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A Rare Case of Total Anomalous Pulmonary Venous Connection in an Adult

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Abstract: ***Total Anomalous Pulmonary Venous Connection (TAPVC) is an uncommon congenital heart condition found infrequently among adults. Here, we discuss the case of a 32 - year - old male who presented with a history of progressive dyspnea and fatigue. Diagnostic evaluation revealed TAPVC with abnormal venous drainage into the right atrium, posing significant hemodynamic challenges. Surgical intervention was promptly undertaken to redirect the pulmonary veins to the left atrium, restoring normal blood flow. This case highlights the critical importance of considering congenital heart anomalies in adults presenting with cardiac symptoms, and emphasizes the complexities and specific management strategies required for TAPVC in this age group. Early detection through comprehensive imaging studies and prompt surgical correction are crucial in achieving favorable outcomes for adult patients diagnosed with TAPVC.***

Keywords: adult tapvc, anomalous venous drainage, pulmonary vein anomaly, adult congenital heart disease, tapvc

1. Introduction

Total Anomalous Pulmonary Venous Connection (TAPVC) is an uncommon congenital heart defect characterized by abnormal connections between the pulmonary veins and the heart. While typically identified and treated during infancy or childhood, occurrences in adulthood are exceedingly rare. This article presents a case study of a 32 - year - old male who presented with progressive dyspnea and fatigue. Diagnostic assessment revealed TAPVC with anomalous venous drainage into the right atrium, presenting considerable challenges in adult cardiology. This case emphasizes the complexities involved in diagnosing and managing congenital heart defects in adults, underscoring the critical role of early detection and appropriate intervention to enhance patient outcomes. The management strategies and clinical implications of TAPVC in adults will be explored to provide comprehensive insights into this infrequently encountered condition.

There are 4 types of TAPVC - Supracradiac (type I), Cardiac (type II), Infracardiac (type III) and Mixed (type IV).

In supracradiac TAPVC in which all 4 pulmonary veins join to form a vertical vein which drains into the right atrium.

In the cardiac (or "heart") type of TAPVC, also known as "intracardiac" TAPVC, the pulmonary veins drain into the right atrium directly or via a common pulmonary vein that connects to the right atrium.

In infracardiac type of tapvc, all four pulmonary veins join to form a common vein, which extended inferiorly below the diaphragm before it drained into the inferior vena cava, a type III total anomalous pulmonary venous return (TAPVR) [1].

In the "mixed" type of Total Anomalous Pulmonary Venous Connection (TAPVC), the pulmonary veins drain into more than one systemic venous structure, combining features from different types of TAPVC.
In this article, we will discuss a case of 32- year old male with supracardiac type of TAPVC (type I).

2. Case Presentation

A 32 - year - old man presented to the cardiology clinic with progressive shortness of breath, severe fatigue, and recent onset of clubbing and cyanosis of the extremities. He had no significant medical history but noted worsening symptoms over several months.

During examination, the patient appeared fatigued, with noticeable clubbing of his fingers and toes. Cyanosis was evident in his nail beds and lips. Auscultation revealed decreased breath sounds and a mild systolic murmur along the left sternal border.

Suspecting a cardiac origin, diagnostic tests were initiated. Electrocardiography showed sinus tachycardia.

Initial radiological investigation done was X - ray which showed enlarged cardiac silhouette, enlarged right hilum and superior mediastinum with 'figure of eight' appearance.

Echocardiography revealed a large atrial septal defect and further radiological investigations were carried out.

Computed tomography (CT) angiography confirmed the diagnosis, delineating the abnormal venous connections. All the four pulmonary veins were seen draining into the right atrium. The blood from right atrium was draining into the right ventricle via Tricuspid valve and to the left atrium via a large Atrial septal defect, which was the cause of cyanosis in this patient. This large atrial septal defect act as a boon in this patient for his survival into adulthood by the flow of oxygenated blood from right atrium to left atrium and then into the systemic circulation. The trachea was seen compressed between the vertical vein anteriorly and aorta

posteriorly which could be the cause of dyspnea in this patient. The findings were seen supporting the need for surgical evaluation. The patient was promptly referred to a cardiothoracic surgeon.

Given the hemodynamic significance of TAPVC, surgical correction was recommended to restore normal pulmonary venous drainage into the left atrium. The procedure was successfully performed, and the patient recovered without complications. Follow - up evaluations, including echocardiography, demonstrated effective surgical correction with improved oxygen levels and resolution of cyanosis.

This case underscores the diagnostic complexities and treatment challenges of congenital heart defects presenting in adulthood. Early recognition of symptoms such as clubbing and cyanosis is critical for identifying underlying cardiac anomalies like TAPVC, highlighting the importance of timely intervention to optimize patient outcomes and improve quality of life.

