

## Case Report

# Percutaneous closure of vertical vein after supracardiac total anomalous pulmonary venous connection repair, using atrial septal defect occluder. A case report

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## ABSTRACT

In patients undergoing surgery for supracardiac total anomalous pulmonary venous connection (SC-TAPVC), not routinely ligating the vertical vein (VV) helps maintain greater hemodynamic stability in the postoperative period, and in many cases, spontaneous closure will be achieved. However, if the VV remains patent, it leads to a pretricuspid shunt with significant pulmonary hyperflow, which in most cases requires surgical or percutaneous closure. We present the case of a postoperative patient with non-obstructive SC-TAPVC and a patent VV, where percutaneous closure was performed using an atrial septal defect occluder.

**Keywords:** Vertical Vein; Septal Occluder Device; congenital heart disease (source: MeSH-NLM).

## Introduction

Total anomalous pulmonary venous connection (TAPVC) is defined as an anomaly in which the pulmonary veins do not connect to the left atrium, where they are directly connected to one of the systemic veins or may drain directly into the right atrium<sup>(1)</sup>.

The frequency of TAPVC is between 1-1.5% of all congenital heart diseases and may be associated with other cardiac anomalies, especially heterotaxy syndrome<sup>(2)</sup>.

Symptoms in these patients can vary, depending on whether it is obstructive or non-obstructive. Patients with non-obstructive TAPVC are usually asymptomatic at birth. However, in the first weeks of life, they may present with cardiomegaly, exertional dyspnea, mild cyanosis, and respiratory difficulty. In contrast, children with obstructive TAPVC will experience respiratory problems, hypoxia, pulmonary hypertension, and rapid progression to cardiopulmonary failure<sup>(1)</sup>.

The repair of supracardiac TAPVC has undergone some modifications, and many surgeons prefer to ligate the vertical vein to prevent residual left-to-right shunts. However, in cases with small left chambers and inadequate compliance of the left atrium, not ligating the vertical vein may improve survival as it provides transient decompression of the right side of the heart for pulmonary hypertension crises in the immediate postoperative period<sup>(3)</sup>. However, this vertical vein may remain patent over time without spontaneous closure, leading to a persistent left-to-right shunt. In many instances, surgical or percutaneous closure of this residual defect becomes necessary.

## Case report

We present the case of a 12-year-old male from Lima, Peru, with a history of non-obstructive supracardiac TAPVC and an atrial septal defect diagnosed at 6 months of age, without a history of hospitalizations or other significant medical events.

The patient was referred to our institution for the first time at the age of 9 years, presenting with dyspnea and mild cyanosis on exertion, with no previous hospitalizations, and an oxygen saturation of 96%. Chest X-ray revealed dilatation of the right chambers with a “snowman” characteristic. Echocardiography showed dilatation of the right chambers, atrial septal defect of 10 mm in diameter; no arrival of the pulmonary veins into the left atrium was observed. Instead, the flow from the pulmonary veins continued with a vertical vein connecting upward to the innominate vein, and no obstructive gradient was detected in this vein. The estimated pulmonary pressure by tricuspid regurgitation was 30 mmHg.

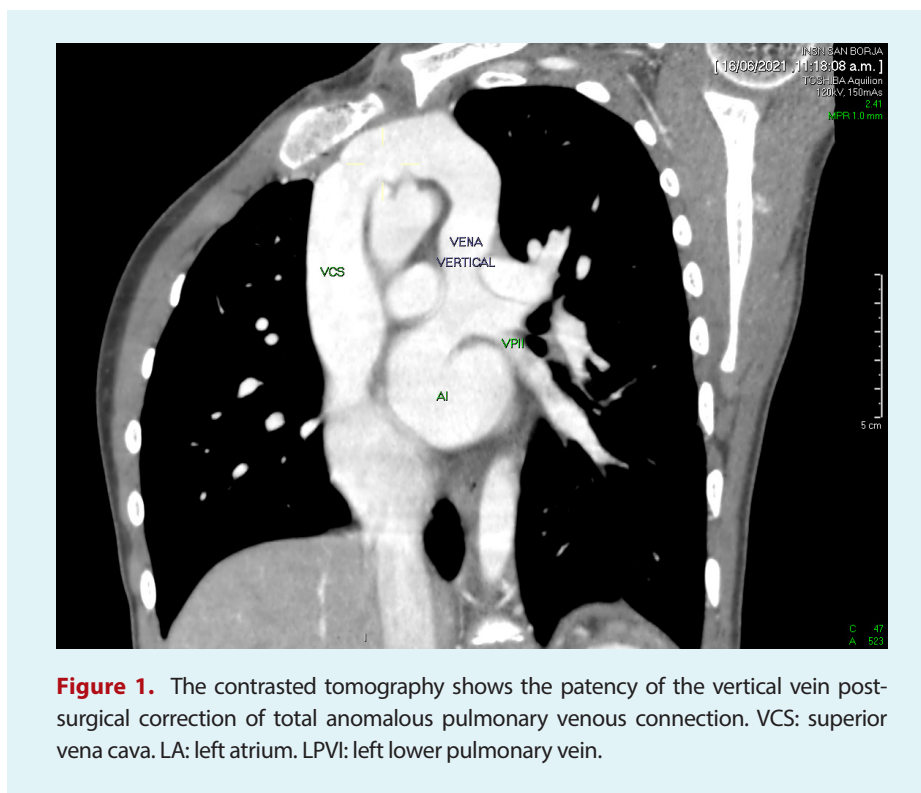
With these findings, the diagnosis of non-obstructive supracardiac TAPVC was confirmed. Medical therapy was initiated with diuretics: furosemide 1 mg/kg every 12 hours and spironolactone 1 mg/kg every 24 hours. Additionally, after analyzing the case, the decision was made to proceed with surgical correction of this malformation.

