

Atrial Septal Defect Presenting with Dyspnoea in the Upright Position

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Abstract

Background: An atrial septal defect (ASD) is a common congenital heart disease in adults. Signs or symptoms such as shortness of breath, especially when exercising, leg fatigue, heart palpitations, stroke, or heart murmur can be detected. However, some patients with ASD present with uncommon symptoms such as dyspnoea with upright position.

Case Summary: We report the case of a patient with ASD who complained of dyspnoea with upright position. Dyspnoea and desaturation were observed immediately after sitting upright. The patient was diagnosed as having platypnoea—orthodeoxia syndrome. We performed transcatheter ASD closure with no compliments, and the desaturation in the upright position was resolved.

Discussion: Patients with ASD present with different symptoms including dyspnoea with upright position. Therefore, dyspnoea in the upright position should not be overlooked in such patients.

Key words: Atrial septal defect, platypnea-orthodeoxia syndrome, dyspnea, transcatheter ASD closure

Introduction

Atrial septal defect (ASD) is a common congenital heart disease mostly women in adults. Commonly, electrocardiogram (ECG) shows atrial fibrillation, right axis and right ventricular hypertrophy in patients with ASD¹⁾. Transthoracic echocardiography shows right heart overload and pulmonary hypertension as well as ASD. Signs or symptoms such as shortness of breath, especially when exercising, leg fatigue, heart palpitations, stroke, or heart murmur can be detected in adulthood²⁾. However, some patients with ASD present with uncommon symptoms. We encountered a patient with ASD who presented with dyspnoea with upright position.

Case

A 79-year-old woman who complained of syn-

cope due to sick-sinus syndrome underwent permanent pacemaker implantation in other hospital one year ago. She didn't take any cardiac medication since then. Although her son who died with acute myocardial infarction, others in family didn't have any cardiac events.

She has been admitted to our hospital with worsening chronic heart failure due to persistent atrial fibrillation and underwent radiofrequency catheter ablation for atrial fibrillation. Then, she was first diagnosed with ASD. Although her heart failure was compensated, she reported worsening desaturation while sitting upright. Her ECG showed atrial pacing and ventricular sensing rhythm with a right axis deviation and complete right bundle branch block (**Fig. 1**). Furthermore, the right heart catheterization shows the pulmonary systemic flow ratio is more than 2.0

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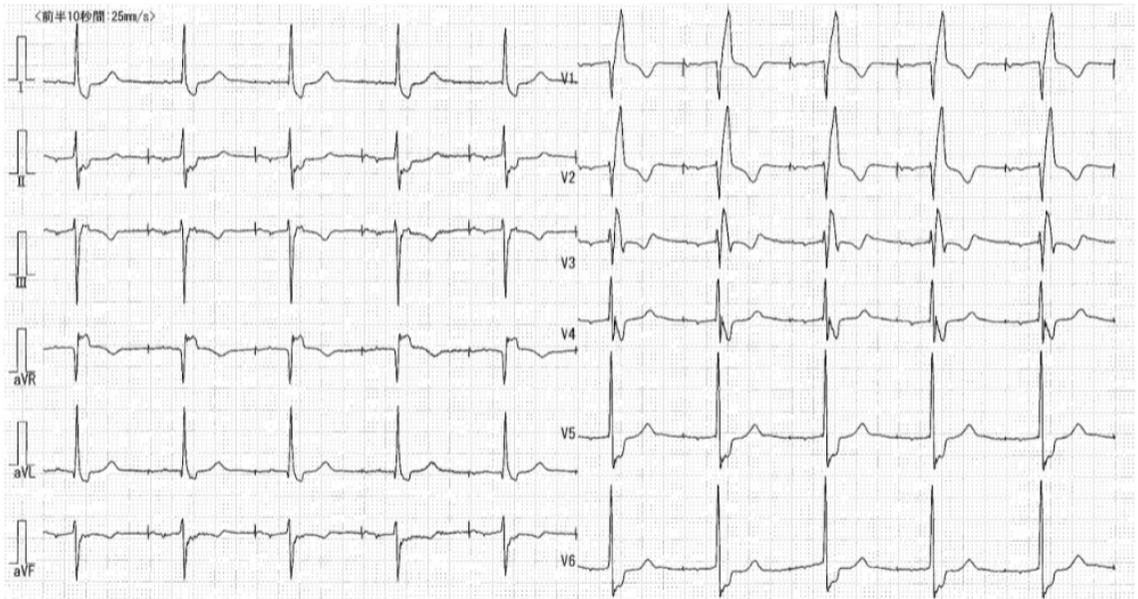


