



CASE REPORT

Staged Surgical Treatment in an Infant with Huge Aortopulmonary Window Presenting with Severe Pulmonary Hypertension and Recurrent Pulmonary Infection

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ABSTRACT

Aortopulmonary window (APW) is a very rare congenital aortic septation defect in which early diagnosis followed by an immediate repair is crucial. Severe pulmonary hypertension may develop even in early infancy as it occurred in our current case. Herein, we report a 4-month-old male who underwent staged surgical closure of a large APW presenting with severe pulmonary hypertension and recurrent pulmonary infection.

Keywords: Congenital Abnormalities, Aortopulmonary Window, Pneumonia, Pulmonary Hypertension, surgical closure

Learning Objective: APW is a very rare congenital aortic septation defect. Early diagnosis followed by timely repair is crucial. APW can present with recurrent pneumonia that is complicated by severe pulmonary hypertension in infants. Although APW is a well-known congenital heart defect, determining the best treatment modality may vary for each patient. Surgical closure with leaving a small residual defect can be a treatment choice particularly in infants who could not tolerate complete closure.

INTRODUCTION

Aortopulmonary window (APW) describes an abnormal connection between the ascending aorta and pulmonary trunk because of inadequate development of the aortopulmonary (AP) septum during the embryonic period. The incidence of APW has been reported as 0.15%–1% (1-5). A significant left-to-right shunt during the earlier weeks of the neonatal period results in congestive heart failure, retarded growth, and pulmonary hypertension. Therefore, early diagnosis and immediate treatment of the disease is crucial.

In this study, we present a 4-month-old male with recurrent pulmonary infection and who was surgically treated along with the data acquired from the multiple cardiac imaging modalities. The surgical management of a large APW presented with severe pulmonary hypertension is discussed in the light of the current literature.

CASE REPORT

A 4-month-old male was referred to our hospital with the diagnosis of lower respiratory tract infection (LRTI) and heart failure. His medical history reported that he had been hospitalized in the second and third month of life because of pneumonia. On admission, his body weight and height were measured as 4.8 kg (<3d percentile) and 58 cm (3d-10th percentiles), respectively. His physical examination revealed heart rate as 168/min, respiratory rate as 98/min, and body temperature as 37°C. His blood pressure was measured as 72/48 mmHg. The pO₂ level in room air and with oxygen delivered at a rate of 5 mL/min was detected to be 89% and 99%, respectively. Grade 3/6 systolic ejection murmur was heard over all cardiac auscultation areas, and capillary refill time was found to be 4 s (normal duration <2 s). The results of complete blood count, CRP test, and biochemical analyses were normal. The chest X-Ray revealed increased pulmonary vascular markings and cardiomegaly (Figure 1). Electrocardiography indicated biventricular hypertrophy. 2-Dimensional (2D) transthoracic echocardiography of the patient revealed left atrioventricular dilatations, moderate mitral, tricuspid insufficiency, and increased right ventricular pressure. Furthermore, apical 5-chamber, parasternal short axis, suprasternal views 2D, and color investigation confirmed the diagnosis of APW (Figures 2a-d, Video 1. See corresponding video/movie images at <http://www.erciyesmedj.com>).

The patient was admitted to the intensive care unit. A treatment regimen, including furosemide, spironolactone, digoxin, and antibiotic, was administered. On the second day, mechanical ventilation support was provided because of CO₂ retention and mixed metabolic acidosis. Because of the bidirectional fashion of the shunt and decreased SpO₂ level that was 80%, the patient was considered to have pulmonary hypertensive crisis. Intravenous infusion of prostacyclin (Ventavis®) and oral bosentan (Tracleer®) were initiated. During the observation, the patient was found to be hypotensive and oliguric; first levosimendan (Simdax®) was administered followed by dopamine and

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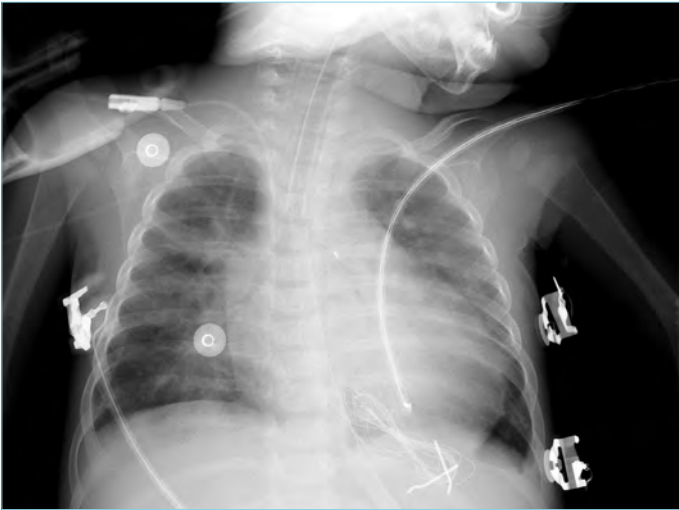