Given the time of presentation of the heart condition, the patient underwent cardiac catheterization before surgery to

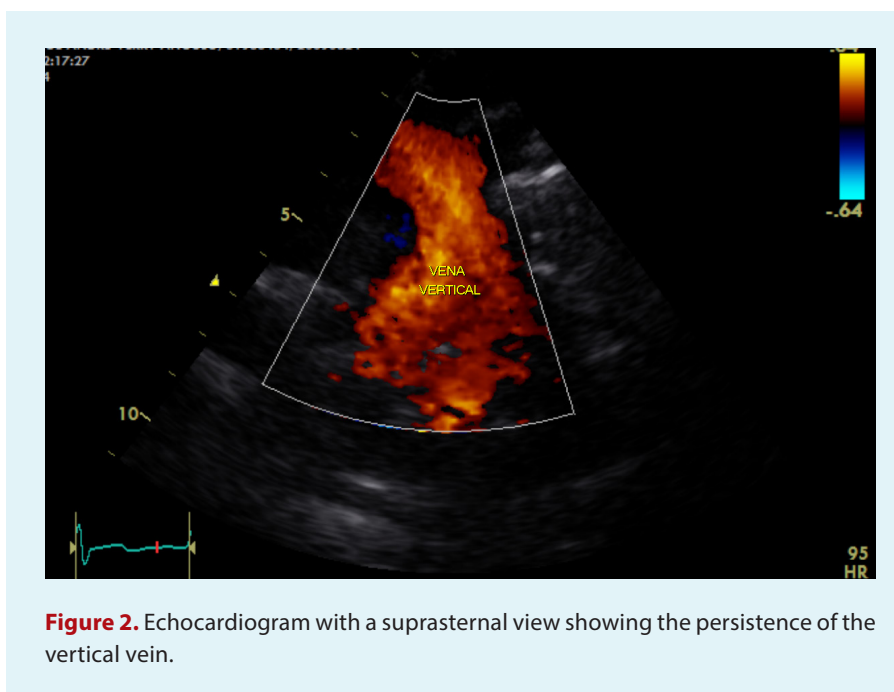
measure cardiac pressures. Mild pulmonary hypertension was found, with a left ventricular end-diastolic pressure (LVEDP) of 10 mmHg, right ventricular end-diastolic pressure of 8 mmHg, pulmonary vascular resistance (PVR) of 1.8 WU.m<sup>2</sup>, and PVR/systemic vascular resistance (SVR) ratio of 0.16. With these values, the patient underwent corrective cardiac surgery with retroauricular correction plus closure of the atrial septal defect using an autologous pericardial patch, leaving the vertical vein patent. The extracorporeal circulation time was 130 minutes, and the clamping time was 84 minutes. Postoperatively, there was no pulmonary hypertension crisis, but there was atypical flutter reverting to sinus rhythm with electrical cardioversion. The patient remained in the cardiovascular postoperative unit for 7 days, then transferred to cardiovascular hospitalization, finally being discharged after 3 days (on the eleventh postoperative day).

In outpatient follow-up, the patient remained with dyspnea New York Heart Association (NYHA) class II and oxygen saturation of 96%. Serial echocardiographic controls showed dilatation of the right chambers, arrival of pulmonary veins to the left atrium without stenosis, and a patent vertical vein without obstructive gradient (**Figure 1**), with no evidence of spontaneous closure. The study is complemented with contrasted cardiac tomography, confirming the patency of the vertical vein (**Figure 2**).

