# Successful endovascular repair of an unusual right-to-left shunt presenting with cerebral ischemia $\stackrel{\text{\tiny{}\%}}{\sim}$



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## ABSTRACT

Ischemic stroke due to congenital cardiopulmonary vasculature anomalies is rare in adults. We report a 54-year-old man with a stroke due to a unique right-to-left shunt who underwent successful endovascular treatment. This patient developed acute onset of right arm weakness with facial droop and aphasia which improved after intravenous thrombolysis. An MRI showed acute cerebral ischemia in the left middle cerebral artery and left posterior cerebral artery distribution. The patient developed recurrent stroke symptoms during agitated saline injection while undergoing a transthoracic echocardiogram which showed right-to-left shunting. Chest CT scan and conventional angiography revealed near occlusion of the superior vena cava. Head and upper limb venous return drained via a large left vertical vein into an anomalous left pulmonary vein into the left atrium. He underwent endovascular surgery to relieve the superior vena cava obstruction and to occlude the source of right-to-left shunt. While rare, congenital cardiopulmonary vasculature when right-to-left shunting is discovered on echocardiography in the setting of ischemic stroke. With large right-to-left shunts, agitated saline should be avoided.

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### 1. Introduction

A cardioembolic source is present in approximately 30% of ischemic strokes. In young adults, cardioembolic stroke may be attributed to a right-to-left shunt, most commonly an atrial septal defect (ASD) or patent foramen ovale (PFO) [1]. In adults, causes of a right-to-left shunt other than a PFO include pulmonary arteriove-nous fistulas and anomalous systemic venous return such as a persistent left superior vena cava with connection to the left atrium [2–5]. We present an adult patient with an ischemic stroke due to a unique cardiopulmonary vasculature anomaly which was successfully repaired endovascularly.

#### 2. Case presentation

A 54-year-old right-handed man with hypertension and a past history of an atrial-septal defect repaired at age 5 developed sudden onset of right-sided arm weakness with right facial droop, dysarthria, and aphasia. Initial head CT scan was negative for hemorrhage. He was administered intravenous tissue plasminogen activator 1 hour after symptom onset with near resolution of his symptoms. The following morning he experienced immediate worsening of his dysarthria with right face and arm weakness during injection of agitated saline for a transthoracic echocardiogram. His symptoms resolved within 30 minutes.

MRI of the brain showed a linear area of restricted diffusion in the left middle cerebral artery distribution and a small focus of restricted diffusion in the left posterior cerebral artery distribution. With a cardioembolic mechanism suspected due to his symptoms

\* This is an open-access article distributed under the terms of the Creative Commons Attribution-NonCommercial-No Derivative Works License, which permits non-commercial use, distribution, and reproduction in any medium, provided the original author and source are credited. during agitated saline in echocardiogram, the radiographic stroke appearance, and minimal vascular risk factors, unfractionated heparin was initiated for anticoagulation. He was then transferred to our institution for further evaluation.

A chest CT angiogram and transesophageal echocardiogram without agitated saline were obtained revealing several anomalies (Fig. 1A, B). The right superior vena cava (SVC) was subtotally occluded. The right head and neck veins drained via a left innominate vein drained into a large left-sided vertical vein, which also drained an anomalous left upper pulmonary vein. The vertical vein entered a dilated, tortuous fistula in the left lower lobe of the lung. The fistula primarily drained into the left atrium creating a significant right-to-left shunt. Upper extremity ultrasound showed no evidence of a venous clot.

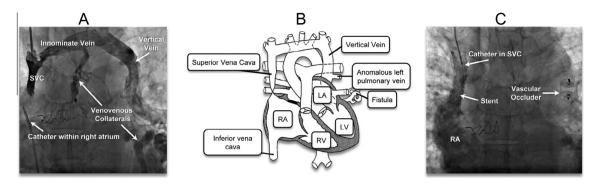
The patient underwent endovascular repair of his anomalous circulation and right-to-left shunting. Balloon dilatation of the stenotic right SVC was performed followed by placement of a Transhepatic Biliary Stent (PALMAZ GENESIS; Cordis, Bridgewater, NJ, USA). Once normal flow was re-established in the SVC, the left innominate vein was entered and an occluder device (Amplatzer Vascular Plug II; St Jude Medical, St Paul, MN, USA) was placed in the left vertical vein just distal to the junction of the anomalous left pulmonary vein. This resulted in a return of normal systemic venous flow with only a small residual left-to-right shunt due to the anomalous pulmonary vein (Fig. 1C). His arterial oxygen saturation rose from a baseline of 88% to 95% at the end of the procedure. He was discharged the following day on clopidogrel and aspirin therapy.

At 6 weeks posthospitalization the patient had a National Institutes of Health Stroke Scale score of 0 and was clinically doing well.

## 3. Discussion

This case demonstrates an adult patient with an ischemic stroke due to right-to-left shunting from an unusual cardiopulmonary

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**Fig. 1.** (A) Angiography of the cardiac and great vessels and (B) diagram demonstrating the preoperative configuration of the venous anomaly. Due to the superior vena cava (SVC) occlusion, injection of dye into the SVC resulted in filling of the innominate and vertical vein to an anomalous pulmonary vein and fistula draining into the left atrium. (C) Angiography demonstrating the stent after placement in the stenotic SVC and a normal flow of dye into the right atrium when injected into the SVC. LA = left atrium, LV = left ventricle, RA = right atrium, RV = right ventricle.

vasculature anomaly. He had a subtotal occlusion of his SVC with venous drainage from his head and upper extremities predominantly passing to the left atrium through a left vertical vein to an anomalous pulmonary vein. The vertical vein and anomalous pulmonary vein were felt to be congenital; however, the SVC occlusion and subsequent development of the fistula were likely acquired. The patient was successfully treated with endovascular repair.

Other cardiopulmonary anomalies have been described in adults associated with left-to-right shunt and stroke, not due to an ASD or PFO. These include persistent left SVC and arteriovenous fistulas [2–5].

While these are uncommon causes of stroke in adults, they should be considered if right-to-left shunting is noted on echocardiography without obvious ASD or PFO. CT angiography and/or conventional angiography should be considered. When a large right-to-left shunt is noted, agitated saline should be avoided to prevent paradoxical air emboli. In addition, intravenous lines should be filtered [6].

Patients should be referred to centers where pediatric and adult cardiology, pulmonary, and neurology expertise is available to determine the best treatment options.

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## **Conflicts of Interest/Disclosures**

The authors declare that they have no financial or other conflicts of interest in relation to this research and its publication.

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