



# Minimally Invasive Surgical Repair of Left-Sided Partial Anomalous Pulmonary Venous Connection: A Case Report

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## Summary

Left side atrial anomalous pulmonary venous connection (PAPVC) is a rare entity. We report one case surgical treated off-pump with minimally invasive approach.

We present a case of left side PAPVC with a non-significant atrial septal defect (ASD) in an asymptomatic 40-year old patient. Angiographic computed tomography (CT) scan has confirmed the diagnosis of vertical vein between left superior pulmonary vein and left brachiocephalic vein, responsible of a significant shunt and right heart dilation. Surgical repair consisted of implantation of the vertical vein into the left appendage through a left mini-thoracotomy and video-assistance. Outcome was event free and patient was discharged on day 4.

Conclusion: Off-pump minimally invasive surgical repair of left side PAPVC is safe, effective and easily reproducible. Possible associated ASD may be treated with hybrid approach and percutaneous closure if indicated.

## Introduction

Left-sided partial anomalous pulmonary venous connection (PAPVC) is a congenital anomaly occurs 10 times less frequently than right-sided PAPVC. Left PAPVC affects the upper lobe drainage twice as commonly as it affects the entire drainage of the left lung. Commonly, the anomalous vein drains via a vertical vein into the innominate vein [1,2]. Isolated PAPVC is often asymptomatic and tends to go unnoticed until adulthood. The development of symptoms and complications from isolated PAPVC depends on the shunt fraction and thus on the number of pulmonary veins anomalously draining to the right heart. Left untreated, long-standing PAPVC predisposes the patient to right-sided volume overload, tricuspid regurgitation (TR), arrhythmias, pulmonary hypertension, irreversible pulmonary vascular disease, right ventricular dysfunction and right ventricular failure [3,4]. A concomitant atrial septal defect (ASD) is present in 80-90 % of cases and patients may have significant symptoms and present in the early childhood [2,4]. We report a patient with a left superior pulmonary vein draining into the brachiocephalic trunk who underwent minimally invasive, video-assisted, off-pump diversion of the anomalous pulmonary vein to the left appendage.

## Case presentation

A 40-year old man, professional football player, totally symptom-free, presented for a systematic clinical assessment regarding sport clearance. Clinical examination was unremarkable, with moderate systolic ejection murmur, common for a sporty person. Doppler echocardiography revealed dilated right atrium (5.3 cm), dilated right ventricle (4.8 cm), mild pulmonary hypertension, small ostium primum ASD (6 mm) without shunt, and sustained Qp/Qs > 2. Right pulmonary vein were seen draining normally into the left atrium with suspicion of left anomalous pulmonary venous connection. Computed tomography (CT) pulmonary angiography aided in defining the anatomy, showing the left superior pulmonary veins drain into a vertical vein to the left brachiocephalic vein (Figure 1). The right heart catheterization confirmed a Qp/Qs of 2.5 with pulmonary pressures recorded as 37/7 (16) mm Hg. Left heart catheterization showed normal left ventricular ejection fraction (LVEF) and absence of significant coronary disease. Indication of surgical correction was decided considering the dilation of right cavities associated with significant left to right shunt, and the high level of sport activity of the patient, according to the American Heart Association/American College of Cardiology (AHA/ACC)

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guidelines for the management of Adult Congenital Heart Disease (ACHD) [3]. Because the associated ASD was small without shunt and accessible to a future endovascular closure if needed, the surgical strategy was focused on the correction of the PAPVC alone with minimally invasive approach and off-pump procedure.