After 2 years post-surgery, due to the persistence of symptoms and the evidence of vertical vein patency, the decision is made to perform percutaneous closure of the vertical vein. Through right and left femoral vein access, a prior hemodynamic study was performed, revealing a QP/QS of 2.9; PVR 0.51 WU.m<sup>2</sup>,



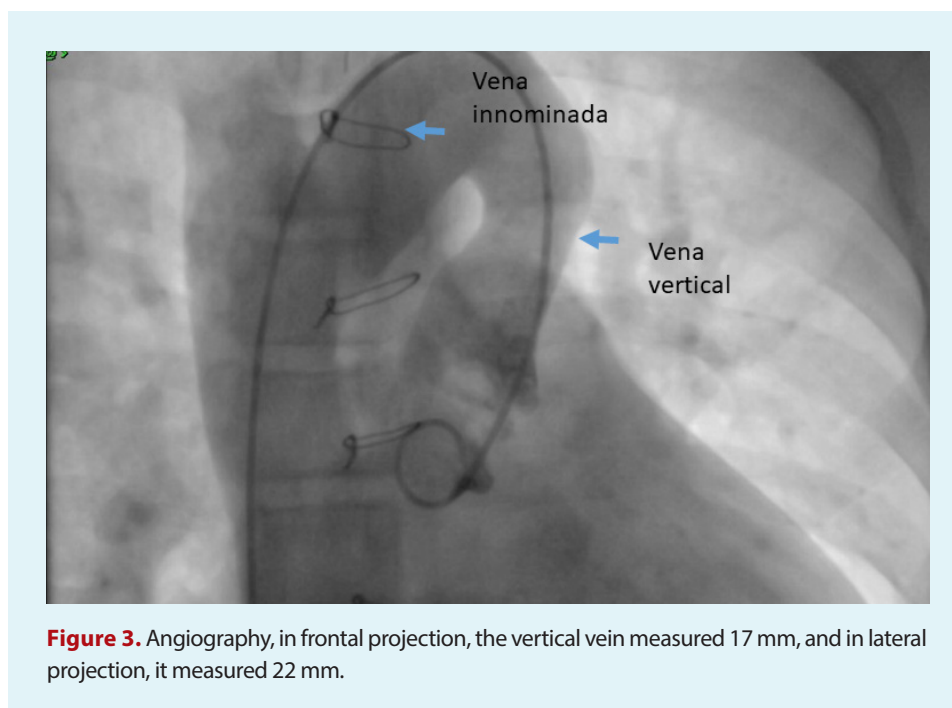
**Figure 1.** The contrasted tomography shows the patency of the vertical vein post-surgical correction of total anomalous pulmonary venous connection. VCS: superior vena cava. LA: left atrium. LPVI: left lower pulmonary vein.

