

Case Report

Postcorrective surgery improvement of nocturnal hypoxemia in a case of partial anomalous pulmonary venous connection and aberrant hepatic vein drainage

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ABSTRACT

Partial anomalous pulmonary venous connection (PAPVC) is a rare congenital anomaly that leads to an anatomical left-to-right shunt. Termination of the intrahepatic inferior vena cava (IVC) with its azygos continuation associated with the hepatic venous connection to the left atrium (LA) is also a rare congenital anomaly that results in an anatomical right-to-left shunt. A 65-year-old male presented with severe dyspnea on exertion and pedal edema. He was further diagnosed at our clinic and was found to have both the aforementioned congenital abnormalities, creating a bidirectional shunt. On further investigation, he was found to have nocturnal hypoxemia on overnight oximetry. The patient was successfully treated via surgical corrections of the congenital anomalies leading to symptomatic improvement as well as the resolution of nocturnal hypoxemia.

KEY WORDS: Congenital heart disease, left-to-right shunt, nocturnal hypoxemia, partial anomalous pulmonary venous connection

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INTRODUCTION

Partial anomalous pulmonary venous connection (PAPVC) is a rare congenital anomaly that leads to an anatomical left-to-right shunt. Another rare congenital anomaly is the termination of the intrahepatic inferior vena cava (IVC) with its azygos continuation associated with the hepatic venous connection to the left atrium (LA). This results in an anatomical right-to-left shunt. Congenital heart disease has been known to be associated with hypoxemia. We present a fascinating case of a patient who had the cardiac anomalies mentioned above that caused a bidirectional shunt. The patient also had nocturnal hypoxemia that resolved after corrective surgery. We discuss the pathophysiologic mechanisms that may lead to nocturnal hypoxemia and postulate the rationale behind its resolution.

CASE REPORT

A 65-year-old man presented to our clinic with dyspnea and swelling of both the legs for 6 months. He had a history of atrial septal defect (ASD) that was repaired at the age of 6 years. Physical examination revealed a pansystolic murmur at the lower left sternal border and pitting edema over his shins. His oxygen saturation on room air was 95%. Complete blood count, cardiac enzymes, and results of the thyroid function tests were within normal limits. Transthoracic echocardiogram (TTE) and transesophageal echocardiogram (TEE) confirmed severe enlargement of the right atrium (RA) and right ventricle (RV) with mildly decreased RV systolic function. On TTE, echocontrast was noted in the left atrial appendage, along with

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prominent flow in the IVC. Estimated right ventricular systolic pressure (RVSP) was 45 mmHg. There was mild-to-moderate tricuspid regurgitation. Left ventricular ejection fraction (LVEF) by echocardiogram was 45%. The injection of agitated saline revealed bubbles on the LA after four-five cardiac cycles, indicating an extracardiac shunt at the intrapulmonary level.

The patient was symptom-limited to significant dyspnea on exertion. He was able to walk 150 m in the 6-min walk distance. The patient underwent an overnight pulse oximetry test that showed low oxygen saturation during the night; the time duration during which oxygen saturation was less than 90% (T 90) was 24.6 min [Figure 1].

A computed tomographic angiogram (CTA) of the chest revealed that the patient had intrahepatic interruption of the IVC with azygos continuation into the superior vena cava (SVC). One of the hepatic veins (HVs) drained into the LA, causing a right-to-left shunt [Figure 2a-d]. In addition, both the right pulmonary veins (PVs) drained into the RA leading to a left-to-right shunt and confirming PAPVC [Figures 3-5]. To better illustrate the concurrent congenital anomalies, an illustration has been provided [Figure 6].

