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Diagnosis and surgical treatment of aortopulmonary window: Our single-center experience

Aortopulmoner pencerenin tanı ve cerrahi tedavisi: Tek merkez deneyimimiz

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ABSTRACT

Background: In this study, we aimed to report our single-center experience in aortopulmonary window and review clinical signs, symptoms, surgical correction techniques, and long-term outcomes.

Methods: We retrospectively reviewed the medical records of a total of 30 patients who were followed with the diagnosis of aortopulmonary window in our hospital between May 1998 and June 2016. The clinical characteristics of the patients, echocardiographic and angiographic findings, surgical treatment outcomes, and medical problems during follow-up were reviewed.

Results: The most common signs and symptoms were murmur, dyspnea, tachypnea, growth retardation, and signs of congestive heart failure. The mean age at the time of surgery was 8.2±14.4 months (7 days to 60 months). Eighteen patients (60%) had additional congenital cardiac anomalies. Eleven patients had simple congenital heart diseases, and seven patients had complex congenital heart diseases. Four patients were unable to be operated due to Eisenmenger syndrome (n=3) and complex congenital heart disease (n=1). No early or late postoperative death was observed. The mean follow-up was 6.4±4.8 years (range, 5 months to 16 years). In addition to aortopulmonary window repair, an additional cardiac anomaly modifying surgical intervention was corrected in nine patients (34.6%). One patient was reoperated for residual aortopulmonary window and another patient for pulmonary stenosis (valvular, supravalvar) after three years. One of these patients underwent pulmonary balloon valvuloplasty after two years. The reoperation rate was 7.7% (n=2) during follow-up.

Conclusion: Aortopulmonary window is a rare cardiac anomaly which may be overlooked by echocardiographic study, and which is amenable for repair with low-surgical risk. It is, therefore, imperative to diagnose and treat this condition, before pulmonary vascular disease develops.

Keywords: Aortopulmonary window; congenital heart disease; pulmonary vascular disease; surgical treatment.

ÖZ

Amaç: Bu çalışmada aortopulmoner pencereye ilişkin tek merkezli deneyimimiz sunuldu ve klinik bulgular, belirtiler, cerrahi düzeltme teknikleri ve uzun dönem sonuçlar incelendi.

Çalışma planı: Hastanemizde Mayıs 1998 - Haziran 2016 tarihleri arasında aortopulmoner pencere tanısı ile izlenen toplam 30 hastanın tıbbi kayıtları retrospektif olarak incelendi. Hastaların klinik özellikleri, ekokardiyografik ve anjiyografik bulguları, cerrahi girişim sonuçları ve takip sırasında tıbbi sorunlar değerlendirildi.

Bulgular: En sık görülen bulgu ve belirtiler üfürüm, dispne, taşipne, gelişim geriliği ve konjestif kalp yetmezliği bulguları idi. Cerrahi sırasındaki ortalama yaş 8.2±14.4 ay (dağılım 7 gün - 60 ay) idi. On sekiz (%60) hastaya ek kardiyak anomali eşlik etmekteydi. On bir hastada basit doğuştan kalp hastalığı, yedi hastada kompleks doğuştan kalp hastalığı vardı. Dört hasta Eisenmenger sendromu (n=3) ve kompleks doğumsal kalp hastalığı (n=1) nedeniyle ameliyat edilemedi. Cerrahi sonrası erken veya geç dönemde ölüm gözlenmedi. Ortalama takip süresi 6.4±4.8 yıl (dağılım 5 ay-16 yıl) idi. Aortopulmoner pencere onarımının yanı sıra, dokuz hastada (%34.6) cerrahi girişimi etkileyen ek kardiyak anomali düzeltildi. Üç yıl sonra bir hasta rezidüel aortopulmoner pencere ve bir diğer hasta pulmoner darlık (valvüler, supravalvüler) nedeniyle yeniden ameliyat edildi. Bu hastalardan birine iki yıl sonra pulmoner balon valvüloplasti uygulandı. Takip sırasında tekrar ameliyat oranı %7.7 (n=2) idi.

Sonuç: Aortopulmoner pencere ekokardiyografik çalışma sırasında atlanabilen ve düşük cerrahi risk ile onarımı olanaklı olan nadir bir kardiyak anomalidir. Bu nedenle, pulmoner vasküler hastalık gelişmeden önce tanı konması ve tedavi edilmesi önemlidir.

