

An uncommon case of congenital thoracic venous anomaly and extracardiac sinus venosus defect in an asymptomatic adult first presenting with brain abscesses: a case report

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Background

Congenital thoracic venous anomalies (CTVAs) with right-to-left shunt constitute an uncommon source of paradoxical embolization in adults. We present a case of a healthy and physically fit individual with a rare asymptomatic anomaly first presenting with brain abscesses after a visit to the dental office; persistent left superior vena cava (PLSVC) without bridging vein, over-riding right-sided superior vena cava (RSVC) connected to the left atrium (LA), and an extracardiac sinus venosus defect.

Case summary

A 29-year-old male presented to the neurosurgical unit due to intracranial abscesses requiring intervention following a visit to his dentist. The abscess cultures isolated bacteria commonly found in the normal oral flora. Transthoracic echocardiography revealed an enlarged coronary sinus consistent with PLSVC. An agitated saline study was performed and raised suspicion of simultaneous extra- and intracardiac shunting. Magnetic resonance angiography confirmed the presence of a PLSVC and revealed an RSVC connected to the LA; however, no intracardiac shunt was evident. Electrocardiogram-gated computed tomography was therefore conducted and discovered the rudimentary remains of the physiologic RSVC forming a connection to the right atrium, explaining the bilateral contrast loading seen on the agitated saline study and diagnosing an extracardiac sinus venosus defect (SVD). The patient recovered and has been referred for surgery.

Discussion

This case illustrates a CTVA and a forme fruste type SVD resulting in a severe complication in a healthy adult. We highlight the diagnostic challenges posed, suggest early usage of agitated saline studies, and discuss the rationale for surgical correction of this patient.

Keywords

Case report • PLSVC • Persistent left superior vena cava • Right-sided superior vena cava • Bilateral superior vena cava • Double superior vena cava • Shunt • Congenital thoracic venous anomaly • Congenital thoracic venous anomalies • CTVA • Sinus venosus defect • SVD • ASD • Not-completed sinus venosus defect • Extracardiac sinus venosus defect • Without interatrial septal defect • Brain abscess • Forme fruste SVD

ESC Curriculum

9.7 Adult congenital heart disease • 2.1 Imaging modalities • 2.2 Echocardiography • 2.4 Cardiac computed tomography • 2.3 Cardiac magnetic resonance

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Learning points

- An embolic event may be the first presentation of a veno-arterial shunt and congenital thoracic venous anomalies should not be overlooked during work-up of these patients.
- Early usage of agitated saline studies from multiple injection sites is useful to guide the need for further imaging in patients presenting with PLSVC.
- Sinus venosus defects may be notoriously difficult to diagnose and may require multimodal imaging to visualize appropriately.

Introduction

Congenital thoracic venous anomalies (CTVAs) constitute quite rare encounters in the world of adult cardiology. The presence of two separate superior vena cavae (SVC) of which the left-sided SVC drains the left upper extremity and neck veins via the coronary sinus (CS) to the right atrium (RA) is termed persistent left superior vena cava (PLSVC). Persistent left superior vena cava is typically accompanied by a right-sided superior vena cava (RSVC) with physiological trajectory; known as a double SVC.^{1–3} Most frequently, adult patients are discovered incidentally during echocardiography or at autopsy when the enlarged CS is evident. The prevalence ranges from 0.3% to 0.5% in the general population,³ increasing to 11% in patients with concurrent congenital heart disease (CHD).¹

The CTVAs constitute a heterogeneous group with multiple possible variants, some of which permit shunting to occur. We describe a very rare case of bilateral SVC without bridging vein, extreme overriding of the RSVC connecting directly to the left atrium (LA), and an extracardiac sinus venosus defect (SVD). To our knowledge, this combination is exceptionally rare in asymptomatic adults without desaturation on room air.^{4–7}

