

Aortopulmonary window in adulthood: Surviving at 22 years without intervention or pulmonary vascular disease

Erişkinde aortapulmoner pencere: Girişimsiz ve pulmoner vasküler hastalık gelişmeden 22 yıl yaşam

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Summary– Aortopulmonary window is a rare anomaly, a communication between the ascending aorta and the main pulmonary artery. Prognosis in the absence of correction is poor, with mortality of around 40% in the first year of life. A case of aortopulmonary window without pulmonary vascular disease in adulthood is described in the present report.

Özet– Aortapulmoner pencere, çıkan aorta ve ana pulmoner arter arasında bağlantının olduğu nadir bir anomalidir. Düzeltme yapılmazsa prognozu kötüdür ve yaşamın ilk yılında mortalite %40 civarındadır. Bu yazıda, pulmoner vasküler hastalık olmaksızın erişkin yaşa ulaşan aortapulmoner pencereci bir olgu sunuldu.

Aortopulmonary window (APW) is a communication between the ascending aorta and the main pulmonary artery. It is usually anatomically large, causing a significant left-to-right shunt that results in congestive heart failure or pulmonary hypertension that can develop rapidly into pulmonary vascular disease (PVD). Prognosis in the absence of correction is poor, with mortality of around 40% in the first year of life. APW is a rare anomaly that accounts for approximately 0.2–0.6% of all congenital heart diseases.^[1] Such an abnormality can be isolated, but may be associated with other cardiac abnormalities in 50% of patients, including arch abnormalities such as coarctation of the aorta, interrupted aortic arch, tetralogy of Fallot, or atrial septal defect.^[2]

Although surgical closure is the primary option in APW, percutaneous therapy may be a good alternative in isolated cases with suitable rims.

CASE REPORT

A 22-year-old woman was referred with history of recurrent upper and lower respiratory infections and

progressive shortness of breath upon minimal exertion. Physical examination revealed a continuous murmur at the left lower sternal border. Laboratory examinations were normal. Cardiomegaly and pulmonary congestion were seen on chest x-ray. Trans-thoracic echocardiography showed moderate dilated left atrium and ventricle, severe turbulent flow in the pulmonary artery, and large defect between the the pulmonary artery and the aorta (Figure 1 a, b). A large aortopulmonary window (APW) (Video 1*) and an elevated pulmonary pressure (50% of systemic arterial pressure) were observed in the catheter laboratory. During right heart catheterization, mean pulmonary artery pressure was 55 mmHg, and systemic blood pressure was 116 mmHg. Qp/Qs ratio was 3.3, and average of main pulmonary artery and distal pulmonary artery oxygen saturation was 92%. Pulmonary vascular resistance was 7.2 Wood units, and systemic vascular resistance was 20.6 Wood units; Pulmonary vascular resistance ÷ systemic vascular resistance was 0.349 Wood units. Application of oxygen significant-

Abbreviations:

APW Aortopulmonary window
PVD Pulmonary vascular disease

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ly reduced pulmonary vascular resistance, and surgery was recommended.

Defect was closed with 8-mm pericardial patch by transaortic approach with median sternotomy and cardiopulmonary bypass. The patient recovered well and her respiratory symptoms decreased. Echocardiography was performed and showed normal left ventricular function without residual aortopulmonary defect. Decreased pulmonary artery pressure (mean: 27 mmHg; no residual aortopulmonary defect in right heart catheterization) with increased exercise capacity and functional class were observed in follow-up. No problems arose during postoperative period.

DISCUSSION

APW is a rare anomaly. Clinical features are grounds for suspicion, and diagnosis can be confirmed with echocardiography. Advances in diagnostic and surgical approaches in recent years have led to earlier interventions with successful outcomes. Surgical closure of APW carries low surgical risk. Most centers recommend transaortic patch closure.^[3] Early surgical closure prevents development of PVD and leads to good immediate and long-term outcomes.^[3] Surgical closure or catheter-delivered devices are recommended in all cases and should be performed as soon as possible to prevent irreversible PVD. Today, early mortality following repair of simple APW is low and depends on the presence of associated lesions, particularly interrupted aortic arch. Surgical closure is the traditional treatment for APW, often with transaortic approach, median sternotomy, and cardiopulmonary bypass, which allow for the correction of associated anomalies. Transcatheter closure is also attempted in cases of APW when the defect is relatively small and adequate space for the margin of the device is available.^[4] While transcatheter closure of APW does not appear frequently in the literature due to the relative rarity of defects with good margins and concomitant presence of associated anomalies, successful transcatheter closure using different devices has occasionally been described in case reports.^[4,5] Amplatzer atrial septal or duct occluders (AGA Medical Corp., Golden Valley, MN, USA) may provide good alternatives in adults