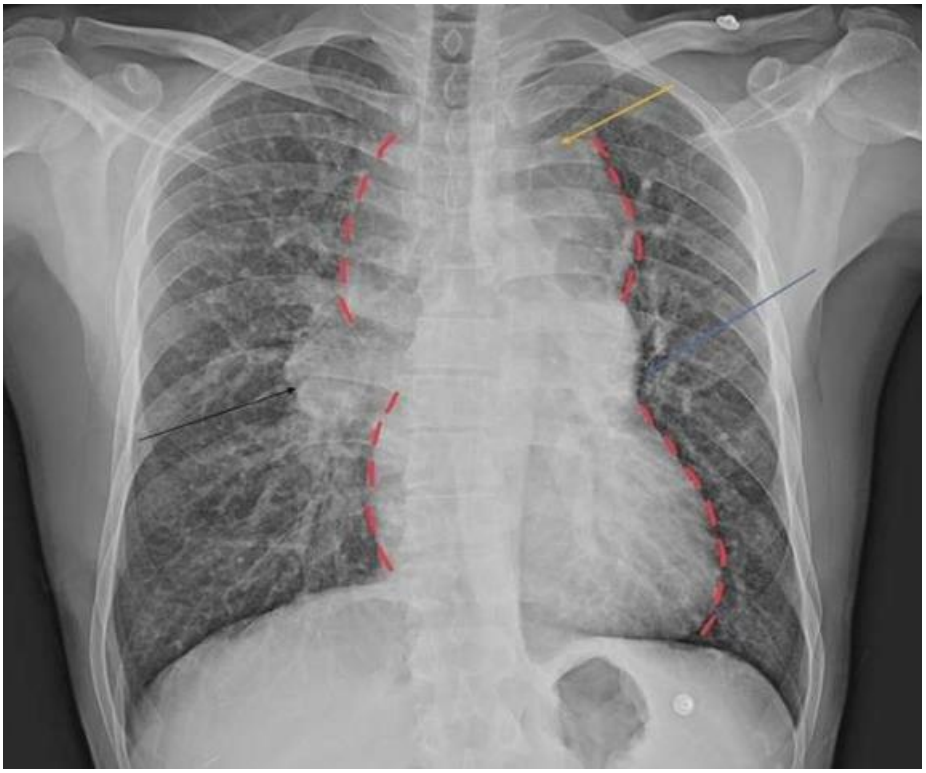

Figure 1: Chest Radiograph

Demonstrates the 'snowman' or 'figure-of-eight' appearance (dotted red outline) characteristic of Total Anomalous Pulmonary Venous Connection (TAPVC). The heart silhouette is enlarged (Blue arrow). The superior mediastinum (yellow arrow) and right hilum (Black arrow) are enlarged.

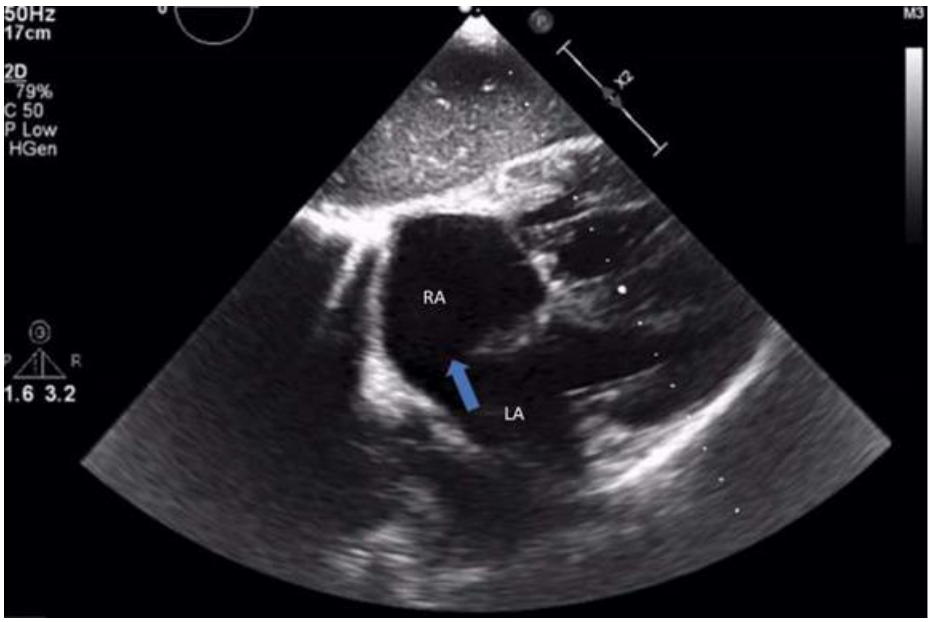

Figure 2: Echocardiography

Depicting an atrial septal defect (ASD) in the heart. The defect is seen as a communication (blue arrow) between the right atrium (RA) and left atrium (LA), allowing blood flow from the right atrium to the left atrium. The chambers are labelled for orientation: RA (right atrium) and LA (left atrium). This visualization confirms the presence of an ASD, facilitating diagnostic assessment and treatment planning for this congenital heart anomaly."