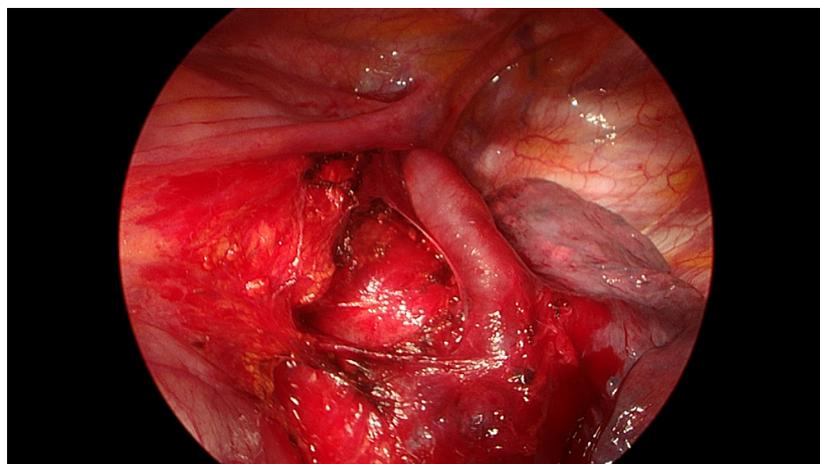
### Surgical technique

Patient was intubated with a double-lumen endotracheal tube for isolation of the left lung. An anterolateral mini-thoracotomy was carried out through the left fourth intercostal space and small retractor was placed. A 30° endoscope was placed through an additional thoracic port in the fifth intercostal space. The left vertical vein was easily visualized and isolated from the hilum to the innominate vein (Figure 2). The left pulmonary artery was exposed. The pericardium was opened posterior to the phrenic nerve and the left atrial appendage (LAA) was

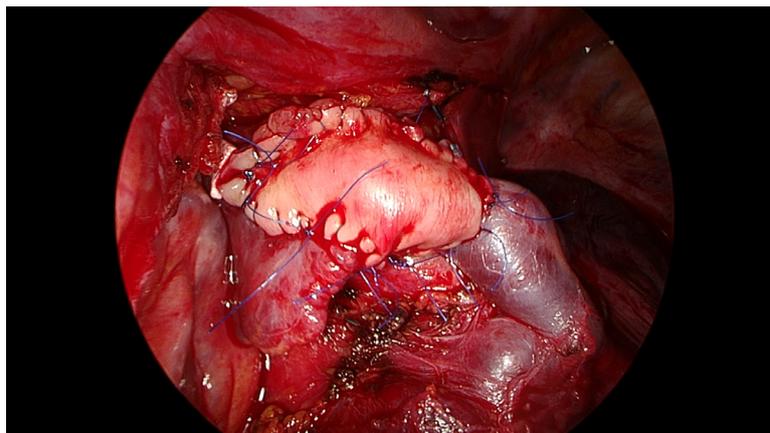
exposed. The anticipated surgical strategy was confirmed with a possible end-to-side anastomosis of the vertical vein into the left appendage. Heparin was given (1.5 mg/kg); the left pulmonary artery was snared; the vertical vein was divided as far as possible and distal part is repaired using 4/0 suture. A side-biting clamp is placed at the base of the LAA, the tip of which is opened and trabeculae trimmed. The back wall of the anastomosis is created using a running 5-0 prolene suture; the anterior walls of the vein and the LAA were spatulated and a large lozenge bovine pericardium patch was inserted with two running 5/0 prolene suture. Deairing of the anastomosis was done and then clamps were removed: the proximal vertical vein, the LAA and the pulmonary artery in sequence (Figure 3). Care was taken to ensure that the anastomosis was not under tension and not purse-stringed. The trans-anastomotic gradient was measured less than 3 mm Hg. The pericardium was left open and the left lung was expanded. Two chest tubes



**Figure 1.** Preoperative angiography CT scan showing the left-sided partial anomalous pulmonary venous connection as a vertical vein toward the left brachiocephalic vein.



**Figure 2.** Operative view showing the anatomy of the vertical vein involved in the left-sided partial anomalous pulmonary venous connection.



**Figure 3.** Operative view showing the anastomosis of the vertical vein into the left appendage with an anterior enlargement with pericardium patch.

were placed and the mini-thoracotomy was closed. Patient was extubated in the OR and then transferred to intensive care unit (ICU); he was discharged home on PO day 4 with an event-free outcome at 6-month follow-up.

### Discussion

Left side PAPVC is a significant anomaly with serious consequences depending on the amount of volume load on the right heart. The hemodynamic similarity between ASD and PAPVC indicates the potential for hypertensive pulmonary vascular disease (HPVD) in the latter and the diagnosis of PAPVC may therefore be considered an indication for surgery on the basis of this potential for HPVD in adulthood. In the current era, symptomatic PAPVC and asymptomatic patients with  $Q_p/Q_s > 1.5$ , right ventricular dilation, mild-to-moderate TR, or early stages of HPVD require surgical treatment in order to prevent the development and progression of right ventricular failure and irreversible pulmonary vascular disease [5].

