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Partial anomalous pulmonary venous connection: Diagnosis, management, and outcomes

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Abstract

Aim: Partial anomalous pulmonary venous connection (PAPVC) is a rare congenital cardiovascular anomaly characterized by one or more, but not all, pulmonary veins draining into the right atrium. This condition can occur in isolation or in conjunction with other cardiac abnormalities such as an atrial septal defect (ASD). In this study we aimed to investigate the clinical presentation, diagnostic approaches, management options, and outcomes of patients with PAPVC.

Materials and Methods: This retrospective study included 140 patients diagnosed with isolated or ASD-associated PAPVC between January 2010 and December 2022.

Results: The median age at presentation was 5 years (1 month to 39 years). Isolated PAPVC was observed in 36 patients (25.7%), while 104 patients (74.3%) had an atrial septal defect (ASD). In our patient cohort, 119 patients (85%) exhibited partially affected anomalous PVs, while the remaining 21 patients (15%) presented with anomalous pulmonary veins involving the entire lung. Right-sided PVs were involved in 116 patients (82.8%), left-sided PVs in 20 patients (14.3%), and both right and left PVs in 4 patients (2.9%). Malposition of primum septum was detected in 8 patients (5.8%), and scimitar syndrome was present in 7 patients (5%). Surgical treatment was performed in 71.4% (n=100) of the patients, while catheter angiography with vertical vein occlusion was performed in 2 patients with dual drainage.

Conclusion: Despite the infrequency and complexity of PAPVC, timely identification and customized surgical intervention can help avert serious complications. The application of advanced imaging technologies in diagnosis and surgical planning is crucial.

Introduction

Partial anomalous pulmonary venous connection (PAPVC) is a congenital cardiovascular malformation in which one or more pulmonary veins do not drain into the left atrium, as they normally do, but instead drain into the right atrium (RA) or a systemic vein [1, 2]. Accounting for a minor proportion of all congenital heart defects, specifically 0.4% to 0.7%, PAPVC is relatively rare and frequently coexists with other anatomical malformations, notably atrial septal defects (ASDs) [2, 3].

The anatomic heterogeneity of PAPVC is apparent, with the anomalous connection mostly involving the pulmonary veins on the right side, this condition being present in 90% of cases [3]. The combination of sinus venosus type ASD and PAPVC accounts for over 80% of such cases [4]. Notably, instances of PAPVC with an intact interatrial septum are exceedingly uncommon, forming less than 3% of the reported cases [3, 4]. Patients with PAPVC exhibit a left-to-right shunt, leading to increased pulmonary blood flow, an essential diagnostic hallmark of this condition [3, 4].

Clinical presentations can vary widely, ranging from asymptomatic to exhibiting significant symptoms, depending on the extent of the abnormal venous connections and coexisting cardiac abnormalities [3, 4]. Potential manifestations include exertional dyspnea, fatigue, and palpitations, all of which can affect an individual's quality of life. Additionally, long-standing left-to-right shunting may cause pulmonary hypertension (PH) and right-sided heart failure, emphasizing the importance of timely diagnosis

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and appropriate intervention in these patients [4]. Scimitar syndrome, a specific variant of PAPVC with the pulmonary veins draining into the inferior vena cava, typically presents early in infancy, particularly when there is coexisting pulmonary vein (PV) obstruction, which may lead to pulmonary arterial hypertension [1, 2, 4]. However, apart from Scimitar syndrome, the presentation of PAPVC is typically late, similar to the usual progression of ASDs [2, 4].

Given the vast heterogeneity of PAPVC, the surgical approach necessitates a comprehensive and individualized plan, reflecting the unique anatomical landscape of each patient's pulmonary venous connections. The ultimate goal of surgical intervention in PAPVC is to restore the aberrant venous connections, preventing left-to-right shunting and potential consequences such as PH and right-sided heart failure [1, 5].

This study investigated the clinical presentation, diagnostic approaches, management options, and outcomes of patients with PAPVC.