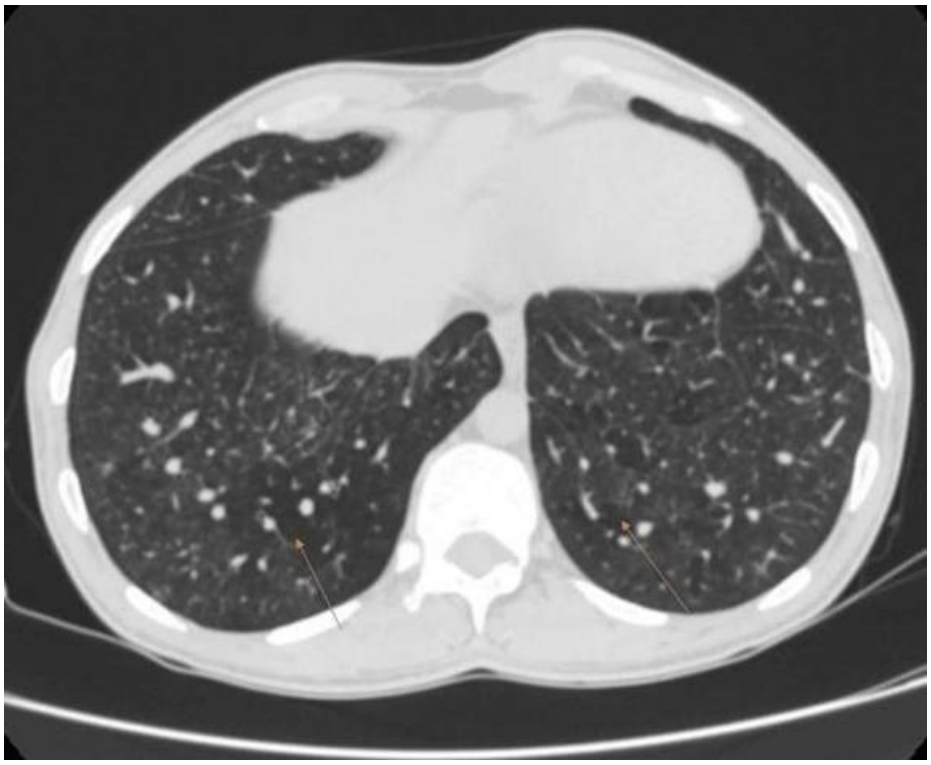

Figure 3: HRCT done for breathlessness.
which showed few mosaic attenuation (orange arrows) in bilateral lung fields which were likely due to airway trapping.

