Journal of
Cardiology & Cardiovascular
Surgery

Case Report

DOI: 10.51271/JCCVS-0061

Total anomalous pulmonary venous connection: mixed variation [2 (1/2)+1 (1/2)] type: bizarre pattern: a case report

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Cite this article: Loona M, Kakkar RVD, Kumar N, Gusian P. Total anomalous pulmonary venous connection: mixed variation [2 (1/2)+1 (1/2)] type: bizarre pattern: a case report. *J Cardiol Cardiovasc Surg.* 2025;3(3):65-67.

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Received: 01/08/2025 ◆ **Accepted**: 25/09/2025 ◆ **Published**: 27/09/2025

ABSTRACT

Among all variants of total anomalous pulmonary venous connection, mixed variety is a rare congenital anomaly. It comprises 3 different patterns including 2+2, 3+1, bizarre variations. These Bizarre variations are uncommon findings in mixed variety. In this particular case, we report surgical correction of such rare variation comprising [2 (1/2) cardiac+1 (1/2) supracardiac] type of pulmonary venous drainage. These variations impose the importance of preoperative delineation of heterogeneous venous drainage and subsequent planning for surgical repair.

Keywords: Mixed total anomalous pulmonary venous connection, total anomalous pulmonary venous connection, TAPVC

INTRODUCTION

The initial description of total anomalous pulmonary venous connection (TAPVC) was given by Wilson in 1798.[1] This particular cardiac malformation is characterized by absence of direct connection of any pulmonary vein to left atrium. Therefore, the presence of atrial septal defect (ASD) is essential with TAPVC for post natal survival.

In 1959, Darling and associates[2] classified TAPVC variants into 4 types based on pulmonary venous drainage:

- Supra cardiac (45-50%)
- Cardiac (25-30%)
- Infracardiac (25-20%)
- Mixed (5-10%)

Mixed variety though uncommon, demonstrates a wide spectrum of anatomical presentation. In 2007, Chowdhury and colleagues[3] have categorized this wide assortment of mixed TAPVC into three general groups based on heterogeneous of pulmonary venous drainage.

- 2+2 pattern,
- 3+1 pattern, and
- Bizarre pattern.

We hereby report, a rare case of mixed TAPVC with bizarre pattern, with [2 (1/2) cardiac+1 (1/2) supracardiac] pulmonary venous drainage type, being operated for total correction.

CASE

A 2-year-old male child presented with complaints of recurrent respiratory tract infections since 2 months of age. On examination, patient had minimal cyanosis with grade 2 clubbing, and ejection systolic murmur. His vitals were unremarkable. X-ray chest revealed cardiomegaly with pulmonary plethora. The echocardiography revealed pulmonary veins opening into coronary sinus and brachiocephalic vein via the vertical vein. For further delineation of the pulmonary vasculature CT cardiac angiography was performed, which depicted left superior pulmonary vein (LSPV) and a branch from left inferior pulmonary vein (LIPV) draining commonly into brachiocephalic vein via the vertical vein, whereas right superior pulmoner verin (RSPV) and right inferior pulmonary vein (RIPV) along with a branch from left inferior pulmonary vein were found to be draining into coronary sinus (CS) (**Figure 1**).

Surgical Procedure

Standard midline sternotomy was performed. Vertical vein was identified, well mobilized and looped. Pericardium was opened. After standard aortic bicaval cannulation, cardiopulmonary bypass (CPB) was commenced. Aorta was cross clamped and heart arrested using delnido cardioplegia and moderate hypothermia was attained. Right atriotomy was contemplated. Coronary sinus unroofing was performed. Right superior and right inferior pulmonary vein along with a branch from left inferior pulmonary vein was found

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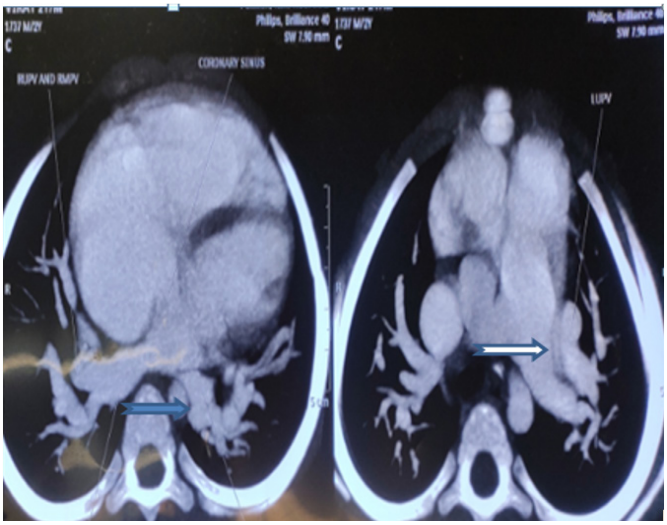