Timeline

Case presentation

A 29-year-old man presented to his local emergency department due to an increasingly severe headache for the past 2 weeks, and new-onset neurological symptoms. A CT scan of the head revealed three right-sided supratentorial lesions (*Figure 1A*). He received betamethasone and was emergently transferred to the neurosurgical unit at Linköping University Hospital. The following morning a head magnetic resonance imaging (*Figure 1B*) was performed and confirmed intracerebral abscesses. He was started on metronidazole and cefotaxime for empiric antibiotic coverage, and the lesions were drained. Blood, urine, and abscess material were collected for culture. *Aggregatibacter Aphrophilus* and *Eikenella Corrodens*, two bacteria commonly found in the normal bacterial flora of the oral cavity were isolated from the abscess cultures.^{8–10}

The patient's past medical history was insignificant. He had not used recreational drugs and did not abuse alcohol. He was a non-smoker but used nicotine pouches. He was of normal weight (body mass index 23.1 kg/m²) and physically fit. He worked as a serviceman for the army and was in active service. On cardiac review, he had never experienced chest pain, palpitations, decreased effort tolerance, cyanosis, or syncope episodes. There was no recent travelling history. Family history revealed premature coronary heart disease

Date	Event
21 days before presentation	Dental scaling
15 days before presentation	Headache onset
1 day before presentation	Neurological symptoms
Day 0	Emergency department/head computed tomography (CT) Suspected intracranial tumours
Day 0	Transfer
Day 1	Head magnetic resonance imaging Confirmed intracerebral abscesses
Day 1	Abscess drainage Cultivated by bacteria commonly found in the oral flora
Day 2	Transthoracic echocardiography Findings of the dilated coronary sinus
Day 4	Transoesophageal echocardiography with agitated saline Confirmation of shunt, suspicion of simultaneous extra- and intracardiac shunting
Day 5	Cardiac magnetic resonance and magnetic resonance angiography Confirmation of persistent left superior vena cava and right-sided superior vena cava draining into the left atrium. However, intracardiac shunting is not evident
Day 15	Cardiac CT with angiography Extracardiac sinus venosus defect discovered

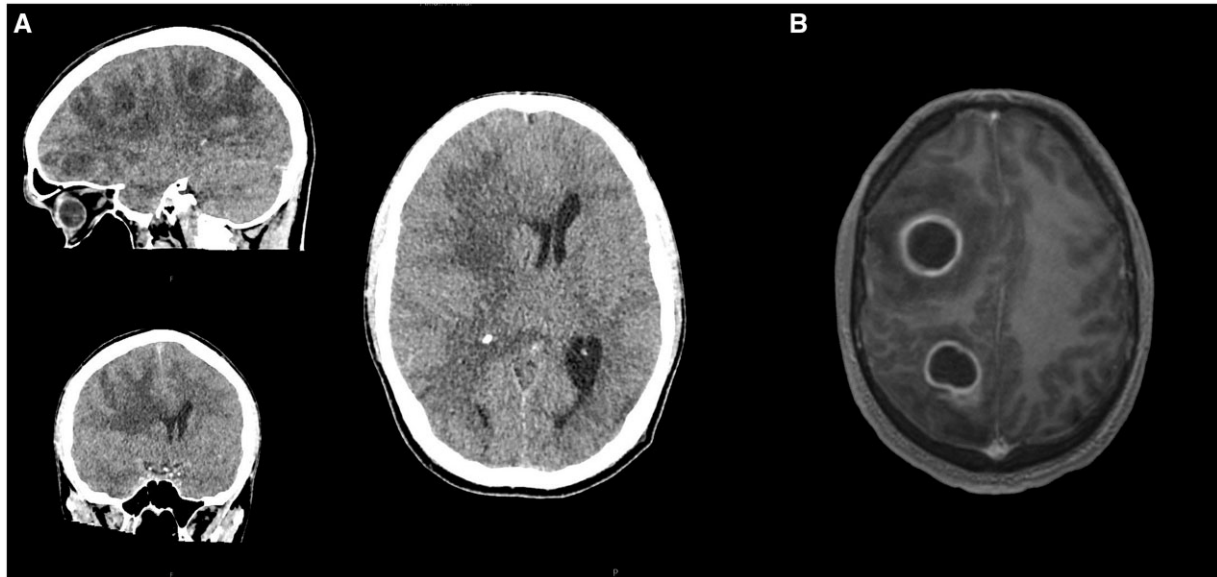


Figure 1 Native computed tomography head scan (A) displaying three supratentorial lesions displaying massive peritumoral oedema and midline shift. Magnetic resonance imaging (B) gadolinium-enhanced T1 sequence.