Anahtar sözcükler: Aortopulmoner pencere; doğuştan kalp hastalığı; pulmoner vasküler hastalık; cerrahi tedavi.

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Aortopulmonary window (APW) is a rare (0.1 to 0.2%) cardiac anomaly which occurs alone or more commonly in combination with other complex congenital heart disorders.[1-7] It is characterized by an abnormal communication between the ascending aorta and pulmonary artery immediately above the two semilunar valves. It develops as a result of a defect of aortopulmonary septation in embryological life.[8-10] Depending on the size of a defect, congestive heart failure, growth retardation, and pulmonary vascular disease (PVD) develops. Diagnosing before developing PVD is life-saving. It is treated by surgical or percutaneous repair before PVD develops. Many studies to date have demonstrated that a defect can be safely repaired by patch closure through transaortic approach.[5,6,11,12] We believe that our study has an important contribution to the existing literature by comprising the largest known series of surgically repaired APW in Turkey, constituting one of the large series in the literature, and representing a single center experience. In our study, we aimed to report admission signs and symptoms, surgical correction techniques, long-term outcomes, and our center's experience in APW.

PATIENTS AND METHODS

The medical data of a total of 30 patients followed for APW at our hospital between May 1998 and June 2016 were retrospectively reviewed. The study protocol was approved by the ethics committee of our university. The preoperative clinical signs and echocardiographic data were analyzed. Indications for catheter angiography were to determine the diagnosis of patients, to evaluate operability in patients with pulmonary hypertension, and to perform vasoreactivity testing, to identify congenital heart defects associated with APW, and to determine treatment modalities. The catheter angiographic data of patients who underwent catheterization were also recorded. Surgical technique and postoperative intensive care data were reviewed. The patients were all followed by echocardiography Additionally, cardiac catheterization and angiography was repeated, if needed, in certain patients. A simple APW was defined as a lesion which was either isolated or accompanied by atrial septal defect (ASD), PDA, or mild-to-moderate valvular regurgitation. A complex APW was defined as a lesion accompanied by more complex lesions such as tetralogy of Fallot, IAA, complete atrioventricular septal defect (CAVSD), or abnormal coronary artery origins. Although several classifications have been defined to date, most commonly used classification is made by Mori et al.[13] It describes three types of aortopulmonary connection: Type 1 is a small defect midway between the semilunar valves and the pulmonary bifurcation; type 2 is a more distal defect, the distal border of which is formed by the pulmonary bifurcation. This type is more commonly associated with aortic origin of the right pulmonary artery; and type 3, a large, confluent defect involving essentially the entire aortopulmonary septum.[13]

Surgical technique

Among patients who were operated on, 17 had APW which was isolated and/or accompanied by simple congenital heart defects. Nine patients had additional congenital heart anomalies affecting the operative course (Table 1). Our youngest patient was operated on at the age of seven days and with a body weight of 2.5 kg. Aortopulmonary window was located proximally (type 1) in 24 patients (Figure 1) and distally (type 3) in two patients. In 34.6% of the patients there were additional cardiac anomalies affecting the course of surgery. All surgically managed patients were operated with cardiopulmonary bypass (CPB) under cardioplegic arrest.

All patients were premedicated with oral midazolam 0.5 mg/kg and hydroxyzine 1 mg/kg. After induction with sevoflurane 8% in oxygen, midazolam 0.1 mg/kg IV, vecuronium 0.15 mg/kg IV, and fentanyl 25 μg/kg IV were administered and sevoflurane was discontinued. Incremental doses of fentanyl 5 μg/kg IV were administered to a total dose of 50 μg/kg IV prior to sternotomy. For anesthesia maintenance, a constant infusion of fentanyl 10 μg/kg/h was started after intubation, and this was continued throughout the procedure. Sevoflurane 0.5% to 1.5% was also administered to assure depth of anesthesia. Vecuronium 0.05 mg/kg IV was repeated during the initiation and rewarming periods of CPB.

