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Pediatric Cardiology

Anomalous origin of one pulmonary artery from the ascending aorta: A rare entity in congenital heart disease

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ABSTRACT

Objective: Anomalous origin of the pulmonary artery from the aorta (AOPA) is a rare cardiac anomaly. This study aimed to define the clinical characteristics, diagnostic features, management, and follow-up of pediatric patients diagnosed with AOPA.

Material and Methods: We retrospectively reviewed children diagnosed with AOPA from 2010 to 2023.

Results: Thirteen children were diagnosed with AOPA over the study period. Seven patients were male (53.8%). At the initial examination, the patients' ages ranged from 11 days to 16 years, and their weights ranged from 3 to 43 kg. The anomalous branch was the right pulmonary artery in 10 patients and the left pulmonary artery in three patients. Five patients underwent catheter angiography, and nine underwent CT. In one patient, the diagnosis was misdiagnosed by the initial echocardiography, and the correct diagnosis was made by CT. Surgical correction of AOPA and associated cardiac abnormalities were performed in five patients. The time of surgery was a median of 2.5 months (range, 27 days–9 months). There were no peri/postoperative in-hospital deaths. Balloon angioplasty was performed in one patient due to significant RPA stenosis in the postoperative 4th year. The median follow-up was 33 months (14–140 months).

Conclusion: AOPA is a rare structural heart disease and can be accompanied by other complex congenital pathologies. Advanced multimodality imaging techniques can be used to not overlook the diagnosis of AOPA. Early diagnosis and prompt surgical repair of AOPA result in excellent survival and avoid the development of pulmonary vascular obstructive disease.

Keywords: Anomalous origin of the pulmonary artery from the aorta, echocardiography, computerized tomography, pulmonary hypertension.

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Bir pulmoner arterin asendan aortadan anormal çıkışı: Nadir bir konjenital kalp hastalığı

ÖZET

Amaç: Pulmoner arterin aorttan anormal çıkışı (AÇPA) nadir görülen bir kalp anomalisidir. Bu çalışma, AÇPA tanısı alan pediatrik hastaların klinik özellikleri, tanısal özellikleri, yönetimi ve izlemini tanımlamayı amaçladı.

Gereç ve Yöntemler: 2010–2023 yılları arasında AÇPA tanısı alan çocuklar retrospektif olarak incelendi.

Bulgular: Çalışma süresi boyunca on üç çocuğa AÇPA teşhisi kondu. Yedi hasta erkekti (%53,8). Hastaların başvuru sırasında yaşları 11 gün ile 16 yaş arasında, ağırlıkları ise 3 ile 43 kg arasında değişmekteydi. Anormal çıkan dal 10 hastada sağ pulmoner arter, üç hastada sol pulmoner arterdi. Beş hastaya kateter anjiyografi ve dokuz hastaya bilgisayarlı tomografi uygulandı. Bir hasta başvuru sırasındaki ekokardiyografik değerlendirmede yanlış tanı almıştı. Hastaya uygulanan bilgisayarlı tomografi ile doğru tanı konuldu. Beş hastada AÇPA ve ilişkili kardiyak anormalliklerin cerrahi olarak düzeltildi. Ameliyat süresi ortanca 2,5 ay (27 gün–9 ay) idi. Ameliyat sırasında/sonrasında hastanede ölüm olmadı. Postoperatif 4. yılda belirgin RPA darlığı nedeniyle bir hastaya balon anjiyoplasti uygulandı. Ortanca takip süresi 33 aydı (14–140 ay).

Tartışma: Pulmoner arterin aorttan anormal çıkışı, nadir görülen bir yapısal kalp hastalığıdır ve diğer doğumsal kardiyak patolojilere eşlik edebilir. AÇPA tanısını gözden kaçırmamak için ileri görüntüleme teknikleri kullanılabilir. AÇPA'nın erken teşhisi ve hızlı cerrahi onarımı, mükemmel sağkalım ile sonuçlanır ve pulmoner vasküler obstrüktif hastalık gelişimini önler.

