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# Transesophageal Echocardiography in a Distal Aortopulmonary Window

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## ABSTRACT

Aortopulmonary window (APW) is a relatively rare cardiac lesion representing approximately 0.2%–0.4% of all cardiac malformation. It is a cardiac abnormality that results from abnormal communication between the proximal aorta and the main pulmonary artery in the presence of two normally separated aortic and pulmonary valves. In the past, the diagnostic and surgical approach to APW was almost always preceded by cardiac catheterization. With recent advances in noninvasive approach and techniques of two-dimensional echocardiography diagnosis of the defect and associated anomalies are facilitated without a cath study. We report a 4-month-old infant with a distal APW who was referred to our center for surgical repair. We emphasize the usage of transesophageal echocardiography as a valuable intraoperative tool which not only confirms the preoperative diagnosis but also helps in assessing the surgical repair of an APW.

**Key Words:** Aortopulmonary, distal, heart failure, pulmonary, window

## INTRODUCTION

Aortopulmonary window (APW) is a rare entity in congenital heart disease with a prevalence of 0.2%–0.4% of all the cardiac lesions. We present an infant with a distal APW who underwent surgical repair of the same and stressed the importance of intraoperative transesophageal echocardiography (TEE) as an invaluable tool that aids in confirming and assessing the adequacy of surgical repair.

## CASE REPORT

A 4-month-old boy, 4.5 kg, presented to our hospital with a history of recurrent upper and lower respiratory infections, feeding difficulties and excessive forehead sweating, progressive tachypnea, and obvious chest retraction at rest and a soft systolic heart murmur. Cardiomegaly and

pulmonary congestion were seen in chest X-ray. His cardiothoracic ratio was near 65% in chest X-ray [Figure 1].

Transthoracic echocardiography showed increased the pulmonary venous return to the left atrium, dilated all four chambers, large distal APW (15 mm × 11 mm), intact interventricular septum. Cardiac catheterization was deferred as the defect was quite evident on two-dimensional echo with an increased pulmonary venous return to the left atrium and the infant being 4 months in age with chest X-ray showing plethoric lung fields and cardiomegaly, surgical repair was planned.

Intraoperative TEE confirmed the defect [Figures 2-5].

The child was subjected to a surgical repair. Median sternotomy approach was used [Figure 6].

Cardiopulmonary bypass (CPB) was instituted after occluding pulmonary arteries with moderate hypothermia

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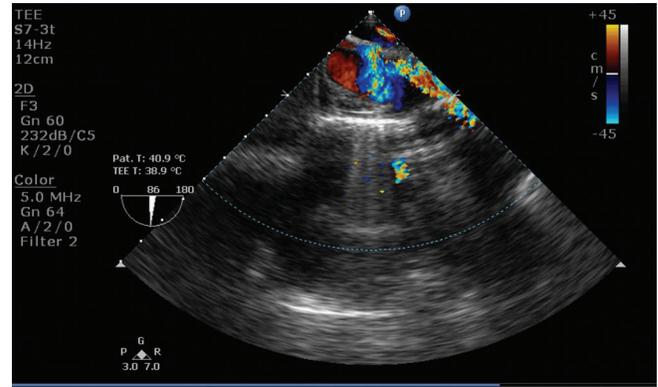
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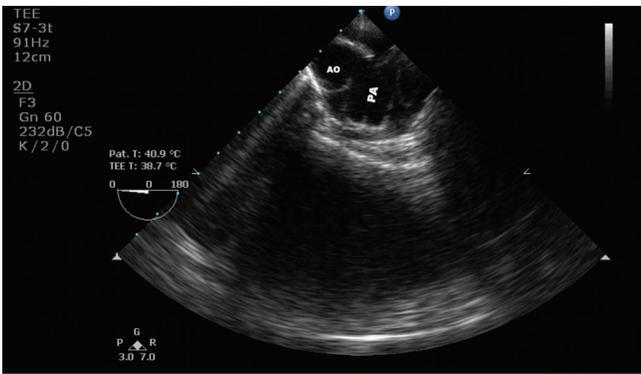
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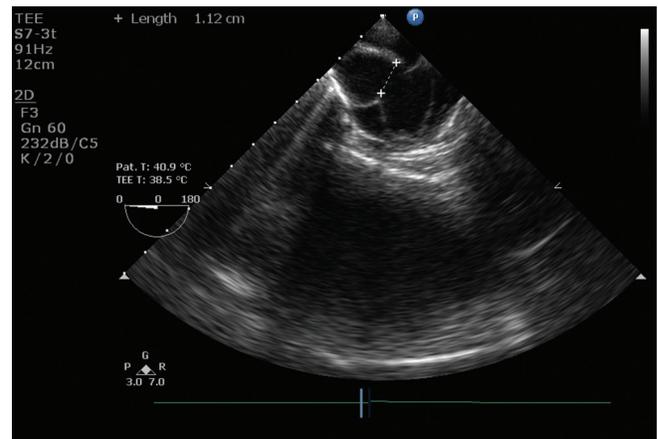
**Figure 1:** Chest X-ray revealing cardiomegaly with dilated pulmonary artery and congested lung fields



