

Surgical Correction of Total Anomalous Pulmonary Venous Drainage in an Adult

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Total anomalous pulmonary venous drainage (TAPVD) is rarely seen in adults, because this congenital heart disease almost always requires surgical treatment in the neonatal period, often on an emergency basis. We report a patient that, despite being diagnosed during childhood, underwent surgical repair at age 25, about one year after his clinical condition worsened.

Total anomalous pulmonary venous drainage (TAPVD) accounts for approximately 2 % of all congenital heart diseases¹, and the co-existence of atrial septal defect (ASD) is virtually mandatory. The anatomical types of this malformation are associated with different degrees of pulmonary venous return obstruction and vasoconstriction of the small circulation. The interaction of these factors results in hypertension and/or increased pulmonary blood flow of varying degree, both of which modulate clinical presentation. Consequently, while some patients are oligosymptomatic, presenting with mild arterial unsaturation, others show severe pulmonary edema².

Darling et al³, proposed a classification of TAPVD into four types:

Type I - supracardiac (45% of the cases) – pulmonary veins connect to the superior vena cava or its tributaries;

Type 2 – infracardiac (25% of the cases) – pulmonary veins drain directly into the inferior vena cava, its tributaries or the portal venous system;

Type III – intracardiac (25% of the cases) – pulmonary veins connect to the venous sinus or right atrium;

Type IV – mixed (5% of the cases) – characteristics of at least two of the above types are present.

In the neonatal period, most children progress to mild cyanosis and congestive heart failure due to pressure and volume overload of the right heart chambers. This scenario often requires, in addition to clinical measures, emergency surgical repair. Operative morbidity and mortality of TAPVD

patients decreased significantly in the last two decades due largely to early diagnosis afforded by advances in Doppler echocardiography and management directed to stabilization and optimization of preexistent clinical conditions.

Very few patients survive to old age without surgical correction⁴⁻⁶, and we found no case in Brazilian literature regarding TAPVD surgical repair in adults. We herein report, however, a patient with supracardiac TAPVD that, although it had been diagnosed during childhood, was corrected only during adulthood, when the worsening of the condition led the patient to seek medical attention.

Case Report

About one year ago, a 25-year-old man with dyspnea on moderate exertion and self-reported history of cardiovascular disease sought out our service. At age 18, he had undergone clinical examination as part of a job pre-admission test and was diagnosed with heart murmur and changes in his extremities and was referred to a cardiologist. However, because he was asymptomatic, he declined treatment.

At admission he was eupneic and in good general status, although marked clubbed fingers and watch-glass nails were present. Mild digital cyanosis was evident. There was supraclavicular venous pulse, best felt in the sitting position, in addition to a fixed split-second heart sound accompanied by systolic-diastolic double murmur in the pulmonary area. The systolic component was moderate, harsh and radiating to the whole precordium, left axilla, and left side of the neck and upper back. The diastolic murmur was soft.

Hemoglobin level was 17.7 g/dl, and hematocrit, 54.1%. Electrocardiogram showed sinus rhythm, right heart overload, and right bundle branch block. Posteroanterior chest X-ray suggested excessive pulmonary blood flow, with enlargement of upper mediastinum, forming the classic “snowman” figure (Fig. 1).

Echocardiographic examination showed *situs solitus*, with the heart in its normal position. In addition to nonrestrictive

Key words

Total anomalous pulmonary venous drainage, congenital heart defect with increased pulmonary blood flow, pulmonary veins, adult.

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ostium secundum ASD, with bidirectional flow favoring right-to-left shunting, a retrocardiac chamber was found where at least three pulmonary veins were connected. From this chamber emerged a large ascending vertical vein, connected to the innominate vein. The latter, as well as the superior vena cava, right heart chambers, and pulmonary artery trunk, were quite dilated. The small left atrium did not receive any pulmonary vein. Ventricular systolic performance was normal, and pulmonary artery trunk flow was rapid (3.2 m/s). Echocardiographic contrast (agitated saline) infused into a systemic peripheral vein confirmed blood flow passing through the interatrial septum, with no contrasting of the posterior chamber and vertical vein, suggesting supracardiac TAPVD.

