

## CASE REPORT

# Delayed bubble, coil and trouble: young stroke as a presentation of paradoxical embolism from previously unrecognised pulmonary arterio-venous malformation (PAVM)

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## Summary

Young stroke patients should be investigated thoroughly to look for cardiac and extra-cardiac sources of emboli. We present a patient who was investigated for a cardiac source of emboli following an ischemic stroke. She was found to have a small patent foramen ovale (PFO), but due to the late appearances of bubbles on the bubble study it was thought that this was an incidental finding. Further investigation confirmed a PAVM was the source of emboli causing her stroke.

### Key Words

- ▶ patent foramen ovale
- ▶ pulmonary arteriovenous malformation
- ▶ contrast echocardiography

## Learning points:

- Paradoxical embolus is a rare complication of a pulmonary arterio-venous malformation (PAVM).
- Young stroke patients should be investigated for intra and extra-cardiac shunts, in particular, patent foramen ovale (PFO), ideally with a bubble study.
- Consider an extra-cardiac source of embolism when bubbles are seen in arriving late into the left heart.

## Background

A pulmonary arterio-venous malformation (PAVM) is an abnormal connection between a pulmonary artery and a pulmonary vein (1, 2). PAVM is a rare clinical condition with 80% being congenital; of these 50–80% are associated with hereditary hemorrhagic telangiectasia (HHT) (1). Clinical symptoms vary depending on the site of the PAVM. These can include epistaxis, dyspnoea haemoptysis, telangiectasias, cyanosis, clubbing (in the

presence of right to left shunt), and gastrointestinal bleeding.

Various imaging techniques can be used to detect a PAVM, with contrast echocardiography being the most sensitive. Though pulmonary angiography remains gold standard especially when therapeutic intervention is being planned, CT is more sensitive at detecting a PAVM and defining anatomical architecture.

Neurological complications are the commonest and are thought to be as a result of paradoxical emboli. Transcatheter embolization is first line treatment with surgery reserved for those who fail embolization or develop complications.

We present a young patient who suffered a stroke as a result of a paradoxical embolus from a previously undiagnosed PAVM.

## Case presentation

A 39-year-old Caucasian optometrist presented with a 2-week history of self-diagnosed unresolved left superior homonymous hemianopia and intermittent right-sided paraesthesia following an episode of typical migraine. The rest of her physical examination was unremarkable. It was felt that this presentation was due to a migrainous infarct of the right occipital lobe.

She had a known history of migraine with aura. Ten years prior she had an episode of sudden onset right-sided paraesthesia which resolved within 24 h with no residual neurological deficit apparently unrelated to her migraine and unexplained.