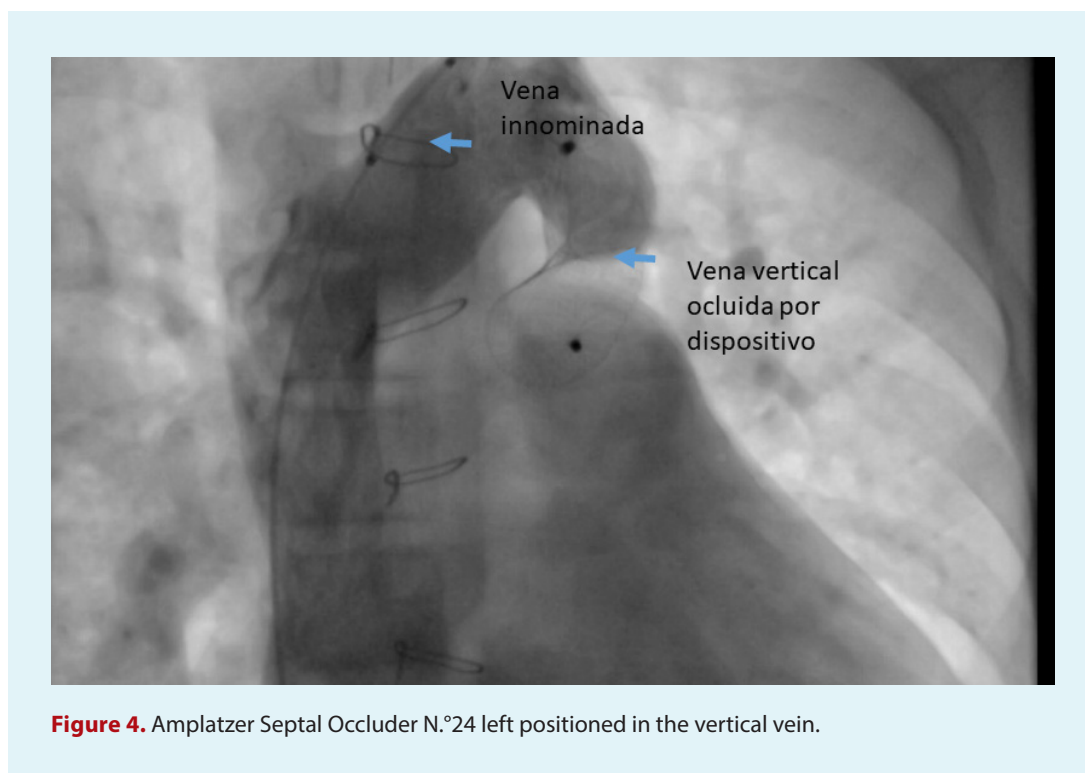


LVEDP: 9 mmHg, mean pulmonary artery pressure (PAP) 11 mmHg. With these values, a right coronary catheter 5 Fr and a hydrophilic guide 0.035" x 150 cm superior vena cava-Innominate vein-vertical vein were introduced. Angiography was performed in 0°/0° and RAO 90°/0° projections, and measurements of the vertical vein were taken, measuring 17 mm in the frontal projection and 22 mm in the lateral projection (**Figure 3, Video 1**). Hydrophilic guide 0.035" x 180 cm was exchanged for an extra-support guide 0.035" x 260 cm and left in position in the right upper pulmonary vein. A 9 Fr long sheath was introduced for an atrial septal defect occluder (Amplatzer Septal Occluder No. 24),

which was positioned in the vertical vein. Control angiography was performed through a 5 Fr Pigtail catheter inserted through the left femoral venous access, ensuring that the jugular confluence with the subclavian and innominate vein was free before releasing the device (**Figure 4**). **Video 2** shows the vertical vein properly occluded.

The patient was discharged 24 hours after the procedure without major complications, with an oxygen saturation of 97%. The echocardiogram at discharge showed no evidence of flow from the vertical vein into the innominate vein. Medical treatment with furosemide 1 mg/kg every 24 hours and spironolactone 1





**Figure 4.** Amplatzer Septal Occluder N.º24 left positioned in the vertical vein.

mg/kg every 24 hours was continued. After 6 months of follow-up, medication was discontinued due to favorable clinical evolution.

## Discussion

TAPVC is a rare heart condition in which the pulmonary veins do not connect to the left atrium, with a mortality rate of 50% by the third month of life, and survival at one year without therapeutic intervention of 20%<sup>(3)</sup>. It is crucial to note that correction is preferably performed at the time of diagnosis, ideally in newborns. If the vertical vein is left patent, spontaneous closure over time is possible. However, in the case of our patient, there was a natural evolution with late correction of the defect.

Most cases of TAPVC are characterized by the presence of a common or retrocardiac pulmonary venous collector, where the pulmonary veins from both lungs converge. This collector, in turn, drains through another vein called the 'vertical vein' into a systemic vein tributary to the right atrium. Depending on the location where the drainage vein, TAPVC is classified as supracardiac (the most common type), cardiac, infracardiac, or mixed forms, with mixed forms being the least common<sup>(5)</sup>.

Correction of the supracardiac TAPVC involves connecting the pulmonary veins to the left atrium, along with ligating the vertical vein. However, some surgeons may choose not to ligate the vertical vein to reduce pulmonary pressure, aiming

to decrease postoperative hypertensive crises and promote postoperative hemodynamic stability<sup>(4)</sup>.

In most cases, the vertical vein spontaneously closes after surgery due to preferential flow to the left atrium and increased compliance. However, there are instances where the non-ligated or partially ligated vertical vein remains patent, leading to a left-to-right shunt<sup>(5,6)</sup>. Kumar suggests that every vertical vein should be closed either surgically or percutaneously, and other researchers recommend vertical vein closure in all supracardiac TAPVC repair surgeries<sup>(4,7)</sup>.

The first reports of percutaneous vertical vein closure were published in 1992, with Hausdorf using a device designed for patent ductus arteriosus closure for this purpose. Subsequently, similar procedures have been performed using various vascular occlusion devices (Vascular Plug I and Vascular Plug II)<sup>(7-9)</sup>. In our case, it was not possible to obtain a different type of device, so the atrial septal defect occluder was used, representing one of the early reported cases.

In conclusion, percutaneous intervention can be considered as part of vertical vein occlusion strategy after supracardiac TAPVC repair surgery with patent vertical vein. In our case, percutaneous closure of the vertical vein was successfully performed using an atrial septal defect occluder since other materials were not available.

### Author contributions:

ACC and KCA: Conceptualization, Writing - Original Draft, Writing - Review & Editing and Investigation.

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