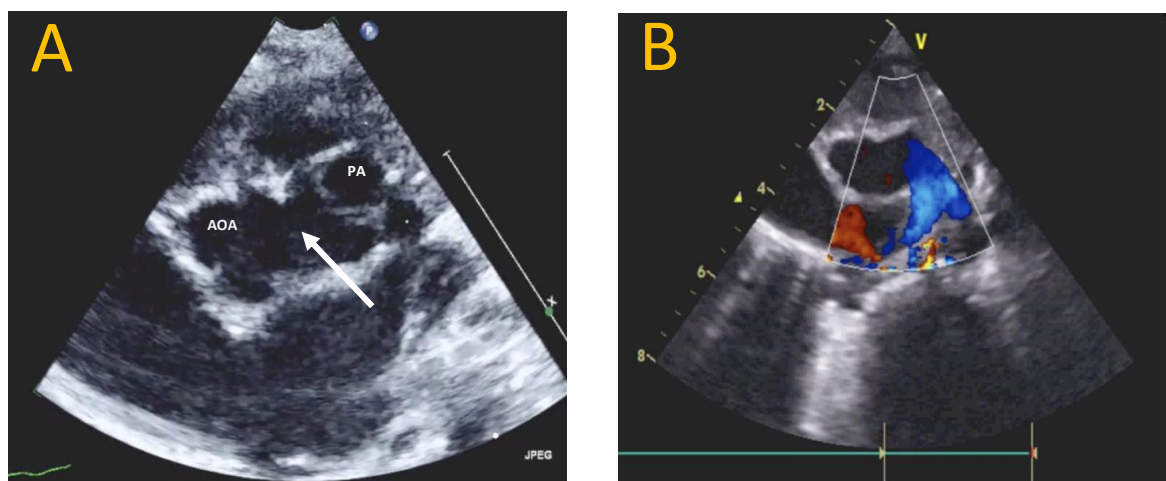


Fig. 1. A: Parasternal short axis echocardiogram of our patient showing the aortopulmonary window between the ascending aorta (AoA) and main pulmonary artery (PA) (white arrow).

B: Unidirectional flow by color Doppler blue flow from the aorta to the pulmonary artery.

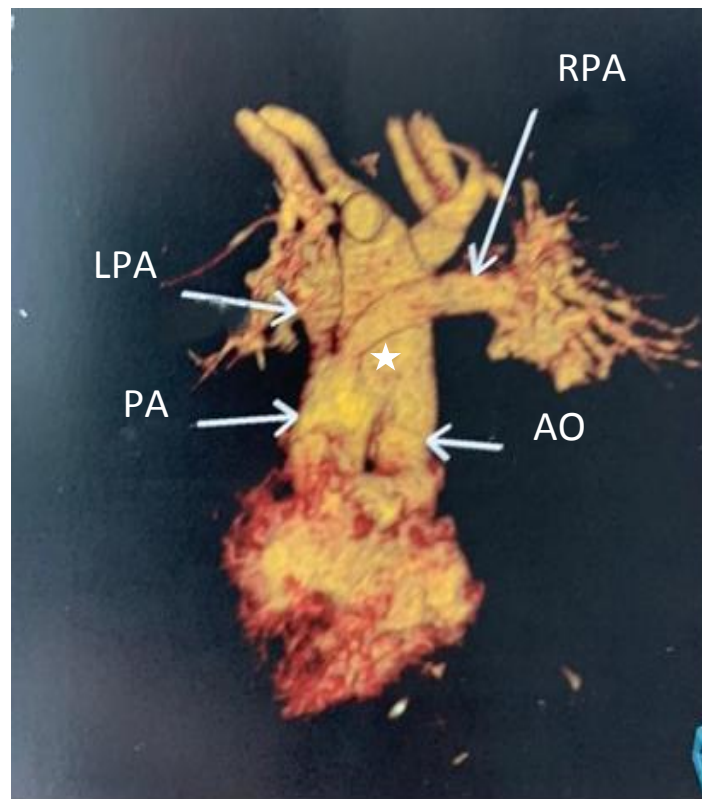


Fig. 2. The three-dimensional CT reconstruction showing the APW (White star).

