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# Persistent Cyanosis Revealing Supracardiac Total Anomalous Pulmonary Venous Return in an Infant: A Case Report

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## Abstract

Total anomalous pulmonary venous return is a rare congenital cardiac malformation and the supracardiac form is the most common subtype. The clinical appearance depends on the presence or absence of venous obstruction, which is a major prognostic factor. We report the case of a 2-month-old infant with persistent cyanosis from birth. On clinical examination, the patient had generalized cyanosis with a peripheral oxygen saturation of 60% on room air. Pulmonary hypertension, dilatation of the right ventricular chambers, and an atrial septal defect with a right-to-left shunt were all shown by transthoracic echocardiography. Thoracic computed tomography angiography confirmed supracardiac total anomalous pulmonary venous return with drainage of the pulmonary venous confluence via a vertical vein into the left brachiocephalic vein associated with hypoplasia of the left cardiac chambers. After the patient was stabilized, surgical intervention was recommended. This case illustrates the diagnostic value of a multimodal imaging approach, where echocardiography and CT angiography work synergistically to define the precise anatomy of supracardiac TAPVR and guide surgical referral. The unfortunate evolution in the postoperative period highlights the still present issue of late presentation and hemodynamic deterioration. It reminds us that the success of a surgical procedure relies not only on technical expertise but also on timely diagnosis and intervention.

## Keywords

Total Anomalous Pulmonary Venous Return, Congenital Heart Disease, Thoracic CT Angiography, Pulmonary Hypertension

## 1. Introduction

Total anomalous pulmonary venous connection (TAPVC) is a rare cyanotic abnormality that accounts for about 1% - 3% of congenital heart disease malformations [1]. It is characterized by an abnormal drainage of the pulmonary veins into the systemic venous circulation, preventing their connection to the left atrium. According to the site of anomalous drainage, TAPVC is generally categorized into supracardiac, cardiac, infracardiac, and mixed types [2].

The most prevalent variety, accounting for around 45% - 55% of cases, is the supracardiac type [3]. Clinical manifestations range from acute neonatal respiratory distress to later non-obstructive forms, depending on whether venous blockage is present or not [4].

Echocardiography is the main diagnostic tool, and cross-sectional imaging is used to improve anatomical characterization [5]. Treatment is surgical and should be performed urgently in cases of obstruction [6].

We report a case of supracardiac total anomalous pulmonary venous return diagnosed in an infant managed at Mohammed VI University Hospital, Oujda, Morocco, highlighting the clinical features and the contribution of imaging in this condition.

## 2. Observation

This was a 2-month-old male infant born to non-consanguineous parents, with no significant past medical history except for transient neonatal jaundice.

The clinical history dated back to birth, marked by the progressive onset of perioral cyanosis associated with feeding difficulties and fatigability during breastfeeding. These manifestations were initially subtle and neglected by the family, but gradually worsened, leading to medical consultation at the age of two months during a routine vaccination visit, where cyanosis was noted and prompted hospitalization for etiological assessment.

On admission, clinical examination revealed a conscious and reactive infant presenting generalized cyanosis predominantly involving the extremities. Hemodynamic assessment showed tachypnea at 60 breaths per minute, tachycardia at 170 beats per minute, and severe oxygen desaturation with peripheral oxygen saturation of 60% on room air. Body weight was estimated at 4.5 kg.

Cardiovascular examination revealed no audible cardiac murmur. Peripheral pulses were palpable and symmetrical, with no signs of right-sided heart failure, particularly no hepatomegaly, peripheral edema, or jugular venous distension. The remainder of the physical examination was unremarkable.

Chest radiography demonstrated moderate cardiomegaly with a cardiothoracic ratio of 0.56, associated with increased pulmonary vascular markings (**Figure 1**).

Transthoracic echocardiography revealed dilation of the right cardiac chambers associated with right ventricular hypertrophy, an atrial septal defect with a right-to-left shunt, and pulmonary regurgitation on continuous-wave Doppler, con-

sistent with severe pulmonary arterial hypertension. It also suggested an abnormal pulmonary venous return (Figure 2).

