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CASE REPORT
Cardiac Imaging

An unusual case of dilated coronary sinus: case report and clinical implications

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Background The presence of a dilated coronary sinus (CS) assessed by transthoracic echocardiography (TTE) is highly suggestive of inferior or superior vena cava (SVC) anomalies, in the absence of a shunt. The most frequent finding is the persistence of a left superior vena cava (LSVC): well-known feature to electrophysiologists. Abnormal inferior vena cava (IVC) drainage is another cause of CS dilatation.

Case summary An 83-year-old woman presented with heart failure symptoms, atrial fibrillation with rapid ventricular rate, and a dilated CS assessed by TTE. Atrioventricular (AV) node ablation was considered given the poor efficacy of a rate control strategy. Cardiac computed tomography (CT) revealed a double SVC with an LSVC draining directly into the dilated CS. Single-lead pacemaker implantation was performed using a right-sided vascular access with no technical difficulties. An aborted AV node ablation procedure was due to the impossibility of getting to the right atrium. Fluoroscopy and CT imaging at second look analysis confirmed the diagnosis of an abnormal IVC with an agenesia of its supra-hepatic segment directly drained into the CS.

Discussion Our clinical case illustrates an unusual and rare double venous abnormality: both LSVC and IVC directly drained into the CS and were responsible for its massive dilatation.

Keywords Left superior vena cava • Inferior vena cava • Coronary sinus • Atrial fibrillation • Atrioventricular node ablation • Cardiac computed tomography • Case report

Learning points

- Dilated coronary sinus (CS) assessed by transthoracic echocardiography (TTE) is highly suggestive of inferior or superior vena cava (SVC) anomalies.
- Abnormalities of both lower and upper venous cava system can be found in a same patient.
- Left-sided SVC draining into the CS can be challenging in lead placement for cardiac devices.
- A dilated CS assessed by TTE should lead to a multimodal imaging approach aimed to provide a comprehensive evaluation of the venous system, more specifically prior to surgical and percutaneous cardiac procedures.

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Introduction

The presence of a left superior vena cava (LSVC) is a rare and generally asymptomatic congenital malformation that represents the most common variation in the anomalous venous return to the heart. It is usually discovered incidentally during central venous catheterization, interventional cardiovascular procedures like pacemaker (PM) or internal cardioverter defibrillator (ICD) lead implantation and non-invasive imaging techniques such as cardiac computed tomography (CT) prior to surgical and percutaneous cardiac procedures. It affects 3–10% of the population with congenital heart disease[1] and 0.3% of the general population.[2] In most cases, bilateral SVCs (with or without a bridging vein) are being observed. A single left-sided superior vena cava (SVC) is a rare and unusual variant with LSVC draining into a dilated coronary sinus (CS).[3] Congenital malformations of the inferior vena cava (IVC) are very sparse, with a prevalence <1%, and include interruption of the IVC, left IVC, and double IVC. Interruption of the IVC is a well-recognized, but uncommon anatomic malformation.[4,5] We hereby report an original case of multiple venous abnormalities with both LSVC and IVC directly draining into the CS.

Timeline

Time	Events
Day 0	Hospitalization for cardiac failure due to permanent atrial fibrillation with fast ventricular rate
Day 1	Echocardiography: major dilatation of the coronary sinus (CS)
Day 5	Failure of the rate control treatment, decision to implant a single-chamber pacemaker (PM) and to perform an atrioventricular (AV) node ablation to control heart rate (HR)
Day 8	Bilateral superior vena cava (SVC) were seen on cardiac computed tomography (CT) scan with the left SVC draining to CS
Day 10	Implantation of a single-chamber PM on the right side
Day 11	AV node ablation failure owing to the right atrium access inability. Cardiac CT scan confirmed an inferior vena cava abnormality. HR control reinforcement

Case presentation

An 83-year-old woman, with a history of arterial hypertension, diabetes mellitus, permanent atrial fibrillation (AF), and cognitive impairment, was referred to our institution for dyspnoea and palpitations. She showed clinical signs of heart failure (peripheral oedema with ankle swelling, pulmonary crepitations, and breathlessness).

