Platypnea-orthodeoxia Syndrome Diagnosed Using Contrast Transesophageal Echocardiography with Simultaneous SpO₂ Monitoring

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Abstract

Platypnea-orthodeoxia syndrome (POS) is a rare condition characterized by interatrial right-to-left shunting that is exacerbated in the upright position. We herein report a 78-year-old woman with POS that remained undiagnosed for 2 years, despite repetitive transthoracic echocardiography (TTE). POS was ultimately diagnosed using contrast transesophageal echocardiography (TEE), which revealed a marked increase in right-to-left shunting in the sitting position, associated with simultaneous desaturation. Therefore, we propose that POS should be considered according to the clinical symptoms, regardless of the repetitive TTE results, and contrast TEE should be performed in both the supine and sitting positions to exclude a diagnosis of POS.

Key words: platypnea-orthodeoxia syndrome, contrast echocardiography, transesophageal echocardiography

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Introduction

Platypnea-orthodeoxia syndrome (POS) is a rare condition characterized by interatrial right-to-left shunting that is exacerbated in the upright position. Although the precise pathology of POS is unknown, this syndrome has been associated with intracardiac structural factors such as patent foramen ovale (PFO) and atrial septal defect (ASD), as well as with extracardiac factors such as aortic root enlargement and elongation (1). However, the diagnosis of POS is very difficult to establish according to transthoracic echocardiography (TTE), which is the imaging technique initially recommended to investigate intracardiac shunting disease. In nearly all case reports of POS, the diagnosis was confirmed using contrast transesophageal echocardiography (TEE).

We herein present a case of POS associated with an aortic root deformity, which took two years to diagnose. Ultimately, the diagnosis was confirmed on contrast TEE with simultaneous monitoring of the peripheral capillary oxygen saturation (SpO₂), which clearly demonstrated the concordance between a postural-related increase in right-to-left shunting and desaturation. The aim of this case report is to highlight the difficulty in diagnosing POS on the basis of TTE imaging alone, as well as the usefulness of contrast TEE.

Case Report

A 78-year-old woman was referred to our hospital due to the onset of severe dyspnea. Her symptoms had begun two years previously, and the cause of the symptoms had not yet been identified, despite performing various investigations including a blood analysis, TTE, spirometry, and chest-enhanced computed tomography (CECT). The patient showed moderate thoracic kyphosis and a marked decrease...
in the SpO2 when assuming an upright position. The results of the physical examination were otherwise unremarkable. The patient’s SpO2 values, measured in various positions and under different conditions, are reported in Table. In addition to a lowering of the SpO2 upon upright sitting, desaturation was also evident in the right lateral decubitus position, which did not improve with oxygen inhalation. Desaturation was not exacerbated by exercise. A blood analysis showed no diagnostic findings to account for the patient’s hypoxia, such as the elevation of brain natriuretic peptide, Krebs von der Lungen-6, or D-dimer levels. Three-dimensional CECT revealed a severely enlarged and elongated ascending aorta, which prominently compressed the right ventricular inflow tract compared with in healthy patients (Fig. 1), without evidence of parenchymal lung disease, pulmonary embolism, or other vascular abnormalities. No intracardiac shunting, prominent Eustachian valve, or atrial septal aneurysms was identified on TTE. However, we considered further investigation of intracardiac shunting to be warranted, as clear visualization of the atrial septum was very difficult on TTE due to the deformed aortic root (Fig. 2a, b), and because hypoxia, which was strikingly resistant to the inhalation of high-concentration oxygen (Table), was consistent with right-to-left shunting. Subsequent contrast TTE using agitated saline revealed intracardiac right-to-left shunting (Fig. 2c, d), and lung perfusion scintigraphy indicated a right-to-left shunt ratio of 11.4%. Considering all these findings, hypoxia due to intracardiac shunting was strongly suspected. However, because of the poor visualization of the atrial septum on TTE, it remained unclear whether it was interatrial shunting.