Figure 1. Chest X-Ray admission; apparent cardiomegaly and increased vascularization indicating pulmonary over-circulation

dobutamine. On the fourth day of treatment, prostacyclin was gradually decreased and discontinued. Subsequently, dopamine and dobutamine infusions were gradually decreased and discontinued on the ninth day. He was extubated the following day.

The size of the APW defect was 9 mm, and the distance to the coronary arteries was 10 mm on a multidetector computed tomography (MDCT) scan (Figure 3a, b). The patient was taken to the catheterization room on day 13 for hemodynamic studies and balloon occlusion testing. Hemodynamic studies were performed. The pressures were measured as following: pulmonary trunk, 75/40 (59) mmHg; aorta, 88/45 (68) mmHg; right ventricle, 78/0/4 mmHg; and left ventricle, 94/0/7 mmHg. The Qp/Qs ratio was calculated as 8.5. The pulmonary vascular resistance was found as 6.4 odds/m², whereas pulmonary/systemic resistance ratio was 0.095. On the left oblique projection, during the contrast medium injection in the left ventricle and ascending aorta, a large APW (9 mm), connecting pulmonary trunk and ascendant aorta, was detected (Figure 4, Video 2. See corresponding video/movie images at <http://www.erciyesmedj.com>). During the

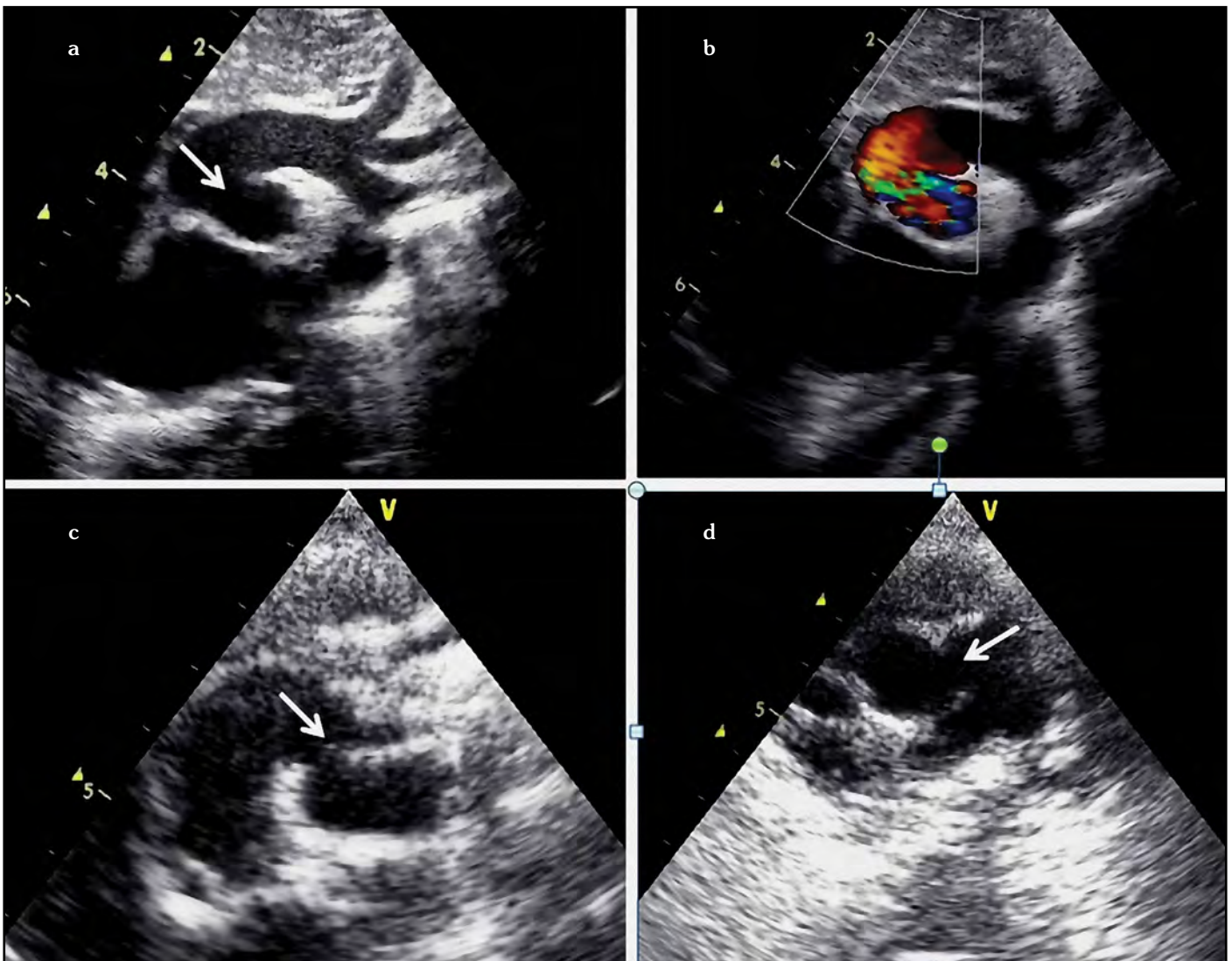


Figure 2. a-d. Large aortopulmonary window marked by arrow 2D (a) and color (b) suprasternal view, modified suprasternal view (c), and parasternal short view (d) in the echocardiography (arrows) (d)

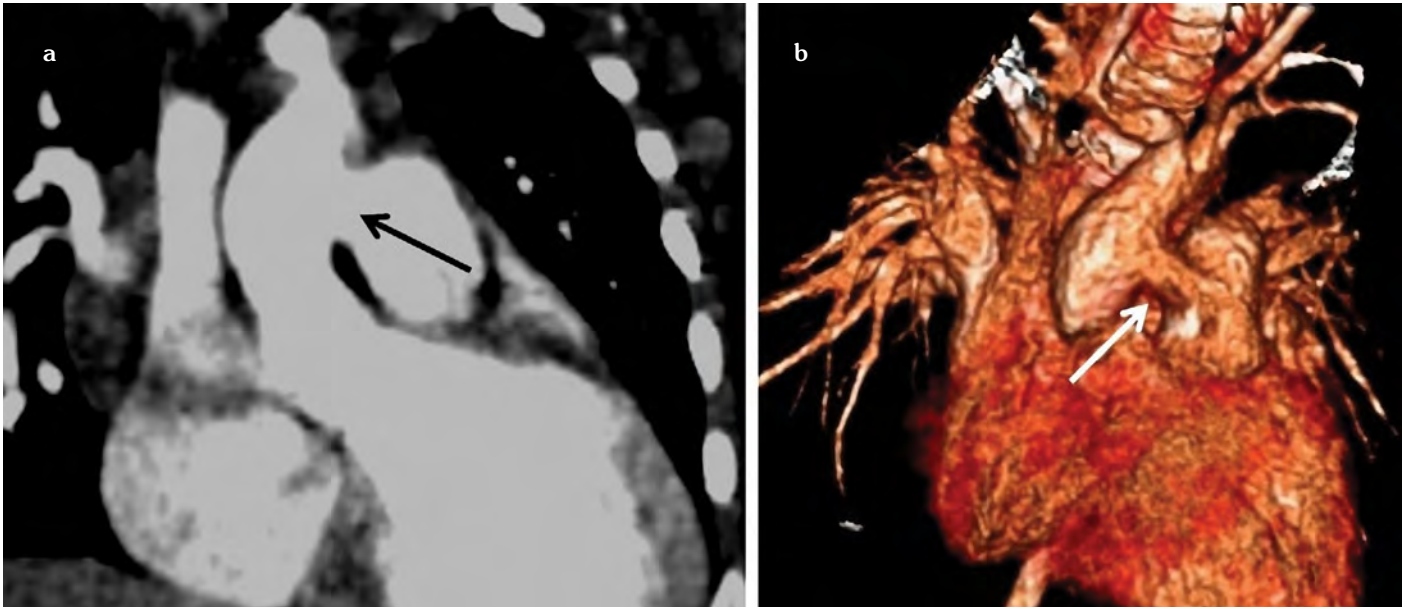


Figure 3. a, b. Coronal contrast enhanced MDCT (a) and volume rendered (b) images revealing APW connecting the pulmonary trunk and ascendant aorta (arrows)