Materials and Methods

Study design and population

This retrospective study included patients diagnosed with isolated or ASD-associated PAPVC between January 2010 and December 2022. The research design for this study was planned according to the principles outlined in the Declaration of Helsinki after obtaining the necessary approval from the local ethics committee (University of Health Sciences, Istanbul Mehmet Akif Thoracic and Cardiovascular Surgery Training and Research Hospital Ethics Committee, November 2022/ 2022.09-64).

Data collection and analysis

Medical records of patients diagnosed with PAPVC were systematically reviewed. Demographic information, clinical details, diagnostic approaches, management modalities, surgical interventions, and follow-up outcome statistics were recorded from the medical files. Patients with PAPVC-related congenital heart disease, apart from secundum and sinus venosus ASD, were omitted from the study.

Diagnostic modalities

All patients in the study underwent comprehensive diagnostic evaluation, including transthoracic echocardiography, computerized tomography (CT), cardiac magnetic resonance imaging (MRI), and cardiac catheterization, as deemed necessary by the treating physicians. The diagnosis of PAPVC was confirmed by at least one of these modalities.

Classification of PAPVC and ASD types

The classification scheme was designed based on the presence and type of ASD as well as the anatomical characteristics and location of the abnormal connections. PAPVC was classified based on the location of the affected pulmonary veins (right-sided, left-sided, or bilateral), the number of pulmonary veins with anomalous connections, and the specific drainage site, including the RA, superior or inferior vena cava, or coronary sinus. Any coexisting cardiac anomalies or syndromes, such as scimitar syndrome, were documented. ASD types were classified based on their anatomical location and characteristics. The categories included secundum ASD, which is characterized by a defect in the central part of the atrial septum, and sinus venosus ASD, which involves a defect near the entry of the superior or inferior vena cava into the RA.

Surgical techniques

Surgical interventions were tailored to each patient's specific anatomy and clinical condition. The surgical techniques employed included direct anastomosis of the anomalous pulmonary veins to the left atrium and intra-atrial baffling. The surgical approach was chosen based on the patient's circumstances.

Follow-up and outcome assessment

Patients were regularly followed up, with clinical evaluations and imaging studies performed as needed to assess their clinical status and identify any potential complications or residual lesions after surgery. The follow-up duration was defined as the time from the initial admission to the date of the last follow-up visit.

Statistical analysis

Descriptive statistics were used to summarize the data, with continuous variables expressed as mean ± standard deviation (SD) or median with range (minimum-maximum), as appropriate. Categorical variables were presented as frequencies and percentages. All statistical analyses were performed using SPSS 17 software (SPSS Inc., Chicago, IL, USA).

Results

Demographic and clinical characteristics

A total of 140 patients were diagnosed with PAPVC at our center. Of these patients, 54.6% (n=77) were male.

Table 1. Demographic and Clinical Characteristics of Patients with PAPVC.

Parameters	Total (n=140)
Male (%)	77 (54.6%)
Age at presentation (Median, Range)	5 years (1 month-39 years)
Weight (Median, Range)	17 kg (2.7– 94 kg)
Recurrent pulmonary infections (%)	12 (8.6%)
Growth retardation (%)	10 (7.1%)
Fatigue (%)	7 (5%)
Palpitations (%)	2 (1.4%)
Isolated PAPVC (%)	36 (25.7%)
PAPVC with ASD (%)	104 (74.3%)
Superior sinus venosus ASD (%)	87 (62%)
Secundum ASD (%)	16 (11.4%)
Superior sinus venosus with secundum ASD (%)	1 (0.7%)

ASD: atrial septal defect; PAPVC: partial anomalous pulmonary venous connection.

Table 2. Anomalies and Diagnostic Techniques of PAPVC.