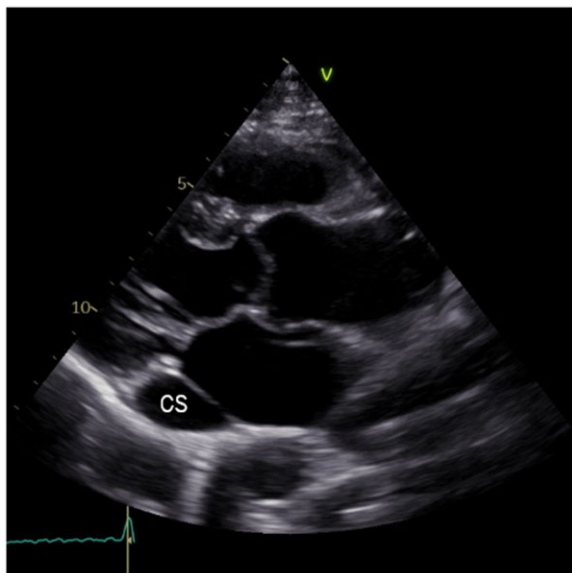
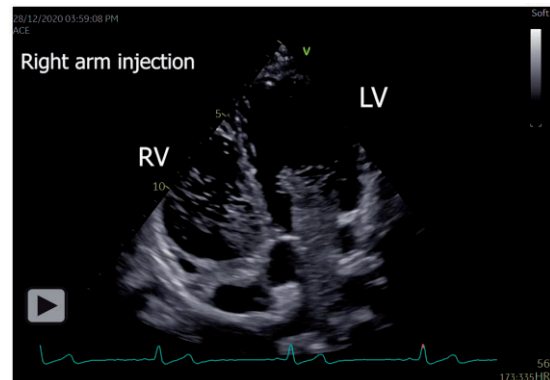


Figure 2 Transthoracic echocardiogram long-axis view. Dilated coronary sinus.



Video 1 Transthoracic echocardiography four-chamber view. Agitated saline is injected via the right cubital vein. Contrast first appears in the left atrium, and with delay passes into the right atrium and ventricle. LV, left ventricle; RV, right ventricle.

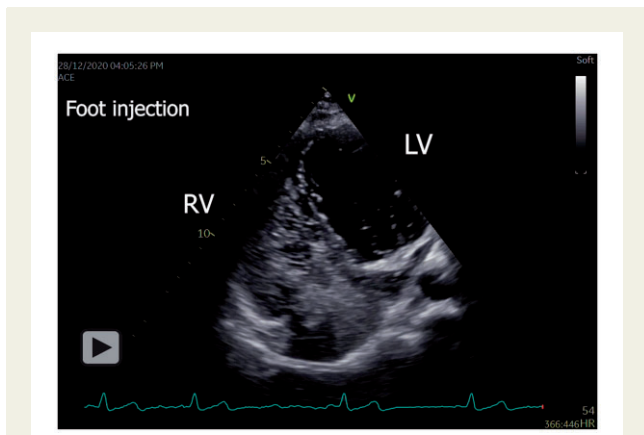
but no sudden death or CHD. His mother had not used medications during pregnancy.

The patient remained afebrile, with normal pulse variability, blood pressure, and oxygen saturation on room air during the entire hospital stay. He displayed no signs of skin, ENT, dental, lung, or GI infection. The patient confirmed a visit to his dentist for scaling 1 week before the onset of the headache. Cardiac auscultation revealed

regular rhythm without gallops or rubs. There was a Grade II/VI pansystolic murmur along the left sternal border increasing with inspiration. No significant jugular distension, hepatomegaly, or hepatojugular reflux was evident. Electrocardiogram (ECG) was normal ([Supplementary material online, Figure S4](#)).

As routine screening, transthoracic echocardiography (TTE) was conducted. The investigation revealed a dilated CS and atypical appearance of the tricuspid valve with moderate insufficiency. To rule out endocarditis or shunt, transoesophageal echocardiography (TOE) was scheduled along with an agitated saline study.

