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CASE REPORT
Congenital Heart Disease

Complications caused by iatrogenic right-to-left shunt after surgical closure of atrial septal defect: a case report

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Background Atrial septal defect (ASD) is a common congenital heart disease. For this condition, surgical treatment can be required depending on the size and type of ASD. This study included a case of a patient who complained of persistent dyspnoea after the surgical treatment for ASD.

Case summary A 16-year-old girl who underwent a surgical patch closure for ASD at the age of 2 years presented to the emergency department and was diagnosed with acute stroke. Since childhood, she had suffered from exertional dyspnoea due to an unknown cause. Transthoracic echocardiography revealed normal chambers size and function and no signs of right heart strain. Transoesophageal echocardiography (TOE) revealed a misplaced interatrial patch from the previous surgery, which allowed the whole blood to flow from the inferior vena cava (IVC) to the left atrium (LA), creating a large right-to-left shunt that resulted in stroke and heart failure. The patient underwent surgical treatment, and her symptoms improved significantly. Six months later, she was doing well without neurological complications and dyspnoea.

Discussion This patient experienced stroke at the age of 16 years and had been suffering from heart failure since childhood. A large right-to-left shunt flow from the IVC to the LA by misplaced interatrial patch was found using TOE, right-sided heart catheterization, and inferior caval venography. This diagnosis should be considered in patients complaining of persistent dyspnoea with hypoxia after the surgical repair of ASD.

Keywords Atrial septal defect • Chronic heart failure • Complication • Hypoxaemia • Postoperative • Case report

ESC Curriculum 2.2 Echocardiography • 6.1 Symptoms and signs of heart failure • 7.5 Cardiac surgery • 9.7 Adult congenital heart disease

Learning points

- Clinical evaluation subsequent to surgical closure for atrial septal defect should assess for signs of embolic events, arrhythmia, and dyspnoea.
- It is important to understand the need of further evaluations to achieve an accurate diagnosis in patients complaining of persistent dyspnoea after surgical treatment for congenital heart disease.

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Introduction

Atrial septal defect (ASD) is a common congenital heart disease in adults.[1] Although device closure has increased over the past few decades, surgical treatment is still an important option for some patients, depending on the size and type of ASD.[2] This study included a case of a patient who complained of persistent dyspnoea after the surgical treatment of ASD, with no causative findings on transthoracic echocardiography (TTE).

Timeline

Fourteen years before admission	Diagnosed with atrial septal defect (ASD). The patient underwent a surgical patch closure
One year before admission	The patient presented to other hospitals with exertional dyspnoea; however, examinations, such as transthoracic echocardiography and chest computed tomography, failed to determine the specific cause of her symptoms
Day 0	The patient was admitted to our hospital with aphasia and right-sided hemiparesis and was diagnosed with cerebral infarction
Day 3	The patient was referred for the investigation of cardioembolic causes Transoesophageal echocardiography was performed, which revealed a misplaced interatrial patch from the previous surgery, creating a large right-to-left shunt
Day 4	The patient underwent right-sided heart catheterization. Inferior caval venography revealed that a contrast medium from the inferior vena cava proceeded through the left atrium and left ventricle into the aorta but not towards the pulmonary circulation
Day 9	Surgery was performed to remove the previous patch and close the original ASD site
Day 14	The patient was discharged from the hospital without any medications. Her symptoms significantly improved
Six months after discharge	The patient could exercise much more than before without shortness of breath. Also, she was doing well without neurological complications

Case presentation

The patient was a Korean female who was diagnosed with ASD at the age of 18 months and underwent surgical patch closure. She regularly visited the outpatient clinics of the cardiothoracic surgery department until she was 5 years old and was later lost on follow-up. When she was a child, she was less active than the other children her age. In her school days, she experienced shortness of breath even during light physical activities such as climbing stairs. Because the dyspnoea persisted, with cyanosis appearing on her lips and nails during activity, she presented to other hospitals at the age of 10 and 15 years; however, examinations, such as TTE and chest contrast-enhanced computed tomography (CT), failed to demonstrate a specific cause of her symptoms.