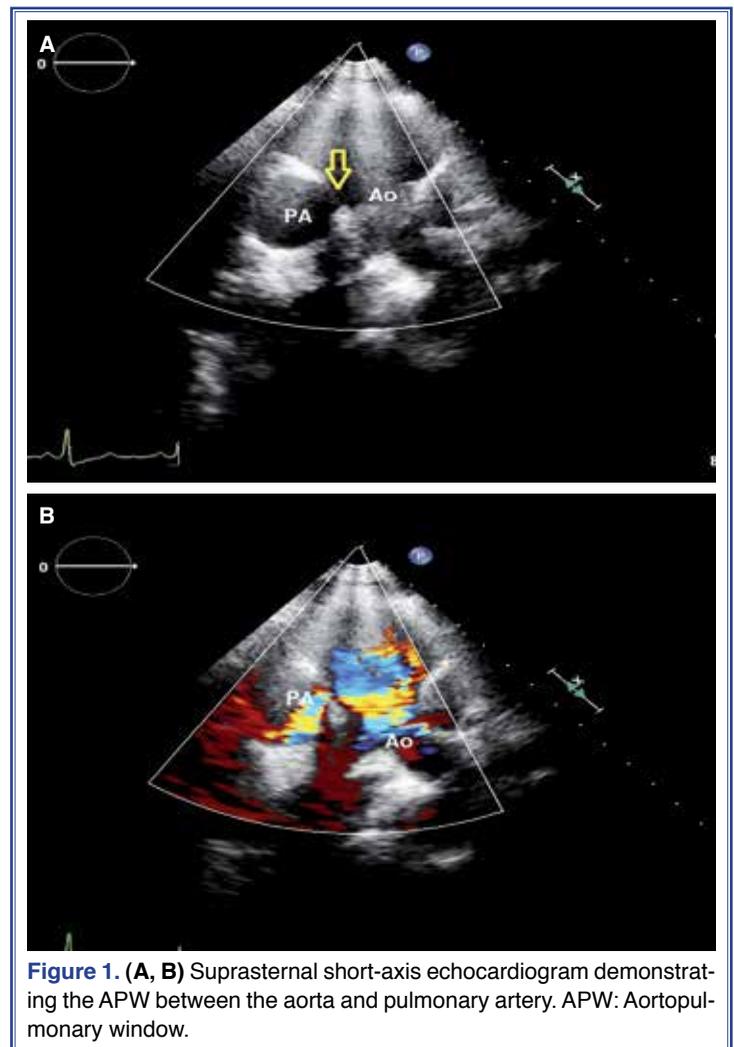


Figure 1. (A, B) Suprasternal short-axis echocardiogram demonstrating the APW between the aorta and pulmonary artery. APW: Aortopulmonary window.

with large defects, though not in infants, as they can protrude into the aorta or the pulmonary artery. In the presence of pulmonary artery hypertension, muscular ventricular septal defect occluder with retention discs on both sides of the defect is recommended to prevent possible embolisation.

Cases of adult patients with untreated APW are very rarely described in the literature. Aggarwal et al. described the surgeries of 6 adult APW patients with reversible pulmonary vascular resistances, and early- or late-term mortality was not reported.^[6] The oldest patient to be described was a man who had reached the age of 32 despite having irreversible PVD (Eisenmenger's syndrome).^[7] That the APW patient had reached adulthood without developing irreversible PVD was a unique feature of the present case.

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**Supplementary video file associated with this article can be found in the online version of the journal.*

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Keywords: Aortopulmonary window; vascular diseases.

Anahtar sözcükler: Aortapulmoner pencere; vasküler hastalıklar.