MDCT: multidetector computed tomography; APW: aortopulmonary window

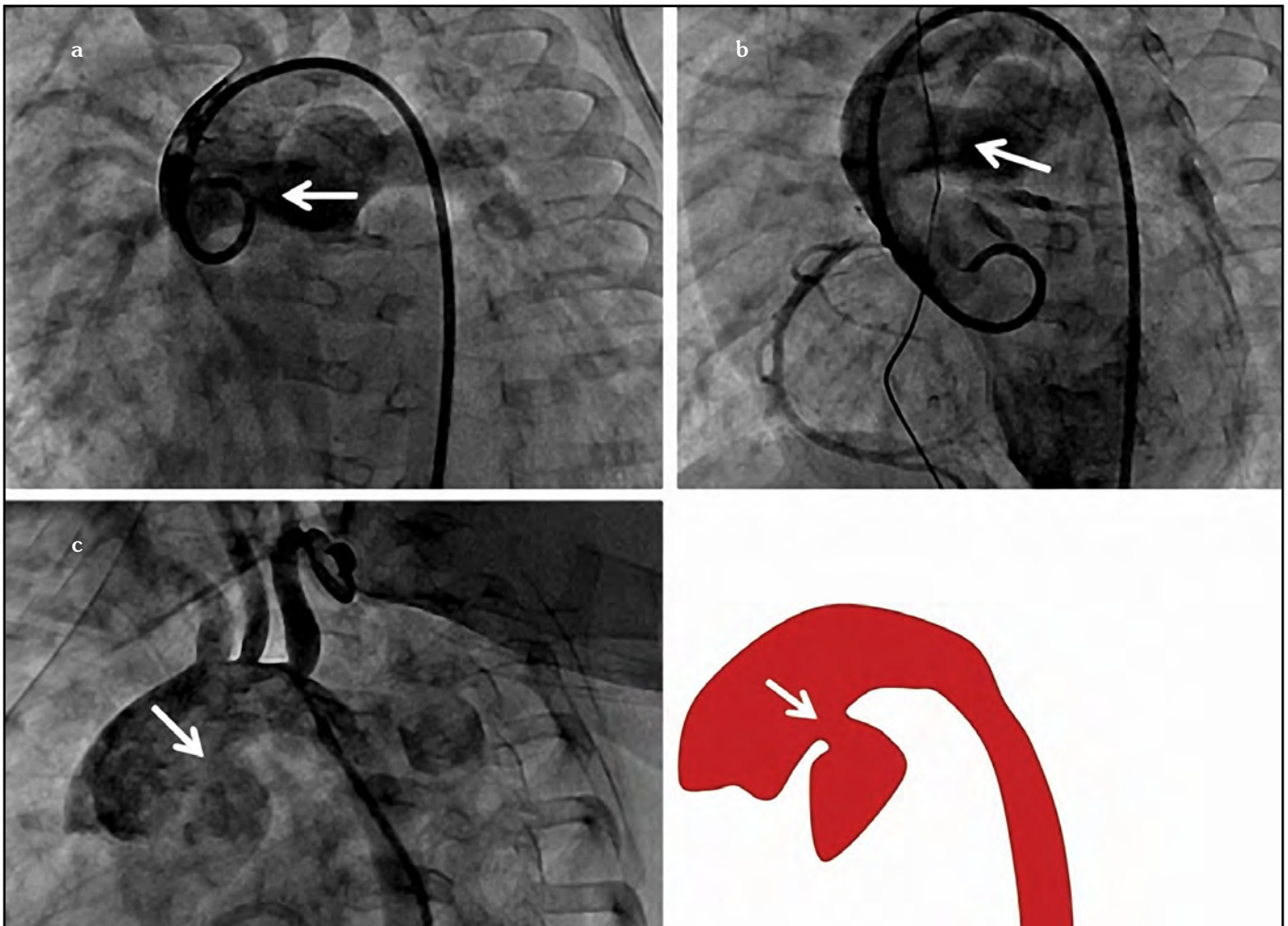


Figure 4. Angiocardiography revealed a huge aortopulmonary window during the ascending aortogram (a), left ventriculogram (b) in the left 40°, and arcus aortogram (c) in the left 60°-20° cranial angulations (arrows)

