

**Case  
Report**

# Double Atrial Septum with Interatrial Chamber Formation and Recurrent Paradoxical Embolism

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**Paradoxical embolism is a common cause of cryptogenic stroke. Cardiogenic origin is often the real culprit of unexplained stroke. We report a rare case of double atrial septum with an interatrial chamber and stenosis of the inferior vena cava orifice, which lead to recurrent paradoxical embolism, and then highlight the clinical importance of this rare type of congenital heart disease.**

**Keywords:** congenital heart disease, double atrial septum, interatrial chamber, paradoxical embolism, echocardiography

## Introduction

Paradoxical embolism is a common cause of cerebral vascular events with the absence of conventional risk factors. Cardiogenic origin, such as patent foramen ovale (PFO), is often the real culprit of unexplained stroke. A double atrial septum is a relatively rare cardiac anomaly, which could also lead to paradoxical embolism. We report a unique case of recurrent paradoxical embolism due to a double atrial septum anomaly with interatrial chamber (IAC) and stenosis of the inferior vena caval (IVC) orifice.

## Case Report

A 41-year-old woman presented to hospital with

recurrent syncope of thirteen years and a recent right hemi-paresis. Decreased myodynamia, elevated muscle tone of the right limbs, right positive Babinski sign, as well as Romberg's sign, were noted during the physical examination. Heart auscultation did not reveal any important information. Magnetic resonance imaging (MRI) and angiography of the brain showed an absence of blood perfusion and encephalomalacia in the left frontal, parietal and insular lobes, which indicated an old ischemic stroke. A further transthoracic echocardiogram led us to suspect a ruptured atrial septal aneurysm with a left-to-right shunt at the atrial level.

Peripheral vascular ultrasound found no arterial atherosclerosis plaque or deep venous thrombus formation. Electrocardiogram confirmed normal sinus rhythm. No other potential source of emboli was confirmed. Thus, a transesophageal echocardiography (TEE) was performed for further evaluation of the heart defect. Instead of a ruptured atrial septal aneurysm, TEE confirmed an unexpected double interatrial septum with persistent IAC formation, which communicated with both IVC and the two atria (**Fig. 1A**). The left atrium (LA) was connecting to the IAC **4/8** through an orifice between two septa (**Fig. 1A**). Rather than being directly connected to the right atrium (RA), the IVC was posteriorly placed and drained into the isolated IAC and communicated with the right atrium with a 5-mm stenotic orifice in the septum