Anahtar Kelimeler: Bilgisayarlı tomografi; ekokardiyografi; pulmoner arterin aorttan anormal çıkışı; pulmoner hipertansiyon.

INTRODUCTION

Anomalous origin of the pulmonary artery from the aorta (AOPA) (also known as hemitruncus arteriosus) is a rare cardiac anomaly accounting for approximately 0.1% of congenital heart disease (CHD) (1). The anomalous origin of the right pulmonary artery (AORPA) is more common than the left one and makes up 70–80% of these cases (2, 3). AOPA can be isolated or frequently accompanied by other cardiac malformations (3). Current ontogenetic theories suggest a partial or complete developmental failure of the fifth and sixth arches or the neural crest cells (4). This anomaly results in a large left-to-right shunt, with one lung receiving blood at systemic pressure from the aorta, and the other receiving all cardiac output from the right ventricle (5). Therefore, early recognition and intervention can prevent pulmonary vascular obstructive disease.

This study aimed to define the clinical characteristics, diagnostic features, management, and follow-up of pediatric patients diagnosed with AOPA.

MATERIAL AND METHODS

We retrospectively reviewed the clinical data of 13 cases of pulmonary arteries originating from the ascending aorta between 2010 and 2023 in our hospital. The study was planned in accordance with the Declaration of Helsinki after obtaining the required approval from the local ethics committee (November 2022/2022.09-65). The pediatric cardiology, cardiac surgery, and radiology databases were reviewed, and cases of AOPA were noted. Clinical records were analyzed for demographic factors, clinical features, site of the anomalous pulmonary artery, associated cardiac anomalies, cardiac catheterization, computerized tomography (CT), operative procedures, and follow-up. The cardiac morphology and associated anomalies were assessed by echocardiography in all patients. Transthoracic echocardiography (TTE) was performed using an iE33 ultrasound system (Philips Healthcare, Andover, Massachusetts) and an EPIQ 7 ultrasound system (Philips Healthcare, Andover, Massachusetts) with an appropriate transducer. Five patients underwent catheter angiography, and nine underwent CT.

Patients were followed regularly for 3–6 months in the outpatient clinic. Follow-up was defined as the time between presentation and the last admission or death.

Statistical Analysis

The statistical analyses were performed using SPSS 25 software (SPSS Inc., Chicago, IL, USA). Categorical variables were expressed as frequencies and percentages. Normally distributed variables were expressed as mean±standard deviation (SD), while non-normally distributed variables were expressed as median value [interquartile range (IQR)].

RESULTS

Of the 13 patients, seven patients were male (53.8%) and six were female (46.2%). At the initial examination, patients' ages ranged from 11 days to 16 years, and their weights ranged from 3 to 43 kg. Three patients (23%) were admitted over the age of 1 year. The main presentation showed signs of congestive heart failure (dyspnea, tachypnea, poor feeding) in 10 patients (76.9%). Cyanosis was the second most common initial symptom (n=3, 23.1%). Detailed information about the patients is presented in Table 1. CT was used for diagnosis in addition to transthoracic echocardiography in nine patients (Fig. 1). In one patient, the diagnosis was misdiagnosed by the initial echocardiography, and the correct diagnosis was made using CT.

Table 1. Details of patients with anomalous origin of the pulmonary artery from the aorta

Patient	Age at admission	Weight (kg) at admission	Sex	Diagnosis	Associated congenital heart disease	Surgery	Outcome
1	14 years	43	F	AOLPA	VSD, PA, mild AR, PFO	No[1]	Survived
2	11 days	3.5	F	AOLPA	PFO	No	Deceased
3	2 months	4	M	AOLPA	VSD, PFO, LPSVC, LPA stenosis, RPA stenosis	No[2]	Survived
4	1.5 months	4.3	M	AORPA	LPA stenosis	No	Lost to follow-up
5	3 years	12	M	AORPA	VSD, PA, moderate AR	No	Lost to follow-up
6	9 months	6	M	AORPA	Large PDA	Yes	Survived
7	1 month	3	M	AORPA	Tricuspid chordae rupture, TR, small ASD	Yes	Survived
8	1.5 months	3.2	F	AORPA	TOF, right aortic arch, small ASD	Yes	Survived
9	16 days	3.5	M	AORPA	Arcus hypoplasia, small ASD	Yes[3]	Deceased
10	1 months	3	F	AORPA	PFO, moderate MR	Yes	Survived
11	4 months	5.1	F	AORPA	Moderate ASD, large PDA, severe TR	Yes	Survived
12	5 months	4.35	F	AORPA	PFO	Yes	Survived
13	16 years	42	M	AORPA	BAV, moderate AR, dilatation of aortic root and ascending aorta, small ASD	No	Lost to follow-up