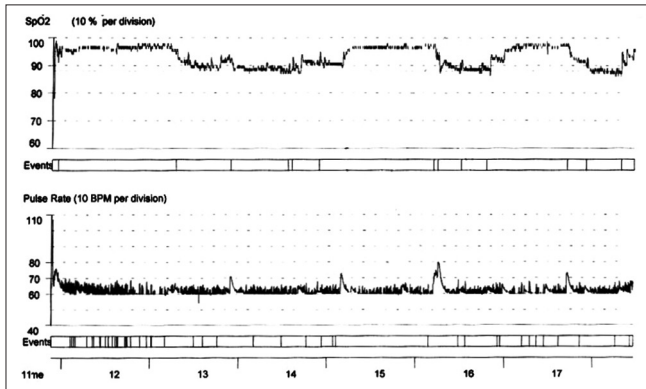


Figure 1: Overnight pulse oximetry prior to corrective surgery

The patient underwent right heart catheterization that was technically difficult due to the complex cardiovascular anatomy. The pulmonary artery systolic and diastolic pressures were 57 mmHg and 17 mmHg, respectively, while the mean pulmonary artery pressure was 33 mmHg. Left ventricular end diastolic pressure was 26 mmHg. The mean right atrial pressure was 20 mmHg. As a result of the long-standing bidirectional shunts, the patient had developed elevated pulmonary pressures and signs of biventricular failure. Subsequently, the patient underwent diversion of the common HV orifices to the RA and correction of the right-sided PV into the LA. This led to elimination of the bidirectional shunts.

Echocardiogram on follow-up at 3 months showed improvement of the LVEF to 60%. There was a marked reduction in the size of the enlarged RA and RV and an improvement in the tricuspid regurgitation. The echocontrast bubble study did not reveal any shunt. The 6-min walk test showed a remarkable improvement to 502 m. The overnight pulse oximetry test repeated on follow-up showed no nocturnal hypoxemia and T90 improved to 0.1 min [Figure 7]. The patient was