Figure 1. Computed tomography (CT) scan of the patient wherein, the blue arrow indicates drainage of left lower pulmonary venous drainage into cardiac chamber and vertical vein and white arrow indicates formation of vertical vein from left upper and branch of left lower pulmonary vein.

draining into coronary sinus through a common chamber underneath (**Figure 2**). Left vertical vein was divided just near to brachiocephalic vein and mobilized followed by rerouting of vertical vein was done to left atrial appendage. A pericardial patch closure of common chamber along unroofed coronary sinus directed to left atrium was done. Right atriotomy was closed. Patient was weaned off form CPB without any event. Total cardiopulmonary bypass time was 124 minutes and cross clamp time was 98 minutes. The patient was extubated after 8 hours postoperatively and maintained 100% saturation. The postoperative period was uneventful and the patient discharged on post-operative day 5th. Patient was being followed up for 12 months on regular intervals without any significant postoperative problems.

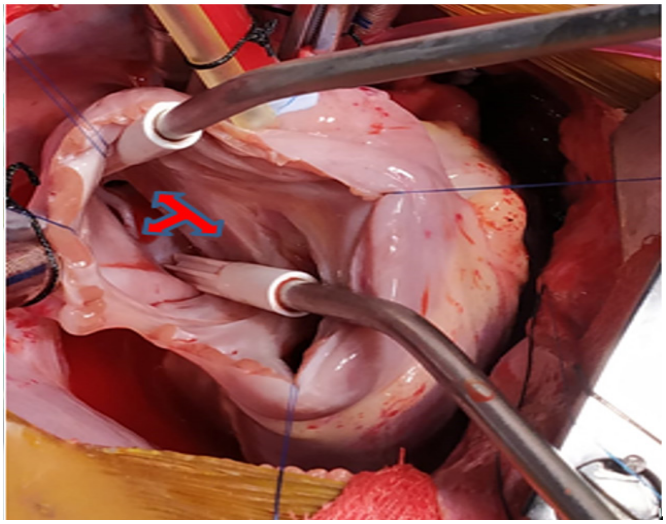

Figure 2. Surgical figure depicting the peculiar anatomy wherein, it indicates formation of vertical vein from left upper and branch of left lower pulmonary vein and the red arrow indicates opening of right upper and lower and branch of left lower pulmonary vein draining into coronary sinus through a chamber.

DISCUSSION

TAPVC is a rare congenital anomaly. Within the spectrum of TAPVC, mixed type variants comprise less than 5-10%.[2] Further as classified by Chowdhury et al.,[3] these mixed variants comprises of three different patterns including 3+1, 2+2 and bizarre with multiple variations of pulmonary venous drainage to heart. Preoperative diagnosis and anatomic delineation remains to be the mainstay for planning surgical treatment of such patients.[5] Echocardiography is a primary choice for diagnosis; however cardiac catheterization provides better results. In non-obstructing pulmonary veins, cardiac catheterization is less likely required, as echocardiography is considered sufficient when three veins are well visualised.[6] Magnetic resonance imaging, CT angiography, and trans-oesophageal echocardiography provides better anatomic delineation of pulmonary venous drainage.[7] Various surgical techniques have been recommended to patients with mixed TAPVC, with individualised approach to variations of 2+2, 3+1, or bizarre pattern.[8-10]

In this case, combined approach for surgical correction were considered, wherein mixed TAPVC with bizarre pattern [2 (1/2)+1 (1/2)] type, having dual LIPV drainage of left inferior lobe as supracardiac and cardiac component were rerouted to drain all the pulmonary venous drainage to left atrium using 'van paragh or mee technique' for (RSPV & RIPV & branch LIPV) and 'posterior approach' for (LSPV& branch LIPV). Surgery carries a high risk of mortality and morbidity, especially in repair of mixed form of TAPVC, having bizarre pattern.

CONCLUSION

A sound characterization of pulmonary venous drainage pattern, preoperative planning and individualization of optimal surgical approach ensures good outcomes.

ETHICAL DECLARATIONS

Informed Consent

The patient's parents have signed the informed consent form.

Referee Evaluation Process

Externally peer-reviewed.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

Financial Disclosure

The authors declared that this study has received no financial support.

Author Contributions

All of the authors declare that they have all participated in the design, execution, and analysis of the paper, and that they have approved the final version.

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