Rare Case of Undiagnosed Supracardiac Total Anomalous Pulmonary Venous Return in an Adult

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A 50-year-old man presented to his cardiologist with a 1-year history of palpitations and exertional chest pressure, dyspnea, and dizziness. His surgical and medical history was unremarkable, including no history of tobacco use, hypertension, or hyperlipidemia. Coronary angiography, transthoracic and transesophageal echocardiography, and cardiac magnetic resonance imaging led to a diagnosis of an anomalous left upper pulmonary vein draining to the innominate vein and a secundum atrial septal defect (ASD). Palpitations correlated with isolated ventricular and supraventricular beats on Holter monitoring. He was referred to our institution for management.

Physical examination revealed a dark-complexioned man with a body mass index of 25.6 kg/m², a blood pressure of 117/77 mm Hg in the right arm, a heart rate of 86 bpm, and an oxygen saturation of 93% on room air. His jugular venous pulsations were normal. His lungs were clear to auscultation. Cardiac examination revealed a right ventricular heave, a normal S1, and a persistently split S2. P2 was not loud, and no murmurs were appreciated. There was no hepatomegaly, peripheral clubbing, or edema.

His ECG showed normal sinus rhythm at a rate of 89 bpm. His QRS axis was 120°, and there was a right bundle branch with a QRS duration of 120 milliseconds.

The outside echocardiograms showed an interatrial communication with bidirectional shunting, a severely dilated right ventricle with mildly decreased systolic function, mild right ventricular hypertension, and a markedly dilated innominate vein. Because transesophageal echocardiography was prematurely terminated as a result of patient intolerance, the pulmonary veins were not visualized.

The outside cardiac magnetic resonance imaging showed a large secundum ASD and severe right atrial and right ventricular dilation. However, on review of the study, imaging of the pulmonary veins was determined to be nondiagnostic but suspicious for supracardiac total anomalous pulmonary venous connection (TAPVC). Therefore, cardiac magnetic resonance imaging was repeated to better delineate the pulmonary venous anatomy before further invasive testing.

Repeat cardiac magnetic resonance imaging showed all 4 pulmonary veins joining a confluence behind the left atrium without evidence of pulmonary vein obstruction (Figure 1A). A large vertical vein arising from the confluence drained to the left innominate vein, resulting in a markedly dilated left innominate vein, superior vena cava, and right atrium (Figure 1B). The right ventricle was severely dilated (right ventricular end-diastolic volume, 216 mL/m² [normal, 86.2±14.1 mL/m²]) with normal systolic function (ejection fraction, 52% [normal, 55.1±3.7%]). Left ventricular size (left ventricular end-diastolic volume, 65 mL/m² [normal, 82.3±14.7 mL/m²]) was normal and function (ejection fraction, 53% [normal, 64.2±4.6%]) was mildly depressed. The estimated ratio of pulmonary blood flow (Qp) to systemic blood flow (Qs) was 3.9:1 by comparison of flows in the pulmonary artery and aorta. A large secundum ASD was present.

Chest radiography (Figure 2A) demonstrated cardiomegaly with widening of the superior mediastinum and a classic snowman appearance, consistent with supracardiac TAPVC. At repeat catheterization (Figure 2B), Qs was estimated to be 2.4 L/min/m² on the basis of a mixed venous saturation (MVO₂) of 67%, derived by averaging the right internal jugular (62%), left subclavian (72%), low inferior vena cava (69%), and distal innominate (63%) saturations, and an aortic saturation of 91%. With the use of a pulmonary vein confluence saturation of 95% and pulmonary artery saturation of 89%, the calculated Qp was 9.6 L/min/m², yielding an estimated Qp:Qs ratio of 4.0:1. The effective deoxygenated component of mixed venous blood going to the lungs (Qe) was 2.1 L/min/m², with a left-to-right shunt of 7.6 L/min/m² and a right-to-left shunt of 0.3 L/min/m². No gradient was detected on pullback from the pulmonary vein confluence. The mean right atrial pressure was 14 mm Hg, and the left ventricular end-diastolic pressure was 12 mm Hg. Mean pulmonary artery pressure was 32 mm Hg. Pulmonary vascular resistance was calculated to be 8.1 indexed Wood units. Coronary angiography revealed no significant epicardial coronary disease.

The patient was referred for surgical repair. At surgery, a transverse incision was made in the pulmonary vein confluence, another was made in the dome of the left atrium, and anastomosis was performed between them. Although the patient had no evidence of preoperative atrial fibrillation, on the basis of data showing that patients undergoing late repair for isolated ASD, another form of left-to-right shunt, remain at risk for late
development of atrial fibrillation, a concomitant low-risk procedure to target arrhythmia was felt to be prudent. Therefore, Cryo Maze lesions were created in the right atrium between the right atriotomy, inferior vena cava, and ASD; between the atriotomy and the coronary sinus; and from the right atriotomy to the tricuspid valve annulus. The ASD was then closed using autologous pericardium. Transesophageal echocardiography confirmed an unobstructed anastomosis of the pulmonary venous confluence to the left atrium and no residual ASD. Immediately after surgery, the patient reported improved energy and resolution of his dyspnea, dizziness, and palpitations.