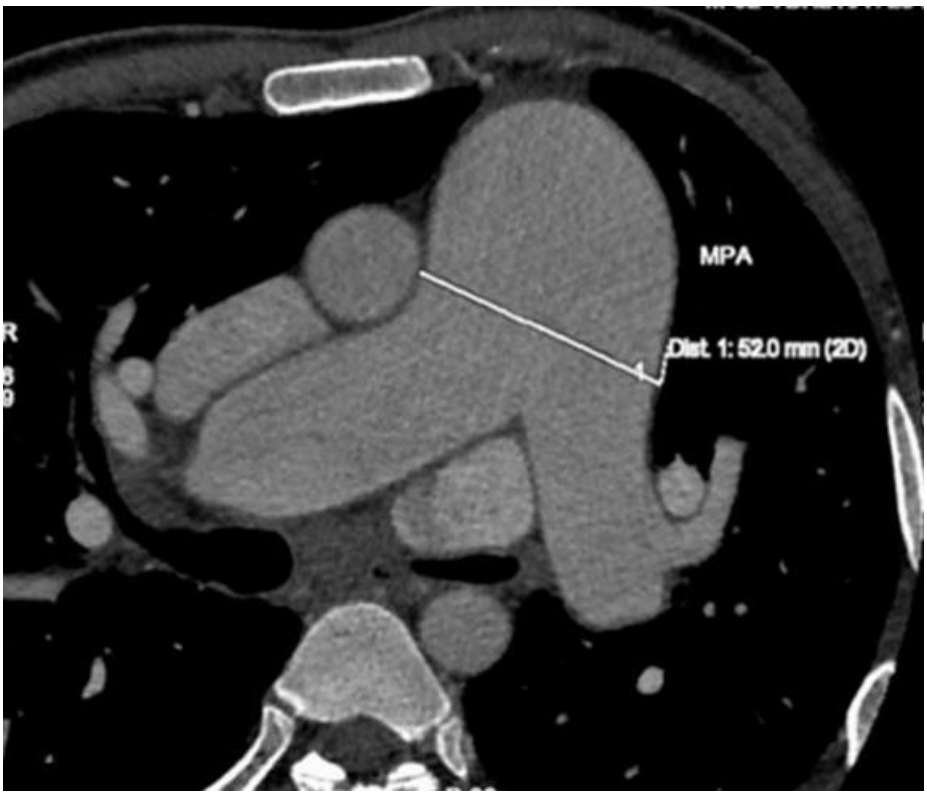

Figure 4: Axial chest CT at the level of pulmonary artery trunkbifurcation
The right pulmonary artery (RPA) and left pulmonary artery (LPA) are dilated. Main pulmonary artery (MPA) is severely dilated and measures 52 mm.

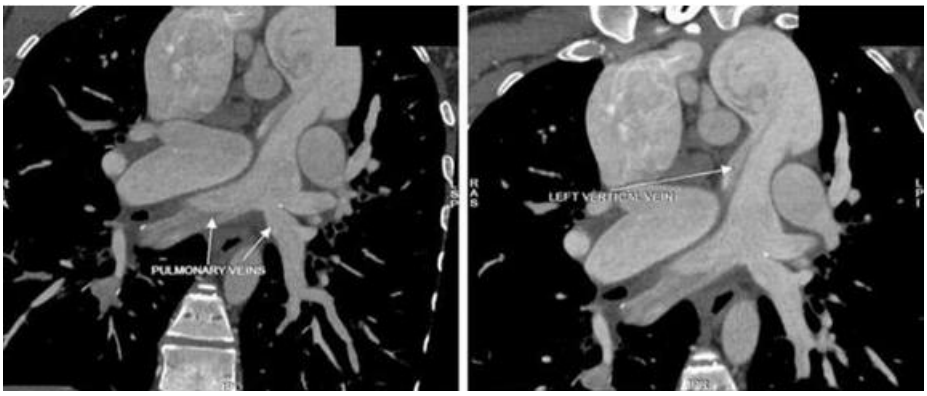

Figure 5: Coronal sections of cardiac CT
(A) Pulmonary veins seen uniting posterior and superior to the left atrial appendage (B) to form a single ascending left vertical vein.

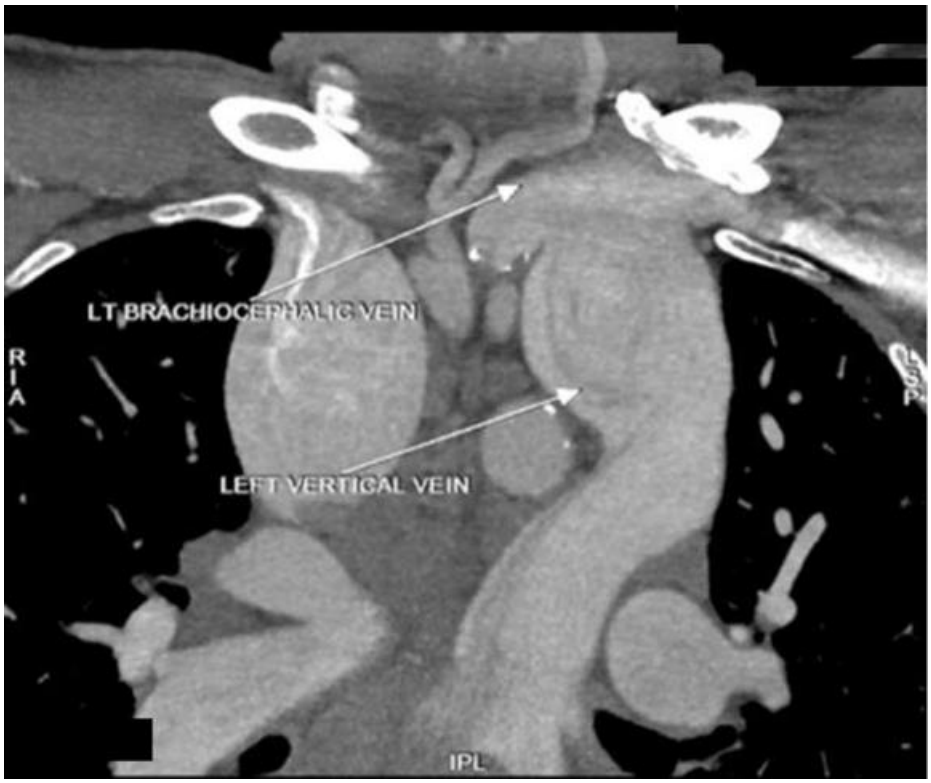

Figure 6: Coronal cardiac CT
The united left vertical vein (innominate vein) which was formed by the confluence of all the 4 pulmonary veins can be seen draining into the left brachiocephalic vein.

