SL Journal of Anesthesia & Critical Care

Case Report

Tetralogy of Fallot with Total Anomalous Pulmonary Venous Connection

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ARTICLE INFO

Received Date: April 25, 2022
Accepted Date: May 23, 2022
Published Date: May 26, 2022

KEYWORDS

Tetralogy of fallot
Total anomalous pulmonary venous connection
Cardiac imaging

Citation for this article: Ajmer Singh and Aashish Jain. Tetralogy of Fallot with Total Anomalous Pulmonary Venous Connection. SL Journal of Anesthesia & Critical Care. 2022; 3(1):119

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ABSTRACT

A 15-month-old baby presented with increasing cyanosis and clubbing. His chest X-ray showed '*coeur en sabot*' and 'figure-of-eight' appearance. The diagnosis of tetralogy of Fallot with total anomalous pulmonary venous connection, an extremely rare combination, was confirmed with echocardiography and computed tomography scan. The significance of correct diagnosis; and potential merits and demerits of various cardiac imaging modalities are discussed in brief.

INTRODUCTION

The association of Tetralogy of Fallot (TOF) with Total Anomalous Pulmonary Venous Connection (TAPVC) is not only uncommon but rare. The incidence of such a combination is reported as 0.25% to 0.34% [1]. The clinical features of this association are likely to be similar to TOF. In the earlier days, the diagnosis was often suspected by echocardiography and confirmed by cardiac catheterization. Computed Tomography Angiography (CTA) and magnetic resonance imaging are now commonly used modalities to delineate pulmonary venous drainage. An early complete anatomic repair of both lesions is desirable for optimal outcomes.

CASE HISTORY

A 15-months-old baby, body weight 8 kg, was admitted to the hospital with increasing cyanosis and clubbing. There was no history of cyanotic spells, failure to gain weight, recurrent respiratory infections, or congestive heart failure. An ejection systolic murmur of III/VI severity was heard over the upper left sternal border. Electrocardiogram showed right axis deviation, right atrial hypertrophy (tall p waves, p-pulmonale), and right ventricular hypertrophy. Chest x-ray revealed right ventricular apex pattern, '*Coeur en sabot*' appearance, 'figure-of-eight' configuration, and normal pulmonary vascular markings (Figure 1). Two-dimensional transthoracic echocardiography with color and spectral Doppler established the diagnosis of tetralogy of Fallot. It showed a large non-restrictive perimembranous Ventricular Septal Defect (VSD) with bidirectional shunt, aortic override of 50%, right ventricular hypertrophy, severe infundibular stenosis, and hypoplastic main pulmonary artery (Figure 2A,2B). In addition, there was supracardiac total anomalous pulmonary venous connection with the pulmonary venous confluence forming a vertical vein and draining into the right atrium through the innominate vein and superior vena cava (Figure 2C). A 9 mm Atrial Septal Defect (ASD) with a right to left shunt was also seen. CTA with a dual-source 256-slice multidetector scanner and volume-rendering technique was consistent with echocardiography findings; and it clearly delineated pulmonary veins

draining into the vertical vein, innominate vein, dilated superior vena cava, and right atrium (Figure 3A,3B). Under standard cardiac monitoring and balanced anesthetic technique, the patient underwent combined repair for TOF and TAPVC under hypothermic Cardiopulmonary Bypass (CPB). The surgical procedure consisted of connection of pulmonary venous confluence to the left atrium through a standard posterior approach, ligation of the vertical vein, dacron patch closure of ASD and VSD, resection of infundibular stenosis, and placement of trans-annular patch with monocusp pulmonary valve. Aortic cross-clamp duration was 90 minutes and the CPB duration was 112 minutes. Weaning from CPB was facilitated with epinephrine 0.07 µg/kg/min and milrinone 0.5 µg/kg/min, and the inotropes were continued for 72 hours. Immediate postoperative course and subsequent stay in the hospital were uneventful.