Anomalies	Total (n=140)
Right-sided PAPVC	
Draining into the SVC (%)	73 (52.2%)
Draining into the right atrium (%)	17 (12.2%)
Draining into the SVC and cavoatrial junction (%)	15 (10.7%)
Draining into the IVC (%)	7 (5%)
Draining into the cavoatrial junction (%)	4 (2.6%)
Left-sided PAPVC	
Draining into the brachiocephalic vein through a vertical vein (%)	14 (10%)
Draining into the coronary sinus (%)	2 (1.4%)
Dual drainage (%)	2 (1.4%)
Bilateral PAPVC	
Draining into the coronary sinus (%)	3 (2.2%)
Draining into both the right SVC and left brachiocephalic vein through a vertical vein (%)	1 (1.4%)
Diagnostic Techniques	
CT scan performed (%)	107 (76.4%)
Catheter angiography performed (%)	41 (29.3%)
Cardiac MRI performed (%)	9 (6.4%)

CT: computerized tomography; IVC: inferior vena cava; MRI: magnetic resonance imaging; PAPVC: partial anomalous pulmonary venous connection; SVC: superior vena cava.

The median age at presentation was five years (1 month to 39 years), and the median weight was 17 kg (2.7– 94 kg). Of these, 12 patients (8.6%) had a history of recurrent pulmonary infections. Ten patients presented with growth retardation, seven reported experiencing fatigue, and two presented with palpitations. Isolated PAPVC was observed in 36 patients (25.7%), while 104 patients (74.3%) had an atrial septal defect (ASD). Among the ASD cases, 16 (11.4%) were of the secundum type, while 87 (62%) had superior sinus venosus defects, and one patient had a secundum and superior sinus venosus defect (Figure 1). Demographic and Clinical Characteristics of Patients are demonstrated in Table 1.

A single PV was anomalously connected in 65 patients (46.4%), two PVs were involved in 63 patients (45%), and three PVs were affected in 12 patients (8.6%). In our patient cohort, 85% (or 119 patients) displayed partially anomalous pulmonary veins, while the other 15% (21 patients) manifested abnormal pulmonary veins affecting the entirety of the lung. In 10 patients (7.1%), all the PVs of the right lung were anomalously connected, while in 11 patients (7.9%), all the PVs of the left lung were affected. Right-sided PVs were involved in 116 patients (82.8%), left-sided PVs in 20 patients (14.3%), and both right and left PVs in 4 patients (2.9%). Malposition of primum septum was detected in 8 patients (5.8%), and scimitar syndrome was present in 7 patients (5%). Figure 2 demonstrates the detailed characteristics of PAPVC.

The most prevalent anomaly observed in this study was right PAPVC draining into the superior vena cava (SVC), accounting for 52.2% of cases (n=73). The second most common type was right PAPVC draining into the RA, found in 12.2% of patients (n=17). It was followed by right PAPVC draining into the SVC concomitant with the cavoatrial junction in 10.7% (n=15) and, left PAPVC draining into the brachiocephalic vein through a vertical vein in 10% of cases (n=14). In left-sided PAPVC cases, PVs were anomalously drained into the coronary sinus in two patients, and dual drainage was seen in two patients. Computed tomography was performed in 107 patients (Figure 3), catheter angiography in 41 patients (Figure 4), and cardiac MRI in 9 patients. Drainage sites of PAPVC and diagnostic techniques are indicated in Table 2.

Management and follow-up

Surgical treatment was performed in 71.4% (n=100) of the patients, while catheter angiography with vertical vein occlusion was performed in 2 patients with dual drainage. Among patients with isolated PAPVC and a Qp/Qs ratio >1.5 in hemodynamic studies, 20 underwent surgical treatment, and four patients awaiting operation. Surgery was performed in 80 PAPVC patients with associated ASD (Figure 5). The median age at the time of surgery was 5.7 years (8 months to 39 years).