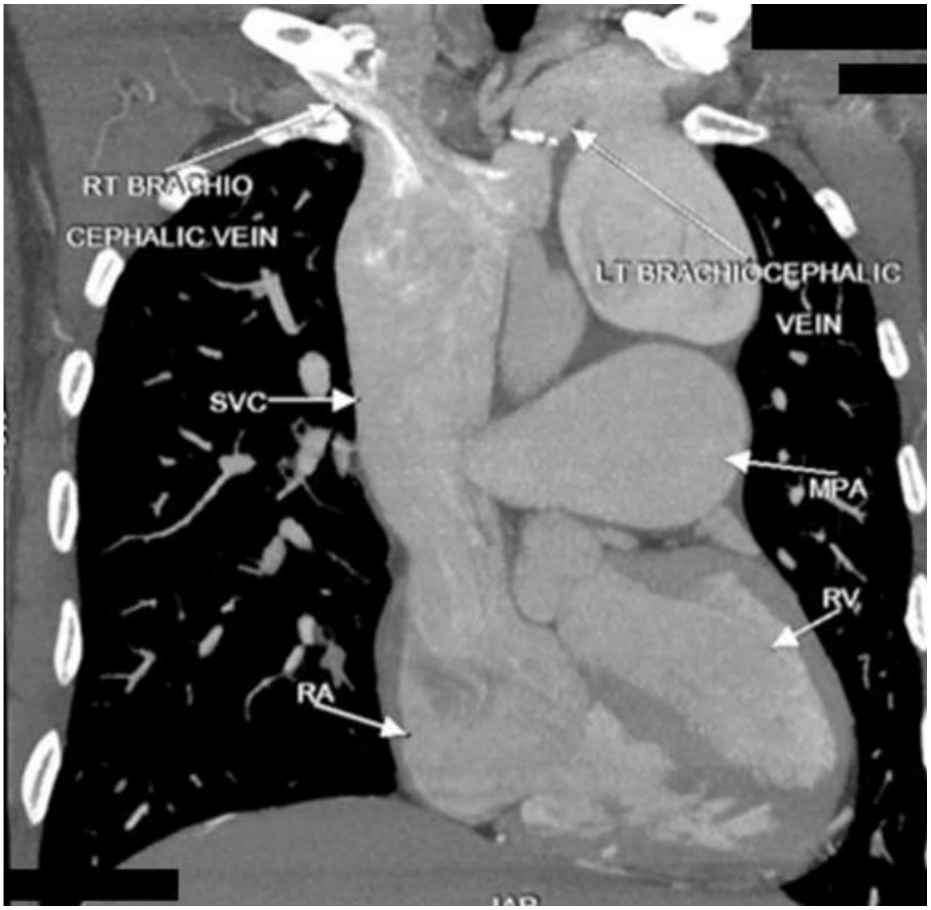

Figure 7: Coronal cardiac CT

The left brachiocephalic vein can be seen joining the (SVC) superior vena cava which is finally draining into the (RA) right atrium.
MPA - Main pulmonary artery
RV - Right ventricle

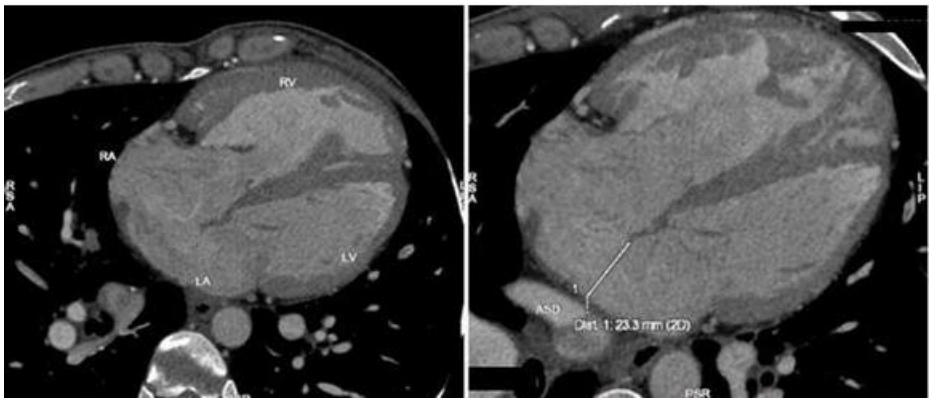

Figure 8: Axial cardiac CT with 4 chamber view

A large ASD (white line) is visualised measuring 23 mm. Right ventricle (RV) and Right atrium (RA) are dilated with evidence of Right ventricular (RV) hypertrophy.
LA - Left atrium
LV - Left ventricle