TAPVC is a rare congenital heart defect, occurring in 0.05 to 0.09 per 1000 live births and accounting for roughly 1.5% of children with congenital heart disease. TAPVC results from failure of union between the common pulmonary vein and the developing left atrium with persisting communication between the pulmonary and systemic veins.

TAPVC is commonly categorized into 4 types based on the site of pulmonary venous drainage into the systemic circulation: supracardiac, cardiac, infracardiac, and mixed. Supracardiac TAPVC, as in the described case, is the most common, accounting for nearly half of all cases. In this type, the pulmonary veins coalesce into a common channel behind the left atrium from which the blood flows through a vertical vein into the left innominate vein and back to the right side of the heart. All blood entering the left atrium does so through an ASD. In cardiac TAPVC, the second most common type, the pulmonary veins drain into the right atrium directly or through the coronary sinus. In infracardiac TAPVC, after the pulmonary veins join behind the left atrium, the blood drains inferiorly into the portal venous system before entering the right atrium through the hepatic veins.

Clinical presentation depends largely on the degree of obstruction to pulmonary venous drainage and on the magnitude of the left-to-right shunt. Patients with obstructed TAPVC generally present very early in life with cyanosis, tachypnea, and respiratory distress and require urgent intervention. Patients with unobstructed TAPVC are usually less ill but present with a cardiac murmur, heart failure, or cyanosis. Reported cases of patients remaining undiagnosed into their sixth decade of life are rare. In most reported cases of surgical repair after 50 years of age, patients either refused surgery earlier in life or were not offered surgery. Our patient is particularly unusual in that he survived to 50 years of age before he was recognized to have significant heart disease.

The most common presenting symptoms in adulthood are cyanosis, dyspnea on exertion, and congestive heart failure. This likely reflects a gradual increase in the magnitude of right-to-left shunting across the ASD as right ventricular compliance decreases relative to that of the left ventricle. Important pulmonary vascular disease resulting from longstanding pulmonary overcirculation must be ruled out.

Hemodynamic assessment in patients with anomalous pulmonary veins is uniquely challenging and relies on multiple assumptions, leaving ample opportunity for error. In estimating \( MV_{O_2} \), we attempt to sample venous return sufficiently “upstream” from the shunt to ensure freedom from shunted blood. Averaging these values to determine \( MV_{O_2} \) ignores the relative contribution of each vessel to the true mix because this would be difficult, if not impossible, to measure; we assume, therefore, equal contribution from each source. Likewise, estimation of \( Qp \), \( Qs \), and pulmonary vascular resistance depends on accurate measure of \( MV_{O_2} \) adding further potential for error. In our case, all venous saturations fell within a range of ±5%, making the range of error relatively small.

In most cases, once the diagnosis of TAPVC is made, surgical repair is indicated. Postoperative outcomes have improved with time, with important predictors of poor outcomes, including younger age at repair, pulmonary venous obstruction before repair, individual pulmonary vein size, univentricular heart, and associated cardiac lesions. Although surgical repair of TAPVC in adults is rare, case reports suggest that a favorable outcome is possible if not likely. Surgery should be considered at a center with expertise in adult congenital heart disease after medical management of the patient’s symptoms and thorough assessment of their hemodynamics and comorbidities.

The most important complication after surgical repair of TAPVC is the development of pulmonary vein stenosis, which occurs in 5% to 19% of cases. Postrepair pulmonary vein stenosis carries a high mortality, with a 3-year survival of 58.7% after initial surgery; outcomes in patients undergoing surgical or catheter reintervention are equally poor.

In summary, when determining the cause of right heart dilation in an adult, all the pulmonary veins must be evaluated. Although TAPVC is very rarely discovered in adulthood, this case demonstrates that in situations with unobstructed pulmonary venous circulation and adequate right-to-left shunting to maintain a satisfactory cardiac output and to prevent pulmonary overcirculation, late diagnosis may occur and surgical repair may still be possible.

Disclosures

None.

References


Figure 1. A, Oblique view laying out the course from the pulmonary venous confluence (*) to the right superior vena cava (†). B, Three-dimensional volume-rendered magnetic resonance angiography showing a vertical vein (‡) draining into the right superior vena cava (†) via the innominate vein.

Figure 2. A, Chest radiograph demonstrating the classic snowman appearance of supracardiac total anomalous pulmonary venous connection. B, Still-frame cardiac angiogram with a pigtail catheter advanced through the inferior vena cava, right atrium, right superior vena cava, and innominate vein into a vertical vein that extends along the left mediastinum.
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In the article by Wu et al, “Rare Case of Undiagnosed Supracardiac Total Anomalous Pulmonary Venous Return in an Adult,” which was published in the September 30, 2014 issue of the journal (Circulation. 2014;130:1205–1207), a correction is needed.

On page 1207, the Figures 1A and 1B were incorrect. The correct version of Figure 1 appears below.

![Figure 1](image)

**Figure 1.** A, Oblique view laying out the course from the pulmonary venous confluence (*) to the right superior vena cava (†). B, Three-dimensional volume-rendered magnetic resonance angiography showing a vertical vein (‡) draining into the right superior vena cava (†) via the innominate vein.

This correction has been made to the current online version of the article.