During follow-up, no early or late deaths were observed, and no reoperations were necessary, demonstrating favorable outcomes for these patients. In the early postoperative period, one patient developed non-sustained supraventricular tachycardia (SVT), and another patient experienced a Mobitz type 2 atrioventricular (AV) block, which resolved during follow-up. One patient presenting with preoperative sick sinus syndrome underwent simultaneous PAPVC surgery and the implantation of a permanent epicardial pacemaker. Additionally, one patient necessitated the implantation of a permanent pacemaker following the surgical correction of PAPVC and ASD. One patient was observed to have mild SVC

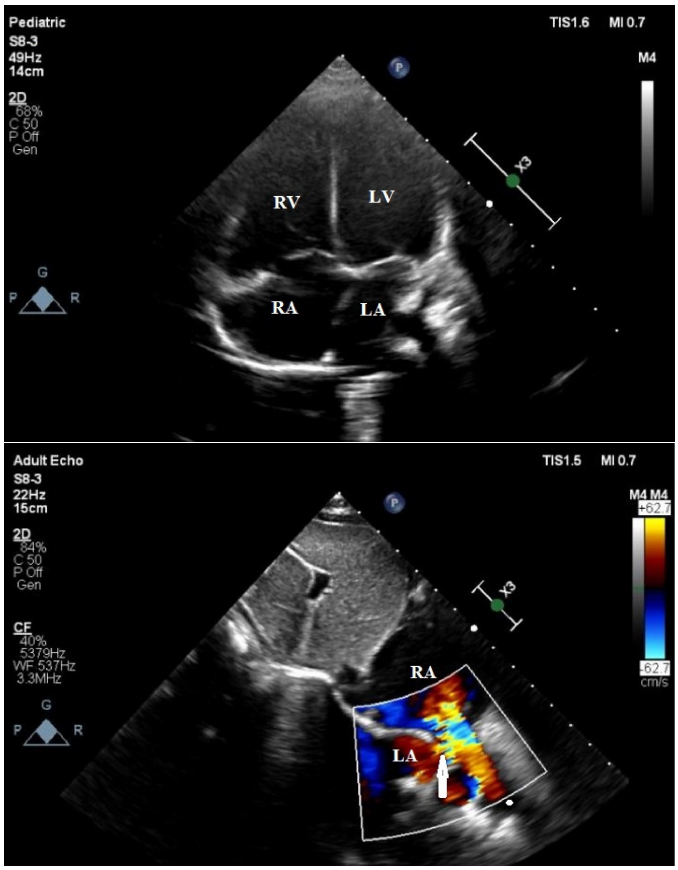

Figure 1. Transthoracic echocardiography four-chamber view (a) shows an enlargement of the right heart chambers, and subcostal sagittal view (b) demonstrates the superior sinus venosum atrial septal defect (white arrow).

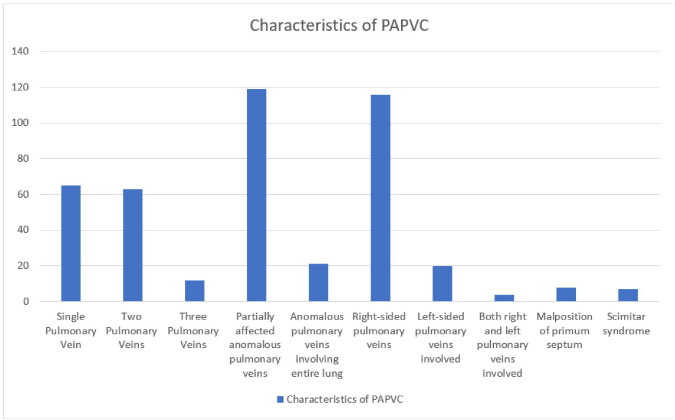

Figure 2. The graphic demonstrates the characteristics of partial anomalous pulmonary venous connection (PAPVC) patients.

stenosis at the first postoperative month. In our study, while one patient was still receiving medical treatment for significant PH, surgical treatment was planned after a positive vasoreactivity test was detected in the last catheter angiography of a patient who was admitted at four months and received anti-PH therapy for approximately 10 years for significant PH.