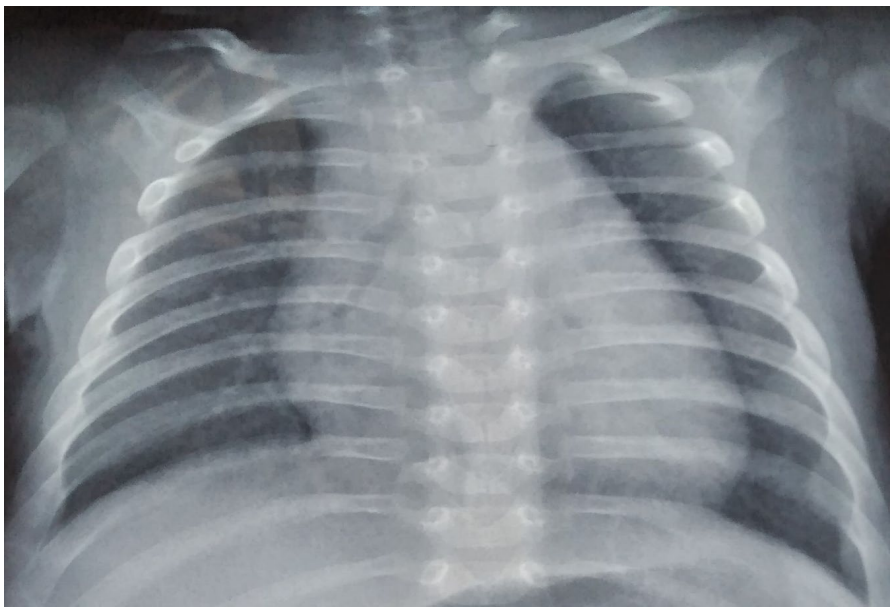

Figure 1. Chest radiograph showing moderate cardiomegaly associated with pulmonary overcirculation in an infant with supracardiac total anomalous pulmonary venous return.

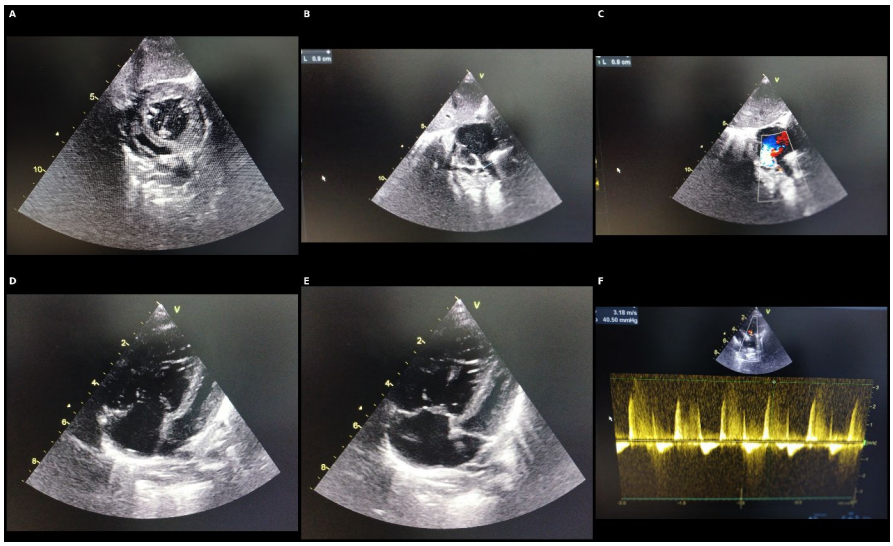


Figure 2. Transthoracic echocardiography findings in our patients with supracardiac total anomalous pulmonary venous return. (A): Parasternal short-axis view showing right ventricular hypertrophy. (B): Echocardiographic image showing a 9 mm atrial septal defect (ASD). (C): Color Doppler demonstrating a right-to-left shunt across the ASD. (D), (E): Apical four-chamber views showing dilation of the right cardiac chambers. (F): Continuous-wave Doppler demonstrating pulmonary regurgitation consistent with severe pulmonary arterial hypertension (PAH).

Thoracic CT angiography further characterized the vascular anatomy and confirmed supracardiac total anomalous pulmonary venous return, characterized by

the confluence of the pulmonary veins into a vertical vein draining into the left brachiocephalic vein (Figure 3).