The TOE ruled out valvular vegetation, and no shunt was evident utilizing colour Doppler. Agitated saline was injected via the central venous catheter located in the right internal jugular vein; contrast appeared in LA first with a short delay before evident in the RA. To obtain a wider field of observation, the investigation was converted from TOE to TTE (Video 1). The finding raised suspicion of an extracardiac R–L shunt with a simultaneous intracardiac shunt. The same



Video 2 Transthoracic echocardiography four-chamber view. Agitated saline is injected via a venous line on the dorsum of the right foot. Contrast is only seen in the right atrium and ventricle. There is a minimal amount of residual contrast present in both ventricles from the previous injection. LV, left ventricle; RV, right ventricle.

result occurred when utilizing the right cubital vein for injection. However, when agitated saline was injected via a venous line on the dorsum of the right foot, the contrast was only observed in the RA (Video 2). To further evaluate the shunt, cardiac magnetic resonance (CMR) and magnetic resonance angiography were scheduled.

Cardiac magnetic resonance confirmed normal cardiac size and function. The tricuspid valve was found to have an atypical appearance presenting elongated leaflets; however, it did not display tethering of the septal leaflet or displacement associated with Ebstein's anomaly, or any other diagnostic pathology. Increased trabeculation of the right ventricle was observed but was non-diagnostic for non-compaction cardiomyopathy.

Magnetic resonance angiography confirmed the presence of a bilateral superior vena caval system without a bridging vein. The PLSVC (Figure 3A) connected to the CS which drained in the RA. The RSVC presented an anomalous trajectory directly to the LA (Figure 3B). An intracardiac connection between the RSVC and the RA could not be identified utilizing this modality and no CS defect was evident (Supplementary material online, Figure S1) An R–L shunt involving the venous drainage of the right upper extremity and right-sided neck veins was confirmed.

As the extracardiac shunt in isolation could not explain the findings of the agitated saline study, the patient was scheduled to return for an ECG-gated cardiac CT (CCT) with angiography after a prolonged course of I.V. antibiotic therapy at his local hospital.

Upon return, the CCT uncovered a 5-mm vessel branching off of the RSVC to form a connection with the RA revealing the extracardiac sinus venosus defect (Figure 4A–C; Video 3).

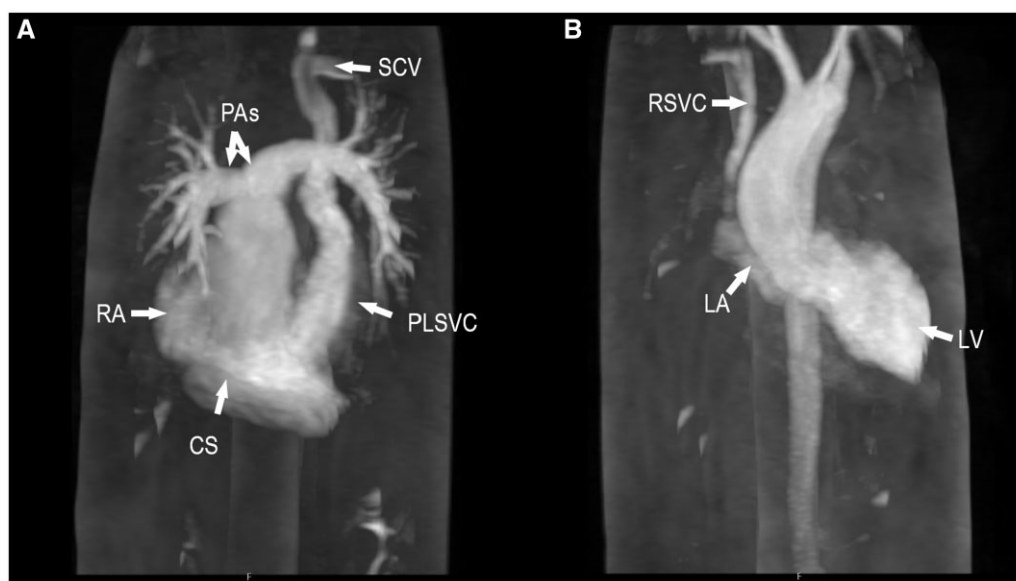


Figure 3 (A,B) Magnetic resonance angiography. (A) Contrast injected in the left cubital vein reveals contrast flow via the subclavian vein, persistent left superior vena cava, coronary sinus, right atrium, right ventricle into the pulmonary arteries. (B) Contrast injected in the right cubital vein reveals contrast flow via the right-sided superior vena cava into the left atrium and left ventricle. The right-sided superior vena cava connects to the left atrium at the location of the right superior pulmonary vein ostium. No bridging vein and no coronary sinus defect are visible.