At the age of 16 years, the patient presented to the emergency department with aphasia and right-sided hemiparesis. Neurological examination revealed recovered sensory and motor function but with persistent aphasia. Physical examination showed the cyanosis of the lips and nails. The heart sounds were regularly rhythmic, with no murmurs or gallops. Her blood pressure was 121/75 mmHg, pulse rate was 79 beats/min, and respiratory rate was 20 breaths/min; however, her oxygen saturation was low (89% at room air). The brain magnetic resonance imaging (MRI) revealed a cerebral infarction at the inferior territory of the left middle cerebral artery (*Figure 1*). Considering her young age and low probability of large-vessel atherothrombotic stroke on cerebrovascular CT angiography, she was referred for investigating cardioembolic causes.

In this setting, because the patient had a history of surgical repair for ASD, the initial differential diagnosis was leaning towards paroxysmal atrial tachyarrhythmia or paradoxical embolism caused by the remnant ASD.

The initial electrocardiography (ECG) and 24 h ambulatory ECG monitoring did not confirm any arrhythmias; moreover, TTE revealed normal chamber sizes and contractile functions of the right and left ventricle (LV; Supplementary material online, *Videos S1 and S2*). No sign of volume or pressure overloading of the right heart was observed, which might indicate remnant ASD shunt. No causative valvular abnormalities were observed but only a trace of tricuspid regurgitation and pulmonary regurgitation. The definite interatrial shunt

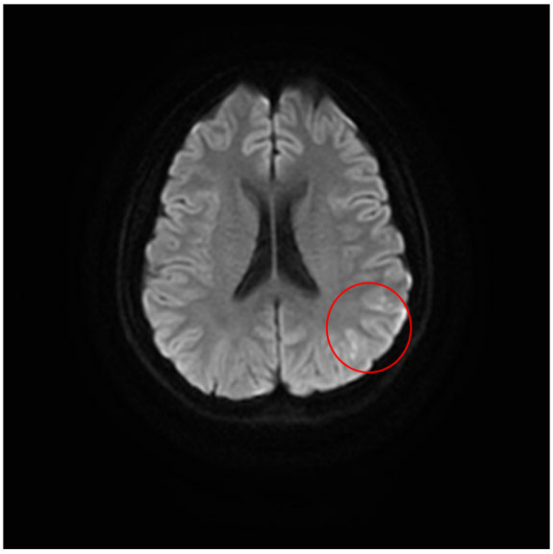


Figure 1 Brain magnetic resonance image (axial diffusion-weighted) showing multiple high-signal-intensity lesions at the inferior territory of the left middle cerebral artery (red circle), which indicates an acute cerebral infarction in that territory.

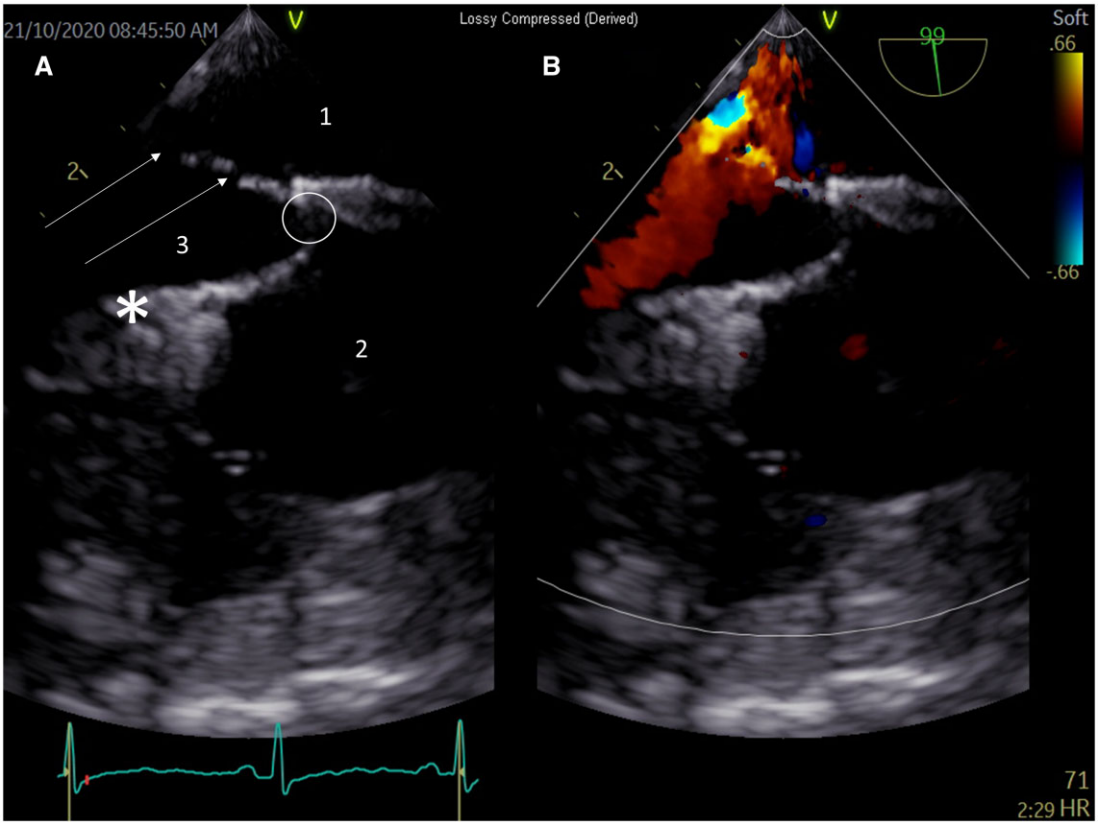
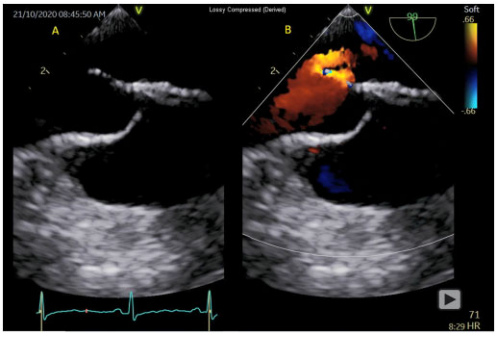