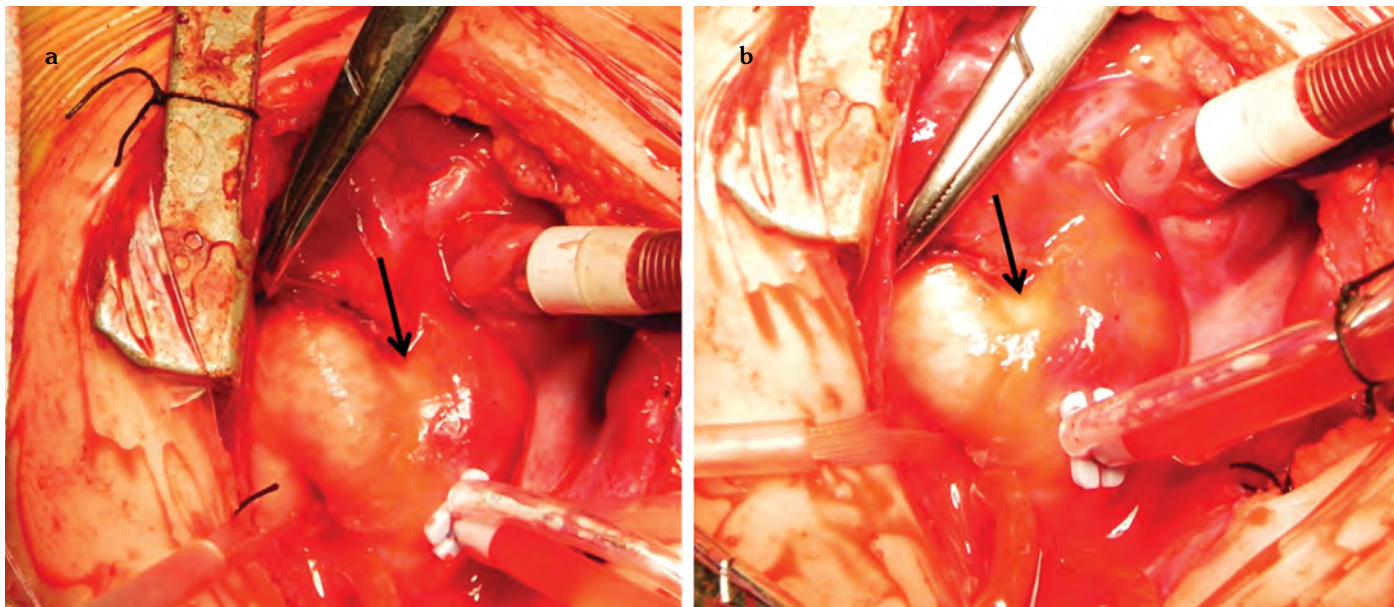


Figure 5. a, b. Defect seems after median sternotomy (arrows)

balloon testing, bradycardia and hypotension appeared. Therefore, we considered that the defect was not suitable for the transcatheter total occlusion.

The following day, APW was closed with a surgical procedure by leaving a small residual defect. APW was observed through a median sternotomy under cardiopulmonary by-pass (Figure 5), and using a polytetrafluoroethylene (PTFE) patch, a residual hole in the middle via aortic punch was left. The patient was extubated on the postoperative second day. A 2.5-mm residual shunt was observed on the patch in the postoperative echocardiography. No complication was observed, and the patient was discharged on the postoperative seventh day.

During the follow-up period, the mitral and tricuspid valve insufficiencies resolved, whereas the right ventricular pressure decreased. A pressure gradient of 24 mmHg was detected by continuous wave Doppler over the residual defect. Currently, the patient is 7-month-old and has been under anti-congestive medication. He has been asymptomatic, and no pneumonia recurred post the surgery. The elective transcatheter closure of the residual defect has been planned.

DISCUSSION

Aortopulmonary window emerges during the embryonic period as an abnormal connection between the ascending aorta and pulmonary trunk over the two normally developed semilunar valves. Currently, there are various classifications of APWs. Mori et al. (2) classified APWs as proximal, distal, and intermediate on the basis of morphological features, whereas Richardson et al. (4) described three main types of APW. A simple APW is in question if it is not accompanied with any other cardiac defect (such as ASD, a small PDA and PFO). If the event of accompanying cardiac defect is more significant, such as transposition of the great arteries, Fallot's tetralogy, and aortic arch abnormalities, APW is considered to be

complex (5). This classification of APW is essential for the treatment of the defect. The features of the defect in our case were consistent with an intermediate, type 1 simple APW; however, contrary to the description of type 1 APW, the size of the defect was fairly large.