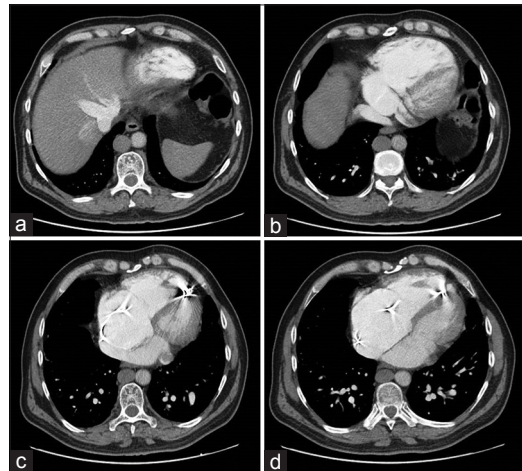


Figure 2: (a-d) CTA images of the HV

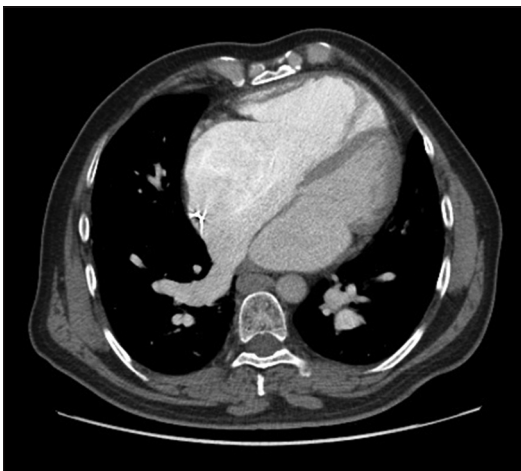


Figure 3: CTA image of the right inferior PV

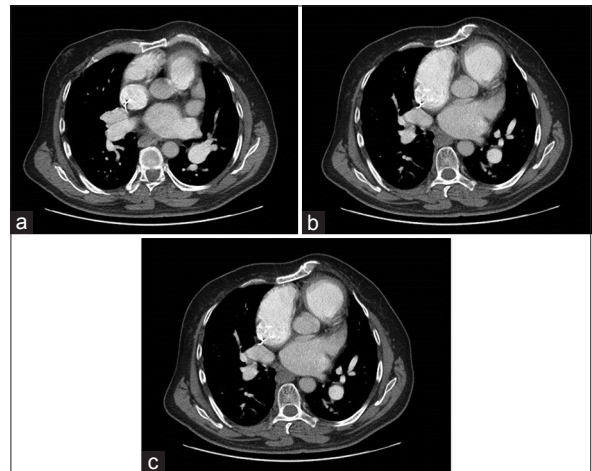


Figure 4: (a-c) CTA images of the right superior PV

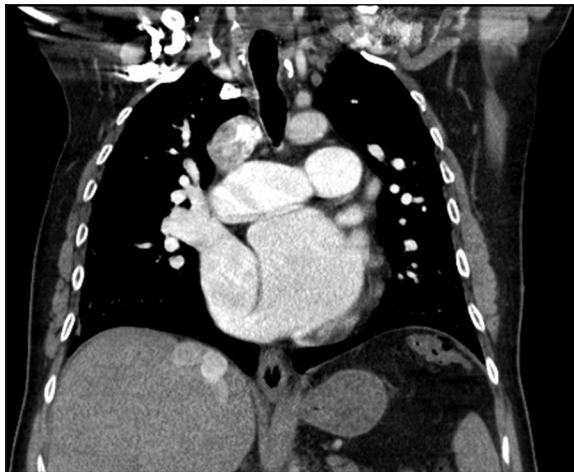


Figure 5: Coronal section - CTA image of the superior PVs

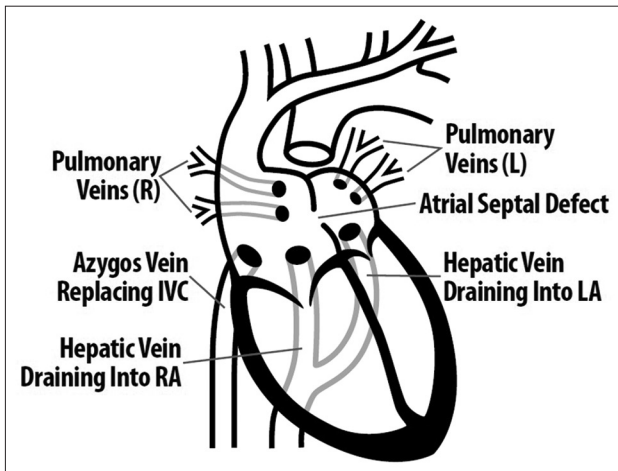


Figure 6: Illustration of concurrent congenital anomalies

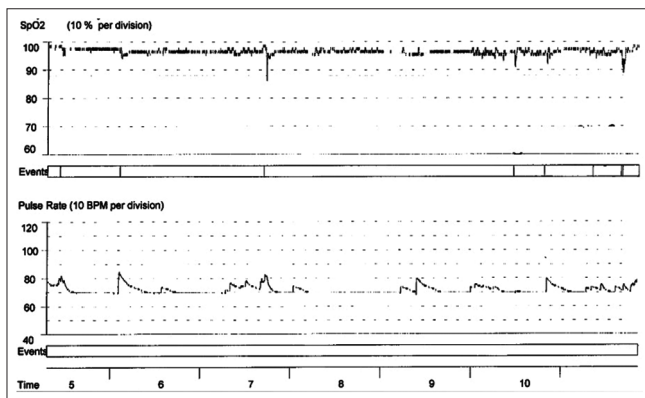


Figure 7: Overnight pulse oximetry after corrective surgery

discharged and continues to remain asymptomatic on follow-up.

DISCUSSION

PAPVC is a rare congenital condition with a reported incidence of 0.2% in a series based on computed tomographic (CT)

evaluation.^[1] In PAPVC, some of the PVs connect to the RA or to one of its tributaries such as the SVC rather than the LA.^[1] It is associated with ASD in up to 82% of the patients.^[2] Most commonly, the right PV drains in to the SVC or the RA.^[2] During early development, a primitive common PV arises from the dorsal wall of the LA. The pulmonary venous plexus (PVP) retains connections with cardinal veins that develop into the SVC and the portal system. PAPVC results when the PV fails to connect with the PVP, leading to a persistent communication between the PVP and the cardinal vein. This persistent systemic venous connections act like a left-to-right shunt. Patients are typically acyanotic and these anomalies can remain asymptomatic and undiagnosed for several years.^[3] However, untreated patients can develop a progressive rise in pulmonary vascular resistance due to an elevated pulmonary arterial blood flow and can develop signs of right heart failure (RHF). Pulmonary hypertension can develop when 50% or more of the pulmonary blood flow returns anomalously.^[4]

Congenital HV drainage to the LA is a rare abnormality with few reported cases in the literature.^[5] Clinically, it results in venous admixture with oxygenated blood that causes hypoxemia that is refractory to supplemental oxygen. To the best of our knowledge, anomalous hepatic venous return, along with azygos continuation of the IVC is extremely rare.^[6-8] The anomalous connection between the HVs directly to the LA and RA can be postulated due to persistent left vitelline veins that are normally absorbed into the liver. The absence of the hepatic portion of the IVC may be explained by the failure of formation of the right subcardinal-hepatic anastomosis that results in the atrophy of the right subcardinal vein, leading to azygos continuation of the IVC. This results in the development of a right-to-left shunt. The surgical correction involves excision of the HV from the LA and connection to the RA, thus ensuring normal venous return to the RA.^[8]