Figure 2 Transoesophageal echocardiography image in a projection of the caval axis. The colour flow comparison between the two-dimensional image (*A*) and its corresponding colour flow Doppler image (*B*) is represented by the left and right images, respectively. The imaging reveals a right-to-left shunt flow from the inferior vena cava to the left atrium through multiple fenestrations connected to the inferior vena cava at the interatrial septum. The inferior margin of the surgically placed interatrial patch was shifted to the right, excluding the inferior vena cava orifice from the right atrium (1 = left atrium; 2 = right atrium; 3 = inferior vena cava; arrows = multiple fenestrations; asterisk = inferior margin of the incorrectly positioned surgical patch; circle = superior margin of the incorrectly positioned surgical patch).

flow was also not clearly identified (Supplementary material online, *Videos S3 and S4*). To further investigate the cardiac source of embolism, transoesophageal echocardiography (TOE) was performed, which revealed the right-to-left shunt flow from the inferior vena cava (IVC) to the left atrium (LA) through the defect in the original membranous septum; moreover, the inferior margin of the surgically placed interatrial patch was shifted to the right, excluding the IVC orifice from the right atrium (RA). This made the entire venous flow from the IVC drain directly into the LA through the original interatrial septal defect (*Figure 2* and *Video 1*). An agitated saline test was performed using TOE through a vein in the upper extremity. This confirmed that the blood flow proceeded to the right ventricle through the superior vena cava and RA and no connection between the RA and IVC was observed (Supplementary material online, *Video S5*). Finally, the patient underwent right-sided heart catheterization, which demonstrated a 71.6% IVC saturation, 92.8% LA saturation, and 91.3% LV saturation, and the mean IVC and LA pressures were 2 and 4 mmHg, respectively. Inferior caval venography demonstrated that a contrast medium from the IVC proceeded through the LA and LV into the aorta but not towards the pulmonary circulation (*Figure 3* and *Videos 2 and 3*).



Video 1 Transoesophageal echocardiography in a projection of the caval axis. The colour flow comparison between the two-dimensional image and its corresponding colour flow Doppler image is represented by the left and right images, respectively. The imaging shows the right-to-left shunt flow from the inferior vena cava to the left atrium through multiple fenestrations connected to the inferior vena cava at the interatrial septum. The inferior margin of the surgically placed interatrial patch was shifted to the right, excluding the inferior vena cava orifice from the right atrium.

