Persistent left superior vena cava connected through the left upper pulmonary vein to the left atrium: an unusual pathway for paradoxical embolization and a rare cause of recurrent transient ischaemic attack

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Ischaemic stroke, especially in the younger population, is an important cause of morbidity and mortality. When compared with the older population, the underlying aetiology of stroke in the young includes higher rates of cardioembolic disease and congenital heart anomalies. Paradoxical embolism may be an important cause of ischaemic cerebral events, which has to be ruled out in patients with no other evident stroke aetiology. A persistent left superior vena cava (PLSVC) that drains into the left atrium is a very rare congenital anomaly occurring in postnatal life and may be the cause of embolic events such as ischaemic stroke with potentially devastating consequences. In this article, we report the diagnosis and successful endovascular repair of this anomaly. This case report also aims to highlight the importance of close collaboration between neurologists, cardiologists and radiologists needed for accurate identification of stroke aetiology in young patients.

Keywords
- Ischemic transient ischaemic attack
- Paradoxical embolization
- Persistent left superior vena cava
- Coil embolization

Introduction

Ischaemic stroke, especially in the younger population, is an important cause of morbidity and mortality. When compared with the older population, the underlying aetiology of stroke in the young includes higher rates of cardioembolic disease and congenital heart anomalies. A persistent left superior vena cava (PLSVC) that drains into the left atrium is a very rare congenital anomaly occurring in postnatal life and may be the cause of embolic events such as ischaemic stroke with potentially devastating consequences. In this article, we report the diagnosis and successful endovascular repair of this anomaly.

Case report

A 25-year-old woman with a history of transient ischaemic attack presented to an emergency department with acute right hemiparesis and mild tactile hypoesthesia. On admission, her National
Institutes of Health Stroke Scale score was 3. Magnetic resonance imaging (MRI) of the brain and cervical spine showed no visible pathology including diffusion-weighted images and MRI angiography. All symptoms resolved completely within 3 h from the onset and thrombolysis was not performed. A follow-up ultrasound examination of the carotid and vertebral arteries as well as transcranial colour-coded ultrasound examination found no pathology. Ten years earlier, the patient had had one short episode of transient speech problems mimicking expressive aphasia. The use of oral contraceptives, smoking, heterozygous methylenetetrahydrofolate reductase mutation (AV 223) with mild hyperhomocysteinemia and mild superficial thrombophlebitis of the left forearm following a minor skin injury were identified as possible risk factors for venous thromboembolism. The other primary and secondary laboratory and clinical thrombophilic risk factors were excluded.

Transthoracic (TTE) and transoesophageal (TEE) echocardiography was used to exclude a possible cardiac origin of cerebral embolism. Intracardiac thrombus and tumour were excluded. A patent foramen ovale was detected but with only a minimal left-to-right shunt without flow inversion after provocative manoeuvres and a bicuspid aortic valve was documented. At rest and during the Valsalva manoeuvre, no right-to-left shunt and no microbubble contrast agent (agitated saline) in the left atrium were detectable. The TEE finding suggested that this was most probably a case of paradoxical embolism in cerebral circulation due to an evident right-to-left shunt with intermittent left upper extremity position-dependent flow via the PLSVC. Its whole course was not visualized but we found its suspected inflow to the left atrium through the left upper pulmonary vein (Figure 1).

Computed tomography of the chest revealed a PLSVC originating from the left brachiocephalic vein (white arrow) and draining (yellow arrow) into the left upper pulmonary vein, which leads directly to the left atrium (Figure 2).

This finding was confirmed by venography (Figure 3). The patient underwent successful percutaneous endovascular occlusion of the proximal part of the PLSVC using coil embolization. The procedure was indicated for demonstrated superficial thrombophlebitis of the left forearm (even without definite ultrasound demonstration of thrombotic masses in the upper extremity venous system) and a known chain of events of a recent neurological deficiency onset. The patient was discharged from the hospital with no neurological deficit. Another venogram showed no visible flow of a contrast agent through the PLSVC. Repeated TTE using a contrast agent

![Figure 1](https://example.com/figure1.png)

**Figure 1** Contrast echocardiography (Supplementary data movie clips). (A, movie clip S1a) After injecting agitated saline into the right antecubital vein, there was a normal sequence of opacification of the right heart chambers with no penetration of microbubbles into the left heart chambers. (B, C, movie clips S1b,c) In the apical four-chamber and midesophageal bicalv transoesophageal views of the patient lying on the left side, synchronous saturation of the left and right heart chambers was noticeable after applying a contrast agent into the left antecubital vein. There was no evidence of echocontrast agent flow from the region of the non-dilated coronary sinus into the left or right atrium. (D, movie clip S1d) Transoesophageal echocardiography examination focused on the pulmonary vein revealed a clear inflow of the contrast agent into the left atrium through the left upper pulmonary vein (white arrow). LA, left atrium; RA, right atrium; IAS, interatrial septum; SVC, superior vena cava; AoV, aortic valve; LUPV, left upper pulmonary vein.
injected into the left antecubital vein when lying on the left side with the elevated left upper extremity showed no apparent penetration of microbubbles into the left heart chambers. During a follow-up of more than 18 months, no recurrent transient ischaemic attack was observed in the patient. She reported no problems and gave birth to her first child without complications.