Cardiac catheterization confirmed the diagnosis and

demonstrated the confluence of all four pulmonary veins at the retropericardial chamber. (Fig. 2).

Gradients between RA/LA and RV/PT were 5 mmHg and 17 mmHg, respectively (Tab. 1). Total pulmonary vascular resistance was calculated at $150.40 \text{ d.s.cm}^{-5}$ and the right-to-left "shunt" at 6.8 l/min, with pulmonary-to-systemic flow ratio of 1.8. Pulmonary-to-systemic vascular resistance ratio was 0.17.

The patient was operated on by median sternotomy, with bicaval to ascending aorta cardiopulmonary bypass at 22°C . The retropericardial ascending vein was ligated and, under low blood flow perfusion and antegrade cold blood cardioplegia, the retrocardiac chamber was anastomosed to the left auricle and atrium through a posterior approach. The atrial septal defect (ASD) was closed by direct suture. Circulatory arrest was not necessary. A catheter was introduced up to the pulmonary artery through the right atrium. Postoperative course was uneventful. Seven months after surgical repair, the patient is in functional class I.

Discussion

TAPVD, by itself, is incompatible with life, and requires ASD coexistence to allow oxygenated blood supply to the left chambers. The smaller the ASD, the greater the right atrial pressure and the lower the blood supply to the left atrium and the cardiac output. In most cases, as the septal defect is non-restrictive, pulmonary blood flow is determined mainly by the pulmonary-to-systemic vascular resistance ratio and ventricular chamber compliance ratio².

Surgical repair is indicated as soon as diagnosed. In type II, or infracardiac TAPVD, pulmonary venous drainage is usually obstructed at its intra-abdominal course. As a result of high capillary pressure, pulmonary edema occurs, leading to severe hypoxia and metabolic acidosis, often characterizing a surgical emergency in the neonate, since there is no conservative palliative treatment. A similar situation may be seen in type I, when the ascending draining vein is pinched as it crosses between the left pulmonary artery and ipsilateral bronchus. Curiously enough, pulmonary venous hypertension proximal to the obstruction site increases the preexistent pulmonary hypertension secondary to high pulmonary flow and worsens the vascular narrowing, resulting in a positive feedback mechanism that eventually leads to death.

In the non-obstructive forms of TAPVD, surgery may be elective while still in the neonatal period or during the first six months of life, before irreversible pathological changes in the vascular pulmonary bed occur. A hemodynamic study was indicated for this patient primarily to evaluate the vascular pulmonary bed physiology and check the course of one of the pulmonary veins, which was not clearly demonstrated by echocardiography.

This case is unique, because the asymptomatic course of the disease until about one year before his surgery is a rare exception to the tragic statistics related to TAPVD, the mortality of which, when surgical repair is not performed, is estimated at 50% during the first three months of life and 80% within the first year. Yet, at least one case was reported



Fig. 1 - PA chest radiography. The classic "snowman" appearance is evident.