1: Negative vasoreactivity; 2: The family did not consent to the surgery; 3: Pulmonary artery banding; AR: Aortic regurgitation; ASD: Atrial septal defect; BAV: Bicuspid aortic valve; F: Female; M: Male; LPSVC: Left persistent superior vena cava; PA: Pulmonary atresia; PDA: Patent ductus arteriosus; PFO: Patent foramen ovale; MR: Mitral regurgitation; TOF: Tetralogy of Fallot; TR: Tricuspid regurgitation; VSD: Ventricular septal defect.

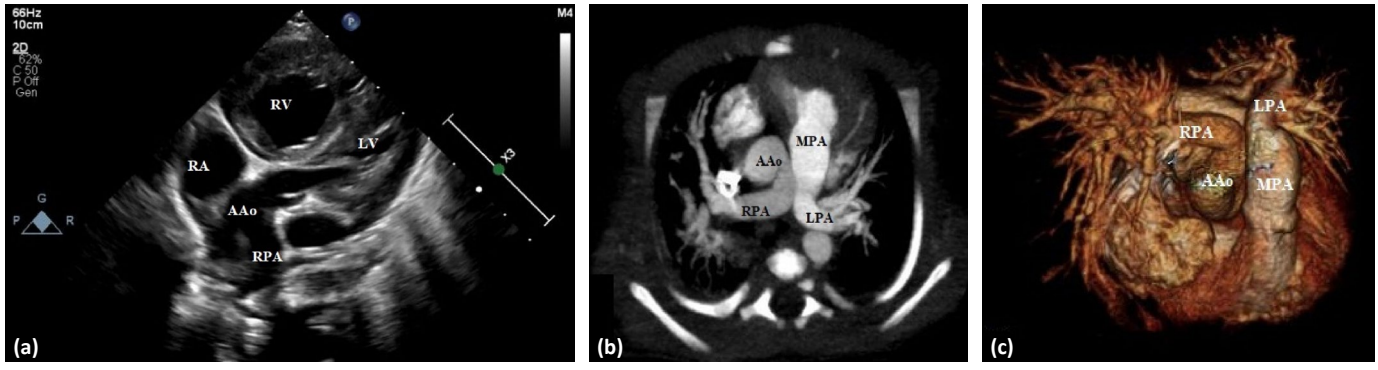

Figure 1. (a) Transthoracic echocardiography from the apical five-chamber view shows the anomalous origin of the right pulmonary artery from the ascending aorta. **(b)** 2D computed tomography shows the anomalous origin of the right pulmonary artery from the aorta in the axial view. The main pulmonary artery continues directly as the left pulmonary artery. **(c)** 3D computed tomography shows the anomalous origin of the right pulmonary artery from the aorta.

Aao: Ascending aorta; LV: Left ventricle; LPA: Left pulmonary artery; MPA: Main pulmonary artery; RA: Right atrium; RPA: Right pulmonary artery; RV: Right ventricle.