After sternotomy, CPB was established with aorto-bicaval or single atrial cannulation. After snaring and controlling the branch pulmonary arteries before perfusion, PDA was ligated and closed in isolated APW and/or concomitant simple congenital heart defects. In complex heart defects concomitant APW, surgical management was modified for these pathologies. The pulmonary branches were closed and cold crystalloid cardioplegia was infused by the aortic root cannula and diastolic cardiac arrest was achieved. The defects were closed with bovine pericardial patches by aortotomy in the listed early in the series. The division of the defect and the primary repair of the aorta and pulmonary artery were performed in one patient. The division of the defect and the patch repair of the aorta and pulmonary artery with two separate patches tailored

Table 1. Clinical characteristics of the patients and cardiac examination

Diagnoses	Gender	Diagnoses age	Surgical age	Current age	Surgery	Sur-Wt (kg)	Type of APW catheter		Qp/Qs	PAP/AoP (mean) mmHg		PVRI/SVRI (IU)		Diagnostic methods	Follow-up time	Outcome
APW	F	3.5Y	3.5Y	18Y	Division repair patch	12	Type 1	+	1.6	56	66	5.8	13.4	Echo, cath	14.5Y	Alive
APW	F	4M	5M	15Y	Division primary suture	5.3	Type 1	+	4.6	53	54	3.3	16.2	Echo, cath	14.5Y	Alive
APW	F	8D	1.5M	4Y	Repair patch	4.6	Type 1	+	4.23	55	68	2.1	18	Echo, cath	4Y	Alive
APW	M	1.5M	3M	9Y	Repair patch	3.7	Type 1	+	4.16	40	50	2.7	21	Echo, cath	9Y	Alive
APW	M	1M	1.5M	3.5Y	Repair patch	4.5	Type 1	+	1.83	54	60	2.6	20.5	Echo, cath	3.5Y	Alive
APW	M	2M	3.5M	2.5Y	Repair patch	4.5	Type 1	+	-	60	52	-	-	Echo, cath	2.4Y	Alive
APW	M	4.5M	5M	3.5Y	Repair patch	5.5	Type 1	-	-	-	-	-	-	Echo	3.5Y	Alive
APW	F	6D	2.5M	6M	Repair patch	3.5	Type 1	+	5.9	44	46	-	-	Echo, cath	6M	Alive
APW	M	4D	1M	13Y	Repair patch	6	Type 1	+	5.3	54	55	0.9	9.2	Echo, cath	3Y	Alive
APW	F	3D	2M	2Y	Repair patch	3.5	Type 1	+	-	-	-	-	-	Echo, cath	2Y	Alive
APW	F	1.5Y	1.5Y	16Y	Repair patch	12	Type 1	+	1.5	27	57	7.8	-	Echo, cath	14.5Y	Alive
APW, ASD	M	15D	1M	3.5Y	Repair patch	3.6	Type 3	+	2.2	50	55	8.2	22	Echo, cath	3.5Y	Alive
APW, ASD	M	5M	5.4M	3Y	Repair patch	5.2	Type 1	-	-	-	-	-	-	Echo	3Y	Alive
APW, ASD	M	1M	1.5M	5.5Y	Repair patch	4	Type 1	+	3.67	50	60	3.7	21	Echo, cath	5.5Y	Alive
APW, ASD	M	6D	7.5M	5.5Y	Repair patch	6	Type 1	+	4.2	47	60	2.5	22.6	Echo, cath	5Y	Alive
APW, MI	F	3M	3.4M	3.5Y	Repair patch	5	Type 1	+	6.14	50	58	2.8	18.4	Echo, cath	3Y	Alive
APW, DSM	F	2.5Y	2.6Y	18Y	Repair patch	14	Type 1	+	1.87	71	81	8.4	19.4	Echo, cath	16Y	Alive
APW, VSD, RRA	F	1M	3.6M	3Y	Repair patch	4.5	Type 1	+	-	42	66	-	-	Echo, cath	3Y	Alive
APW, VSD	M	25D	1M	6M	Repair patch	2.5	Type 1	-	-	-	-	-	-	Echo, CTA	6M	Alive
APW, PDA, PLSVC	F	5D	5M	8Y	Repair patch	5	Type 1	+	6.6	53	68	2.6	21	Echo, cath, MRI	8Y	Alive
APW, PDA, ARCAPA	M	7D	13D	10Y	Repair patch	3.4	Type 1	+	2.3	30	50	0.9	-	Echo, cath	10Y	Alive
APW, ASD, ALCAPA	M	5D	2M	3Y	Repair patch	4.5	Type 1	-	-	-	-	-	-	Echo	3Y	Alive
APW, LPBS, EA	M	2.5M	3M	2.5Y	Repair patch	3.1	Type 1	+	6.9	41	60	2.5	22.7	Echo, cath	4Y	Alive
APW, VSD, ALPB	M	5Y	5Y	14Y	Repair patch	16	Type 1	+	3.56	48	57	3	17.6	Echo, cath	9Y	Alive
APW, IAA, PDA	M	6D	7D	5Y	Repair patch	3.1	Type 1	-	-	-	-	-	-	Echo	5Y	Alive
APW, IAA, PDA	M	3M	3.5M	15Y	Repair patch	4.2	Type 3	+	0.66	48	46	4.7	8.6	Surgery, cath	15Y	Alive
APW, RAI, CAVSD, DOLV, CA, IAA, PDA, MCA	F	1M	-	-	No surgery	-	Type 1	+	-	-	-	-	-	Echo, cath	-	No follow-up
APW, IAA, PDA, ES	F	15Y	-	-	No surgery	-	Type 1	+	1.7	55	-	21.6	-	Echo, cath	-	No follow-up
APW, VSD, PH, ES	F	17Y	-	-	No surgery	-	Type 1	+	3.3	82	80	5.8	18.9	Echo, cath	-	No follow-up
APW, ES	F	8.5Y	-	15.5Y	No surgery	-	Type 1	+	1.26	83	83	31.4	49.6	Echo, cath	7Y	Alive