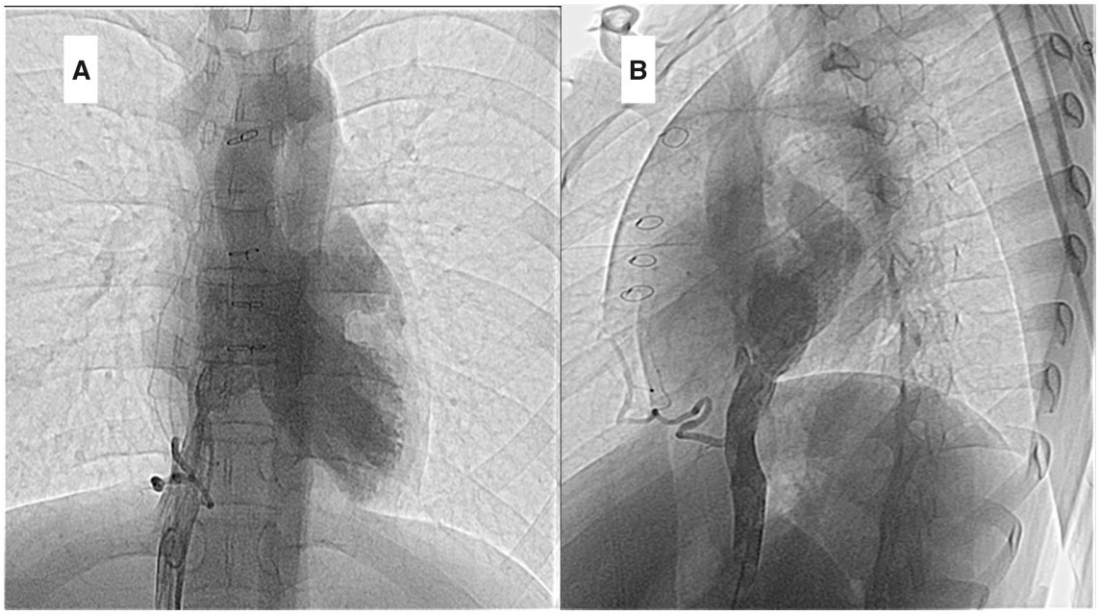
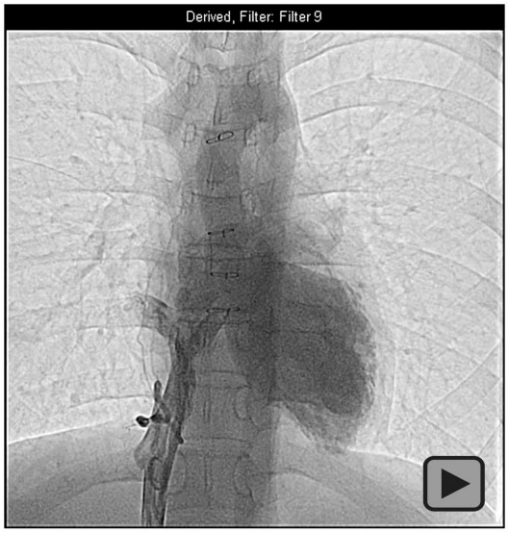
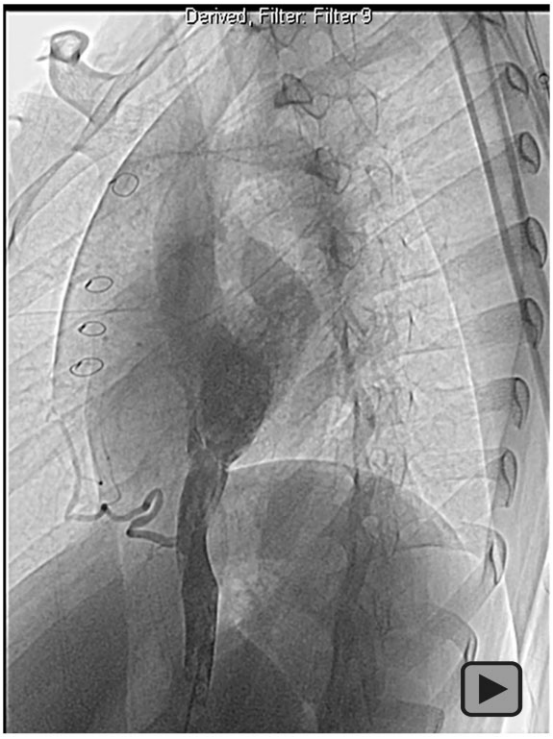


Figure 3 Inferior caval venography image. The image shows that the contrast medium started from the inferior vena cava to the left atrium and left ventricle into the aorta. (*A*) Anteroposterior view. (*B*) Lateral view.