PAPVC may be found in adults incidentally on imaging or may present with symptoms of dyspnea. The workup includes TTE, TEE, and CTA. CTA and gadolinium-enhanced magnetic resonance (MR) angiography accurately identify PAPVC, along with the course, number of anomalous veins, and associated cardiovascular defects.^[9] Surgical repair of PAPVC separates the pulmonary and systemic venous returns and carries low mortality.^[10] Indications include the presence of RHF, significant left-to-right shunt, and pulmonary/systemic ratio exceeding 1.5 or recurrent infections.^[3] Therapeutic options for the repair of PAPVC vary with the type of anomaly^[10] and coexistence of other cardiac defects. Surgical repair of this anomaly includes diversion of the PV into the LA instead of the RA. Our patient underwent both procedures that resulted in the successful resolution of his right-to-left shunt as well as left-to-right shunt. His dyspnea and exercise tolerance improved that included an improvement in the 6-min walk distance.

An intriguing phenomenon noted in our patient was the correction of nocturnal hypoxemia after surgery. To the

best of our knowledge, improvement of the nocturnal hypoxemia after the correction of anatomical shunts has not been reported in the literature.

The mechanisms of nocturnal hypoxemia in such patients are unclear but we can certainly put forward a few explanations for this phenomenon observed in our patients. It is well-established that heart failure is associated with central sleep apnea.^[11] Thus, the nocturnal hypoxemia in our patient could be related to central sleep apnea. Our patient had significant improvement in LVEF after surgery. It is certainly plausible that with improvement in the left and right ventricular functions, his central sleep apnea resolved that in turn improved his nocturnal dyspnea.

Nocturnal hypoxemia has been previously noted in patients with congenital heart disease.^[12] The phenomenon of nocturnal desaturation could also be attributed to the increased right-to-left anatomical shunting due to increased venous return in the supine position and worsening ventilation/perfusion (V/Q) mismatch.^[12] It is possible that the correction of the hepatic venous connection can lead to the resolution of the anatomical right-to-left shunt and VQ mismatch, thereby possibly improving nocturnal hypoxemia.

Patients with PAPVC in the long term can develop pulmonary hypertension.^[9] Rafanan *et al.*^[13] have also reported nocturnal desaturation in patients with pulmonary arterial hypertension. In addition, Schulz *et al.*^[14] observed Cheyne-Stokes respiration associated with higher mean pulmonary arterial pressure (mPAP) and reduced right ventricular function. It would be certainly fair to propose a similar mechanism of resolution of nocturnal hypoxemia in our patient since he had significant improvement in biventricular function after corrective surgery. Another possible etiology of nocturnal hypoxemia is obstructive sleep apnea (OSA). Although OSA has been known to be associated with heart failure,^[15] resolution of the hypoxemia after surgery makes this diagnosis less likely.

We also believe that nocturnal hypoxemia improved in our patient as the pulmonary arteriovenous malformations (PAVMS) improved after corrective surgery. The echocontrast bubble study showed eradication of the extracardiac shunt detected on prior echocardiogram after surgery. The presence of PAVMS has been well-described in patients with right-to-left shunts such as anomalous HV and LA connection^[6] and are known to regress after corrective surgery.^[6,7] Srivastava^[16] suggested that the common denominator in the development of PAVM in a right-to-left shunt is the diversion of the normal hepatic venous blood flow away from the pulmonary circulation. Such PAVMS have also been reported in patients with the classic Glenn shunt.^[17] In our case, it is likely that PAVM occurred with interruption of the IVC and HV drainage into the LA. The PAVMS regressed after corrective diversion of the hepatic venous blood to the pulmonary venous circulation. We believe that further studies are necessary to fully evaluate nocturnal oxygen status in patients with acyanotic adult congenital heart disease.

CONCLUSION

Our case report contributes to the sparse literature on this unusual combination of congenital anomalies. It emphasizes the need to consider PAPVC and the HV anomaly as the causes of bidirectional shunt that may lead to biventricular failure. It demonstrates that noncyanotic adult congenital heart diseases can be associated with nocturnal hypoxemia. The reversal of the shunt and corrective surgery may lead to the resolution of the hypoxemia, improvement in hemodynamics and symptom relief and also nocturnal hypoxemia.

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Conflicts of interest

There are no conflicts of interest.

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