PAP: Pulmonary arterial pressure; Sur-Wt: Surgical weight; APW: Aortopulmonary window; PVRI: Pulmonary vascular resistance index; SVRI: Systemic vascular resistance index; D: Day; M: Month; IU: International unit; Y: year; ASD: Atrial septal defect; MI: Mitral insufficiency; DSM: Discrete subaortic membrane; VSD: Ventricular septal defect; RRA: Right renal agenesis; CTA: Computed tomography angiography; PDA: Patent ductus arteriosus; PLSVC: Persistent left superior vena cava; MRI: Magnetic resonance imaging; ARCAPA: Anomalous right coronary artery from the pulmonary artery; ALCAPA: Anomalous left coronary artery from the pulmonary artery; LPBS: Stenosis of the left pulmonary branch; EA: Esophageal atresia; ALPB: Absence of left pulmonary branch; IAA: Interrupted aortic arch; RAI: Right atrial isomerism; CAVSD: Complete atrioventricular septal defect; DOLV: Double outlet left ventricle; CA: Common atrium; MCA: Multiple congenital anomaly; ES: Eisenmenger syndrome; PH: Pulmonary hypertension.

for the aorta and the pulmonary artery defects were performed in another patient. For the patients of later in our series who constituted the majority of the study population, the defect was opened from its anterior aspect and closed with a single layer of bovine pericardial patch held in place by a running polypropylene suture. Care was taken to preserve the coronary artery and pulmonary branch origins. In patients with ventricular septal defect (VSD), the defect was closed with a patch after the completion of APW repair. In patients with interrupted aorta, the APW repair followed the completion of aortic reconstruction. In a patient with the left coronary artery originating from the pulmonary artery and having a short intramural course, coronary reconstruction was performed and the defect was closed in a way that the bovine pericardial patch with the left coronary artery remaining on the aortic side.

Besides general measures to avoid pulmonary hypertensive crises, three patients with elevated pulmonary vascular resistance (PVR) were given intravenous iloprost (ILOMEDIN® 20 mcg/1 mL Bayer) began right before weaning from CPB and continued till extubation with a dose titrated 2-8 ngr/kg/min.

No early or late postoperative death was observed. Postoperative transthoracic echocardiographic examination was performed in all patients. The defects were completely closed and the retrograde diastolic flow pattern was eliminated in all, but one patient. No aortic or pulmonary valve stenosis or regurgitation was observed.

Statistical analysis

Statistical analysis was performed using the PASW version 17.0 software (SPSS Inc., Chicago, IL, USA). Descriptive statistics were expressed in mean ± standard deviation (SD), and frequency. A p value less than 0.05 was considered statistically significant.