## Investigation

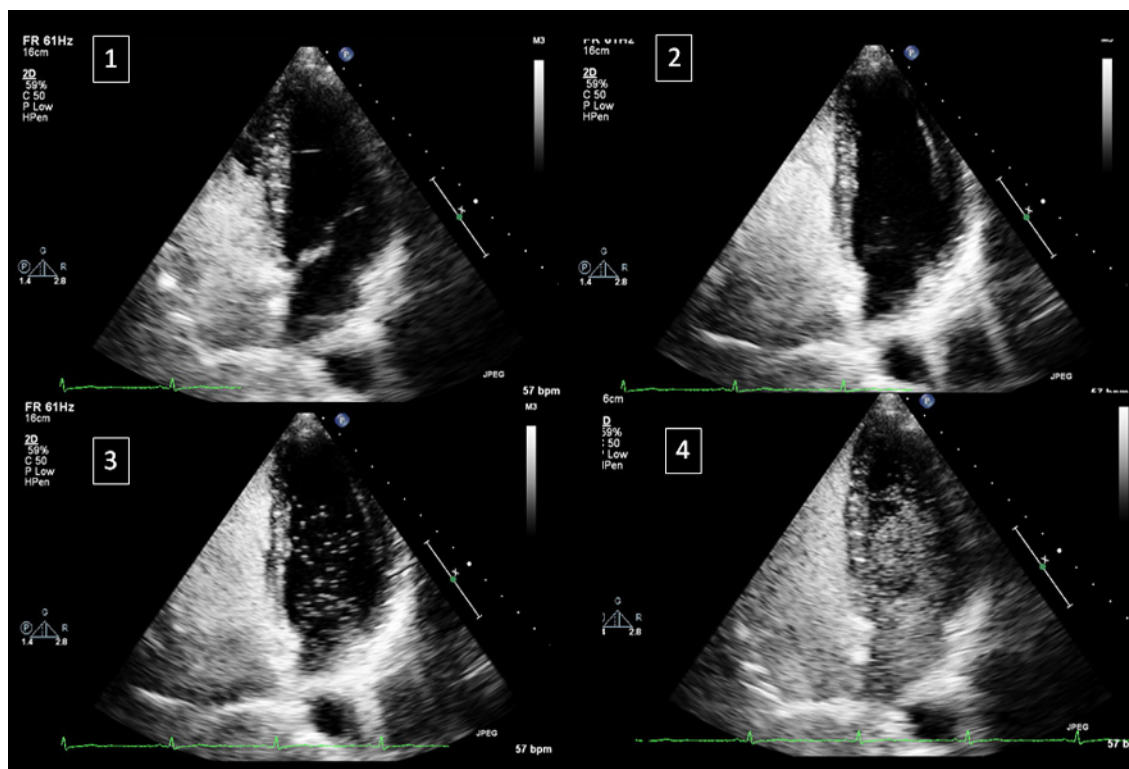
MRI of the brain confirmed a right medial occipital infarct. Thrombophilia and autoimmune screening were unremarkable and a 24-h cardiac monitor showed sinus rhythm with no arrhythmias. She was started on aspirin (75 mg once daily).

To exclude a cardiac source of embolism, a transthoracic echocardiogram (TTE) with agitated saline was performed. This demonstrated a large volume of bubbles within the left heart after two cardiac cycles (Fig. 1 and Video 1) with Valsalva. Trans-oesophageal echocardiogram (TOE) demonstrated a small patent foramen ovale (PFO) (Fig. 2), which was thought to be an incidental finding because of the delay in appearance of bubbles in the left heart.

### Video 1

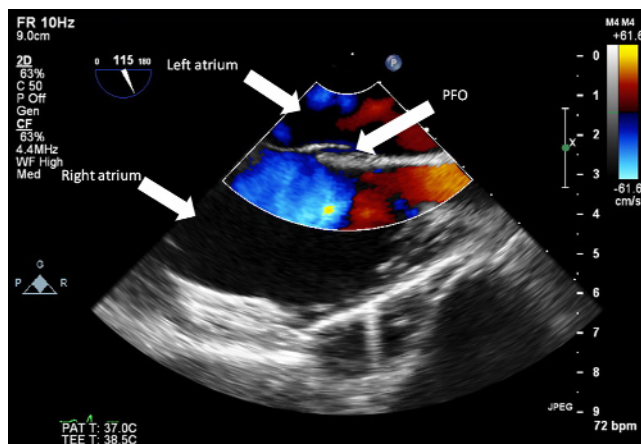
Apical four chamber with agitated saline, demonstrating delayed bubbles in the left heart. View Video 1 at <http://movie-usa.glencoesoftware.com/video/10.1530/ERP-20-0001/video-1>.

A CT study of the chest demonstrated a peripheral PAVM in the left lower lobe, measuring



**Figure 1**

Apical four chamber with agitated saline, demonstrating transit from right to left heart across four cardiac cycles.