**Discussion**

A PLSVC is the most common anomaly involving central venous return in the thorax with an estimated prevalence of $\sim 0.3\%$ in the general population and 4–10\% in patients with congenital heart disease.$^{1,2}$ Most commonly (90\%), the PLSVC drains through the coronary sinus into the right atrium and this form is not associated with increased risks inherent with a right-to-left shunt such as embolic cerebrovascular events.$^2$ Rarely ($< 10\%$), the PLSVC drains into the left atrium, typically through an unroofed coronary sinus and very seldom directly or through the pulmonary vein.$^3$ This form is characterized by the presence of a right-to-left shunt associated with a relatively high risk of air or septic embolism$^{4,5}$ or thromboembolism in the systemic circulation.$^6$ The suggested pathogenetic mechanism for brain

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**Figure 2** Computed tomography of the chest. Reconstructed images using volume rendering (A) and a two-dimensional image (B) demonstrates that the persistent left superior vena cava originates from the left brachiocephalic vein (white arrow) and drains (yellow arrow) into the left upper pulmonary vein, which leads directly into the left atrium. Ao, aorta; PA, pulmonary artery.

**Figure 3** Venography (A) and chest X-ray after catheter closure of the persistent left superior vena cava (B). (A) After application of a contrast agent, the persistent left superior vena cava can be seen, originating from the left brachiocephalic vein (white arrow) draining into the left upper pulmonary vein at the confluence with the pulmonary vein (yellow arrow), from where the flow is seen as a negative contrast (darker endovascular content) is apparent. Furthermore, there is an evident of normal venous inflow into the right atrium through the superior vena cava. (B) The position of the coil under the left clavicle is visible after the catheter closure procedure (red arrow).
ischaemia and stroke is paradoxical embolism through a right-to-left shunt, with the possible embolic source in the left upper extremity veins.

As to the diagnosis, a simple diagnostic procedure such as contrast TTE with a contrast agent injected into the left upper extremity venous system should be used. It may reveal simultaneous left and right atrium opacification consistent with an intracardiac right-to-left shunt. Contrast TEE (with obligatory left antecubital vein contrast agent application) is the favoured screening modality for cardiac sources of systemic embolism and should be performed routinely in all young patients without an evident cause of ischaemic stroke. Right upper extremity intravenous contrast agent application should not be used to detect the PLSVC. Moreover, the contrast agent should be injected into the left upper extremity venous system with various positions of the extremity (elevated/along the body) as well as the body (lateral/supine). This is because the flow through the PLSVC may be position dependent. With the exception of cardiac thrombus, spontaneous echo contrast, vegetations and tumours, other pathological findings such as a right-to-left shunt through an atrial septal defect, patent foramen ovale, pulmonary arteriovenous malformations and congenital anomalies of thoracic venous circulation such as a PLSVC connected through the left upper pulmonary vein to the left atrium may be identified as a pathway for paradoxical embolization.

Endovascular approaches to the treatment of patients with a symptomatic PLSVC and right-to-left shunt using the Amplatzer occluder4–6 or coil embolization7 are only sporadically reported in the literature. To our knowledge, there has been no previous report of similar endovascular treatment using coil embolization of the proximal part of a PLSVC for symptomatic right-to-left shunt manifested as a recurrent transient ischaemic attack.

In conclusion, paradoxical embolism may be an important cause of ischaemic cerebral events, which has to be ruled out in patients with no other evident stroke aetiology. For diagnostic purposes, we recommend the use of contrast TTE and TEE with contrast agent application through the left arm peripheral intravenous line, which makes it possible to ascertain the presence of a right-to-left shunt. Nevertheless, computed tomography of the chest is recommended for a PLSVC with atypical left atrial drainage confirmation. Consequent endovascular occlusion of the PLSVC is feasible and safe and can be performed with minimal procedural risk. If transcatheter closure is not possible or effective, surgical treatment is necessary. If this cause of paradoxical embolism is not taken into consideration, the first manifestation of this clinical entity could be underestimated, increasing the likelihood of ischaemic stroke recurrence with potentially disabling or fatal consequences. This case report also aims to highlight the importance of close collaboration between neurologists, cardiologists and radiologists needed for accurate identification of stroke aetiology in young patients.

Supplementary data
Supplementary data are available at European Journal of Echocardiography online.

Conflict of interest: none declared.

References