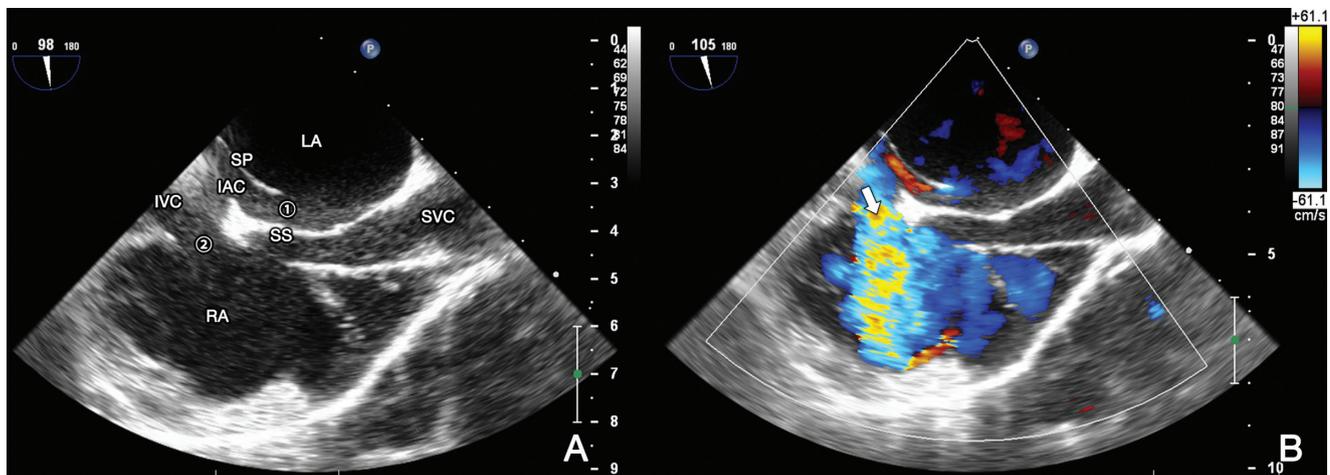
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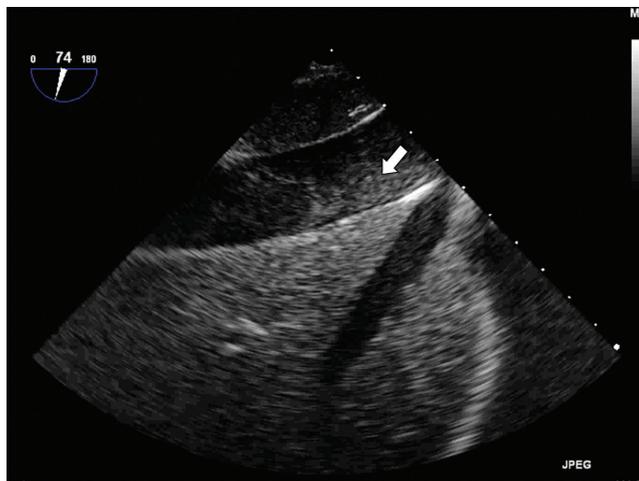
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**Fig. 1** TEE mid-esophageal bicaval view. (A) Double atrial septum enclosing an interatrial chamber (IAC). (B) Blood flow from inferior vena cava (IVC) restricting by stenosis of IVC orifice (arrow). LA: left atrium; RA: right atrium; SP: septum primum; SS: septum secundum; SVC: superior vena cava. ①: orifice between IAC and LA; ②: orifice between IVC and RA.



**Fig. 2** TEE showing blood stasis in the proximal part of inferior vena cava (IVC) (arrow).

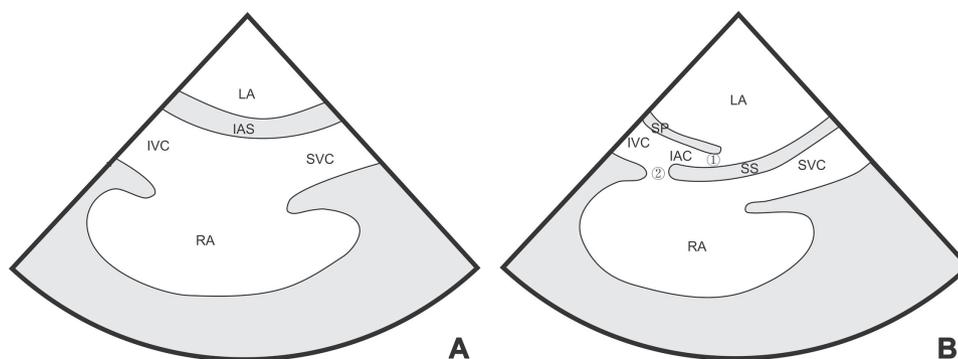
secundum (**Fig. 1A, B**). Blood flow was restricted when drained into the RA (**Fig. 1B**), and partial static blood flow in the IVC was also noticed by TEE, which largely facilitated the thrombus formation (**Fig. 2**). The TEE diagrams illustrated a comparison between the normal structure and this anomaly in the bicaval view (**Fig. 3**). Contrast echocardiography indicated a bidirectional shunt at the atrial level. Cardiogenic paradoxical embolism was then confirmed for this patient.

Surgical repair under cardiopulmonary bypass was performed. Stenosis of the IVC orifice was relieved and the shunt was closed. She was discharged home after 7 postoperative days without complications.

