Heart Failure Due to Anomalous Pulmonary Vein Connection Associated with Atrial Septal Defect

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Introduction

Anomalous pulmonary vein drainage (APVD) is a rare disease, accounting for nearly 1 to 3% of congenital heart diseases.¹ It may be total, when all PVs are involved, or partial, when only some PVs drain anomalously into one or more systemic veins or directly into the right atrium (RA).^{1,2} This condition occurs alone or in association with an interatrial septal defect, more often sinus venosus interatrial communication (IAC) (which accounts for nearly 5% of all IAC cases).³

Patients with this condition may remain asymptomatic until adulthood. However, overload of right chambers may trigger symptoms, especially reduced functional capacity and progressive dyspnea, in addition to the development of pulmonary hypertension (PH) and ventricular dysfunction, which is the most severe disease scenario.³ Recognizing these symptoms and establishing early diagnosis and treatment is crucial to avoid disease progression and its prognostic implications.

We present a case of heart failure (HF) due to partial anomalous pulmonary vein drainage (PAPVD) associated with IAC with reverse remodeling after surgical correction in adulthood.

Clinical case

A previously healthy 36-year-old patient was admitted to the emergency department with dyspnea that started nearly 15 days before and progressively worsened with time, evolving to dyspnea at rest, associated lower limb (LL) edema and orthopnea. On admission, the patient presented with tachydyspnea, blood pressure of 100x60 mm Hg, heart rate of 96 bpm, respiratory rate of 26 irpm, saturation of 93% on oxygen 4 L/min via a nasal cannula. Examination revealed the presence of pathological jugular vein engorgement and

hepatojugular reflux. Pulmonary auscultation showed crackles up to the bilateral middle third. Furthermore, we observed propulsive apex beat and palpable right ventricle (RV) by means of hooking palpation, with systolic murmur 3+/6+ more evident in the left paraesternal region (accessory aortic focus) irradiating to the entire precordium, as well as rhythmic heart sounds with loud B2. Abdominal assessment revealed painful and palpable liver located 4 cm below the right costal margin, presence of bowel sounds, and no peritoneal signs. Finally, the patient presented with LL edema 3+/4+.

Chest X-ray showed significant cardiomegaly, with increased apex beat and increased pulmonary artery trunk (PT), enlarged pulmonary hilum, and changes in vascular network, suggestive of congestion (Figure 1A). Electrocardiogram revealed sinus rhythm with right axis deviation, right bundle branch block, signs of biventricular and right atrium (RA) overload (Figure 1B). Laboratory tests did not show relevant changes.

Transthoracic echocardiogram showed PH (pulmonary artery systolic pressure [PASP] of 85 mmHg), major right chamber dilatation with RV dysfunction, left ventricle (LV) systolic dysfunction, grade 2 diastolic dysfunction, moderate pulmonary insufficiency, and major tricuspid insufficiency (Table 1). These findings were confirmed by transesophageal echocardiogram, which also revealed the presence of wide sinus venosus IAC, measuring 38 mm in its greatest diameter.

Chest computed tomography angiography (CTA) was requested to investigate the possible causes of PH, showing cardiomegaly involving the right chambers, negative results for pulmonary thromboembolism but with signs of pulmonary artery hypertension (dilated PA trunk measuring 5.2 cm) (Figure 1C).

On cardiac magnetic resonance, significant biventricular dilation and dysfunction (LV ejection fraction [EF] of 19% and RVEF of 20%) were found, as well as dilated RA and possible right inferior APVD for RA and PT dilatation (Figure 1D).

Initial clinical measures were taken to compensate the patient using intravenous diuretics and oral vasodilators. Subsequently, right heart catheterization was conducted, revealing wide sinus venosus IAC, right superior pulmonary vein with drainage in the superior vena cava, and right inferior pulmonary vein with drainage in the RA roof, RA oxymetric jump, HP (PASP 53 mmHg, pulmonary vascular resistance [PVR]: 3.04 woods), pulmonary-systemic flow ratio (Qp/Qs): 3.79/1, and PVR/systemic vascular resistance (SVR) 0.11.

The patient underwent surgical correction consisting of atrioseptoplasty with redirection of pulmonary vein blood flow,

Keywords

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Figure 1 – Complementary tests. A: Chest X-ray. B: Electrocardiogram. C: Chest computed tomography angiography (blue arrow: entrance of the right pulmonary vein into the right atrium). D: Magnetic resonance imaging (red arrow: entrance of the pulmonary vein into the right atrium).

Table 1 – Evolution of echocardiographic and magnetic resonance findings before the procedure and 4 months and 1 year after treatment

	Magnetic resonance imaging	Baseline echocardiogram	Echocardiogram 4 months after correction	Echocardiogram 1 year after correction
Right atrium	Dilated	143 mL/m ²	Increased	36 mL/m ²
Left atrium	27 mm	39 mm	39 mm	39 mm
Left ventricle	739x591 mL	46 mm	42 mm	44 mm
RV functional assessment	EF 19%	FAC 25%	FAC 31%	FAC 24%
Left ventricle	178x141 mL	50x44 mm	50x37 mm	50x36 mm
LVEF	20%	26%	51%	54%
Pulmonary insufficiency	Unvalued	Moderate	Moderate	Moderate
Tricuspid insufficiency	Unvalued	Massive	Moderate	Mild to moderate
Pulmonary trunk	52 mm	39 mm	40 mm	43 mm
PASP	CIA	85 mmHg	57 mmHg	46 mmHg
Other findings	Possible right inferior APVC to RA	IAC 38 mm		

VD: ventrículo direito; FEVE: fração de ejeção do ventrículo esquerdo; PSAP: pressão sistólica da artéria pulmonar; FE: fração de ejeção; DAVP: drenagem anômala da veia pulmonar; AD: átrio direito; FAC: fração de alteração da área do ventrículo direito; CIA: comunicação interatrial.

with good evolution. He received hospital discharge and was prescribed with enalapril 5 mg 12/12 h (maximum tolerated due to hypotension), bisoprolol 10 mg/d, spironolactone 25 mg/d, and furosemide 40 mg/d. The patient returned to the outpatient clinic 4 months after the procedure, asymptomatic, with follow-up echocardiogram showing reverse remodeling (LVEF of 51%) and moderate pulmonary and tricuspid insufficiency. These findings were maintained at late follow-up with a new echocardiogram 1 year after the procedure, with

LVEF of 54%, moderate pulmonary insufficiency, and mild to moderate tricuspid insufficiency.