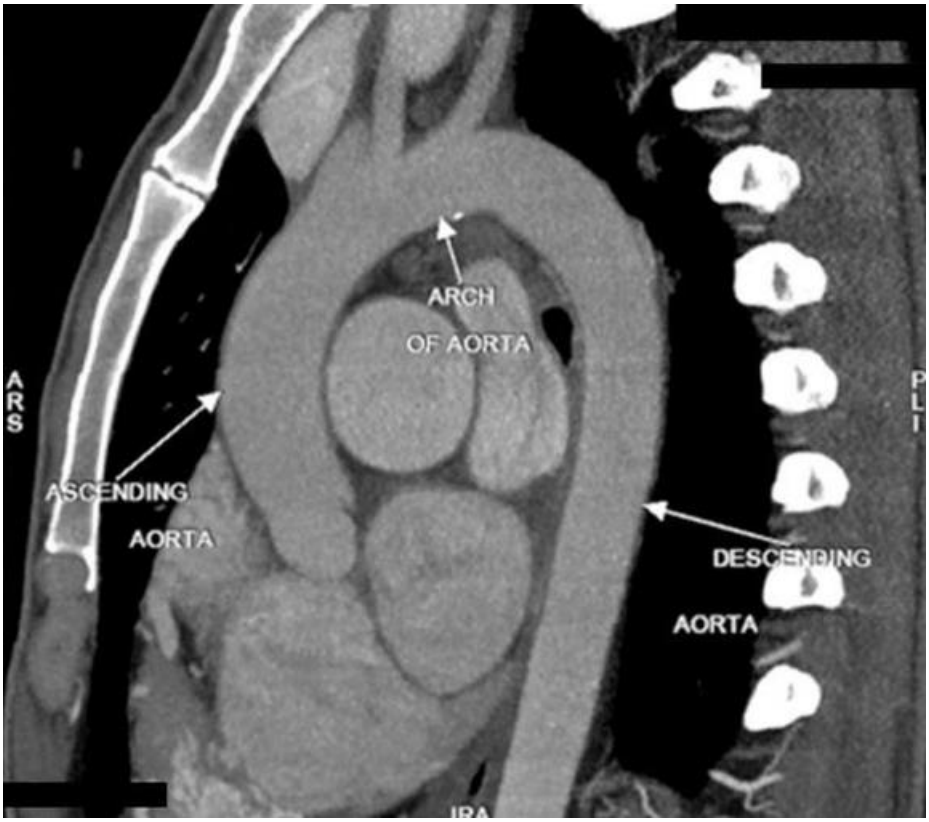

Figure 9: CT thorax sagittal view
The aortic root, ascending aorta, arch of aorta and descending thoracic aorta are normal in caliber