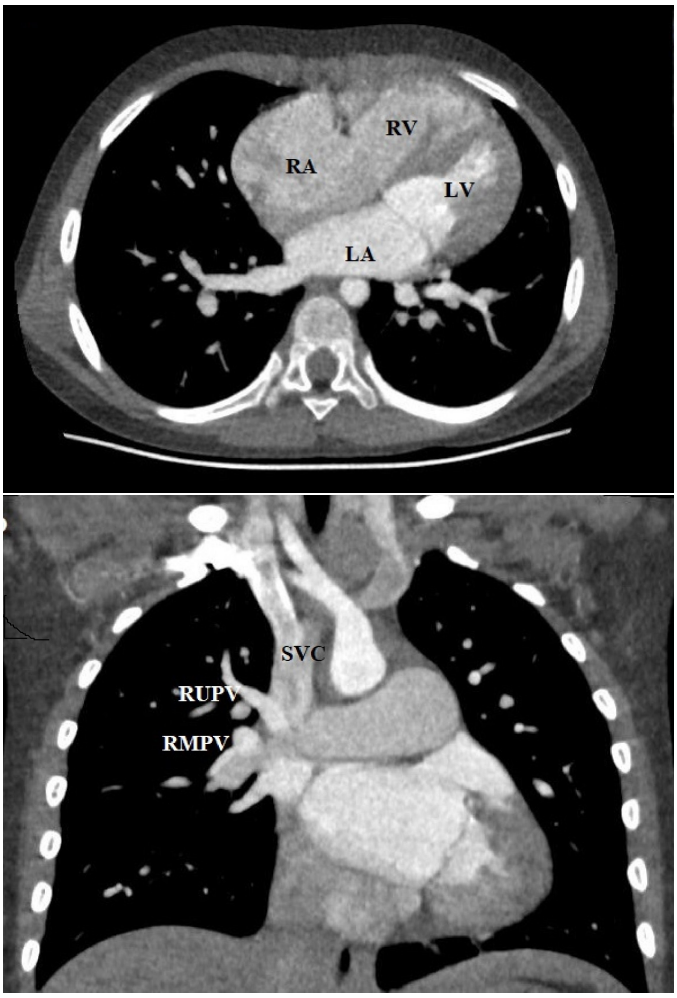

Figure 3. Computerized tomography images show an enlargement of the right heart chambers at the axial view (a) and anomalous pulmonary venous connection of the right upper and middle pulmonary veins to the superior vena cava at the coronal view (b). LA: left atrium; LV: left ventricle; RA: right atrium; RUPV: right upper pulmonary vein; RMPV: right middle pulmonary vein; RV: right ventricle; SVC: superior vena cava.

The mean follow-up duration was 33.7±35.7 months (6 to 149 months).

Discussion

This manuscript comprehensively analyzes the clinical presentation, diagnostic evaluation, management, and outcomes of patients diagnosed with PAPVC at a single centre. Our study adds to the existing literature on this uncommon congenital cardiovascular anomaly and provides valuable insights into best practices for its management.

Our study appears to align with prior studies examining PAPVC, particularly regarding demographic distribution. Notably, the preponderance of male patients, accounting for 54.6%, reflects a subtle male predominance also noted in existing literature [2]. Moreover, the broad age range at the time of diagnosis confirms the known diverse age presentation of PAPVC, reaffirming that this condition can

