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Arteriovenous fistula in persistent left superior vena cava scenario

Fístula arteriovenosa en escenario de persistencia de vena cava superior izquierda



Dear Editor,

Persistent left superior vena cava (PLSVC), although rare, is the most common thoracic venous anomaly.¹ It results from persistence of the embryonic left anterior cardinal vein and is considered a normal variant.² The clinical significance depends mostly on the drainage site, and in 80%–90% of cases the drainage occurs to the coronary sinus (CS).³ We present a case of PLSVC incidentally detected after dysfunctional maturation of an arteriovenous fistula (AVF).

A 68-year-old man with stage 4 chronic kidney disease of undetermined etiology with a left radiocephalic AVF created 11 years back but never used for dialysis access. He also has atrial fibrillation (AF) that was diagnosed three years after AVF creation. Aneurysmal and serpiginous dilatation was found throughout the fistulous tract, with evidence of collateral circulation and edema of the left upper limb. Transthoracic echocardiography (TTE) showed normal left ventricular function, enlargement of left and right atrium, and a markedly dilated CS (33–35 mm of transverse diameter) (Fig. 1A). The presence of a PLSVC draining into the right atrium through a volume-overloaded CS was confirmed by use of saline contrast (“bubble study”) echocardiography and by magnetic resonance imaging (Fig. 1B). Since the CS dilatation caused by volume overload due to draining of the PLSVC was presumably aggravated by the left sided AVF and the unknown risk of rupture in these cases, the arteriovenous access was ligated.

PLSVC occurs in approximately 0.3%–0.5% of the population.² Left superior vena cava (SVC) commonly coexists with right SVC, and it drains into the right atrium through the CS with no major hemodynamic effect.^{1,3} The clinical significance of PLSVC also depends on the accompanying

anomalies. The most common associated congenital heart defects are single ventricle, atrioventricular septal defect, and tetralogy of Fallot.^{2,3} This anomaly should be suspected whenever a dilated CS is found on TTE and diagnosis can be confirmed by use of saline contrast echocardiography.¹

Although PLSVC is mostly asymptomatic, the creation of an AVF in the left upper limb not only increases the amount of blood drained into the CS – which under normal conditions corresponds to only 20% of the total venous drainage³ – but also increases the pressure at the level of this vascular bed. The enlargement of the CS, which can reach the aneurysmatic level, may cause compression of the sinus/atrioventricular node and His bundle, leading to cardiac arrhythmias such as atrial and ventricular fibrillation.^{2–6} In fact, PLSVC plays a considerable role in induction and maintenance of AF.⁷ This fact could explain the development of AF after AVF construction in our patient. An atrioventricular flow obstruction, with consequent decreased cardiac output, may also occur secondary to the compression of the left atrium by the dilated CS.⁶ In this scenario, the patient can develop symptoms of cardiac failure, particularly when there is a high-flow AVF draining into CS. Otherwise, hemodynamic alterations with development of more turbulent flow can cause intimal hyperplasia and lead AVF stenosis⁵ or compromise AVF maturation.

There were only found four case reports of a PLSVC coexisting with a left sided AVF^{5,6,8,9} (Table 1), in two main databases (PubMed and Google Scholar). All cases were accidental findings. AVF was being used for dialysis access in four patients – in two of those there was a high-flow AVF,^{6,9} and in the other two a stenosis in AVF.^{5,8} Only one patient was symptomatic, presents with dyspnea on exertion. In this case, there was a high-flow AVF (Qa 3000 mL/min) which, presumably, would be causing an overload of volume and pressure in the CS justifi-

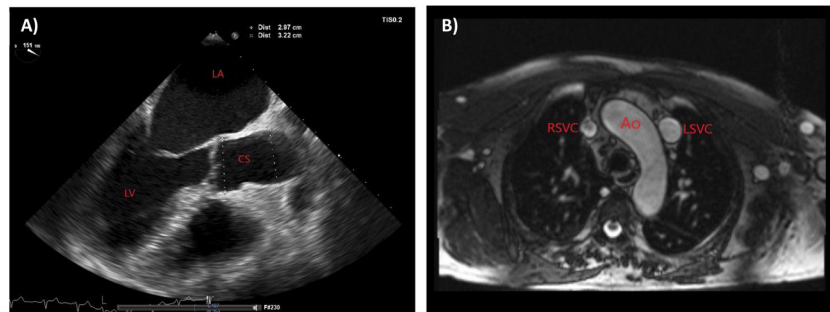


Fig. 1 – Imaging findings. (A) Transthoracic echocardiography showing a markedly dilated coronary sinus, and enlargement of left and right atrium. (B) Magnetic resonance imaging in transversal view showing a bilateral superior vena cava with a right superior vena cava and a persistent left superior vena cava. Ao, aorta; CS, coronary sinus; LA, left atrium; LSVC, left superior vena cava; RA, right atrium; RSVC, right superior vena cava.

Table 1 – Summary of previously reported cases of a PLSVC coexisting with a left sided AVF.