Fig. 1. Electrocardiogram of the case
 The ECG demonstrates atrial pacing and ventricular sensing rhythm with right axis deviation and complete right bundle branch block.
 ECG; Electrocardiogram

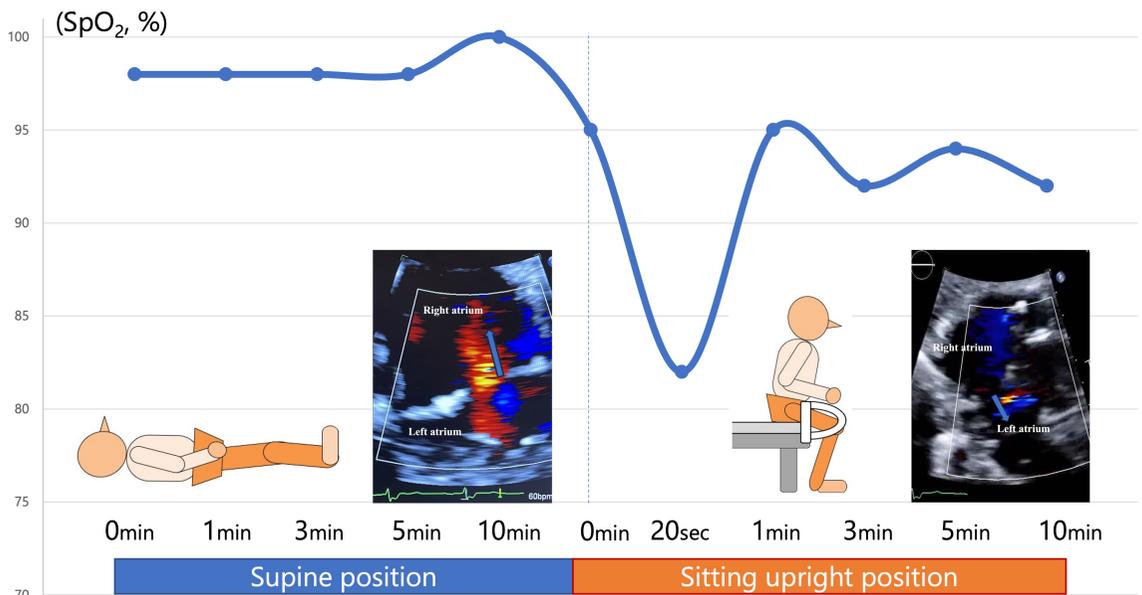


Fig. 2. Desaturation pattern before ASD closure
 Preoperatively, desaturation is observed immediately after assuming the upright position. Color Doppler echocardiography reveals a left-to-right shunt in the supine position and a right-to-left shunt in the upright position through the ASD.
 ASD; atrial septal defect