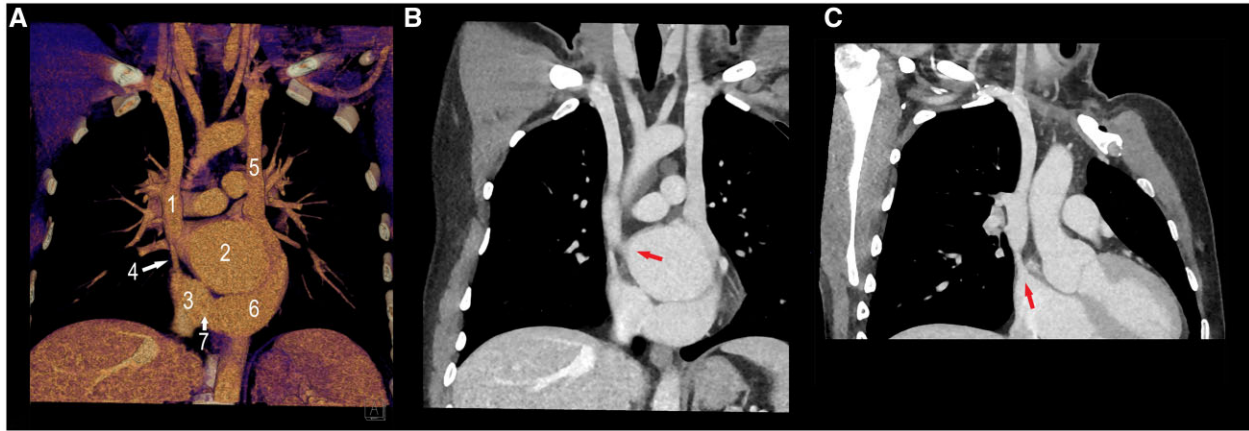
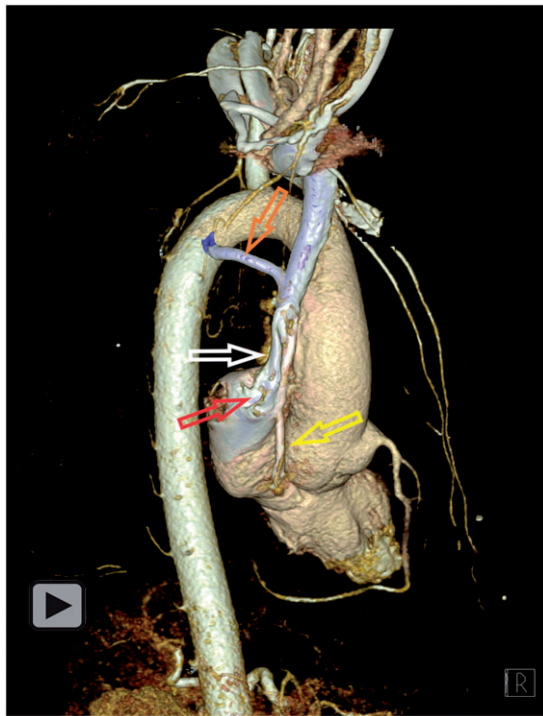


Figure 4 (A–C) Computed tomography angiography (A) 3D reconstruction. 1. Right-sided superior vena cava 2. Left atrium 3. Right atrium 4. Sinus venosus defect 5. Persistent left superior vena cava 6. Coronary sinus 7. Coronary sinus ostium. (B) Frontal view of the sinus venosus defect. The right-sided superior vena cava connects (arrow) to the left atrium at the level of the right upper pulmonary vein ostium. (C) Slightly oblique view of the sinus venosus defect following the trajectory of the remnant right-sided superior vena cava (arrow) into the right atrium.