RESULTS

The mean age at the time of diagnosis was 22.5±52 months (range, 3 days to 17 years). The mean age at the time of surgery was 8.2±14.4 months (range, 7 days to 60 months), and the mean body weight was 5.6±3.2 kg (range, 2.5 to 16 kg). The mean follow-up was 6.4±4.8 years (range, 5 months to 16 years). The most common signs and symptoms were murmur, dyspnea, tachypnea, growth retardation, and signs of congestive heart failure. Eighteen patients (60%) had additional congenital cardiac anomalies. A complex congenital cardiac disease accompanied APW in seven patients (23%) (Table 1). The diagnosis was made by echocardiography in 19 patients; catheter angiography was performed in addition to echocardiography in eight patients, while computed tomography angiography (CTA) was needed in addition to echocardiography in

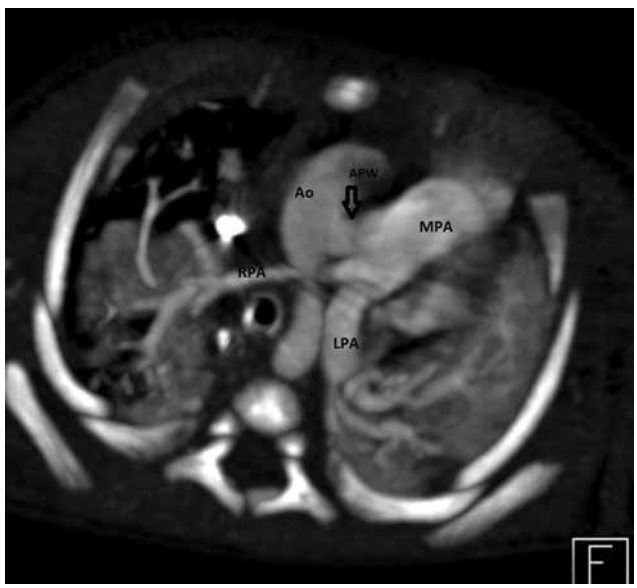

Figure 1. Aortopulmonary window is seen on computed tomography angiography.

Ao: Aorta; APW: Aortopulmonary window; MPA: Main pulmonary artery; LPA: Left pulmonary artery; RPA: Right pulmonary artery.

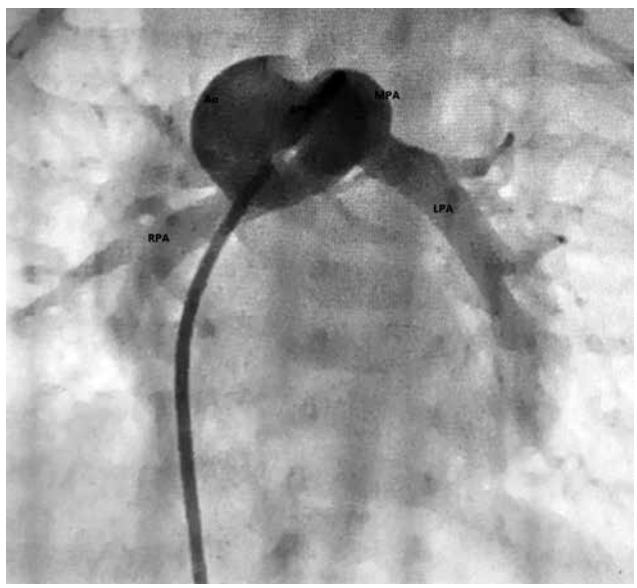

Figure 2. Catheter angiography shows that the catheter passes from the main pulmonary artery to the ascending aorta through the aortopulmonary window. Angiography performed ascending aorta shows that the contrast passes from the aorta to the pulmonary artery through the aortopulmonary window.

Ao: Aorta; APW: Aortopulmonary window; MPA: Main pulmonary artery; LPA: Left pulmonary artery; RPA: Right pulmonary artery.