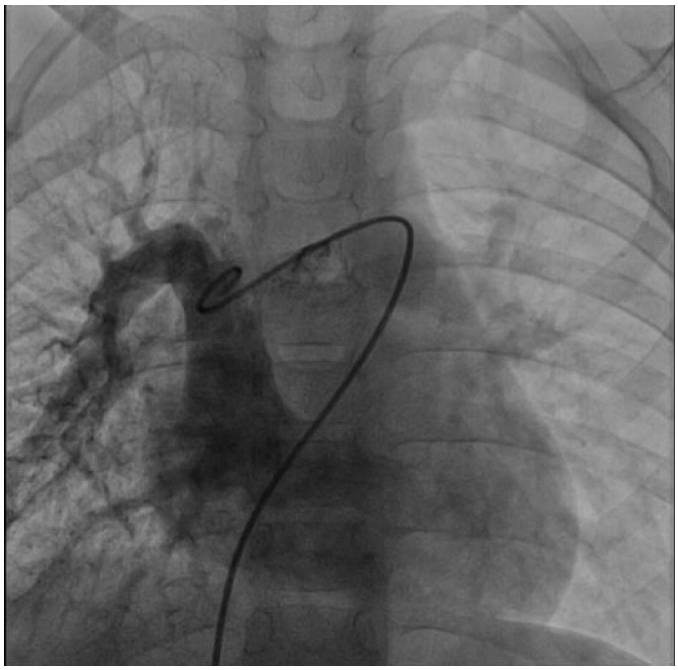

Figure 4. Catheter angiographic image demonstrates the venous phase, anomalous pulmonary venous connection of right upper and middle pulmonary veins to the right atrium via superior vena cava.

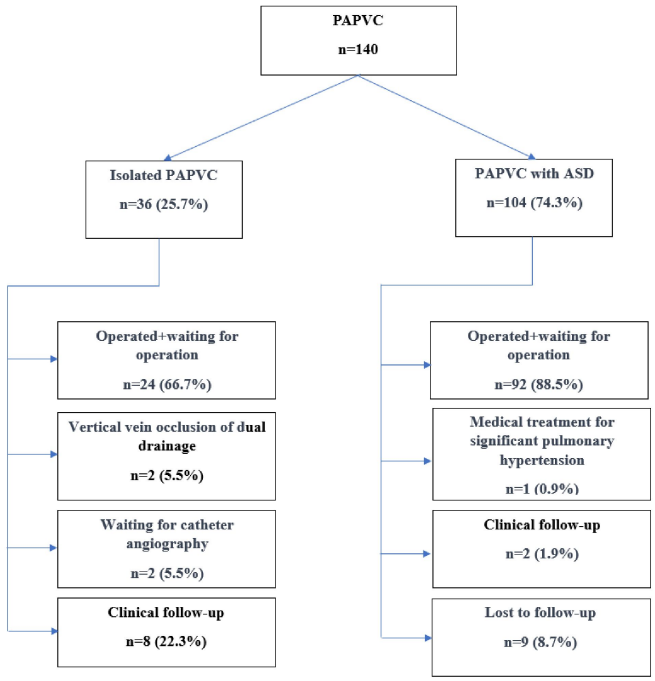

Figure 5. Management of partial anomalous pulmonary venous connection (PAPVC) patients. ASD: atrial septal defect.

manifest at any age and is not restricted to a particular age group [1, 2].

Consistent with prior reports [4, 6], our results show that most patients had PAPVC associated with ASD. However, a minority of patients (25.7%) presented with isolated PAPVC, a crucial consideration for clinicians as the manifestations and treatment approaches can vary between these two categories.

The present study found that out of the entire cohort, 46.4% (n=65) patients displayed a single PV anomalously connected, 45% (n=63) had two PVs involved, and an additional 8.6% (n=12) had three PVs affected, consistent with reports in previous research [1, 2]. These results support the existing understanding of the disease's clinical heterogeneity [3, 4]. Meticulously assessing all echocardiographic windows is essential to ensure successful visualization of the confluence of all pulmonary veins into the left atrium. Furthermore, particular attention should be devoted to detecting right or left-sided vertical veins.

Of the patient population, 85% (n=119) exhibited partially affected anomalous PVs, leaving the remaining 15% (n=21) with anomalous PVs involving the entire lung. This distribution of PV involvement further underscores the variable presentation of this condition, ranging from isolated veins to whole-lung involvement [1, 2]. The study detected a prevalence of anomalous connections in the right lung, with all the PVs of the right lung being anomalously connected in 7.1% (n=10) of patients. Meanwhile, 7.9% (n=11) had all the PVs of the left lung affected. Right-sided PVs were implicated in 82.8% of the cohort (n=116), whereas left-sided PVs were involved in 14.3% (n=20), with both right and left PVs being involved in a minor proportion of patients (2.9%, n=4). This observation aligns with the existing literature, which shows a general predisposition towards right-sided involvement [5, 7, 8, 9]. The most common anomaly observed was right PAPVC draining into the SVC, followed by right PAPVC draining into the RA.