	Left AVF	Access complications	PLSVC drainage site	Congenital heart defects	Management
Orija A, Rajan J, and Degenhard A. ⁵	Brachio basilic	Stenosis	SC	None	No data
Wolf M, and Scott B. ⁶	Radiocephalic	High flow AVF	SC	None	Ligation of left AVF and construction of a right AVF
Hsu LF, Jais P, Keane D, et al. ⁸	Radiocephalic	Stenosis	SC	None	Maintenance of left AVF as HD access
Musci RL, De Michele L, D'Agostino C, and Colonna P. ⁹	No data	High flow AVF	SC	Atrial septal defect	ASD closure; no data about AVF

ying the patient's symptoms. This was the only case where the AVF was ligated and a new AVF was constructed.⁶

In summary, end-stage kidney disease patients are particularly vulnerable to complications from the PLSVC. Not only the coexistence of this anomaly with a left sided AVF may be a problem, but also central venous catheterization through the PLSVC into CS can cause potentially fatal complications (hypotension, perforation, cardiac tamponade, and cardiac arrest). For this reason, clinicians must be especially alert to this anomaly when planning a dialysis access. Besides the ultrasonographic vascular mapping, a TTE performed with attention to the CS might be useful.

Although this issue was addressed for the first time in the 2019 KDOQI guidelines for vascular access, there are no recommendations for how to act in a PLSVC coexisting with a left sided AVF. Guidelines only remember the possibility of this anomaly occurring when choosing the internal jugular veins for catheterization.

Considering the possible hemodynamic effects on CS dilatation and AVF maturation, it seems wise to avoid a left sided AVF in the presence of a PLSVC. However, further studies are warranted given the lack of sufficient data on this common and potentially complex finding.

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A rare case of PD-related cardiac tamponade after cardiac surgery

Un caso clínico raro de tamponamiento cardíaco tras de cirugía cardíaca asociada a la técnica de diálisis peritoneal



Dear Editor,

Regarding a rare complication secondary to the peritoneal dialysis (PD) technique, our team is sending you this letter to disclose an unusual clinical presentation among the clinicians.

This case report describes a 48-year-old man with end-stage-renal disease secondary to primary focal and segmental glomerulosclerosis on to PD since October 2022. The patient presented with a previous history of arterial hypertension, diabetes mellitus and hyperuricemia. One month after starting PD, he was admitted in the nephrology ward with malaise, fever and dry cough, which started four days before. Blood analysis showed elevated levels of inflammatory markers and blood cultures were positive for *Enterococcus faecalis*. Transthoracic echocardiography revealed multiple aortic vegetations in the right coronary cuspid with valvular perforation and severe aortic regurgitation which led to a diagnosis of infective bacterial endocarditis. After three weeks of intravenous ampicillin administration, the patient's condition deteriorated with acute pulmonary edema. He was transferred to the cardiac surgery department and underwent aortic valve replacement. Automated PD was re-started three days postoperatively with mild hematic effluent drainage. A computerized tomography (CT) excluded active intra-abdominal bleeding. Two weeks later, he presented with recrudescence of fever and worsening of inflammatory markers despite antibiotic therapy with Piperacillin/Tazobactam. At this time, blood cultures revealed *Klebsiella oxytoca* bacteriemia, a new contrast-enhanced CT scan showed mediastinal collections and a diagnosis of post-surgical mediastinitis was presumed.

Regardless improvement of infectious parameters, the patient evolved with hypotension and peritoneal drainage with daily variable ultrafiltration volumes was observed. Echocardiography was performed revealing *de novo* pericar-

dial effusion with signs of cardiac tamponade and, on day twenty-five after the surgery, the patient underwent evacuating pericardiocentesis. Evaluation of pericardial fluid for glucose revealed a concentration of 2000 mg/dL, superimposed with that of peritoneal fluid, which led to a diagnosis of an iatrogenic peritoneal–pericardial leakage after pericardiocentesis. After two days of PD suspension the patient has restarted the technique, but extremely variable peritoneal ultrafiltration volumes persisted. Reevaluating echocardiography revealed recrudescence of severe pericardial effusion. A CT peritoneography with iodinate contrast infusion through the peritoneum was performed and confirmed peritoneal–pericardial leakage (Fig. 1).

The pericardial effusion resolved with definitive PD suspension.

Peritoneal–pericardial leakage is a rare complication.¹ Only 9 cases have been described in the literature.² A history of cardiovascular interventions or multiple abdominal surgeries, which may be responsible for a breach; the presence of a congenital anomaly favoring tissue fragility with the occurrence of fistulas and hernias; history of malnutrition; long-term immunosuppression; previous peritonitis and excessive or too rapid increase in the volume of infusion represent the main risk factors for this condition.³ The clinical presentation varies from mild symptoms, such as cough and thoracalgia, to more severe clinical scenarios with dyspnea or hemodynamic instability in a cardiac tamponade setting.⁴

The presentation of our patient was exceptional, as he presented with variable ultra-filtration volumes with true ultrafiltration failure rarely observed. Biochemical analysis with glucose measurement of the pericardial fluid and radiologic investigations such as peritoneography with injection of iodinated contrast by CT or by magnetic resonance and peritoneal scintigraphy allow for a definitive diagnosis, as was the case of our patient.⁵