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LETTER TO THE EDITOR

WILEY

Persistent left superior vena cava draining into the left superior pulmonary vein in a scuba diver: A case report and literature study

1 | INTRODUCTION

A persistent left superior vena cava (PLSVC) is the most common congenital variation in the thoracic venous system, affecting around 0.5% in the general population and around 3.0% in patients with congenital heart disease.[1,2] The most common associated cardiac anomalies are atrio-ventricular septal defects, conotruncal malformation, bicuspid aortic valve, and cor triatriatum.[1,2] In the majority of cases, a PLSVC does not cause any hemodynamic disturbances or complaints and is an unexpected finding during pacemaker implantation, introducing central venous catheters or cardiovascular imaging for other reasons.[3,4] However, a PLSVC can have important consequences for scuba diving. We present a case of a scuba diver with a PLSVC draining in the left atrium via the left superior pulmonary vein, which was discovered after a diving injury.

2 | CASE REPORT

A 44-year-old man was referred to our outpatient clinic to examine whether there was any cardiac pathology present (such as a patent foramen ovale [PFO]) contributing to his diving injury. He had no relevant medical history, did not use medication, and was previously without complaints. He was an experienced diver (*Dive Master)* and had completed about 180 previous dives without any problems.

His last dive was the deepest of that day with a depth of 40 m. There were no abnormalities in the preparation. The descent took four minutes, and after a bottom time of nine minutes, a normal ascent with decompression stops was performed without problems. He finished the dive slightly tired, but without any other complaints. Ten minutes after leaving the water, he started to feel dizzy and nauseous. On the mainland, he needed support while walking due to heavy dizziness, nausea, and disorientation. He was transported by helicopter to a hyperbaric oxygen chamber. After several sessions of hyperbaric oxygen therapy, his complaints disappeared.

The physical examination at our outpatient clinic a few months later revealed no abnormalities besides obesity (body mass index 30.0 kg/m^2). The electrocardiogram was normal. A computed tomography (CT) scan of the thorax revealed an apical bulla in the lower lobe of the right lung (not shown here). A magnetic resonance imaging (MRI) of the cerebrum showed no abnormalities. Transthoracic echocardiogram (TTE) showed good left and right ventricular function with normal atrial and ventricular dimensions and minimal aortic valve regurgitation. No dilated coronary sinus was seen (Figure 1). Because a PFO could not be ruled out, a transesophageal echocardiogram (TEE) with saline contrast and Valsalva maneuver was performed, revealing a PFO with flow.

However, after injecting intravenous agitated saline contrast in the *left* arm, the contrast was first seen in the left atrium (Figure 2). After intravenous contrast injection in the *right* arm, the contrast was first seen in the right atrium (as expected) with flow to the left atrium via the PFO (Figure 3). The existence of a PLSVC was suspected, and a cardiac MRI was performed. This confirmed the presence of a PLSVC without dilatation of the coronary sinus. The PLSVC drained into the left atrium via the left superior pulmonary vein, which is a rare anatomical variation (Figure 4). Based on the stroke volume difference between the left and right ventricle, there was no evidence for significant shunting.

In conclusion, three abnormalities were found in this diver after a vestibular decompression illness: a PLSVC draining in the left atrium via the left superior pulmonary vein, a PFO, and a bulla in the right lung. The symptoms of this patient were probably caused by right-to-left shunting, either via the PLSVC or PFO. Since the presence of a lung bulla is incompatible with diving, the PLSVC and PFO were also treated conservatively.

3 | DISCUSSION

3.1 | Embryonic development

During early embryonic development, the thoracic venous system consists of two pairs of veins: the anterior cardinal veins for cranial venous drainage and the posterior cardinal veins for caudal venous drainage. These four veins drain