The malposition of the septum primum is a rare malformation that results in either partial or total anomalous pulmonary venous drainage with a typical pulmonary venous connection to the left atrium, depending on the septal displacement [10]. In our study, septum primum malposition was detected in a small portion of the patients (5.8%, n=8), emphasizing the complex morphological variations associated with [10]. Furthermore, scimitar syndrome was present in 5% (n=7) of patients, highlighting the diversity of clinical presentations and the importance of accurate diagnosis for managing this disease [2, 4, 11].

As emphasized in scholarly literature, the diagnostic approach for these patients necessitates a comprehensive strategy, underscored by applying an array of imaging modalities, encompassing CT, catheter angiography, and cardiac MRI [5, 9, 12]. This approach aligns with the importance of meticulous preoperative planning in managing such cases. Advanced imaging techniques, such as three-dimensional reconstruction, can provide critical information beneficial in tailoring surgical planning, thus potentially enhancing postoperative outcomes, as suggested by Guariento et al. [9].

A cautious and conservative strategy is typically recommended in managing patients presenting with PAPVC yet remaining asymptomatic, exhibiting a low shunt fraction, and showing no signs of right ventricular dysfunction. Such an approach necessitates careful monitoring to track the progression of the condition. The emergence of symptomatology, the enlargement of the right ventricle,

or the manifestation of a significant left-to-right shunt, determined by a Qp/Qs ratio exceeding 1.5, are substantial factors warranting the consideration of surgical intervention [1, 13]. Most of our study cohort (82.8%) required surgical intervention due to hemodynamically significant left-to-right shunts, affirming the role of surgery in managing PAPVC. Clinical progress was monitored through regular follow-ups for 10 of 140 patients (7.1%), underlining the importance of continuous evaluation in this patient population.

The risk of developing irreversible pulmonary hypertension is a serious concern for patients with untreated PAPVC. Pulmonary hypertension can lead to significant morbidity, as supported by Sahay et al. [14], highlighting the necessity for timely intervention in this condition. Within the scope of our research, one patient was receiving medical treatment for severe pulmonary hypertension. Another patient, admitted at four months of age and treated for significant PH for nearly ten years, was also scheduled for surgical intervention after a positive vasoreactivity test. The results reported by Alsoufi et al. [2] show encouraging outcomes following surgical intervention for PAPVC, including a substantial decrease in the risk of irreversible pulmonary hypertension, thus underscoring the benefits of surgery in PAPVC patients.

The absence of early or late deaths among the patients who underwent surgery, coupled with no necessity for reoperations, is a promising outcome and seems to echo previous studies that also reported favorable results following surgical correction of PAPVC [2, 4]. It is also noteworthy that complications were relatively rare and manageable, reinforcing the overall safety of the surgical approach for PAPVC [12].

However, the incidence of postoperative arrhythmias, specifically non-sustained SVT and Mobitz type 2 AV block, is a matter of concern. These observations highlight the importance of close follow-up and management of arrhythmias in the postoperative period, as suggested by Hatipoglu et al. [12]. In some cases, the implantation of a permanent pacemaker may be necessary, such as in patients with pre-existing sick sinus syndrome or postoperative AV block [15]. Our experience suggests that such cases need a coordinated approach, and the availability of an electrophysiology department can be crucial.

Finally, the mean follow-up duration was substantial, contributing to the study's strength. Long-term follow-up is critical in assessing the durability of surgical results and potential long-term complications, an aspect highlighted in previous literature [12].

Limitations

This study has certain limitations. Its monocentric design may limit the generalizability of findings to varied healthcare settings and populations. The retrospective model, while beneficial in some respects, may introduce bias. The potential for selection bias, a common challenge in observational studies, exists as our cohort is comprised of patients seeking medical attention, possibly not reflecting the broader PAPVC patient demographic. Despite these limitations, we believe this study offers essential insights into PAPVC management, and further research is needed for validation.