The anomalous branch was the right pulmonary artery in 10 patients and the left pulmonary artery in three patients. While the other pulmonary artery was in continuity with the main pulmonary artery originating from the right ventricle in 11 cases, there was no continuity between the right ventricle and the main pulmonary artery in two patients (Patients 1 and 5). Of these, a 14-year-old female patient (Patient 1) was admitted with a complaint of cyanosis and underwent catheter angiography, and was diagnosed with ventricular septal defect (VSD), the absence of main pulmonary artery (MPA) and right pulmonary artery (RPA) (pulmonary atresia), and anomalous origin of the left pulmonary artery (AOLPA). Catheter angiography revealed severe pulmonary hypertension (PH); therefore, anti-PH therapy was initiated. The other two patients (Patients 5 and 13) were diagnosed at admission to the outpatient clinic once, and then they did not consent to further evaluation and were lost to follow-up.

Surgical correction of AOPA and associated cardiac abnormalities were performed in five patients. The time of surgery was a median of 2.5 months (range, 27 days–9 months). A 9-month-

old infant (Patient 6), who had AOPA accompanied by a large patent ductus arteriosus (PDA), suffered from a pulmonary hypertensive crisis in the early postoperative period. He was weaned from mechanical ventilation on postoperative day seven and discharged on the 31st day. In the remaining four patients, the postoperative process was uneventful. There were no peri/postoperative in-hospital deaths. Balloon angioplasty was performed in one patient (Patient 10) due to significant RPA stenosis in the postoperative 4th year. At the last follow-up, there was mild stenosis in the RPA. A five-month-old girl (Patient 12) was admitted to our hospital with a history of RPA re-implantation at 11 days old. After the first operation, an embolectomy was performed due to proximal RPA thrombosis. RPA disconnection was seen on echocardiography upon admission. Catheter angiography was performed, and contrast injection showed a tiny passage to the RPA. Then, a 4.5x12 mm bare-metal coronary stent was deployed into the proximal RPA. At follow-up, a patent RPA stent was demonstrated by TTE.

A 2-month-old boy (Patient 3) diagnosed with AOLPA, VSD, LPA stenosis, RPA stenosis, patent foramen ovale, and left persistent superior vena cava underwent catheter angiography and CT for the assessment of cardiac morphology. Re-implantation of the left pulmonary artery to the MPA was planned; however, the family did not consent to the surgery. Patient 4, who was diagnosed as AORPA by TTE and CT, did not consent to further evaluation and was lost to follow-up.

Two patients (Patients 2 and 9) died due to critical conditions when they were transferred to our hospital from other institutes. Because of their poor status, one patient could not be operated on, and one underwent palliative pulmonary artery banding.

The median follow-up of the study was 33 months (range, 14–140 months).

DISCUSSION

Anomalous origin of one pulmonary artery from the ascending aorta is a rare congenital anomaly (6–8). As the lungs receive total cardiac output from the right ventricle in patients with AOPA, pulmonary hypertension and congestive heart failure occur within one year after birth. If this abnormal circulation is not corrected in a timely manner, early development of pulmonary vascular obstructive disease can be expected. For this reason, early diagnosis and timely surgery are incredibly critical to prevent inoperability of the patients (9, 10). Prifti et al. (11) showed that 63.6% of children undergo surgery within 6 months after birth, mainly in the first month. Unfortunately, three of our patients were diagnosed after age one, which can be defined as a late diagnosis. In the current literature, the mean age at the operation was reported as 17.2 months. Our six patients underwent surgery at a median of 2.5 months, ranging from 27 days to 9 months. However, no matter how early the diagnosis is made, patients can sometimes die without being operated on. Two of our newborn patients died before corrective surgery due to critical conditions when they were transferred to our hospital from other institutes.

The current literature reports that the anomalous origin of the right pulmonary artery is 4–8 times more common than the anomalous origin of the left pulmonary artery. Cho et al. (12) showed that eight patients presented with right AOPA, and four presented with left AOPA. Similar to this data, the anomalous branch was the right pulmonary artery in 10 of our patients and the left in the remaining three patients. Although AORPA is more common, Tetralogy of Fallot (TOF) is most frequently associated with a left-sided anomalous origin (13).