one patient and magnetic resonance imaging (MRI) was needed in addition to echocardiography in another patient (Figures 1-4). The diagnosis was made at the time of IAA and PDA repair in one patient. Catheter angiography was performed in 25 patients. Among patients undergoing surgical treatment, the mean flow ratio (Qp/Qs) was 3.7 (0.6-6.9); mean pulmonary artery pressure was 48.6 (27-71) mmHg; mean aortic pressure was 58.4 (46-81) mmHg, mean PVR was 3.7 IU (0.9-8); and mean systemic vascular resistance (SVR) was 18.1 IU (8.6-27.8 IU). Three patients were not eligible for repair due to irreversible PVD, and a patient because of an accompanying complex congenital cardiac disease and multiple congenital anomaly (MCA). The ages of the patients at the time of diagnosis were 8.5 years, 15 years, 17 years, 1 month respectively. Among three patients who developed PVD, catheter angiography revealed a bidirectional shunt through APW, a high pulmonary vascular resistance index, and a negative vasoreactivity testing with iloprost. The coronary artery anomaly was corrected in two patients; VSD was closed in three patients; and cardiac pathologies accompanied by interrupted aorta were repaired in two patients. In a patient, moderate mitral regurgitation accompanying isolated APW resolved without any intervention within the first six months after APW repair. A patient was operated on for esophageal atresia accompanying APW and left pulmonary artery stenosis at the age of 10 days. Ten patients were extubated within the first 24 hours, 12 patients within the first 48 hours, and four patients within the first 72 hours. No patient suffered from prolonged intubation and/or pulmonary hypertensive crisis. In our study, two (7.7%) patients were reoperated. One patient was reoperated for residual APW and one patient for pulmonary stenosis (valvular, supravalvular) after three years. The other two interventions were aimed at relieving the additional cardiac pathologies accompanying APW. One patient with isolated APW whose defect was repaired by the division of the defect and the repair of the aorta and pulmonary artery was reoperated three years after the initial surgery. In addition, one patient who was repaired with bovine pericardial patch for isolated APW at 18 months of age developed pulmonary stenosis that was reconstructed with a patch three years after the initial surgery. Subsequent surgical interventions for these two patients were associated with primary surgery, while reoperation for other conditions was associated with additional cardiac anomalies accompanied by APW. One patient who underwent patch repair for APW and VSD developed valvular and subvalvular pulmonary stenosis. This patient underwent pulmonary valvuloplasty 14 months and right ventricular outflow tract reconstruction 20 months after the initial operation. One patient was reoperated for ascending aortic stenosis 13 years after APW, IAA, PDA repair at the age of 3.5 months. Other patients were free from any problems during follow-up.

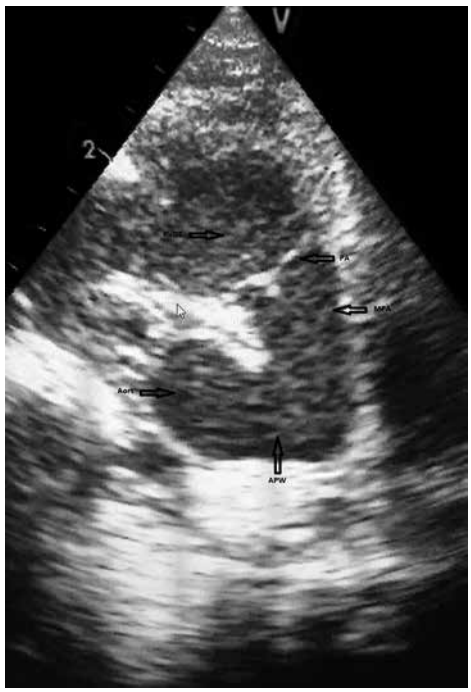

Figure 3. Aortopulmonary window showing in parasternal short axis image on transthoracic echocardiographic examination.

Ao: Aorta; APW: Aortopulmonary window; MPA: Main Pulmonary artery; RVOT: Right ventricular outflow tract; PA: Pulmonary annulus.

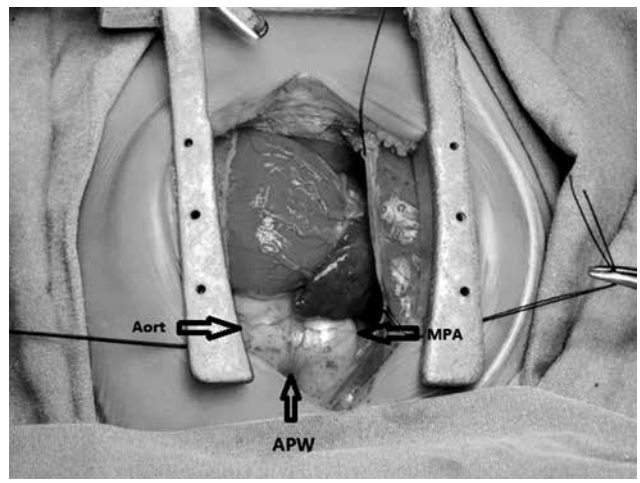

Figure 4. Aortopulmonary window is seen during surgery.