Conclusion

Despite this congenital heart defect's rarity and morphological diversity, early detection and appropriate surgical intervention tailored to each patient's unique anatomy can potentially prevent severe complications. The utilization of comprehensive imaging techniques in diagnostic and surgical planning stages has proven to benefit significantly. However, further research is necessary to confirm these findings and improve the treatment and care of PAPVC patients.

Ethical approval

The study was approved by the University of Health Sciences, Istanbul Mehmet Akif Thoracic and Cardiovascular Surgery Training and Research Hospital Ethics Committee (November 2022/ 2022.09-64).

References

1. El-Kersh K, Homsy E, Daniels CJ, Smith JS. Partial anomalous pulmonary venous return: A case series with management approach. Respir Med Case Rep. 2019;27:100833.
2. Alsoufi B, Cai S, Van Arsdell GS, et al. Outcomes after surgical treatment of children with partial anomalous pulmonary venous connection. Ann Thorac Surg. 2007; 84: 2020-6.
3. El Bardissi AW, Dearani JA, Suri RM, Danielson GK. Left-sided partial anomalous pulmonary venous connections. Ann Thorac Surg. 2008;85:1007-14.
4. Fragata J, Magalhães M, Baquero L, et al. Partial anomalous pulmonary venous connections: surgical management. World J Pediatr Congenit Heart Surg. 2013;4:44-9.
5. Seller N, Yoo SJ, Grant B, Grosse-Wortmann L. How many versus how much: comprehensive haemodynamic evaluation of partial anomalous pulmonary venous connection by cardiac MRI. Eur Radiol. 2018;28:4598-606.
6. Pavy C, Gavira N, Maminirina P, Baron O. Right partial anomalous pulmonary venous connection to the superior vena cava following the Warden procedure. J Card Surg. 2018;33:565-9.
7. Mathis L, Crethers D, Buckman B, et al. Partial Anomalous Pulmonary Venous Connection Repair: Customized Approach and Outcomes. Pediatr Cardiol. 2021;42:1064-73.
8. Suzuki K, Iwata Y, Hiramatsu T, et al. Mid- to long-term surgical outcomes of partial anomalous pulmonary venous connection. Gen Thorac Cardiovasc Surg. 2021;69:27-31.
9. Guariento A, Cattapan C, Chemello E, et al. 3D reconstruction for preoperative planning of partial anomalous pulmonary venous return. Kardiol Pol. 2021;79:1271-3.
10. Cuttone F, Hadeed K, Lacour-Gayet F, et al. Isolated severe leftward displacement of the septum primum: anatomic and 3D echocardiographic findings and surgical repair. Interact Cardiovasc Thorac Surg. 2017;24:772-7.
11. Al-Muhaya MA, Alkodami AA, Khoshhal S, et al. Transcatheter occlusion of the vertical vein in a partial anomalous pulmonary venous connection with dual Drainage, case series with literature review. Int J Cardiol Heart Vasc. 2021;37:100889.
12. Hatipoglu S, Almogheer B, Mahon C, et al. Clinical Significance of Partial Anomalous Pulmonary Venous Connections (Isolated and Atrial Septal Defect Associated) Determined by Cardiovascular Magnetic Resonance. Circ Cardiovasc Imaging. 2021;14:e012371.
13. Stout KK, Daniels CJ, Aboulhosn JA, et al. 2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease: A Report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines [published correction appears in J Am Coll Cardiol. 2019 May 14;73(18):2361-2362]. J Am Coll Cardiol. 2019;73:e81-e192.
14. Sahay S, Krasuski RA, Tonelli AR. Partial anomalous pulmonary venous connection and pulmonary arterial hypertension. Respirology. 2012;17:957-63.

15. Alkady H, Elnaggar A, Eldegwy M, Nosair A. The Warden procedure for partial anomalous pulmonary venous connection in children: 10 years experience in 65 cases. Cardiol Young. 2020;30:1486-9.