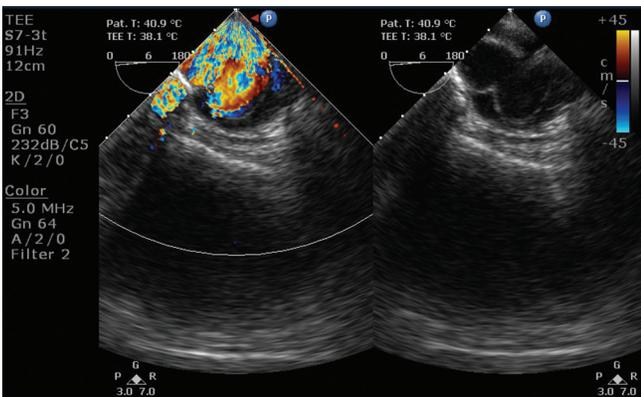
**Figure 2:** Mid-esophageal view at 86° with probe turned to the right revealing a patent foramen ovale shunting left to right



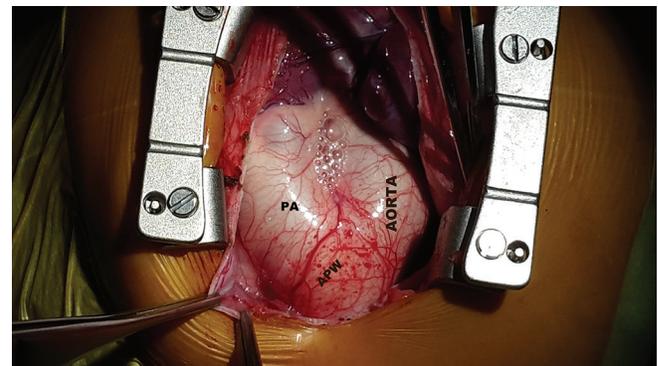
**Figure 3:** Upper esophageal view at 0° revealing the aortopulmonary window extending till the bifurcation of the pulmonary artery



**Figure 4:** Upper esophageal view with a transesophageal echocardiography demonstrating the aortopulmonary window measuring 11.2 mm



**Figure 5:** Color Doppler (left) showing turbulent flow in pulmonary artery across the aortopulmonary window



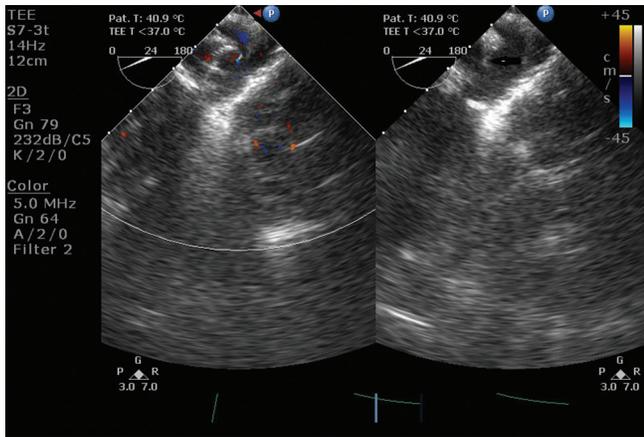
**Figure 6:** Intraoperative snapshot of the aortopulmonary window from head end

using two venous cannulas and a single arterial cannula positioned in the aorta distal to the defect. Aortic cross clamp applied. Antegrade cold blood cardioplegia was injected. The approach for APW repair was transaortic with Gore-Tex patch closure. The patent foramen ovale (PFO) was closed. Postoperative TEE confirmed the adequacy of repair [Figure 7].