APW: Aortopulmonary window; MPA: Main Pulmonary artery.

DISCUSSION

Aortopulmonary septum is formed by the development and fusion of bilateral truncal cushions. Hence, truncus arteriosus is divided into two arterial structures. During embryological development, the interruption of the development of distal aorto-pulmonary septum due to developmental anomalies of the right and left conotruncal structures gives rise to APW.[8-10] High systemic-to-pulmonary shunt at the arterial level is the basic mechanism underlying the pathophysiological changes.[8-10] In case of APW, increased pulmonary blood flow, congestive heart failure, and pulmonary hypertension are the major pathophysiological issues. Additionally, the condition may pose a risk for infective endocarditis. Congestive heart failure and pulmonary hypertension were also frequently seen in our cases. However, we did not observe any case of infective endocarditis preoperatively and postoperatively. The communication between the aorta and the pulmonary artery is best visualized at the high parasternal short axis on transthoracic echocardiography. In addition to echocardiographic examination, catheter angiography, CTA and MRI provide valuable information for diagnosis. Of the patients, 19 were diagnosed by echocardiographic examination. To confirm the diagnosis, catheter angiography was performed in addition to echocardiography in eight patients, CTA examination in one patient, and MRI examination in one patient. One patient received the diagnosis while being operated on for IAA and PDA. The defect was unable to be identified by echocardiographic examination in three patients with Eisenmenger syndrome. These patients were diagnosed during catheter angiography to identify the etiology of pulmonary hypertension. We believe that the diagnosis of APW can be difficult by echocardiography, and catheter angiography should be performed in patients with elevated pulmonary artery pressure and established PVD. Backer and Mavroudis[6] reported a mean preoperative PVR of 5.4 U/m^2. In our series, the mean preoperative PVR and the mean SVR were 3.8 U/m^2 and 18.1 U/m^2, respectively. Additional cardiac anomalies existed in 65% of patients in the study by Naimo et al.,[14] 63% of patients in the study by Backer and Mavroudis[6] and 61% in the study by Demir et al.[2] Although an additional cardiac anomaly accompanied APW in 60% of our patients, 23% of these were congenital complex cardiac anomalies (Table 1).

Early surgery or transcatheter treatment should be performed before PVD develops in patients with APW.[1-4] Surgical repair is commonly performed for the treatment of APW with favorable outcomes.[1,15-17] Complications of surgical repair such as aortic or pulmonary artery stenosis, residual APW, and aneurysmal aortic dilatation are possible, albeit rare.[1] Rare cases of percutaneously closed APW have been also reported.[18-24] Complications of percutaneous closure include impaired coronary circulation, aortic or pulmonary valvular insufficiency, aortic or pulmonary artery stenosis, and residual APW.[22] In our clinic, 26 patients were treated surgically. Additionally, other congenital heart defects accompanying APW were also repaired. A mean age at the time of surgery of 40 days was reported by Naimo et al.,[14] 52 days by Chen et al.,[7] 3.6 months by Backer et al.,[6] and 4.3 months by Naik et al.[3] In our series, the mean age at the time of surgery was 8.2±14.4 months (range, 7 days to 60 months). The major reason of the older age at the time of surgery in our series was the delayed diagnosis of our patients.

In the present study, surgical treatment was performed after one year of age in four patients. In these patients, there was no significant time interval between the referral age, diagnosis age, and surgery age (surgery age: 3.5 years, 1.5 years, 2.6 years, and 5 years, respectively). The main reason for the late age of surgical treatment in these patients was the late admission to the hospital in the region where they were located. These patients were directed to surgical treatment after being diagnosed at the center where they were present. Cardiac catheter angiography was performed preoperatively in these four patients. Pulmonary artery pressure, PVR, and SVR were measured for PVD. Besides general measures to avoid pulmonary hypertensive crises, three patients with elevated PVR were given intravenous iloprost (ILOMEDIN® 20 mcg/1 mL Bayer) began right before weaning from CPB and continued till extubation with a dose titrated 2-8 ngr/kg/min. No pulmonary hypertensive crisis was observed in the early postoperative period after surgery.