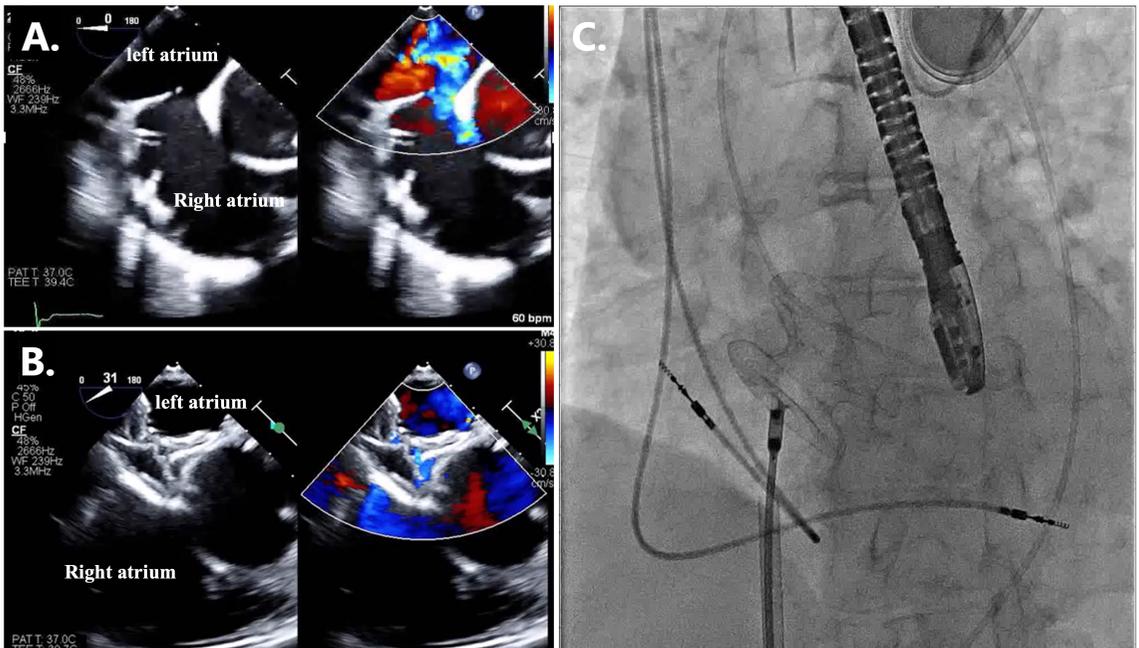


Fig. 3. Transthoracic echocardiographic and fluoroscopic images during ASD closure
 The ASD identified by transesophageal echocardiography (A) was successfully closed (B, C) using a 24-mm Amplatzer septal occluder.
 ASD; atrial septal defect

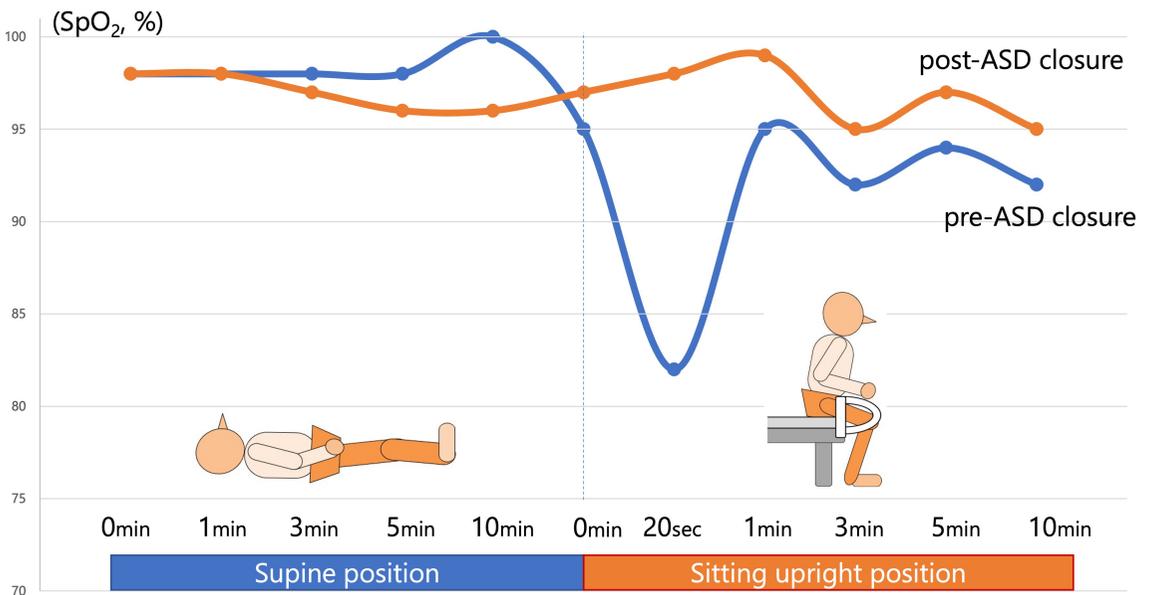


Fig. 4. Desaturation pattern before and after ASD closure
 Preoperatively, desaturation was observed immediately after assuming the upright position. Postoperatively, no significant desaturation was observed following ASD closure.
 ASD; atrial septal defect