Baseline electrocardiogram registered an AF with rapid ventricular rate (RVR) (150 b.p.m.). The plasma concentration of B-type natriuretic peptide was elevated to 500 ng/L ($N < 100$ ng/L).

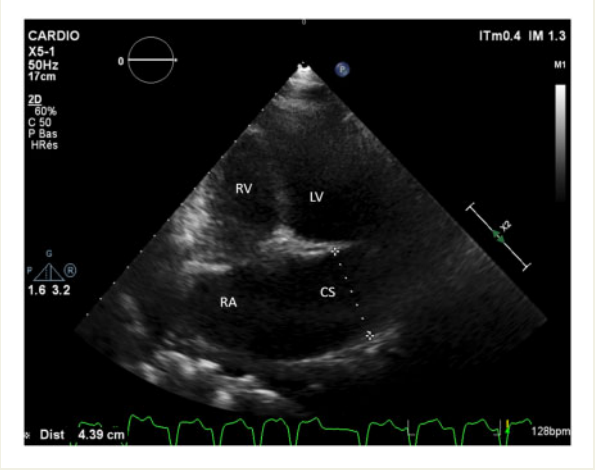


Figure 1 Transthoracic echocardiography, apical four-chamber view showing a dilated coronary sinus measured at 44 mm. CS, coronary sinus; LV, left ventricle; RA, right atrium; RV, right ventricle.

Transthoracic echocardiography showed a mild systolic dysfunction with a left ventricular ejection fraction of 45–50% and an extremely dilated CS (*Figure 1*).

Initial heart rate (HR) control in the management of this AF with RVR consisted of Atenolol doses up to 100 mg daily and Digoxin (0.125 mg per day) but our patient failed to respond to such pharmacological approach with an RVR at 140 b.p.m. The anticoagulant therapy was Apixaban (non-vitamin K antagonist oral anticoagulant) 5 mg twice daily.

After scanning the patient's characteristics and comorbidities, the benefits of either pharmacological and/or electrical cardioversion were sought inconsistent with the complexity of AF [permanent AF, dilated left atrium (63 mL/m^2), age, cognitive decline, agitation and/or intolerance with trans-oesophageal echocardiogram (TOE) guided cardioversion]. Atrioventricular (AV) node ablation was then considered the best strategy for our elderly patient in whom pharmacologic rate control had been unsuccessful.

Given the dilated CS, a cardiac CT was performed to better depict the vascular anatomy before implanting a single-chamber PM. It revealed a bilateral SVC system with an LSVC draining directly into the dilated CS (*Figure 2*). The volume of the CS was estimated on cardiac CT ~50 mL.

A right-sided implantation was therefore preferred with a PM implanted without difficulty from a right subclavian venous access and the lead further positioned on the interventricular septum. The AV node ablation procedure was attempted but was unsuccessful because of difficulty accessing the right atrium (RA). The ablation catheter could not get into the RA and actually entered into the CS at every attempt. Contrast injection confirmed that the IVC directly flowed into the CS (*Figure 3*).

Given the poor general condition of the patient in our case (reduced autonomy and cognitive decline), an upper extremity venous access was not attempted. Heart rate control was intensified using Nadolol (80 mg per day) instead of Atenolol and this translated into a suboptimal control at rest with HR ~90/min and during mild exercise (walking). Clinical improvement was rapidly observed with

decreasing signs of heart failure, and the patient was discharged home after 2 weeks.

At second look analysis, CT scan confirmed the diagnosis of an abnormal IVC with an agenesia of its supra-hepatic segment and that the IVC drained directly into the CS (*Figure 4 and Video 1*). There was no azygous continuation in this case.

To conclude, our patient presented an unusual and rare double venous abnormality: both LSVC and IVC directly drained into the CS and were responsible for its massive dilatation (Video 1).

