

Atypical Mixed Total Anomalous Pulmonary Venous Connection: A Case Report

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Abstract:

A 2 years old boy presented to us with a history of repeated respiratory tract infections and bluish discoloration of tongue, lips and figure tips for last 18 months. Echocardiography and Computed tomography (CT) angiogram revealed total anomalous pulmonary venous connection (TAPVC) mixed type (supracardiac and cardiac);all pulmonary veins drain into a common

chamber behind left atrium (LA) and left lower pulmonary vein (LUPV) drains to vertical vein and common chamber both. The patient underwent rerouting of pulmonary veins and vertical vein ligation above the drainage of LUPV.

Key words: TAPVC, mixed type

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Introduction:

The occurrence of multiple drainage sites in total anomalous pulmonary venous connection (TAPVC) has important implications in preoperative diagnosis and surgical treatment. Although the surgical outcome of total anomalous pulmonary venous connection (TAPVC) has improved, repair of mixed-type TAPVC is still technically challenging.¹⁻⁴ The pattern of the pulmonary venous drainage has a wide variety, most of the patients had confluence of three pulmonary veins (major drainage) and a single pulmonary vein connected to the systemic vein independently (minor drainage).² Here, we report a case where the minor drainage has dual connection both with vertical vein and common chamber.

Case Report:

A 2 years old boy presented to us with a history of repeated respiratory tract infections and bluish discoloration of tongue, lips and figure tips for last 18 months. He has history of

hospitalization for respiratory distress at the age of 7 months. Physical examination revealed cyanosis (SPO₂ 85%), grade I clubbing and systolic murmur (grade 3/6) at pulmonary area. A routine chest X-ray showed cardiomegaly, widening of superior mediastinum and pulmonary plethora with normal viscera-bronchial situs with gastric air bubble on the left side and a liver shadow on right side. In echocardiography, the patient had situs solitus and levocardia; TAPVC mixed type (supracardiac and cardiac), and all pulmonary veins drain into a common chamber behind left atrium (LA) and LUPV drains to vertical vein and common chamber both (Figure 1). CT angiogram revealed mixed TAPVC and confirmed the communication between vertical vein and common chamber (Figure 2). With these investigations, the patient was taken up for surgery.

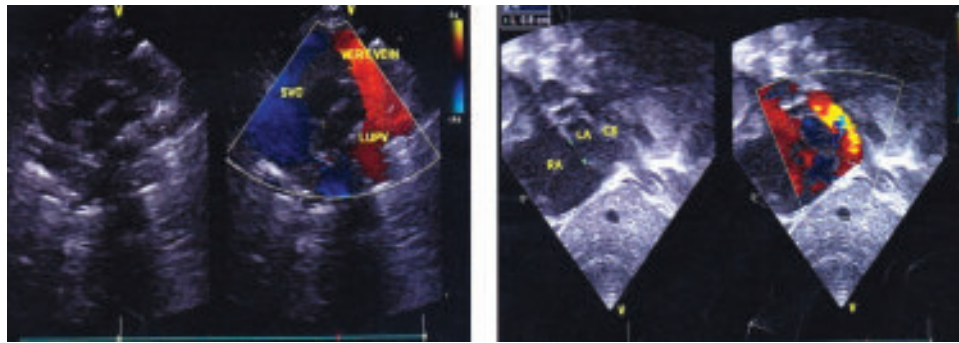
After induction, and establishment of standard invasive and noninvasive monitoring median sternotomy was done. Anatomy was checked preoperatively. Extra pericardial vertical vein was dissected free up to the common chamber and LUPV drainage site. An incision is made in the right atrium parallel to the atrioventricular groove. The large coronary sinus is identified, which is the point of the anomalous pulmonary venous connection. The membrane of the foramen ovale is excised to provide a large opening in the atrial septum. The common wall of the coronary sinus and left atrium is incised by placing scissors through the foramen ovale. The incision is made well back into the left

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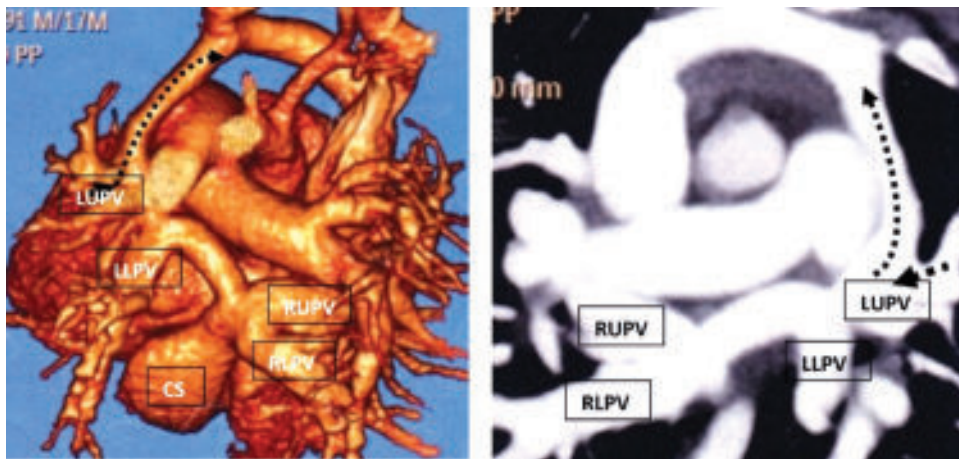
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A B
Fig.-1 (a,b): Preoperative Echocardiography