When surgery is indicated, median sternotomy and cardiopulmonary bypass (CPB) support has been the standard approach in all types of PAPVC repair [3,6]. However, minimally invasive approach is technically feasible in many patients with right side PAPVC even if a significant ASD is associated [3,7]. In left side PAPVC with intact atrial septum or with hemodynamically insignificant ASD/ patent foramen ovale (PFO), the minimally invasive approach with mini-thoracotomy and without the use of CPB is advantageous option as it was done in our case report [8]. In the largest series reported from the Mayo clinic (9), 50% (13/27) of left side PAPVC were treated with an off-pump minimally invasive approach with similar results than median sternotomy approach. Nevertheless, an obvious limiting factor is the presence of concurrent cardiac anomalies, most commonly significant ASD, that require repair implying median sternotomy and CBP with bicaval cannulation. When left side PAPVC are found to coexist with an ASD, a hybrid approach with endovascular closure of the ASD followed by a minimally invasive surgical repair of the left PAPVC may be a suitable strategy and an appropriate option; a multidisciplinary approach is requested to evaluate the best strategy [9]. There is no evidence that minimally invasive approach has an impact on postoperative complications including atrial fibrillation,

complete heart block or pulmonary venous obstruction [10]. Nowadays, the robotic assistance with minimally invasive approach is an option; its added value is not demonstrated so far [11]. The enlargement of the anastomosis with pericardium patch and the importance of gradient free anastomosis must be emphasized; Intraoperative echocardiography and measurement of anastomotic gradients are mandatory with anastomosis taken down and reconstructed in gradient greater than 4 mm Hg. Regardless the surgical technique used, the postoperative outcome is favourable with fast recovery [7,10]. An empiric postoperative anticoagulation is usually instituted to prevent thrombosis from suture lines in a low-flow state of the pulmonary venous circulation. Our case confirms that when performed correctly left PAPVC repair with minimally invasive approach and off-pump procedure provides excellent outcome.

### Conclusion

Off-pump minimally invasive surgical repair of left side PAPVC is safe, effective and easily reproducible. Possible associated ASD may be treated with hybrid approach and percutaneous closure if indicated.

### References

- Walsh MJ, Ungerleider RM, Aiello VD, Spicer D, Giroud JM. Anomalous pulmonary venous connections and related anomalies: nomenclature, embryology, anatomy, and morphology. *World J Pediatr Congenit Heart Surg.* 2013;4:30-43.
- Türkvatan A, Güzeltaş A, Tola HT, Ergül Y. Multidetector Computed Tomographic Angiography Imaging of Congenital Pulmonary Venous Anomalies: A Pictorial Review. *Can Assoc Radiol J.* 2017;68(1):66-76.
- Majdalany DS, Phillips SD, Dearani JA, Connolly HM, Warnes CA. Isolated partial anomalous pulmonary venous connections in adults: twenty-year experience. *Congenit Heart Dis.* 2010;5:537-45.
- Fragata J, Magalhães M, Baquero L, Trigo C, Pinto F, Fragata I. Partial anomalous pulmonary venous connections: surgical management. *World J Pediatr Congenit Heart Surg.* 2013;4:44-9.
- Stout KK, Daniels CJ, Aboulhosn JA, et al. 2018 AHA/ACC Guideline for the Management of ACHD. *Circulation.* 2019;139:e698-e800.
- Bobylev D, Breyman T, Boethig D, Ono M. Surgical repair

- of partial anomalous pulmonary venous connection shunting from left atrium to innominate vein.
7. J Cardiothorac Surg. 2013;8:100.
  8. Waqar T, Ansari ZA, Raza Baig MA. Outcome after surgical repair of partial anomalous pulmonary venous connection. Pak J Med Sci. 2016;32(6):1386-1389.
  9. Ammannaya GKK, Mishra P, Khandekar JV. Left sided PAPVC with intact IAS-Surgically managed with vertical vein anastomosis to LA appendage: A rare case report. Int J Surg Case Rep. 2019;59:217-219.
  10. ElBardissi AW, Dearani JA, Suri RM, Danielson GK. Left-sided partial anomalous pulmonary venous connections. Ann Thorac Surg 2008;85:1000-14.
  11. Graham G, Dearani JA, Mathew J, et al. Partial anomalous pulmonary venous connection with intact atrial septum: early and midterm outcomes. Ann Thorac Surg. 2022;S0003-4975(22)00663-4.
  12. Onan B, Aydin U, Kadirogullari E, Onan IS, Sen O, Kahraman Z. Robotic repair of partial anomalous pulmonary venous connection: the initial experience and technical details. J Robot Surg. 2020;14:101-107.