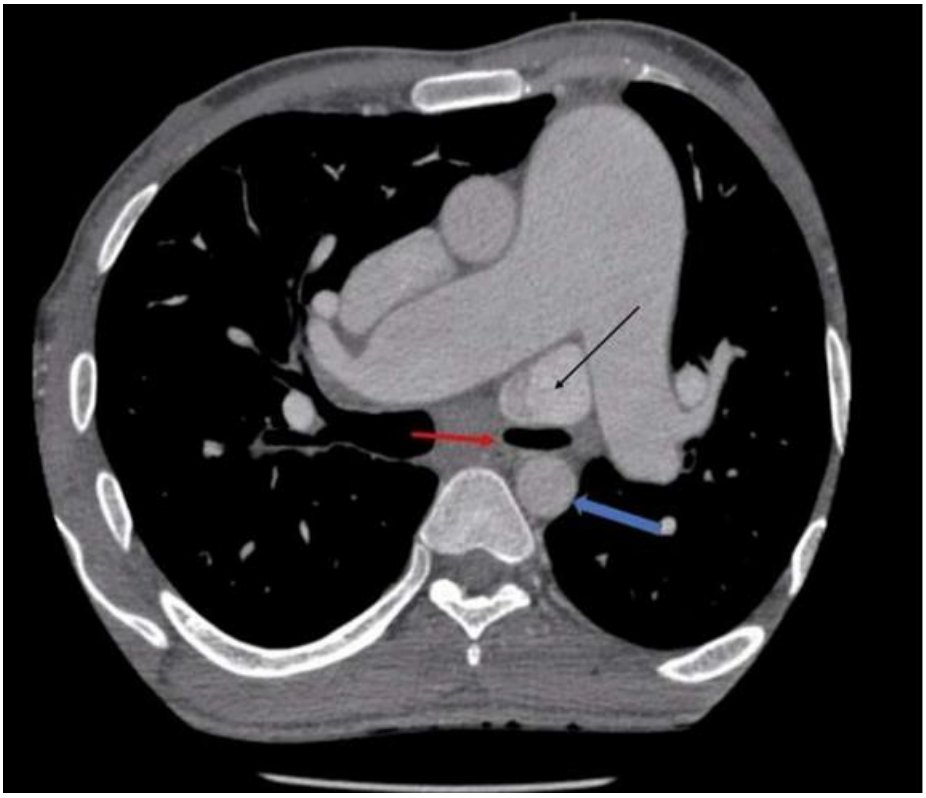

Figure 10: Axial Non-enhanced CT thorax at the level of bifurcation ofpulmonary artery and trachea
The trachea can be seen compressed (red arrow) between the vertical vein (black arrow) anteriorly and aorta (blue arrow) posteriorly. This could be one of the factor contributing to difficulty in breathing.

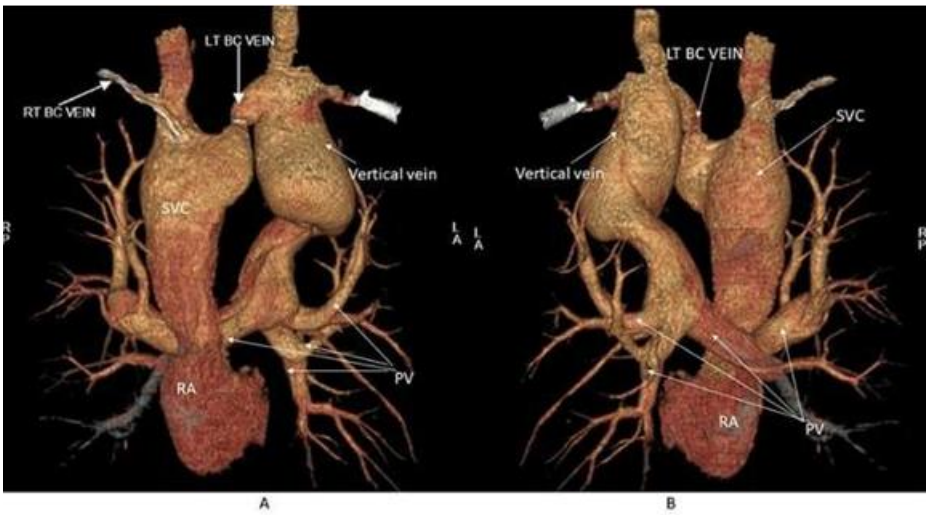

Figure 11: Volume rendering from a helical thoracic CT angiographyscan show anterior (A) & posterior (B) views of all the 4 pulmonary veins (PV) uniting to form Left ascending vertical vein (innominate vein) which is joining the left brachiocephalic vein. The left brachiocephalic vein (BC vein) is draining into the superior vena cava (SVC) which is finally draining into the (RA) right atrium. The left vertical vein and superior vena cava (SVC) appear grossly dilated.

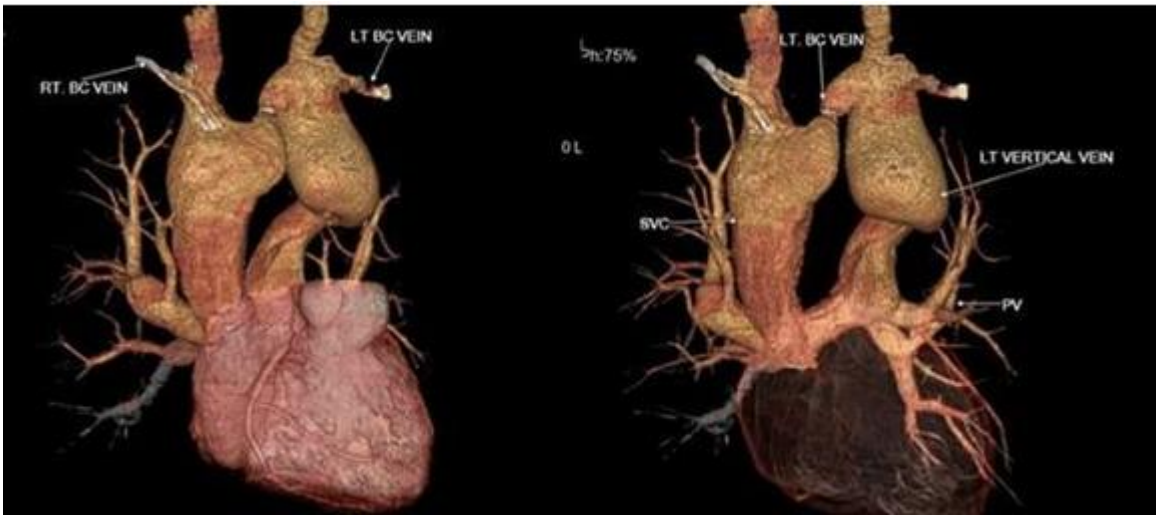

Figure 12: Cardiac 3D CT anterior view with all four chambers and Pulmonary veins (PV) with features as described in figure 11 (A) & (B).