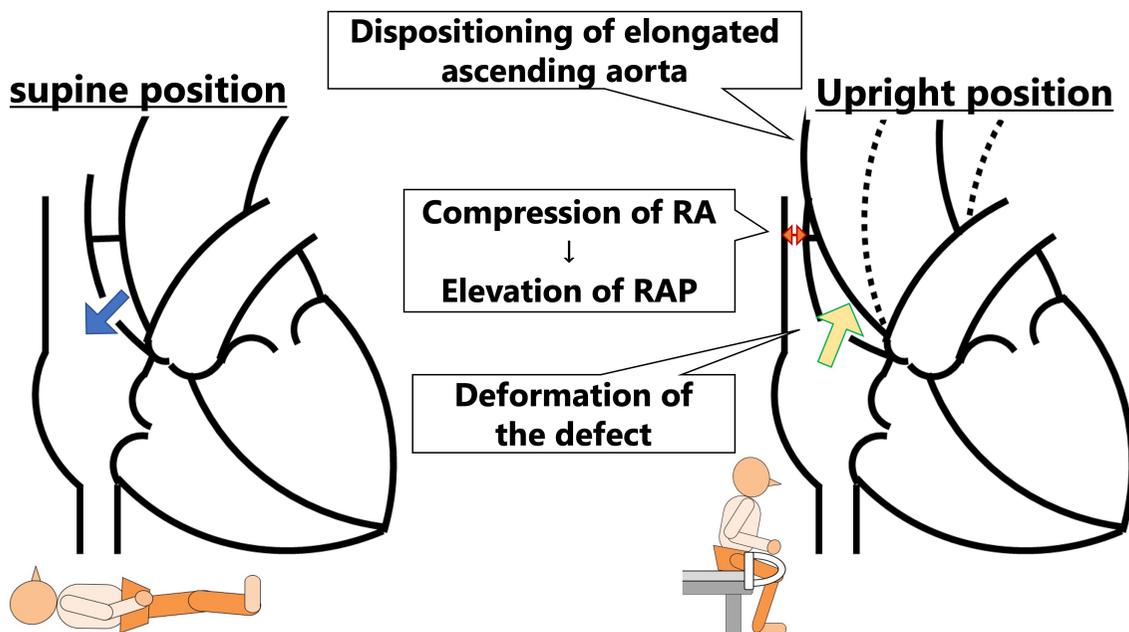


Fig. 5. Schematic representation of the cause of POS in this case
 In the sitting position, the elongated ascending aorta repositions, compressing the right atrium and elevating right atrial pressure, which leads to deformation of the defects.
 POS: platypnoea—orthodeoxia syndrome.

and desaturation while sitting upright was proven by the decreasing partial pressure of oxygen in blood gas evaluation (Fig. 2). The echocardiography shows R-L shunt when it's upright position (Fig. 2). We performed transcatheter ASD closure with a 24-mm Amplatzer septal occluder (Fig. 3), and the desaturation in the upright position was resolved (Fig. 4).

Discussion

We encountered a patient with ASD who presented with dyspnoea in the upright position, also known as the platypnoea—orthodeoxia syndrome (POS). POS is characterized by dyspnoea and oxygen desaturation induced by an upright position and relieved by recumbency. Since a patent foramen ovale shows near symptoms in its general population, POS is mostly missed inpatients with a patent foramen ovale (PFO)³. Generally, the pathogenesis of POS involves anatomical factors such as interatrial communication, and functional factors such as chronic obstructive pulmonary disease, ascending aorta elongation, and pulmonary embolism.⁴ POS is primarily reported in association with PFO due to its morphology⁵. However, there have also been reports of POS associated with ASD⁶, and improvement in

symptoms has been demonstrated following ASD closure. This case followed a similar course, making it a valuable example where simultaneous monitoring of SpO₂ changes and echocardiographic findings led to a definitive diagnosis. In our patient, positional change of the ascending aorta while sitting in an upright position expanded the inter-atrial communication, temporarily allowing venous blood from the inferior vena cava to flow into the left atrium (Fig. 5). Therefore, this case of POS improved with ASD closure. Although a slight decrease in oxygen saturation, possibly due to residual neuroregulatory dysfunction, was still observed post-closure, there was a clear overall improvement in oxygen saturation, and the symptoms were consistently alleviated after ASD closure. We successfully treated a 79 years old patient with ASD who presented with dyspnoea with upright position using a transcatheter closure device. Patients with ASD have various symptoms and sometimes present with unusual symptoms. Therefore, POS should not be overlooked in patients with ASD.

Conclusion

Patients with ASD have varied symptoms and sometimes present with unusual symptoms. There-

fore, platypnoea—orthodeoxia syndrome should not be overlooked in patients with ASD.

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Author contributions

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Conflicts of Interest

M. Izumo and S. Kuwata are consultants for Abbott vascular and Edwards Lifesciences. The other authors have no conflicts of interest to declare.