

## Aortopulmonary Window: A Two Cases Report and Literature Review

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### Abstract

### Case Report

Aortopulmonary window (APW) is a rare congenital cardiac anomaly. These malformations originate an important left-right shunt with congestive heart failure in the first days or months of life, and early development of severe pulmonary hypertension.<sup>7</sup> Until the present, only about 300 cases have been published; for the most part as isolated reports.<sup>3-5</sup> We report our experience with surgical treatment of APW in 2 consecutive patients in the first years of life. Two infants respectively 4 years old male, 6-month-old male have been referred to our hospital with a medical history of dyspnea, tachypnoea without cyanosis and history of recurrent pulmonary infections. There was no perinatal illness in both infants. An APW diagnostic was performed by echocardiography and type I was identified. The anatomical defects have been repaired surgically using synthetic patch and autologous pericardial patch according to the anatomical type. Additional gestures were also performed. The surgical procedures were uneventful. Post-operative course was favourable in one infant with best post-operative recovery. However one infant deceased in day 10 post-operative due to sepsis chock. In conclusion surgical repair is the gold standard of the treatment of APW with good results. Due to the irreversible pulmonary vascular disease this malformation should be diagnosed early and repaired at the diagnosis.

**Keywords:** Aortopulmonary window, surgical repair, Embryological anomaly.

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## INTRODUCTION

Aortopulmonary window (APW) is a rare congenital cardiac anomaly accounting for 0.1% of all cardiac defects in the autopsy study reported by Abbott [1]. Since it was first described in 1830 [2], until the present, only about 300 cases have been published; for the most part as isolated reports [3-5]. Half of the patients have other more or less complex associated cardiac defects (complex windows) that make the diagnosis difficult.

Different modalities of APW exist and several classifications have been proposed. The classification most often used is that of Mori et al. [6] which divide them in type I, or proximal (70%) (The defect is circular, located in a zone equidistant between the sigmoid valve plane and the pulmonary bifurcation); type II, or distal (25%) (Of spiral form, it affects the trunk and origin of the right pulmonary artery), and type III (5%) (Complete defect of the aortopulmonary septum).

In general, these malformations originate an important left-right shunt with congestive heart failure

in the first days or months of life, and early development of severe pulmonary hypertension [7].

APW has been infrequently reported in patient's younger than 1 year and to the best of our knowledge, surgical treatment of this anomaly has been only sporadically attempted in this age group [7, 8]. In this article, we report our experience with surgical treatment of APW in 2 consecutive patients in the first years of life.