3. Discussion

The case of a 32 - year - old male presenting with symptoms of shortness of breath, fatigue, cyanosis, and clubbing highlights the clinical complexity of Total Anomalous Pulmonary Venous Connection (TAPVC), a rare congenital heart defect. In this individual, the combination of symptoms, including cyanosis and clubbing, suggested chronic hypoxia, which prompted a thorough diagnostic workup.

Various imaging modalities played a crucial role in confirming the diagnosis of TAPVC. Transthoracic echocardiography provided initial insights, revealing a large atrial septal defect (ASD) with enlarged right atrium. Radiographs showed 'snowman' or 'figure of eight' appearance. These findings were corroborated by computed tomography (CT), which detailed the precise anatomical abnormalities, including the anomalous connection of pulmonary veins and the enlarged right atrium. These imaging studies were instrumental in guiding clinical decision - making and planning for surgical intervention.

Research indicates that the survival rates of individuals with Total Anomalous Pulmonary Venous Connection (TAPVC) have significantly improved in recent decades, owing to advancements in medical and surgical interventions. Historically, untreated cases of TAPVC in infancy and childhood have been associated with high mortality rates (~80%) [2]. However, in our case study, the patient survived into adulthood without requiring medical or surgical intervention, primarily due to the simultaneous presence of a large atrial septal defect (ASD). This defect facilitated both oxygenated and deoxygenated blood flow from the right atrium to the left atrium. The resulting cyanosis and clubbing were attributed to the circulation of partially oxygenated blood into the systemic circulation.

The classification of Total Anomalous Pulmonary Venous Connection (TAPVC) includes four types: Supracardiac, Cardiac, Infracardiac, and Mixed, based on how the pulmonary veins drain. In our case involving a 32 - year - old male, the TAPVC is classified as supracardiac. Here, all four pulmonary veins converge to form a left ascending vertical vein, which then connects with the superior vena cava (SVC)

before draining into the right atrium [3]. The supracardiac variety is the most common of the TAPVC [4]. The left ascending vertical vein is also known as innominate vein. CT imaging plays a crucial role in accurately identifying the specific type of TAPVC due to its ability to visualize detailed anatomical structures.

Managing Total Anomalous Pulmonary Venous Connection (TAPVC) in adults presents distinct challenges when compared to pediatric cases, primarily due to delayed diagnosis and potential complications stemming from prolonged strain on the right heart chambers. The primary approach to treatment continues to be surgical correction, which focuses on reestablishing normal circulation by redirecting pulmonary venous flow to the left atrium, thereby enhancing oxygenation and cardiovascular function.

Furthermore, the uncommon occurrence of TAPVC in adults underscores the importance of heightened awareness among healthcare providers. It is crucial for clinicians to promptly consider and accurately diagnose TAPVC in patients who present with unexplained hypoxia or cardiovascular symptoms.

4. Conclusions

In conclusion, managing Total Anomalous Pulmonary Venous Connection (TAPVC) in adults presents unique challenges, including delayed diagnosis and potential complications arising from prolonged cardiovascular strain. Advanced imaging techniques like CT are crucial for accurately diagnosing TAPVC type and guiding surgical interventions, which remain pivotal in restoring normal pulmonary venous circulation and enhancing oxygenation. Our case study of the 32 - year - old male underscores the importance of early detection and comprehensive care in achieving favorable outcomes. Ongoing research and clinical vigilance are vital to refining diagnostic methods and optimizing treatment strategies for adults affected by this complex congenital heart anomaly. By raising awareness and advancing treatment standards, we aim to improve quality of life and prognosis for individuals living with adult - onset TAPVC.

The characteristic important features for long - term outcome without operation are a large atrial septal defect as in this case, absence of pulmonary venous obstruction and a normal left ventricular end - diastolic pressure.

Additional Information

Disclosures

Human subjects: Consent was obtained or waived by all participants in this study.

Conflicts of interest: In compliance with the ICMJE uniform disclosure form, all authors declare the following:

Payment/services info: All authors have declared that no financial support was received from any organization for the submitted work.

Financial relationships: All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work.

Other relationships: All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

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