Many surgical techniques have been developed for the treatment of APW and its variations. Simple ligation is not recommended for the surgical repair of APW. Many studies have described the patch closure of a defect through aorta or pulmonary artery. It is important to clearly demonstrate semilunar valves, coronary artery origins, and pulmonary artery branches that are in close neighborhood of APW to avoid possible surgical complications. Backer and Mavroudis[6] used the division and primary repair technique for the first 16 cases of their series and found a mortality rate of 37%. They used the transaortic patch closure technique in the last six

patients of their series and observed no mortality. They reported no aortic or pulmonary artery stenosis for a mean eight years of surveillance in patients treated by transaortic patch closure. In the same series, a patient was operated at the age of three years when PVR was 11 U/m^2, and developed and died from PVD 26 years after surgery.[6] Naimo et al.,[14] in a 43-patient series, observed a mortality rate of 6.7% for simple APWs and 18% for APWs with additional cardiac anomalies, which were repaired through aorta (86% direct suture, 16% patch closure) or pulmonary artery (51% direct suture, 49% patch closure) under CPB. All of our patients were operated on with CPB and under cardioplegic arrest. Several different methods were used for the first patients of our series; however, in all patients operated on in the last 10 years, the defect was opened from its center and repaired with a single patch without unfavorable outcomes. Division of the defect and primary repair of the aorta and pulmonary artery were performed in a patient. Division of the defect and repair of the aorta and pulmonary artery with a bovine pericardial patch were carried out in another patient. The defect was opened from its anterior aspect and closed with a bovine pericardial patch for the last patients in our series who constituted the majority of the study population. In surgically treated patients, we did not experience pulmonary hypertensive crisis at the early postoperative period. No PVD or death occurred in any of our surgically managed patients during follow-up. The majority of the mortality and morbidity of such patients in the literature have been related to accompanying cardiac defects.

The reoperation rate is low for simple APW.[6,25,26] The reoperation rate has been reported higher (15 to 32%) for APWs accompanied by complex congenital cardiac anomalies.[6,25,27] In a study comprising 43 patients followed for 10 years, Naimo et al.[14] reported a residual APW six months after surgery in a patient; the authors reported reoperation due to the stenosis of pulmonary artery bifurcation six days after the initial surgery in another patient. In the aforementioned study, aortic balloon dilatation was performed 13 years after surgery in a patient with repaired IAA accompanying APW; another patient with repaired tetralogy of Fallot accompanying APW was operated with balloon dilatation of pulmonary valve nine years after surgery.[14] In a study by Backer and Mavroudis[6] one patient was reoperated for subaortic stenosis at the age of 10; a patient was reoperated for residual APW two months after surgery; a patient underwent arcus aorta revision at six months; a patient was reoperated with homograft arch augmentation at nine month; and a patient was operated with pulmonary valve replacement twice, one at the age of two and the other at the age of nine, after tetralogy of Fallot repair accompanying APW. In addition, in a series of 10 cases with a mean follow-up time of 34 months, Chen et al.[7] reported a premature death in a patient and reoperations in three other patients. In our study, two patients (7.7%) were reoperated. A patient was reoperated for residual APW and a patient for pulmonary stenosis (valvular, supravalvular). The other two interventions were aimed at relieving the additional cardiac pathologies accompanying APW.

In conclusion, aortopulmonary window should be definitely considered in the differential diagnosis in patients with congestive heart failure and unexplained pulmonary hypertension. Catheter angiography is necessary in conjunction with echocardiography, particularly for patients with unexplained left ventricular dilatation and equalized pulmonary and systemic pressures. Echocardiographic examination is usually sufficient for diagnosing isolated APW in patients younger than six months. In case of accompanying complex pathologies, catheter angiography and thoracic computed tomography angiography may be needed, in addition to echocardiography, for a detailed preoperative evaluation.

Based on our study results, we suggest that aortopulmonary window can be safely closed at every age, including newborns without wasting time, as it may lead to congestive heart failure, growth retardation and most importantly pulmonary vascular disease in the early period. In our study, no pulmonary hypertensive crisis and/or any surgical complications in the early postoperative period were observed in our surgical repair patients. In addition, no pulmonary vascular disease or death was observed in any patients after long-term follow-up. As reported in the literature, the majority of recurrent surgical interventions were associated with additional cardiac defects in our patients. However, as this study is retrospective and there are missing data about the patients who were lost to follow-up, further large-scale studies are needed to establish a conclusion.

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