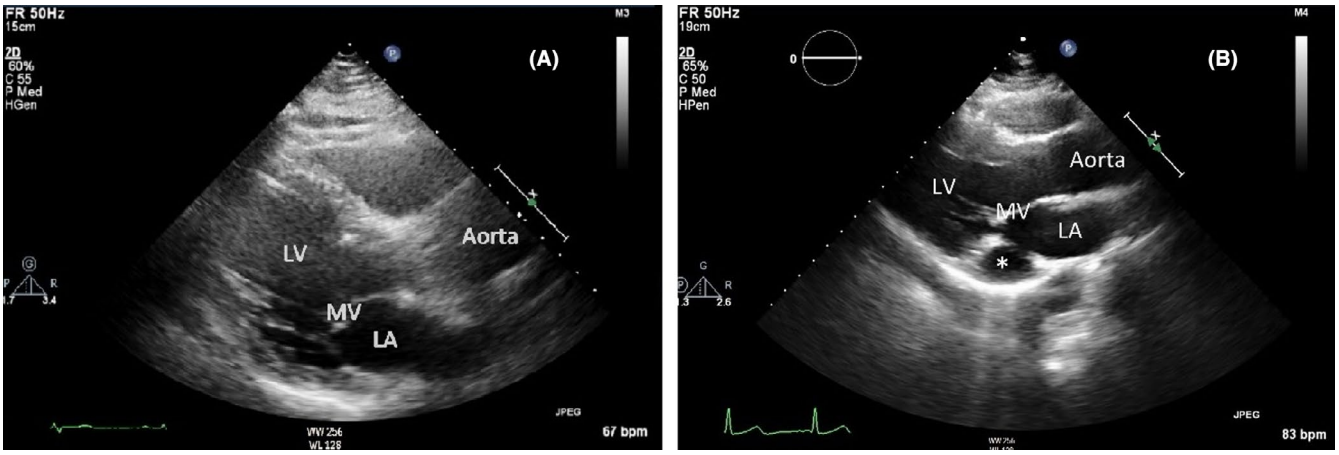

FIGURE 1 A, Transthoracic echocardiogram, parasternal long axis view. No dilated coronary sinus was seen in this patient. B, Dilated coronary sinus (asterisk) on the parasternal long axis view in a different patient. LA, left atrium; LV, left ventricle; MV, mitral valve

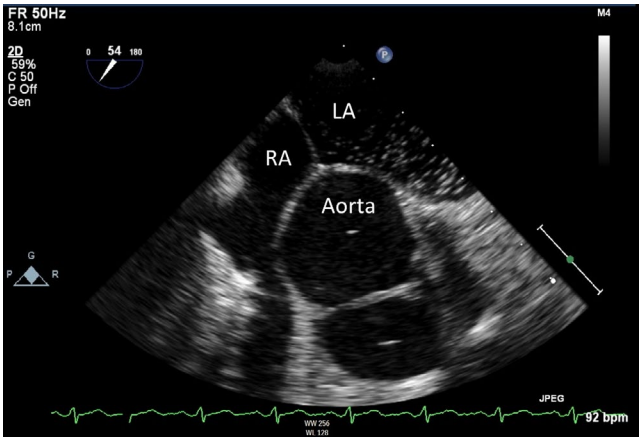

FIGURE 2 Transesophageal echocardiogram showing saline contrast bubbles arriving in the left atrium first after intravenous injection in the left arm. LA, left atrium; RA, right atrium

into the left and right common cardinal veins, returning the venous blood to the heart. During the 8th week of gestation, the left brachiocephalic (innominate) vein develops, connecting the two anterior cardinal veins. The right-sided vena cava superior will originate from the caudal part of the right anterior cardinal vein, while the caudal part of the left anterior cardinal vein normally obliterates and forms the ligament of Marshall. If this part remains open, a PLSVC is formed.[5-7]

In up to 90% of cases of PLSVC, the right anterior cardinal vein also remains patent, resulting in a bilateral superior vena cava with or without a communicating vein in between (Figure 5A,B).[5] Rarely, the right anterior cardinal vein degenerates, leaving only a PLSVC with an absent right superior vena cava (Figure 5C). In 80%-90%, the PLSVC drains into the right atrium through the coronary sinus that is often dilated. However, in about 10% the PLSVC drains into the left atrium via a direct opening, an unroofed coronary sinus, or the left superior pulmonary vein and causes a right-to-left shunt (Figure 5D,E).[5,6,8] Left atrial drainage via the left superior pulmonary vein is very rare and has, to our best knowledge, not been described in a scuba diver before.