It should be kept in mind that the coexistence of AOPA and structural heart disease may occur. In our study, atrial septal defects were the most common cardiac pathology accompanying AOPA. Similar to our data, Dong et al. (14) reported that 33 of 52 cases were associated with patent ductus arteriosus, and 20 cases were associated with atrial septal defect. In the study reported by Cho et al. (12), they had two patients with absent pulmonary valve syndrome, two with arch hypoplasia, and one with MAPCA on the contralateral side. Although other structural heart diseases, such as ventricular septal defect and aortic abnormalities, are seen together with AOPA, complex congenital cardiac diseases can also be encountered. In these patient groups, AOPA can easily be overlooked when evaluating other components of complex heart disease with echocardiography. Therefore, it may be necessary to perform advanced imaging methods such as CT or cardiac MRI. Cho et al. (12) indicated that, more recently, they used CT angiograms, which are being used to image the branch pulmonary artery and confirm suspected cases in their daily practice. One of our patients was initially misdiagnosed as having a transposition of the great arteries with a posterior aorta during the first echocardiographic evaluation. AOPA was diagnosed after assessment by a more experienced pediatric cardiologist and confirmed by cardiac CT. In support of this recommendation, Dong et al. (14) reported that three patients were misdiagnosed as CT was not performed before surgery.

In previous reports, hospital mortality in patients with AOPA has been reported in the range of 0 to 21% (15, 16). In our study, none of the five patients who underwent surgery died in the postoperative period. As expected, postoperative pulmonary hypertension crises can be observed in these patient groups. In fact, a postoperative pulmonary hypertension crisis, which was managed medically, was detected in one of our patients. Similar to our data, Cho et al. (12) reported a postoperative pulmonary hypertensive crisis in one of their patients. In the postoperative period, deep sedation and, if necessary, nitric oxide can be used to prevent pulmonary hypertension crises in patients who have undergone surgery for AOPA.

The need for reintervention in patients who have had surgery for AOPA has been reported to be between 12.5% and 36% (17). Although total surgical correction is performed in infancy, reinterventions may be required in patients during follow-up. In our study, two patients underwent cardiac catheterization due to critical branch pulmonary artery stenosis, and stent implantation was performed. Cho et al. (12) reported that one of the

twelve patients required reoperation for the left pulmonary artery 14 years after the initial surgery, and two underwent catheter-based interventions. In the follow-up of this patient group, it can be challenging to evaluate pulmonary arteries with classical echocardiographic windows. To determine the reintervention requirements of these patients, advanced imaging methods should be performed in suspected cases.

Study Limitations

There are several limitations to this study. Our analysis was conducted in a single center and retrospectively with a relatively small number of patients.

CONCLUSION

AOPA is a rare structural heart disease and can be accompanied by other complex congenital pathologies. Advanced multimodality imaging techniques can be used to avoid overlooking the diagnosis of AOPA. Early diagnosis and prompt surgical repair of AOPA result in excellent survival and prevent the development of pulmonary vascular obstructive disease.

Ethics Committee Approval: The Mehmet Akif Ersoy Thoracic and Cardiovascular Surgery Training and Research Hospital Clinical Research Ethics Committee granted approval for this study (date: 08.11.2022, number: 2022.09-65).

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

Conflict of Interest: No conflict of interest was declared by the authors.

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Authorship Contributions: Concept – FSŞ; Design – FSŞ, PA; Supervision – AH, SH; Data collection and/or processing – P.Arslan; Analysis and/or interpretation – FSŞ, PA; Literature review – OY, HK; Writing – FSŞ, SUA; Critical review – PA, AG, SH.

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Çıkar Çatışması: Yazarlar çıkar çatışması bildirmemişlerdir.

Mali Destek: Yazarlar bu çalışma için mali destek almadıklarını beyan etmişlerdir.

Yazarlık Katkıları: Fikir – FSŞ; Tasarım – FSŞ, PA; Denetmele – AH, SH; Veri Toplanması ve/veya İşlemesi – P.Arslan; Analiz ve/veya Yorum – FSŞ, PA; Literatür Taraması – OY, HK; Yazıyı Yazan – FSŞ, SUA; Eleştirel İnceleme – PA, AG, SH.

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