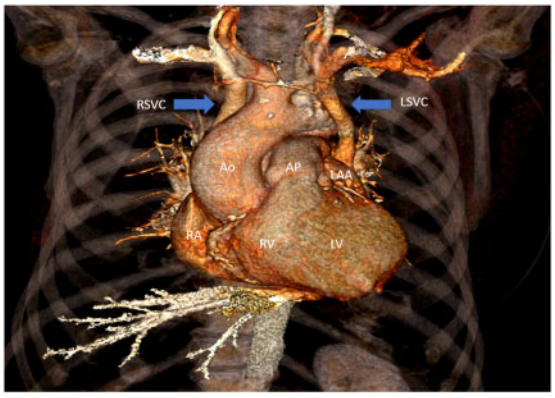


Figure 2 A three-dimensional reconstruction of a cardiac computed tomography scan clearly showing a double superior vena cava. Ao, aorta; AP, pulmonary artery; LAA, left atrial appendage; LSVC, left superior vena cava; LV, left ventricle; RA, right atrium; RSVC, right superior vena cava; RV, right ventricle.

Discussion

The presence of an LSVC draining into the CS is a well-recognized feature that can be challenging in lead placement for cardiac devices and therefore particularly acknowledged by electrophysiologists. LSVC persistence is thought to result from the failure of involution of the left anterior and left common cardinal veins.[6] It may be suspected in case of CS dilatation assessed by echocardiography[7] and multimodal cardiac imaging such as CT angiography or cardiovascular magnetic resonance imaging is recommended to further characterize the anatomy of the venous system. A right-sided vascular access for the device (PM or ICD) implantation is preferred in case of an associated right SVC.[8] Less common causes of dilated CS (without shunt) include:[9]

- an interrupted IVC with hemiazygous continuation to an LSVC. In this case, veins from the upper and lower parts of the body drain into the CS resulting into a major dilatation;
- hepatic veins connecting directly to the CS.[10]

The interruption of the IVC has a prevalence of 0.6–2.0% of all congenital heart diseases and 0.3% in the general population.[11] Inferior vena cava interruption occurs when the hepatic and prerenal segments of the developing IVC fail to fuse into a continuous channel. In such circumstances, the interrupted IVC continues as the azygous vein (right IVC) or in case of a left IVC as the hemiazygous vein.[12] These malformations have been reported in rare situations of visceral situs abnormalities, heterotaxy syndrome, typically the left isomerism type of heterotaxy or polysplenia.[13] Haswell and Berrigan[14] further described three routes for the interrupted left IVC and the existence of a hemiazygous vein in each case.