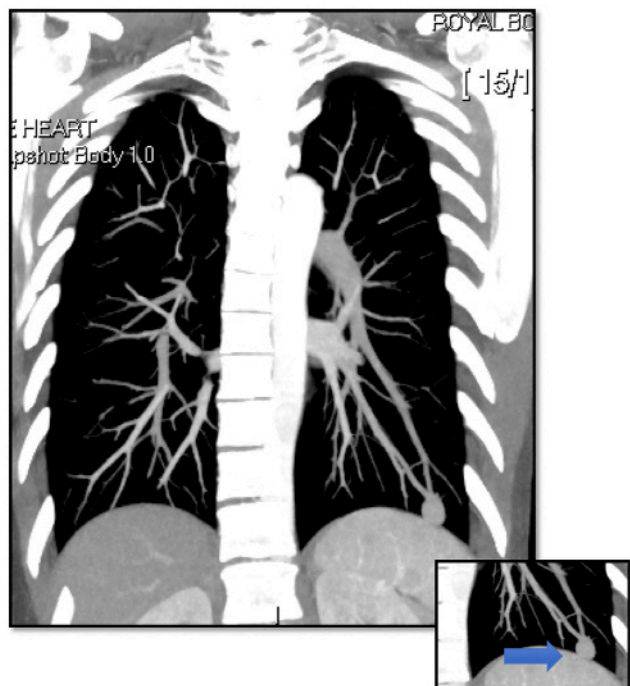


**Figure 2**  
Transoesophageal bi-caval view.

14×20×17 mm (Fig. 3). A pulmonary angiogram confirmed a small left basal AVM.

### Treatment and outcome

She underwent successful embolization of the PAVM with Amplatzer vascular device (St. Jude Medical). Since her embolization, she has had no further neurological symptoms or migraines. Repeat TTE with agitated saline



**Figure 3**  
CT chest: confirms pulmonary-arterial venous malformation (PAVM) in left lower lobe.

showed no residual right to left shunting with and without Valsalva. She remains well.

### Discussion

Cerebral infarction is a rare but recognised presentation of an undiagnosed PAVM. This complication results from a right to left shunt from a pulmonary artery to a pulmonary vein (3).

The most common complications of PAVM affect the CNS (20–60%). These include migraine 43%, transient ischemic attack 37%, stroke 18% and abscess 9% (1, 2, 3, 4). The most likely mechanism for this is a paradoxical embolism across the PAVM as demonstrated by this case.

Pulmonary angiography is the gold standard for diagnosis (1, 2, 3, 4, 5). It identifies a PAVM and defines the angioarchitecture of pulmonary vasculature needed for therapeutic embolization or surgical resection. Contrast enhanced CT is a valuable tool in diagnosing PAVM and is preferred to MRI (1, 2). Bubble echocardiography is useful at detecting intra-cardiac shunting and shunt fraction. Both TTE and TOE are sensitive in detecting cardiac shunting but often do not provide anatomical detail as to the cause (2, 3). Classically in PAVM there is a delay of 3–8 cardiac cycles before bubbles are seen in the left heart (1).

First line treatment is embolization with either a coil or balloon technique. Surgery is indicated in patients who fail embolotherapy, develop bleeding despite embolotherapy, intra-pleural rupture or those who have lesions that are not amenable to embolization (5). Current recommendation is to correct the PAVM without treating the associated PFO, as functional closure of the PFO is seen on follow-up. This suggests that as well as causing an ischaemic stroke, a PAVM may be responsible for development of a PFO (5).

Young stroke should be investigated with echocardiogram, ideally with a bubble study looking for a right to left shunt. In patients where bubbles appear in the left side of the heart but there is a clear delay, then an extra-cardiac source of embolism should be sought.

#### Declaration of interest

The authors declare that there are no conflicts of interest that could be perceived as prejudicing the impartiality of this case report.

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### Patient consent

Written informed consent for publication of their clinical details and clinical images was obtained from the patient.

### Author contribution statement

T S and J H drafted this work. R S and J K were the responsible physicians for this patient's care and guided revisions of this work.

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