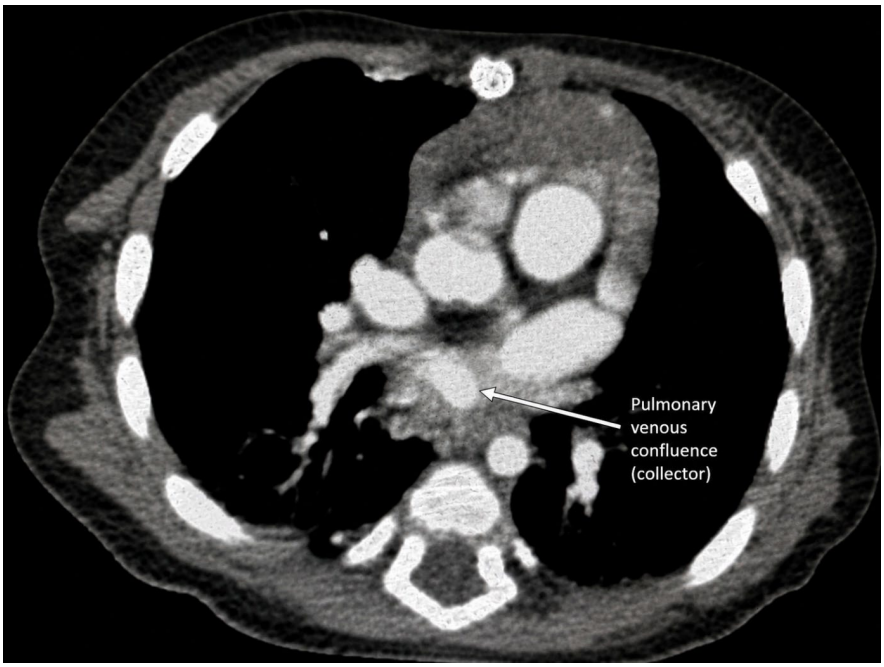


Figure 3. Thoracic CT angiography demonstrating the pulmonary venous confluence (collector).

The examination also demonstrated hypoplasia of the left cardiac chambers, cardiomegaly predominantly involving the right chambers, and ectasia of the inferior vena cava. Bilateral ground-glass pulmonary opacities were also noted, suggesting associated interstitial lung involvement.

Laboratory investigations revealed thrombocytosis at 516,000/mm$^3$. Coagulation studies showed a hemostatic disorder with decreased prothrombin time at 58% and prolonged activated partial thromboplastin time.

At the end of the diagnostic workup, a diagnosis of supracardiac total anomalous pulmonary venous return with significant hemodynamic impact was established.

The patient received symptomatic medical management including diuretic therapy with furosemide and vitamin K supplementation.

Initial evolution was marked by relative clinical stabilization despite persistent hypoxemia, leading to transfer to a specialized cardiac surgery center for definitive management. Unfortunately, the patient died during the postoperative period.

## 3. Discussion

TAPVR represents a diagnostic and therapeutic emergency, particularly in neonatal forms [3]. In our observation, early cyanosis associated with severe oxygen desaturation was consistent with the classical presentation of symptomatic forms. This clinical presentation is related to systemic and pulmonary blood mixing, re-

sulting in persistent hypoxemia [2].

From a pathophysiological perspective, the absence of a connection between the pulmonary veins and the left atrium results in drainage of oxygenated blood into the systemic venous circulation, leading to obligatory blood mixing within the right cardiac chambers. Survival therefore depends on the presence of an interatrial shunt allowing blood to reach the systemic circulation [2]. This mechanism also explains the right heart volume overload observed on echocardiography [2] [5].

In supracardiac forms, drainage through a vertical vein into the brachiocephalic vein is usually non-obstructive. However, obstruction may occur at different levels of the venous pathway, particularly at the level of the vertical vein or its connections, thereby worsening the prognosis by inducing pulmonary venous hypertension and impaired gas exchange [2] [4]. This distinction between obstructive and non-obstructive forms is essential, as it determines the clinical presentation and the urgency of therapeutic management [4].

Radiologically, chest radiography may demonstrate cardiomegaly associated with increased pulmonary vascular markings, as observed in our case [6]. In some non-obstructive supracardiac forms, a characteristic "snowman sign" may be observed, although this finding is rare in infants [7].

Echocardiography remains the first-line imaging modality, allowing rapid functional and morphological assessment. However, its limitations in the detailed evaluation of pulmonary venous connections are well documented, particularly in cases of complex anatomy or limited acoustic windows [8].