Video 3 Animation of the computed tomography angiography 3D reconstruction. Still image: Orange arrow—Azygos vein. White arrow—Sinus venosus defect. Red arrow—right-sided superior vena cava connection with the left atrium. Yellow arrow—rudimentary right-sided superior vena cava connection with the right atrium.

Discussion/anatomy

This patient presents a remarkable combination of congenital venous anomalies; bilateral SVC without a bridging vein, over-riding RSVC, and SVD.

- PLSVC—left-sided subclavian and jugular veins connect to the PLSVC which drain into the RA via the CS.
- RSVC—right-sided subclavian and jugular veins connect to the RSVC which present extreme overriding to the LA at the location of the right upper pulmonary vein ostium.
- Collateral circulation of the caval veins occurs to a lesser extent via the azygos vein and small subcarinal veins but there is no bridging vein ([Supplementary material online, Figure S5](#)).
- Sinus venosus defect—A 5-mm-wide vessel branches off of the RSVC to form a communication with the RA at the physiological site of the SVC, this is proposed to be the remnant remains of the physiological RSVC constituting an extracardiac sinus venosus defect.
- Pulmonary circulation—Physiologic pulmonary venous drainage into the LA. The right upper pulmonary veins pass caudally to the pulmonary artery ([Supplementary material online, Figure S2](#)). The pulmonary veins drain at their physiologic location in the LA ([Supplementary material online, Figure S3](#)).

Conclusion

Persistent left superior vena cavae in isolation is often a benign and incidental finding that requires no further work-up. However, when a patient presents with signs or symptoms of a possible complication due to a shunt, further investigation is indicated. In our case, the patient presented with brain abscesses, a common complication of R–L

shunt which was why we opted for an early saline study.⁷ The flow pattern discovered later guided the continued need for specialized cardiac imaging to finally reveal the sinus venosus defect in addition to the veno-arterial shunt from the right upper body. Utilizing a saline study early was not only a low-cost initial screening measure but was the key to pursuing the correct diagnosis.

After diagnosing a shunt, the venous access should be limited to none-shunting locales if possible to minimize the risk of iatrogenic embolic events. We relocated all access points during the investigation and treatment to either the lower or left side of the body as well as utilized air filters for the IV lines.⁷

We find it remarkable that despite the shunting venous return from the right upper extremity and neck veins into the systemic circulation, the patient did not desaturate at rest and was able to maintain a physical job as an active serviceman. The CMR did not reveal haemodynamic decompensation due to overload and exercise stress testing later performed revealed normal oxygen saturation even while performing exercise. As the patient has been successfully treated for his abscesses and made a full recovery, he has been referred for corrective surgery after a heart team discussion. However, the lack of apparent haemodynamic decompensation somewhat questions the need for corrective surgery at this stage. The patient will undergo a complementary CMR with flow measurement before meeting with the surgeon. Prophylactic corrective surgery may be indicated as the embolic event likely occurred due to veno-arterial seeding during the dental scaling. However, the event was provoked and a conservative approach offering prophylactic antibiotics may be feasible initially. The patient's preference will greatly influence the decision for surgical correction.

The SVD described in our case is situated atypically high and is constituted of two separate well-formed vessels. Only a few such cases of forme fruste SVD have ever been previously described.¹¹ This finding strengthens the theory of the SVDs closer relationship to veno-venous malformations rather than a defect of the atrial septum.¹¹ Up to 85% of patients diagnosed with a superior SVD have concomitant partial anomalous pulmonary venous drainage (PAPVD) of the right upper pulmonary veins, which may prove a useful finding to pursue this diagnosis; however, this was not evident in our case.^{11,12}

Lead author biography



Staffan Pettersson is an Linköping University trained internal medicine specialist and is currently working clinically at Kungälv's sjukhus, Sweden. He has a special interest in teaching and is the current program director of the internal medicine program.

Supplementary material

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

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 the submission and publication of this case, including images, has been obtained from the patient in line with COPE guidance.

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Conflict of interest: None declared.

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