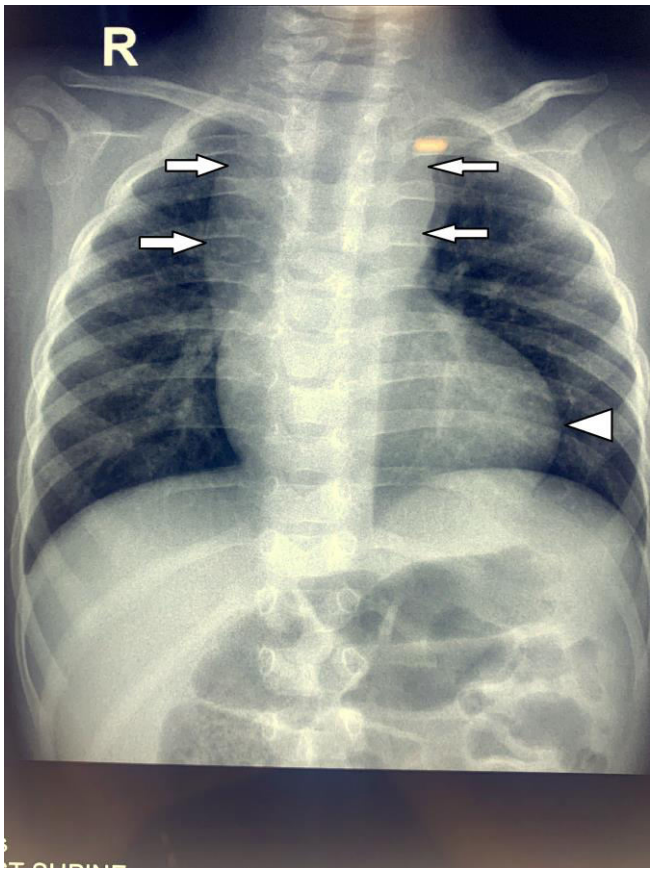

Figure 1: Chest X-ray demonstrating 'figure of-eight' configuration (arrows), and 'coeur en sabot' appearance (arrow head).

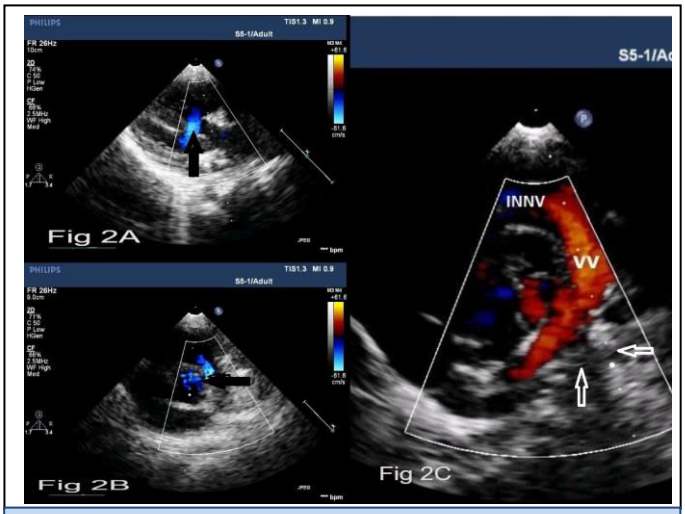

Figure 2: Two-dimensional transthoracic echocardiography showing perimembranous ventricular septal defect (arrow) in parasternal short-axis view (Figure 2A), infundibular stenosis (arrow, Figure 2B), and pulmonary venous confluence (arrows) draining into vertical vein (VV) and Innominate Vein (INNV) in suprasternal short-axis view (Figure 2C)