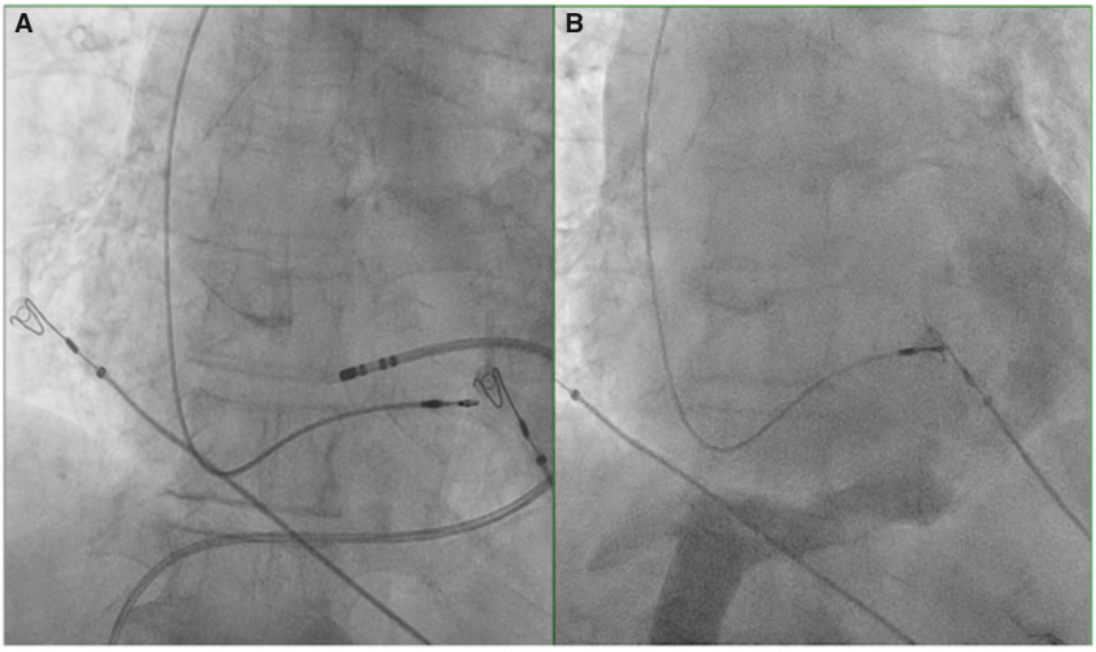


Figure 3 (*A*) Fluoroscopic view of the ablation catheter located in the coronary sinus. (*B*) Contrast injection confirming that the inferior vena cava flowed directly into the coronary sinus.

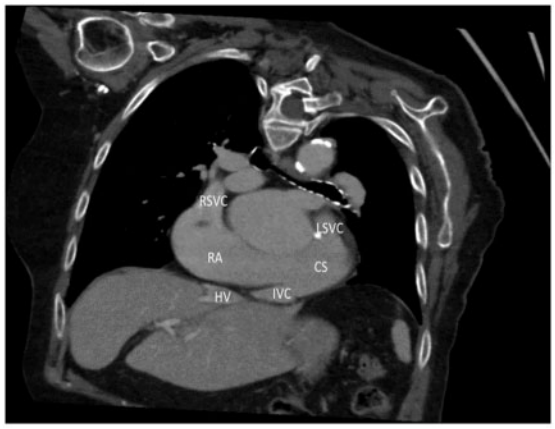
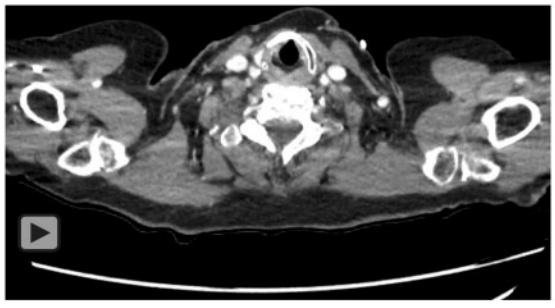


Figure 4 Cardiac computed tomography scan oblique coronal view showing the termination of the inferior vena cava and hepatic veins in the coronary sinus. CS, coronary sinus; HV, hepatic veins; IVC, inferior vena cava; LSVC, left superior vena cava; RA, right atrium; RSVC, right superior vena cava.



Video 1 Video of the cardiac CT scan axial images showing LSVC and IVC directly drained into the CS.

An abnormal hepatic veins connection to the CS has rarely been described in the literature. Indeed, only 14 cases have been reported up to 2015.[15] In patients with interruption of the IVC, the hepatic veins usually drain directly into the RA. Sometimes, the hepatic veins may connect with a residual IVC that eventually drains into the RA, pulmonary veins, or the left atrium.[10] Our case is the first to report these multiple systemic vein abnormalities without azygous continuation.

Conclusion

Our case illustrates the coexistence of two rare malformations of the venous system with on one hand an interruption of the hepatic segment of the IVC that drained directly into the CS without azygous continuation, and on the other hand the presence of an LSVC also draining into the CS. These two variants were together and synergistically responsible for the dilated phenotype of the CS. These malformations can impact both lead implantation and catheter ablation procedures.

A dilated CS assessed by echocardiography constitutes an important diagnostic clue for anomalous systemic venous return and should lead to a cross-sectional imaging approach aimed to provide a comprehensive evaluation of the venous system.

Lead author biography

Thomas Cardi is a third-year fellow in cardiology, specialized in electrophysiology and sports cardiology. He is working at the University Hospital in Strasbourg, France.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

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Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: None declared.

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