Thoracic CT angiography currently represents an essential diagnostic tool. It provides precise three-dimensional visualization of vascular structures, facilitating identification of the drainage site and detection of potential obstructive areas [4]. In our case, it confirmed a supracardiac form draining into the left brachiocephalic vein. Cardiac magnetic resonance imaging may also be used, particularly for hemodynamic assessment, although its use remains limited in neonatal practice [9] [10].

Our patient was diagnosed to have non-obstructive supracardiac TAPVR on thoracic CT angiography which demonstrated drainage through a vertical vein to the left brachiocephalic vein with no evidence of pulmonary venous narrowing. This distinction is clinically relevant since obstructive forms usually present with severe neonatal respiratory distress and pulmonary oedema, while non-obstructive forms may present later with progressive cyanosis and signs of pulmonary overcirculation as in our patient [3] [4]. The absence of radiological evidence of pulmonary venous obstruction did not prevent the delayed diagnosis of the severe pulmonary hypertension which, together with the delayed surgical management, probably contributed to the poor post-operative outcome [11].

The associated findings, including left heart hypoplasia, bilateral ground-glass pulmonary opacities, and coagulation abnormalities, reflect the hemodynamic consequences of TAPVR and the severity of the patient's clinical condition.

The differential diagnosis mainly includes other causes of neonatal cyanosis, particularly cyanotic congenital heart diseases such as transposition of the great arteries, pulmonary atresia, and truncus arteriosus. Differentiation relies on imaging findings and intracardiac flow analysis [12].

Recent literature data indicate that the prognosis of TAPVR depends on several determining factors, including the presence of pulmonary venous obstruction, patient weight at the time of surgery, severity of pulmonary hypertension, and delay in diagnostic and therapeutic management [11] [13]. Other prognostic factors have also been identified, such as prematurity, associated cardiac anomalies, and early postoperative complications [13] [14].

Furthermore, advances in pediatric cardiac surgery and intensive care management have significantly improved survival, with rates currently exceeding 85% - 90% in specialized centers [11] [15]. Nevertheless, morbidity remains mainly related to pulmonary vein stenosis, a serious complication that may occur after surgery [15] [16].

The treatment of TAPVR is exclusively surgical and consists of restoring a normal anatomical connection between the pulmonary veins and the left atrium while eliminating the abnormal venous drainage pathways [16]. The timing of surgery depends on the obstructive nature of the lesion and the patient's clinical condition, with obstructive forms requiring urgent intervention [13].

Recent technical advances, particularly the sutureless repair technique, have shown promising results by reducing the risk of postoperative pulmonary vein stenosis and improving long-term outcomes [16]. The unfavorable postoperative outcome observed in our patient illustrates the severity of TAPVR in infants and underlines the prognostic impact of delayed diagnosis, pulmonary hypertension, and hemodynamic impairment despite current surgical advances.

Our observation highlights the importance of early diagnosis in improving prognosis, while emphasizing the essential role of multimodal imaging in diagnostic confirmation and anatomical characterization of the malformation. It also illustrates the variability in the clinical presentation of supracardiac TAPVR, which may delay management and influence outcomes.

## 4. Conclusions

Supracardiac total anomalous pulmonary venous return remains a rare entity characterized by marked anatomical and clinical variability, which may make its identification challenging. Through this observation, we highlight the rich clinical spectrum of this malformation as well as the challenges it poses in clinical practice, particularly in cases of delayed diagnosis.

This case also emphasizes the value of detailed clinical descriptions in enriching the existing literature, especially for atypical or poorly documented forms. It therefore contributes to a better understanding of the clinical presentations and pathophysiological implications of this anomaly, indirectly helping to improve diagnostic and therapeutic strategies.

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## Ethics Statement

Informed consent for publication was obtained from the patient's parents. All patient data were anonymized, and no identifiable information is included in this report.

## Author Contributions

All authors contributed to the conception and design of the study, data acquisition, analysis and interpretation, manuscript drafting, and critical revision. All authors reviewed and approved the final manuscript and agreed to be accountable for all aspects of the work. Abdeladim Babakhouya, Maria Rkain, Aziza El Ouali, and Sara Anane supervised the work.

## Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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