Discussion

PAPVD usually occurs during fetal development, secondary to failure of one or more pulmonary veins of right lung upper lobe in connecting to the left atrium.^{2,4} Physiologically, it produce a shunt from left to right, similar to an atrial septal

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defect, allowing for the already oxygenated blood to circulate again to the lungs, resulting in excessive pulmonary blood flow. In 80 to 90% of the cases, drainage occurs to the right and is associated with atrial septal defects, more commonly sinus-venosus IAC.^{2,3,5}

Many adults with PAPVD are accidentally diagnosed by chest images taken for any other indication, either with CTA in an emergency service to rule out pulmonary embolism, during cardiac catheterization for coronary intervention, or as part of an assessment of post-radiofrequency catheter ablation.^{6,7} Most patients with isolated PAPVD are asymptomatic.^{2,5,8} However, when this condition is associated with IAC, left-to-right (L-R) shunt occurs, which is responsible for increased right chamber flow and pressure leading to the remodeling of these chambers and of pulmonary vasculature and culminating in symptoms and in PH.^{5,8} Since many patients remain asymptomatic up to adulthood, early diagnoses eventually occur as an incidental finding in less than 1% of the cases.^{2,5,8} In the remaining cases, diagnostic suspicion based symptoms directs the investigation.

The presence of IAC alone may not be responsible for the onset of symptoms. The magnitude and direction of flow depend on defect size and on the filling pressure of each ventricle.⁹ As a rule, the septal orifice needs to measure at least 10 mm, have a Qp/Qs > 1.5/1.0, or promote right chamber dilatation in order to be considered a hemodynamically-significant L-R shunt.⁹

The main diagnostic test for PAPVD is transthoracic echocardiogram.^{3,5} However, in cases when this condition is associated with IAC, transesophageal echocardiogram, magnetic resonance imaging, or even right heart catheterization, may be required to elucidate the diagnosis. In addition to diagnosis, invasive hemodynamic assessment allows to calculate Qp/Qs ratio, an important information for therapeutic planning.^{2,3}

The treatment of most PAPVD cases consists of clinical follow-up.^{2,3,8} Conversely, there is an increase in morbidity and mortality rates when cases of significant IAC do not receive early treatment. Patient's age and pulmonary pressures at the time of correction are the main outcome predictors.^{9,10} In both conditions, when there are symptoms, enlarged RV, or significant increase in L-R shunt (Qp/Qs > 1.5), surgical correction is mandatory and may be responsible not only of symptom resolution^{2,3,8} but also of reducing the likelihood of late arrhythmia, HF, and PH complications. The risk of arrhythmias increases after the age of 40 in non-operated patients, with atrial flutter, being more frequent up to the age of 60 years, and atrial fibrillation becomes predominant after this age.^{6,7}

The short- and long-term results after surgical repair of PAPVD are excellent, and the reported complications rates are low.^{6,7} The association between reverse remodeling and improved clinical outcomes is well established in the literature. Therapies capable of promoting EF increase and reducing ventricle sizes are associated with lower mortality in HF.¹¹

In the correction of heart diseases with L-R shunt, remodeling is expected after shunt surgical correction. The reduction in RA and ventricle volumes may occur as early as 24 hours after the procedure, but is generally more evident after 6 month and may extend to up to 18 months after the procedure. There is an expected reduction in right chambers, in tricuspid reflux, and in pulmonary pressures, as well as in improvement in LVEF, as observed in the present clinical case. However, the magnitude of remodeling is inversely proportional to patient's age at the time of closure, which reinforces the need for urgently starting treatment soon after symptom onset and diagnosis.^{9,12}

Potential complications include stenosis or obstruction of pulmonary or systemic veins, residual IACs, or new atrial arrhythmias.^{6,7} Although rare, PH represents a special situation and, if present in an adult patient with PAPVC, may not undergo remission after repair. Advanced cases with RVP > 8 woods or inversion of right-to-left shunt are contraindicated for surgical treatment.^{2,3,6-8}

In the present report, the patient started to present with HF symptoms in adulthood, few days before admission. An etiologic investigation found APVD and IAC with important hemodynamic repercussion showing a significant increase in right chambers and in left-to-right shunt (Qp/Qs of 3.79), in addition to PH (PASP of 85 mmHg) and bilateral dysfunction. Surgical correction and pharmacological treatment of HF promoted clinical and echocardiographic improvement in an early and sustained fashion, reinforcing the importance of investigation and treatment of patients with this condition.

Author Contributions

Conception and design of the research: Bonatto MG; Acquisition of data, Analysis and interpretation of the data and Writing of the manuscript: Bonatto MG, Freitas AKE; Critical revision of the manuscript for intellectual content: Bonatto MG, Freitas AKE, Rocha LSO, Moura LAZ; Doctor responsible for the patient: Collatusso C; Assistance in imaging methods: Torres RA, Blume GG.

Potential Conflict of Interest

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Ethics approval and consent to participate

This article does not contain any studies with human participants or animals performed by any of the authors.

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