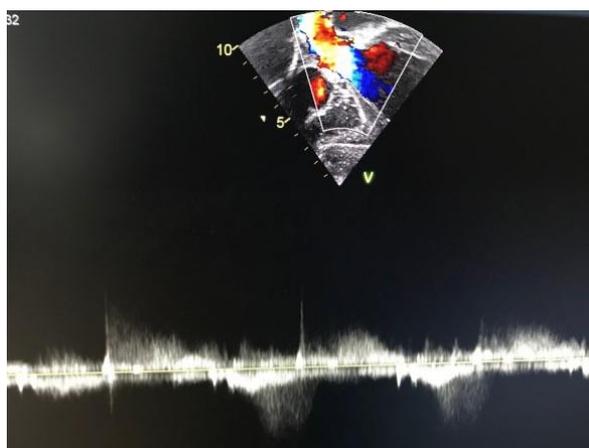
## CASE REPORTS

### Case 1

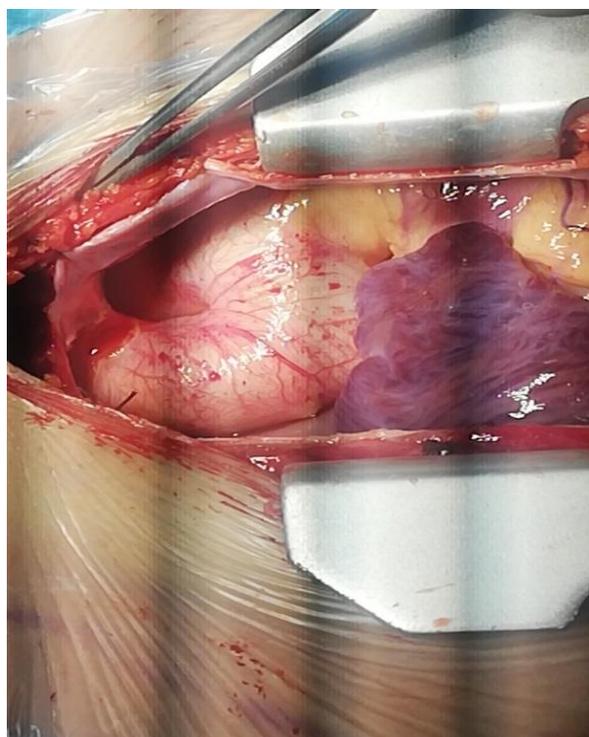
A 4 years old male infant has been referred to our hospital with a medical history of dyspnea and tachypnoea since 6 months old. He also had history of recurrent respiratory infections without cyanosis. He was born with 38-weeks of gestational age and 2.7 kg of body weight without significant perinatal illness. On examination, the blood pressure and the pulse rate were 90/60 mmHg and of 112 beats per minute respectively, the weight was 12 Kgs and the saturation was 89%. The cardiac examination found a systolic murmur of moderate intensity located in the upper left parasternal area.

Echocardiography was performed (Figure 1), and an APW was identified with 17 mm of length and a bidirectional shunt. The pulmonary artery was highly dilated. We could not perform right heart catheterisation.

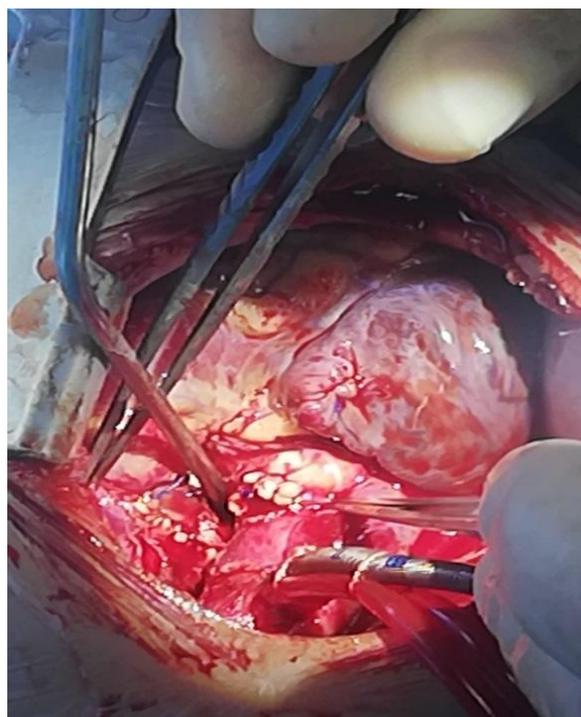
We performed the surgery through median sternotomy with cardiopulmonary bypass without cardiac arrest. After pericardiotomy, we found a type I APW (figure 2) with a patent ductus arteriosus. Through a longitudinal aortotomy, the defect measuring 17 mm of diameter was closed with a 'teflon' patch (figure 3).The patient made an uneventful recovery and remained well 6 months after surgery.



**Fig-1 : Doppler Echocardiographic view of the APW type I**



**Fig-2: Intra-operative view pulmonary artery dilation**



**Fig-3: Intra-operative view of aorta and pulmonary artery defects closure using teflon patch**

## Case 2

A 6-month-old male infant has been referred to our hospital with a medical history of dyspnea since 1 month old, without cyanosis or history of recurrent pulmonary infections. On examination, the blood pressure and the pulse rate were 85/60 mmHg and of 145 beats per minute respectively. The cardiac examination was unremarkable except for sinus tachycardia and breathing sounds was equal bilaterally. Echocardiography was performed, and an APW was identified with 12 mm (figure4) of length and good left ventricle function, no ductus arteriosus or any order cardiac abnormalities associated.