## Discussion

Ischemic stroke in the absence of conventional risk factors for cerebral vascular diseases is known as cryptogenic stroke. Paradoxical embolism is a common cause of this type of stroke. It may due to venous thromboembolus transit from the right to left cardiac chamber via the inter-ventricular or interatrial (e.g. PFO) communications, or pulmonary arteriovenous malformation.<sup>1)</sup> A double atrial septum is a rare type of cardiac anomaly, which was reported by Seyfert and Javois.<sup>2,3)</sup> Roberson further summarized the echocardiogram feature of the double atrial septum anomaly with a persistent interatrial space.<sup>4)</sup> Cohen and Palinkas also described the anomaly of an atrial septum primum deviation, which might present as a double atrial septum in certain cases.<sup>5,6)</sup> Although the majority of patients were asymptomatic, paradoxical embolism was a common clinical 5/8 manifestation for this type of anomaly.

Different from a previously reported double atrial septum anomaly, this unique case featured an isolated IAC connecting the IVC to both atria and a sequent restricting the venous return from IVC to right atrium (**Fig. 3**). Static blood in the IVC caused by restriction (**Fig. 3**) could be assumed to be the origin of the thromboembolism. A potential shunt between the IAC and LA could make it easy for emboli to transit into the left-sided chamber and cause a paradoxical embolism. This explains the symptoms and signs presented in this patient. As to the anatomical nature of this defect, it could not be a cor triatriatum dexter because there is no subdivision of the



**Fig. 3** Schematic mid-esophageal bicaval view. (A) Normal connection of IVC and RA, and interatrial septum (IAS) separating LA and RA. (B) In this case, the interatrial chamber (IAC) was bordered with SP and SS, and connected LA, IVC and RA. LA: left atrium; RA: right atrium; SP: septum primum; SS: septum secundum; SVC: superior vena cava. ①: orifice between IAC and LA; ②: orifice between IVC and RA.

RA. A large eustachian valve with a PFO could also be ruled out because the “membrane” emerged from the posterior of the IVC and the true eustachian valve remnant should be noted on the anterior rim of the IVC orifice (**Fig. 1A**). We speculated that the origin of this anomaly was due to malposition of the septum primum, which failed to fuse with the septum secundum during embryological development while the IVC was posteriorly placed and somehow overriding the interatrial septa, leaving a persistent space as a passage between the two atria and the IVC.

Clinical awareness of a disease of this kind should be noted. Unlike normal congenital heart diseases, malformation in the atrium level can often be misdiagnosis via transthoracic echocardiography. A TEE is often necessary.

## Conclusion

The rarity of this case comprised at least four aspects including double atrial septum, interatrial chamber as a common pathway of blood flow from IVC, RA and LA, stenosis of the IVC orifice, and recurrent paradoxical embolism. To the best of our knowledge, this is the first case reported of 6/8 this kind of anomaly.

## Disclosure Statement

None.

## References

- 1) Maron BA, Shekar PS, Goldhaber SZ. Paradoxical embolism. *Circulation* 2010; **122**: 1968-72.
- 2) Javois AJ, Roberson DA. Unusual atrial septal anatomy resulting in an interatrial chamber: the true triatrial heart? *Pediatr Cardiol* 2007; **28**: 224-8.
- 3) Seyfert H, Bohlscheid V, Bauer B. Double atrial septum with persistent interatrial space and transient ischaemic attack. *Eur J Echocardiogr* 2008; **9**: 707-8.
- 4) Roberson DA, Javois AJ, Cui W, Madronero LF, Cuneo BF, Muangmingsuk S. Double atrial septum with persistent interatrial space: echocardiographic features of a rare atrial septal malformation. *J Am Soc Echocardiogr* 2006; **19**:1175-81.
- 5) Cohen MS, Weinberg P, Coon PD, et al. Deviation of atrial septum primum in association with normal left atrioventricular valve size. *J Am Soc Echocardiogr* 2001; **14**: 732-7.
- 6) Pálincás A, Nagy E, Czakó L. Deviation of the atrial septum primum predisposing to local thrombus formation. *J Am Soc Echocardiogr* 2011; **24**: 935.e3-5.