A B
Fig.-2 (a,b): CT angiogram

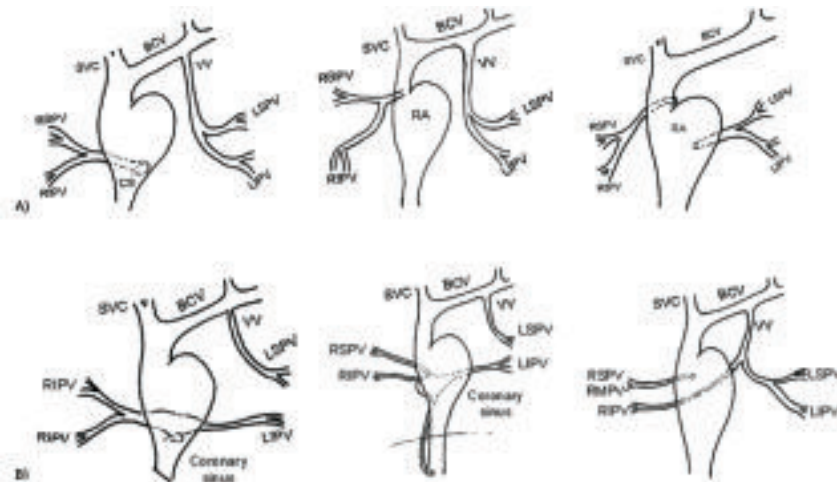


Fig.-3 (A, B): A: bilateral and symmetrical connections (“2+2” pattern of drainage); B: bilateral and asymmetrical connections (“3+1” pattern of drainage; BCV, Brachiocephalic vein; CS, coronary sinus; LIPV, left inferior pulmonary vein; LSPV, left superior pulmonary vein; RA, right atrium; RIPV right inferior pulmonary vein; RSPV, right superior pulmonary vein; SVC, superior vena cava; VV, vertical vein.

atrium, to allow free drainage of the coronary sinus into the main portion of the left atrium. The atrioventricular node and bundle of His should be anterior to the incision and protected from injury. An appropriately sized pericardial patch is fashioned from the anterior pericardium to close the atrial septum. Finally, ligation of vertical vein was done just above the insertion of LUPV at vertical vein under standard Cardiopulmonary bypass (CPB) and cooled to 30°C. Postoperative recovery was uneventful.

Discussion:

Mixed type of TAPVC is a rare cardiac anomaly, the frequency of which is 5% to 10% of the patients with TAPVC in the literature.⁴ Although the surgical outcome of TAPVC correction has improved, but repair of mixed TAPVC is still technically challenging. In most cases of TAPVC, echocardiography is sufficient for diagnosis. Some authors have stated that cardiac catheterization may be unnecessary if three veins are well visualized and there is no clinical evidence of obstruction.⁴ Apical and subcostal 4-chamber echocardiographic views usually best identify individual pulmonary veins and their confluence.^{5,6}

Mixed TAVCs are allocated into three categories. Category I: Bilateral & symmetrical connection, “2+2” pulmonary venous drainage pattern (Figure 3a); category II: Bilateral & asymmetrical connection, “3+1” pulmonary venous drainage pattern (Figure 3b) & category III: Bizarre anatomic variants.¹

In this case of mixed TAPVC, the left upper pulmonary vein is left unrepaired, because it has a dual communication between vertical vein and common chamber. Though, we left it untreated, it can drain easily into common chamber. Kasama and colleagues reported two stage approach for mixed TAPVC due to single connection of LUPV to vertical vein.⁷ Delius and colleagues stated that if there is evidence that the isolated anomalous pulmonary vein of mixed TAPVC is obstructed, anastomosis to the left atrium is mandatory, but the vein may be left uncorrected if it is not obstructed⁴. So, if LUPV had no dual connection, it can be left untreated. In our case the patient is lucky enough due to dual connection of LUPV.

Conclusion:

Mixed type of TAPVC is a rare cardiac anomaly and has a wide variety in pulmonary venous anatomy. There seems to be a limitation in diagnosis by echocardiography only; CT angiogram is a good alternative and recommended when mixed-type TAPVC is suspected.

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