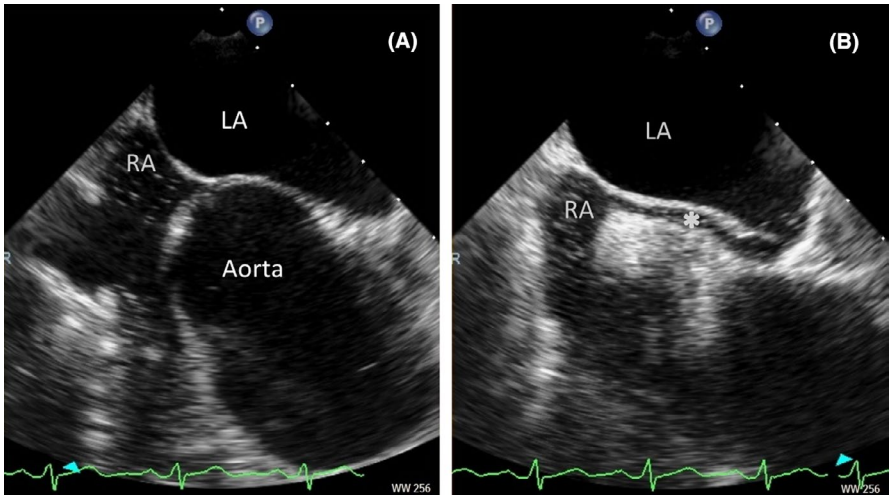

FIGURE 3 A, Transesophageal echocardiogram showing saline contrast bubbles arriving in the right atrium first after intravenous injection in the right arm. B, Saline contrast bubbles crossing over from right to left atrium via the patent foramen ovale (asterisk). LA, left atrium; RA, right atrium