We performed the surgery through median sternotomy with cardiopulmonary bypass without cardiac arrest. After pericardiotomy, a type I APW was identified with some length, from which the anomalous Right coronary artery (RCA) originated clearly. The RCA origin was closer to the aorta than the main pulmonary artery. A small patent ductus arteriosus has been individualized and ligated. After careful dissection and clamping we separated the pulmonary artery from the aorta through the APW in order to have enough tissue for the suture without damaging the RCA orifice. The aortic side of the APW has been closed by a double continuous suture with 5-0 prolene. The main pulmonary artery has been reconstructed by an autologous pericardial patch (figure 5). Postoperatively, the echocardiography showed a good left ventricle function, without signs of hypokinesia. The postoperative outcome has been complicated by a re-intubation at day 2 due to a respiratory distress. The clinical and biological examinations concluded in a

severe pulmonary infection. However the patient died at postoperative day 10 due to septic shock.



**Fig-4: Intra-operative view of the APW**



**Fig-5: Intra-operative view of aorta Defect closure using autologous pericardial patch**

## DISCUSSION

APW can result from incomplete fusion (type I), poor alignment (type II), or total absence (type III) of the right and left conotruncal cushions, which normally complete conotruncalseptation between weeks 5 and 8 of intrauterine life [9]. Until the moment, to our

knowledge, only 3 cases of APW associated with TGV have been reported: [10, 11] the first two died, as did our patient, which is the fourth case communicated in the literature.

Both the type of associated lesion and size of the APW condition the clinical manifestations of patients. The diagnosis must be suspected in cases of early heart failure with signs of significant left-right shunt, such as dilation of the left cavities, particularly the left atrium, and/or functional mitral insufficiency with a morphologically normal valve [12] associated with severe early pulmonary hypertension. The continuous cardiac murmur characteristic of the disease is auscultated in fewer than half of the cases, as occurred in our cases.

The electrocardiographic and radiological findings are non-specific, which is why echocardiography has an important role in diagnosis. The cross-sectional parasternal planes over the aortic valve plane, coronal subcostal plane of both outflow tracts, suprasternal longitudinal and upper parasternal planes are used.

Some false positives are found, particularly when using equipment with scant lateral resolution, because there may be an artificial echo-loss phenomenon (dropout) in the region of the aortopulmonary septum, due to the alignment of the septum in the direction of the lateral resolution of the transducer [13]. To differentiate this phenomenon from true APW, aside from exploring the septum in several planes, some authors resort to the T sign. This sign is nothing other than the greater refringence that the edge of the true defect acquires which is perpendicular to the rest of the septum and adopts a T image.

The Doppler colour study is of inestimable aid and reveals a low-speed bidirectional laminar flow in large, unrestricted defects, with pulmonary hypertension and continuous turbulent flow in the trunk and/or right pulmonary artery. A high-speed flow without pulmonary hypertension is found in small defects.

In our experience, the diagnostic performance of echocardiography was 100%. Negative diagnoses are related mainly to the existence of associated complex anomalies that explain the symptoms of the patient. For this reason, we believe that the diagnosis should be confirmed by cardiac catheterization in all cases.

Due to the rapid development of irreversible pulmonary vascular disease, the malformation should be repaired when diagnosed, preferably before 6 months of age. When total pulmonary vascular resistance at the time of the intervention is  $<8 \text{ U/m}^2$ , the long-term evolution is optimal.

Since the first correction made by gross [14], numerous techniques have been described, with or without ECC, and using a transaortic or transpulmonary patch. The transaortic approach is preferred because it provides better exposure of the window and ostium of the left coronary artery [15]. Matsuki et al. [16] and Meissner [17] later described the use of a pulmonary artery flap, later using autologous pericardium to repair the pulmonary artery. Di Bella et al. [18] also published a similar technique, but without using a patch to repair the pulmonary artery, which was closed using aortic adventitia, which produces excellent results when autologous tissues with normal growth potential are used. Percutaneous closure with favorable results have been reported in specific situations, such as type I, small (3-4 mm) windows not associated with other anomalies, particularly in the origin of the coronary arteries.

Long-term outcome after operation is predominantly dependent on the level of pulmonary resistance at the time of closure of the defect. Regular postoperative control is recommended throughout the growth period in order to detect recurrent stenosis which can be corrected readily, resulting in a good long-term outcome.

## CONCLUSION

Aortopulmonary window (APW) is a rare congenital cardiac anomaly with different anatomical types. Surgical repair is the gold standard of the treatment with good result. Due to the rapid development of irreversible pulmonary vascular disease, the malformation should be repaired when diagnosed.

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