Aortopulmonary window results in substantial volume overloading in the pulmonary vascular bed and left ventricle because of the significant left-to-right shunt similar to large PDAs. Therefore, patients with APW develop clinical signs of heart failure, including tachycardia, tachypnea, dyspnea, cardiomegaly, and systemic venous congestion. Growth retardation and pulmonary hypertension may be associated with APW. In case of a delayed diagnosis, adult patients and even infants may be considered as inoperable because of the development of the pulmonary vascular disease and Eisenmenger syndrome (6).

Although the relationship between recurrent LRTI and congenital heart disease with the left-to-right shunt is well known, the relationship between APW and recurrent LRTI is not well defined in medical literature. The diseases with significant left-to-right shunt result in reduced pulmonary compliance and increased respiratory rate. The increased capillary wedge and left atrial pressure developed secondary to the increased pulmonary flow, lead to pulmonary edema, and ultimately to decreased functional residual capacity, ventilation-perfusion imbalance, and hypoxia. Furthermore, increased lung weight secondary to the alveolar edema results in the decreased pulmonary compliance and lung volume. All these factors contribute to the increased respiratory workload and prevalence of LRTI in heart diseases with a left-to-right shunt leading to pulmonary overloading, including APW. Another mechanism that explains the tendency of infections in these patients is the increased requirement of calorie intake and malnutrition secondary to heart failure (7). Therefore, APW should be considered in the differential diagnosis of recurrent LRTI.

As a conventional diagnostic method, echocardiography has been accepted as an adequate diagnostic tool. However, APW may be occasionally overlooked by mistaking it for artificial vascular drop-

out images. Therefore, further evaluation with the imaging modalities, such as CT or magnetic resonance imaging, may be beneficial for detailed assessment of the anatomy of the defect and determination of the distance to the coronary artery in cases that are considered to be candidates for transcatheter closure. Cardiac catheterization that allows the investigation of the pulmonary vascular disease by hemodynamic assessment as well as transcatheter closure of the defect in the same session has been an important diagnostic and therapeutic modality (2, 8-10). In our case, the diagnosis was made by transthoracic echocardiography. AMDCT was performed to assess detailed description of the anatomy and to determine the distance of the defect to the coronary artery in terms of a probable compression related to the device placed in the defect.

The standard treatment is the surgical closure of the defect in infants with symptomatic APW. The term proximal type is used for the defect having an insufficient inferior rim that is located close to the aortic semilunar valves. Distal type describes the defect with insufficient superior rim. In both types of the defect, surgical closure is usually the choice of treatment. However, as in our case, the intermediate type with sufficient superior and inferior rims is the most suitable case for the application of transcatheter closure, which excludes the risk of surgery and cardiopulmonary bypass (3, 8). A variety of Amplatzer devices have been used for antegrade and retrograde (through an AV loop or without creating an AV loop) transcatheter closure. To our knowledge, the transcatheter closure of the largest APW defect (14 mm) was performed by Srivastava and Radha (8) in a 4-year-old boy. Several surgical methods has been described particularly in huge defects, in premature patients, in Type 1 APW, and in confluent-type defects such as closure by the autologous or PTFE patch under extracorporeal circulation and closure with a patch separating each of the two arteries (3, 5, 8-10). In our case, the defect was closed using a PTFE patch via the transaortic approach.

CONCLUSION

Cases presenting with a history of recurrent pulmonary infection should be meticulously examined for a probable congenital heart disease, and APW should be kept in mind in differential diagnosis. Patients, even as young as a 4-month-old, may present with severe pulmonary hypertension and pulmonary hypertensive crisis in the earlier periods of the disease because of a significant left-to-right shunt. Therefore, supportive treatments aimed for respiratory and cardiac stabilization before the closure should be planned in detail

in pediatric intensive care units. The anatomical features of the disorder should be assessed in detail for the appropriate assignment of the patients to either surgery or transcatheter closure.

Video 1. A large aortopulmonary window was shown by modified suprasternal view in transthoracic echocardiography

Video 2. A large aortopulmoner window was detected on the left oblique prejection during the contrast medium injection into ascending aorta

Informed Consent: Written informed consent was obtained from patients who participated in this study.

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Authors' contributions: Conceived and designed the experiments or case: AÖ, AT, NÜT, BNA. Performed the experiments or case: AÖ, NÜT, BNA, NN. Analyzed the data: AÖ, GK, SD, NN. Wrote the paper: AÖ. All authors have read and approved the final manuscript.

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