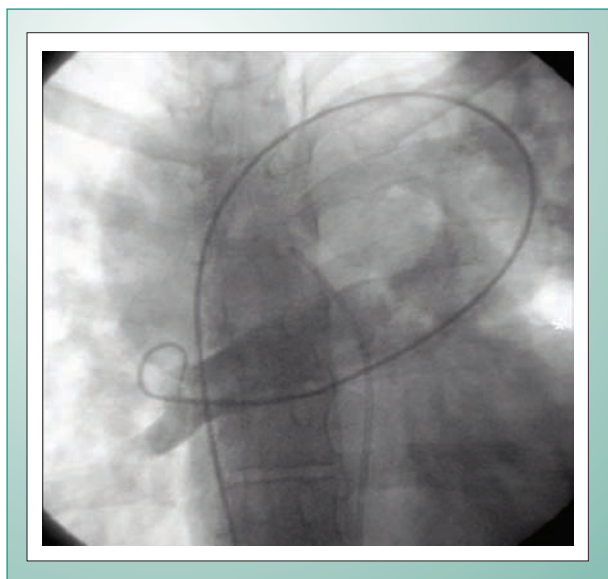


Fig. 2 - Cardiac catheterization. Slight rotated PA view. Contrast injection in the pigtail catheter positioned at the right pulmonary veins depicts the common collecting chamber (*) and ascending vein (**).

Chamber	Systolic blood pressure	Diastolic blood pressure	Mean	PO ₂	PCO ₂	Hb saturation
SVC			5	71.40	33.00	94.40
IVC			5	37.40	34.80	71.80
Low RA			5	31.50	30.70	87.10
Mid RA			5	49.90	30.50	85.50
High RA			5	60.00	34.00	91.00
RV	57		12	58.40	31.00	89.40
PA	40		29	57.90	33.80	89.60
Anomalous PVD			4	98.80	36.60	96.50
LA			10	57.60	32.70	89.80
LV	114		11	51.60	27.90	87.60
Ao(a)	114	83	100	57.60	32.70	89.80

Note: Blood pressure expressed in mm Hg and saturation in percentages.

Table 1 - Hemodynamic Data

of a patient diagnosed at necropsy after surviving for 62 years untreated⁴.

The major factor contributing to the long-term survival of our patient is the absence of obstruction to anomalous drainage, including large interatrial communication and maintenance of vascular resistance and pulmonary pressure within normal limits. Despite obvious digital hippocratism, the increased pulmonary flow and good ventricular function resulted in mild venous desaturation, which may explain the lack of symptoms until adulthood, causing him to decline surgical treatment previously. Fortunately, the patient decided to seek specialized treatment soon after the first symptoms appeared.

Surgical correction of supracardiac TAPVD was performed by ligation of the ascending draining vein of the collecting chamber, at the point of its wide connection with the left atrium, and ASD closure. For surgeons familiar with TAPVD correction in neonates, operative treatment involving adult patients proves to be no more difficult technically and may provide excellent long-term survival⁷. On the other hand, adults with varying degrees of obstruction of the pulmonary venous drainage tend to exhibit more evident and early cyanosis, and lower survival rate owing to changes in the pulmonary arterial tree.

During surgery, the retrocardiac chamber was found to receive the four pulmonary veins. The posterior approach to the anomalous retropericardial structures was facilitated by not placing stay stitches in right border of the pericardium, thus allowing the ventricular mass to be rotated toward the

right for optimal exposure. A well-developed left auricle made it easier to perform a large anastomosis between the retropericardial collecting chamber and the left atrium, thanks to the large opening of both structures in their largest diameter. This way, both incisions were superimposed when the heart was restored to its normal position, preventing distortion of the anastomosis.

Circulatory arrest, albeit considered during surgical planning, was not necessary, because under decreased flow the collection chamber can be opened and aspirated, thus ensuring good exposure for performing the anastomosis.

Some authors suggest that the vertical vein should not be ligated when the left atrium is small and, therefore, poorly compliant to accommodate blood flow after surgical repair⁸. Others⁹ believe that, although this technique may be associated with smoother postoperative course, it has the disadvantage of requiring reintervention to eliminate the left-to-right shunt that may persist in the late postoperative phase.

Pulmonary hypertension is the leading cause of surgical morbidity and mortality, because it may cause severe acute hypoxemia with metabolic acidosis and circulatory collapse, although prompt treatment including mechanical hyperventilation under sedation and curarization, pulmonary vasodilators - such as nitroglycerin, milrinone and nitric oxide - and volume restriction and diuretics may yield satisfactory results^{2, 10}.

Hence the importance of pulmonary artery monitoring, as was done with this patient.

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