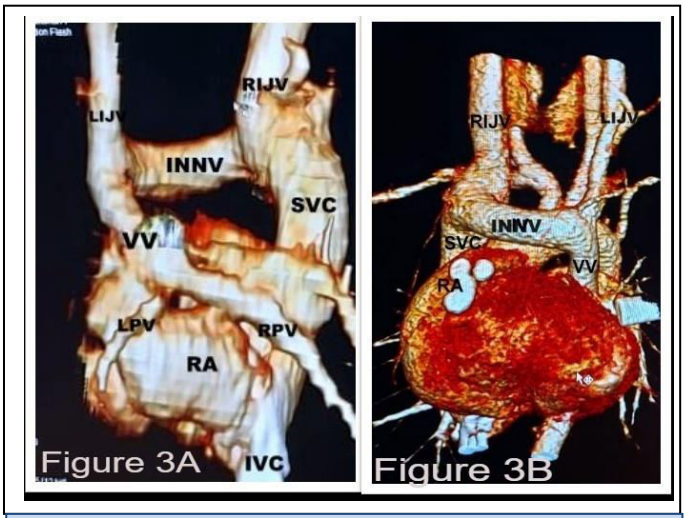

Figure 3: Volume-rendered technique of computed tomography (posterior projection, Figure 3A) demonstrating Left and Right Pulmonary Veins (LPV, RPV) draining into Vertical Vein (VV), Innominate Vein (INNV), Superior Vena Cava (SVC), and Right Atrium (RA).

DISCUSSION

The case described herein shows the importance of correct preoperative diagnosis and its impact on the management in a patient with a rare combination of TOF and TAPVC. The common anomalies associated with TOF include ASD, persistent left superior vena cava, patent ductus arteriosus, pulmonary atresia, additional VSD, and atrioventricular septal defect. The combination of TOF with TAPVC is not only rare but presents a unique challenge in the diagnosis and perioperative management. There are only a few case reports and case-

series reports of TOF with the TAPVC combination. Talwar et al have reported a total of 25 cases of TOF with TAPVC in the entire medical literature before their publication [2]. Clinical presentation of this particular association is generally similar to TOF, and symptoms of TAPVC may never appear because pulmonary stenosis limits pulmonary blood flow and does not allow elevation of pulmonary venous pressure [3]. Rarely, when the pulmonary stenosis is mild, features of TAPVC may dominate. A single-stage correction of both anomalies is recommended if the left ventricle is well developed. Conversely, if the left ventricle is underdeveloped, a two-stage procedure involving correction of TAPVC with a systemic-pulmonary shunt at the first stage and correction of TOF at a later date is advised [4]. If TAPVC is not detected and corrected, it can result in intractable and fatal pulmonary edema, and in difficulty in weaning the patient from CPB [5].Therefore, detailed delineation of correct anatomy is crucial in surgical planning for these patients. The immediate post CPB period can be complicated by small left atrium and left ventricle, and right ventricle failure secondary to pulmonary regurgitation. Left atrial pressure monitoring may be required, as these patients are very sensitive to volume overload. Optimal inotropic support and after load reduction should be considered before discontinuation of CPB [6].

Liu and colleagues have shown that CTA is superior to echocardiography and catheter angiography in the evaluation of TAPVC, especially in patients with complex congenital cardiovascular anomalies [7]. A volume-rendered technique with three-dimensional reconstruction of CTA images helps in better delineation of the anatomy, because of its high spatial and temporal resolution. Moreover, it helps in identifying airway abnormalities and delineating atypical vessels draining into the systemic veins. Cardiac catheterization is limited by its invasiveness, need for sedation/anesthesia, and the risk of radiation exposure. Magnetic resonance imaging and CTA are now playing larger roles in the diagnosis of these anomalies [8,9]. These imaging methods help overcome the potential pitfalls of transthoracic echocardiography, like a suboptimal acoustic window and/or a poor depiction of extracardiac vascular structures.

CONCLUSION

A rare association of TOF with TAPVC is described highlighting the importance of accurate diagnosis, choosing the appropriate modality for investigation, and an individual-based plan of management.

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