The child had an uneventful postoperative course in the hospital with regression of pulmonary hypertension.

## DISCUSSION

An APW is a communication between the pulmonary artery (PA) and the ascending aorta in the presence of two separate semilunar valves. Elliotson first described it in



**Figure 7:** Upper esophageal view at 24° postrepair showing the Gore-Tex patch closure of the defect

1830.<sup>[1]</sup> Mori *et al.* classified the APW as proximal, distal, and total depending on the extent of the lesion.<sup>[2]</sup> Richardson classified APW as simple defects between the ascending aorta and pulmonary trunk (type I), defects extending distally till the bifurcation of PA (type II), and anomalous origin of the right main PA from the ascending aorta with no other aortopulmonary communication (type III).<sup>[3]</sup> Simple APW may be associated with PDA, right aortic arch, and PFO. Complex APW is associated with ventricular septal defect (VSD), interrupted aortic arch, transposition of the great artery, tetralogy of Fallot, or coronary artery anomalies. Our patient had a simple and type II APW.

Transthoracic echocardiography has proved itself as a reliable diagnostic tool. There can be artifactual dropouts with transthoracic echo in the lateral wall of the aorta with normal or aberrant coronaries in the aortic root area which does not happen with a transesophageal echo that can be superior in imaging the defect. There have been reports regarding the role of intraoperative transesophageal echo in complex APW.<sup>[4]</sup> Cardiac catheterization and cineangiography with retrograde aortography are done in infants  $\geq 6$  months of age for evaluating the presence of irreversible pulmonary vascular disease and in case of complex APW for delineating the exact morphology.<sup>[5]</sup>

Closure of APW is indicated in all patients, and it should be performed as soon as possible after diagnosis. Successful repair of an APW was reported from several years ago, and many authors have reported their results. The spectrum of the surgical techniques which evolved with time include simple ligation,<sup>[6]</sup> division and suturing without CPB, division and suturing with CPB, trans-PA closure using CPB,<sup>[7]</sup> trans-window closure (anterior sandwich patch closure), transaortic direct closure;<sup>[8]</sup> to transaortic patch

closure of the defect;<sup>[9]</sup> using CPB and arresting the heart. First successful repair of an APW using CPB was reported in 1957. Putnam and Gross described the transpulmonary approach in 1966.<sup>[6]</sup> Transaortic direct closure of an APW was first reported by Wright *et al.* in 1968.<sup>[8]</sup> In the year 1969, Deverall *et al.* described the first use of a patch to close an APW.<sup>[9]</sup> We had used a Gore-Tex patch to close the defect in our patient by transaortic route, and the PFO was closed.

As the transaortic repair using CPB allows accurate visualization of coronaries arteries and right PA reconstruction using a Dacron patch without distorting either the aorta or the PA, it is the recommended approach for most patients with an APW. The possible occurrence of pulmonary hypertensive crisis should be kept in mind while managing these children in the postoperative period. Recently, isolated successful attempts of transcatheter closure of APW have been reported.<sup>[10-13]</sup>

Intraoperative TEE aids in confirming the diagnosis and is an invaluable tool in detecting additional intracardiac defects which may have been missed by a transthoracic echocardiography. The severe pulmonary hypertension usually associated with APW may mask underlying small shunts in the heart which become evident once we close the APW. TEE is more sensitive in picking up the defects in the prebypass period, as well as postbypass, when it mandates addressing the additional shunts immediately. In addition, TEE in postbypass period confirms the adequacy of repair and detects any residual shunt across the window, direction of PFO shunting indicates the severity of pulmonary arterial hypertension, valvular competence of the semilunar valves, the left ventricular function which can have an afterload mismatch when shunt across into the low resistance pulmonary circulation is suddenly removed, and the delineation of coronaries postrepair in type 1 proximal APW. TEE guides us in the postoperative plan of an APW repair patient, be it simple or complex, the pulmonary hypertension management and left ventricular afterload reduction strategies.

Postoperative mortality depends on the age of the patient at operation, status of preoperative pulmonary vascular disease, and presence of associated intracardiac defects. As the natural history of APW is similar to that of a large VSD or PDA, early recognition is necessary for a successful surgical result before the onset of irreversible pulmonary vascular disease in both simple and complex APW.

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## Conflicts of interest

There are no conflicts of interest.

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