Video 2 Inferior caval venography. It demonstrates that the contrast medium started from the inferior vena cava to the left atrium and left ventricle into the aorta. Anteroposterior view.



Video 3 Inferior caval venography. Lateral view.

The patient's symptoms and signs were determined to be due to iatrogenic IVC to LA drainage resulting from the improper surgical repair of ASD; therefore, we decided to perform a surgical correction. Preoperatively, the cardiac CT images with contrast could not clearly demonstrate the connection from the IVC to LA (Supplementary material online, *Video S6*). A CT scan was performed, including the lower extremities, to determine the possible source of embolism. The CT scan results indicated that the bilateral lower extremities had no evidence of deep vein thrombosis and other solid organs were also unremarkable; moreover, she was not taking any

medications and had no family history of venous thromboembolism, and no coagulation disorders were detected.

In the operating room, it was confirmed that the previous patch for ASD was placed between the RA and IVC orifice. This prevented the progression of the blood flow from the IVC to the RA and instead caused the unsaturated venous flow to directly proceed into the LA. Thus, the previous patch was removed, and a repatch closure of the interatrial septal defect near the IVC was performed. An intraoperative TOE was performed immediately and showed no residual shunt flow.

After a favourable postoperative course, the patient was discharged from the hospital without any medications. The surgical correction of the IVC drainage was confirmed in a follow-up CT scan, and her symptoms significantly improved. Six months later, the patient could exercise much more than before without any shortness of breath. She was also doing well without any neurological complications.

Discussion

Atrial septal defect is a common congenital heart defect. The sinus venosus type of ASD is a rare form, accounting for 5–10% of all ASD types.[3] If a large defect is left untreated, prolonged left-to-right shunting and excessive pulmonary blood flow may lead to right-sided heart failure, pulmonary hypertension, and Eisenmenger syndrome.[1] Surgical closure is considered a required treatment.[3] Remnant intracardiac shunt or improper closure of ASD can cause persistent left-to-right shunt flow and subsequent heart failure, which can be a more common complication of intracardiac shunt after surgical correction of ASD. In those cases, the sign of right heart strain is mostly observed. In this case, we cannot detect any sign of volume or pressure overload in the right heart, which made it more difficult to suspect an intracardiac shunt as a diagnosis.

Regarding the patient from this study, although her past surgical records could not be retrieved, it can be presumed that the surgical patch was placed between the RA and IVC by an inappropriate IVC cannulation during surgery. In paediatric surgery, the application of TOE can be more difficult in adults. Cardiac CT or MRI can be alternative diagnostic tools;[4] however, they also have some limitations. Children might require general anaesthesia for these imaging modalities. Echocardiography with agitated saline is a feasible option. The injection of a contrast through the lower extremities can provide a clearer understanding of the anatomical structures of IVC and both atria. If an individual has adequate echocardiographic windows, TTE with saline contrast test may provide a clue for the diagnosis. Unfortunately, this patient had poor acoustic windows, especially the subcostal view. If the diagnosis by TTE is insufficient because of the inadequate visualization of relevant structures, TOE must be considered for visualization of the structures of IVC and both atria.

This patient has been suffering from heart failure for many years and had an acute stroke at an early age. The delayed diagnosis may result in paradoxical embolism and high cardiac output heart failure via right-to-left shunt flow. Long-term regular follow-up after surgical closure is important because of the late onset of atrial arrhythmias and stroke, particularly in adults older than 40 years.[5]

A previous study reported a case of drainage from the IVC to the LA caused by a redundant flap of the septum secundum before the surgical correction of ASD.[6] Moreover, we presented a rare case of a right-to-left shunt of the entire IVC flow that proceeded to the LA because of a surgically misplaced patch.

Conclusions

In patients complaining of persistent dyspnoea after surgical closure of ASD, the possibility of complications due to improper surgical corrections should be considered. Transoesophageal echocardiography can be a useful method to understand the complex anatomy in such cases.

Lead author biography

Myeong Seop Kim, MD, graduated from Kyungpook National University School of Medicine, Daegu, Republic of Korea. He has been a doctor for 2 years in Department of Cardiovascular Medicine at Kyungpook National University Hospital. He is interested in diagnosing and treating patients of heart failure and cardiac echocardiography and pursuing further career in cardiology.

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

Conflict of interest: None declared.

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