3. DISCUSSION

Aortopulmonary window (APW) is an uncommon anomaly, occurring in less than 1% of all persons with a congenital heart disease. It consists of a communication between the ascending aorta and the pulmonary trunk [10]. APW can be classified into 3 types: proximal (type I), distal (type II) and total (type III) defects [11]. Type I APW is the most frequent (70%) and the communication is located in the proximal aorta above the sinus of Valsalva midway between the semilunar valves and the pulmonary bifurcation. Type II (25%), the defect is located in the upper portion of the ascending aorta before the aortic branches with aortic origin of right pulmonary artery. Type III is the rarest (5%) and it's characterized by total absence of the aortopulmonary septum resulting from the combination of proximal and distal defects, simulating a common truncus arteriosus (CTA) [1,12]. About 50% of cases of APW are associated with other defects, and this makes its diagnosis more difficult. According to the literature, the most frequently associated anomalies are aortic arch interruption (15-20%) especially type A, patent ductus arteriosus (11%), ventricular septal defect (8%), right aortic arch (7%), anomalies of the coronary arteries

(8%), tetralogy of fallot (5%), subaortic stenosis (3%), and bicuspid aortic valve (3%) [12,13].

Antenatal diagnosis of aortopulmonary window is possible by fetal echocardiography. However, it can be challenging when this cardiac anomaly is associated with other cardiovascular abnormalities [14]. After birth, the clinical presentation of patients with APW is similar to the one of patients with left-to-right shunts, such as patent ductus arteriosus or ventricular septal defect. Most often, the diagnosis is made when a murmur is discovered during the postnatal or during the exploration of heart failure with polypnea, diaphoresis, hypotrophy and breastfeeding difficulties during the first weeks of life. The existence of cyanosis or other clinical signs may be noted in the forms associated with other cardiac abnormalities [8].

Physical examination demonstrates a tachypneic infant with accessory respiratory muscle use. Cardiac examination reveals an enlarged heart, and like patients with patent ductus arteriosus, the pulses are bounding. A systolic murmur can be heard along the left sternal border; however, unlike patients with a patent ductus arteriosus, a diastolic component to the murmur is rare.

Hepatic congestion and hepatomegaly are developed in proportion to the degree of heart failure. Failure to thrive is concomitant with the degree of heart failure [8,15].

Chest x-ray films reveal cardiomegaly and increased pulmonary vascular markings consistent with increased pulmonary blood flow. The electrocardiographic and radiological findings are non-specific, the reason why echocardiography has an important role in diagnosis. This non-invasive investigation allows to identify the morphological features and typing of the defect, exact location, sizing of defect, and associated cardiac anomalies, but in few cases, hemodynamic study using a cardiac catheterization is required for the assessment of pulmonary artery pressure and pulmonary vascular resistance and in some complex cases for defining the anatomy more clearly [1,2,16,17].

The closure of APW is indicated in all patients and it should be performed as soon as possible after diagnosis to avoid irreversible pulmonary hypertension and pulmonary vascular obstructive disease. The surgical results are satisfactory when the APW presents as an isolated defect and when surgery is performed early. several techniques have been described, with or without cardiopulmonary bypass, including simple ligation, division and suturing, transpulmonary approach, transaortic direct closure, and polyester patch closure via the transaortic approach and the sandwich technique [18]. Currently, some cases of percutaneous closure with favorable results have been reported [14,19,20]. Transaortic approach using a median sternotomy and cardiopulmonary bypass was performed for our patient.

4. CONCLUSION

The diagnosis of APW should be considered in any infant with congestive heart failure and a faltering growth, also when the left atrium is very dilated or there is a severe pulmonary hypertension. Echocardiography is mostly enough for diagnosis, but in few cases, cardiac catheterization can be required for the comprehensive evaluation of associated defects and hemodynamic study. The aortopulmonary septum defect should be closed immediately after diagnosis in all patients with APW to prevent congestive heart failure, pulmonary hypertension and progressive of pulmonary vascular disease. Surgical closure still the primary option, but percutaneous therapy may be

a good alternative in some cases, especially when the defect is small and there are no associated cardiac abnormalities. The outcome of these patients is excellent if correction is performed before complications.

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

CONSENT

As per international standard or university standard, patients' written consent has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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Peer-review history:

The peer review history for this paper can be accessed here:
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