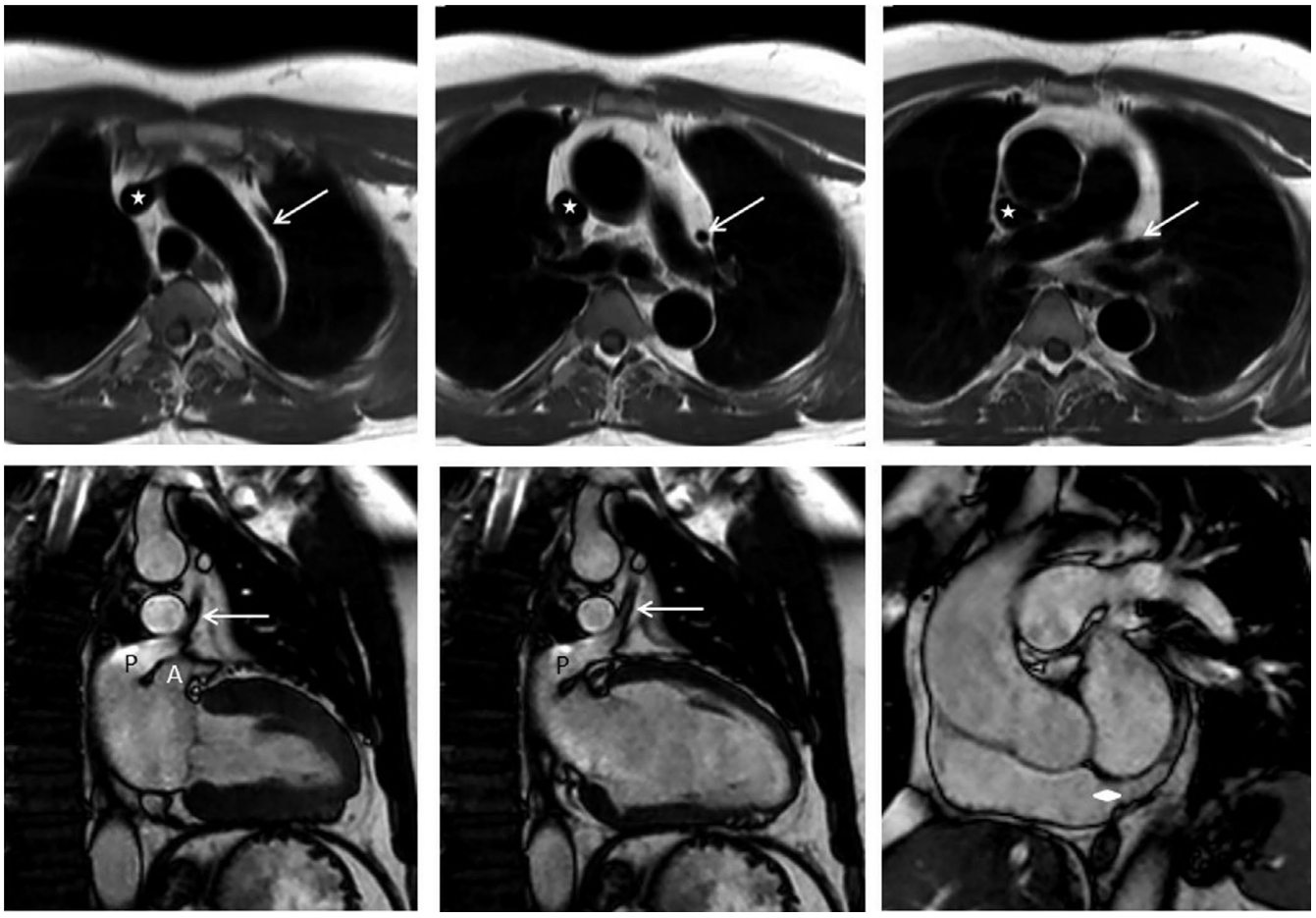

FIGURE 4 Cardiac MRI. Top row: axial T1 black blood images showing the PLSVC (arrow) and the right superior vena cava (star). Bottom row: two chamber cine images of the left ventricle in systolic (left) and diastolic (middle) ventricular phase showing the PLSVC (arrow) draining into the left superior pulmonary vein (P) behind the left atrial appendage (A). A short axis cine image at atrial level (right) showing the connection (diamond) between the non-dilated coronary sinus and right atrium

3.2 | Diagnosis

A PLSVC is often an incidental finding during cardiac imaging, electrophysiological study, or pacemaker or Swan-Ganz catheter placement. An unexpected route of the guidewire after entering via the left subclavian vein is frequently described (descending on the *left* side of the vertebral column before entering the coronary sinus).[1,4,9]

An ECG and plain chest x-ray are not specific, although a paramediastinal bulge or rounded shadow next to the upper left cardiac border may be seen on x-ray.[4,5]

Transthoracic echocardiogram can demonstrate a dilated coronary sinus without elevated right-sided pressures, and sometimes, the PLSVC itself can be seen. However, the absence of these findings on a TTE cannot exclude the diagnosis of a PLSVC, as shown in this case. TEE has an higher sensitivity than TTE for diagnosing PLSVC and associated cardiac abnormalities.[3]

A bilateral contrast TTE or TEE using agitated saline ("bubble study") injected from both left and right peripheral arm veins can confirm the diagnosis of PLSVC.[1,6] Normally, after injecting on either side, the bubbles are first seen in the right atrium, before passing through to the right ventricle. In most cases of PLSVC after injecting in the left arm, the bubbles are first seen in the coronary sinus before being visible in the right atrium. In our case, the bubbles were first seen in the *left* atrium, indicating a right-to-left shunt.

Multislice CT, MRI, and venography can be of additional value in diagnosing PLSVC and obtaining more detailed anatomical information.[1,5]

3.3 | Clinical relevance

In those patients with a significant right-to-left shunt due to the PLSVC, cyanosis can occur and the risk of paradoxical embolism is increased.[10] In scuba divers, this right-to-left shunt increases the risk of diving injuries such as decompression illness (DCI), analogous to a PFO.[11] In these cases, percutaneous closure of the left superior vena cava can be indicated.[12]

Moreover, PLSVC can cause technical difficulties during pacemaker, implantable cardioverter-defibrillator (ICD), or

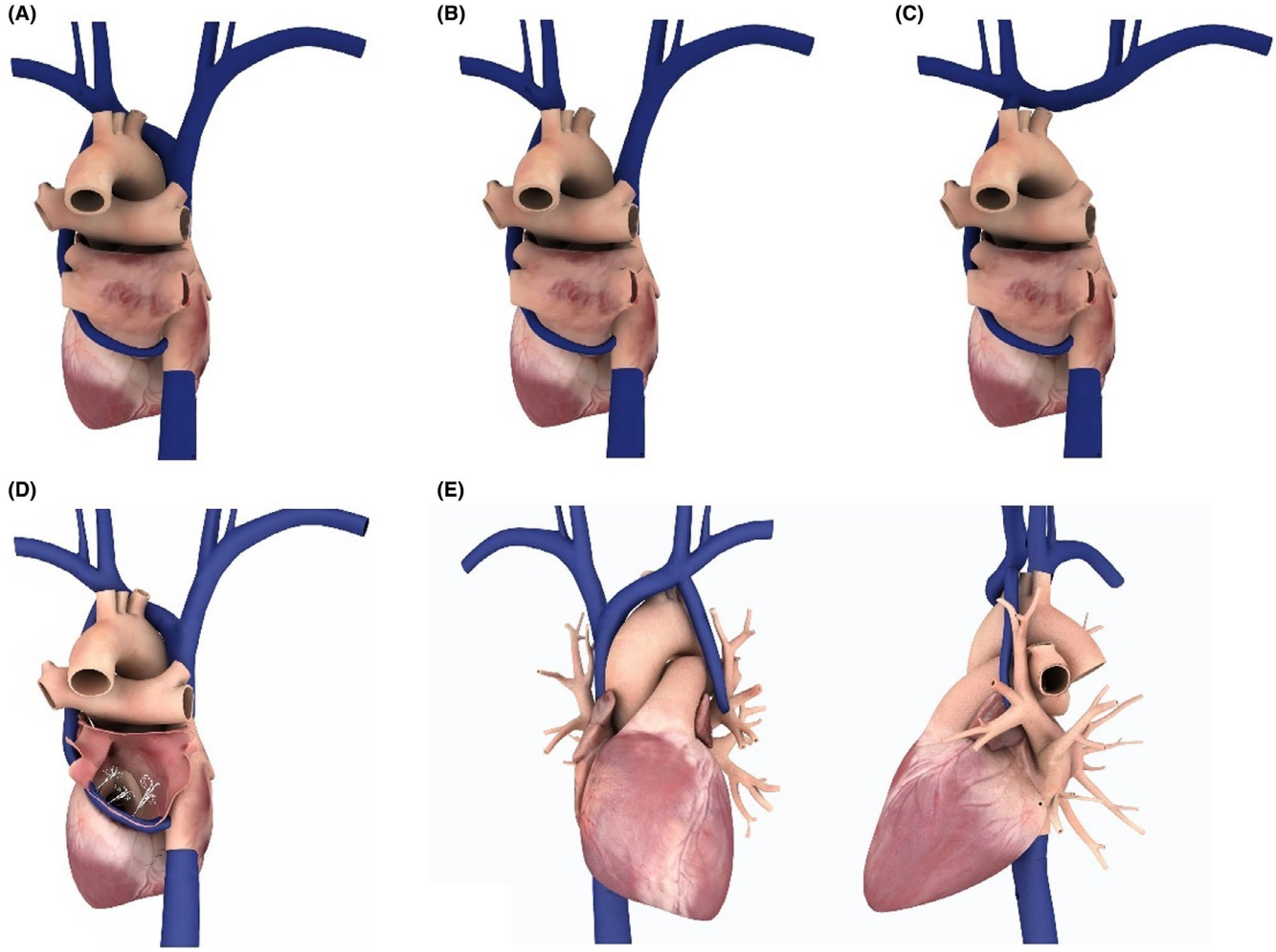

FIGURE 5 Illustration of different variations of a PLSVC viewed from posterior (except for E where anterior and lateral view are showed). R, right; L, left; A, anterior; and P, posterior. A, Bilateral superior vena cava with communicating vein. The PLSVC drains into the coronary sinus. B, Similar to A, but without communicating vein. C, Single PLSVC, draining into the coronary sinus. D, Bilateral superior vena cava with communicating vein. The PLSVC drains into the left atrium via an unroofed coronary sinus. E, Bilateral superior vena cava with communicating vein. The PLSVC drains into the left superior pulmonary vein, as seen in our patient

Swan-Ganz catheter placement, with serious complications such as angina, coronary sinus thrombosis, arrythmia, and cardiac arrest.[1,5] However, the incidence of these complications is low and successful implantations are previously described in patients with PLSVC.[4,5,9]

Because of the frequently associated congenital heart diseases in PLSVC, appropriate diagnostic evaluation should be undertaken. Finally, PLSVC is associated with various cardiac arrhythmias, including atrial fibrillation.[13]

4 | PERSPECTIVE AND RECOMMENDATIONS

A PLSVC can cause a right-to-left shunt, increasing the risk of DCI in scuba divers. Therefore, a PLSVC should be suspected in every diver with a DCI, especially in those without a PFO. A PLSVC can also cause difficulties during a wide range of cardiac interventions, and therefore, all physicians involved should be aware of the possibility of a PLSVC. A normal echocardiogram without a dilated coronary sinus cannot exclude an PLSVC, especially those draining in the left atrium. Moreover, we would recommend to perform contrast injection in both arms during every contrast echocardiogram as only right-sided venous contrast injection cannot exclude a PLSVC.

All patients with PLSVC should undergo evaluation for associated cardiac anomalies, such as atrial of ventricular septum defect, bicuspid aortic valve, or coarctation aortae.

A patient with an asymptomatic PLSVC without shunting, hemodynamic consequences, or associated congenital anomalies, does not have to be restricted in sports participation in general and scuba diving in particular. Echocardiographic follow-up is not necessary.

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