To confirm the possible diagnosis of POS, we proceeded to perform contrast TEE in both the left lateral decubitus and left lateral sitting positions. TEE clearly demonstrated a significant increase in the influx of contrast through the PFO slit in the left lateral sitting position compared with in the left lateral decubitus position. The slit of the PFO was prominently widened to 5.5 mm in the left lateral sitting position (Fig. 3). These findings were consistent with simultaneous SpO2 monitoring performed during the examination (Fig. 4). According to the results of these imaging examinations, the diagnosis of POS was confirmed and the patient was transferred to another hospital for transcatheter occlusion of the PFO, which led to resolution of her dyspnea and hypoxia.

### Discussion

In this case report, we described the clinical presentation and diagnosis of POS in a 78-year-old woman with a two-year history of dyspnea. Using contrast TEE and CECT, a PFO and a severely enlarged and elongated aortic root were identified to be the underlying causes of the POS. According to the findings of this case, we advocate the use of contrast TEE with simultaneous SpO2 monitoring for patients presenting with dyspnea with no clear diagnosis.

POS is a rare condition characterized by interatrial right-to-left shunting, through a PFO or ASD, when the patient is in an upright position. Aortic root deformity (2), vertebral fracture (3), lung surgery (4), and other conditions have been reported as underlying causes of this condition. Eustachian ridge/valve and atrial septal aneurysms are additional risk factors for POS (5).

In the present case, a PFO and a deformed aortic root were identified to be the underlying causes of the POS. The deformed ascending aorta was considered to stretch the atrial septum when the patient assumed an upright position, thereby redirecting the blood flow toward the PFO (6). In support of this mechanical pathway of dyspnea in POS, a previous case report described a transient elevation in the right atrial pressure in the upright position (1). Three-dimensional imaging of this case clearly demonstrated a severely compressed right ventricular inflow tract, which presumably contributed to the patient’s condition.

Although the mechanism of POS is gradually being elucidated, the diagnosis of POS remains difficult, due to the low detectability of the anatomical defects on TTE, which is recommended as the initial step in the investigation of intracardiac shunting disease. According to our case and a review of previously reported clinical cases, we propose the following as reasons contributing to the difficulty in diagnosing POS: first, deformation of the aortic root may not only cause the condition, but also hide the condition itself. On TTE, the severely enlarged aortic root, which is located in front of the atrial septum, may prevent direct visualization of the atrial septum (Fig. 2). Second, POS might be attenuated when a patient is placed in the left lateral decubitus position, which is the standard position for a TTE assessment. Although nearly all previous case reports of POS have described postural-induced changes in the SpO2, or partial pressure of oxygen in the arterial blood (1, 2, 4), the effects of lateral positioning on blood oxygen saturation have not been fully discussed. In our case, the patient showed hypoxia not only in the sitting position, but also in the decubitus position when she twisted her body to the right (Table). In contrast, her hypoxia decreased when she assumed the left lateral decubitus position. Although not confirmed, stretching of the atrial septum and right atrial pressure elevation might be

### Table. The Patient’s SpO2 in Various Situations.

<table>
<thead>
<tr>
<th>Position</th>
<th>FIO2</th>
<th>SpO2</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sitting for a few minutes</td>
<td>Room air</td>
<td>89%</td>
</tr>
<tr>
<td>Supine</td>
<td>Room air</td>
<td>95%</td>
</tr>
<tr>
<td>Right lateral decubitus</td>
<td>Room air</td>
<td>92%</td>
</tr>
<tr>
<td>Left lateral decubitus</td>
<td>Room air</td>
<td>95%</td>
</tr>
<tr>
<td>Prone</td>
<td>Room air</td>
<td>98%</td>
</tr>
<tr>
<td>Sitting (before a 6-min walk)</td>
<td>Room air</td>
<td>84%</td>
</tr>
<tr>
<td>Sitting (after a 6-min walk)</td>
<td>Room air</td>
<td>82%</td>
</tr>
<tr>
<td>Sitting (Post 15 min oxygenation)</td>
<td>98%</td>
<td>75%</td>
</tr>
</tbody>
</table>

FIO2: fraction of inspiratory oxygen

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Innerv...
Figure 1. A coronal section of the original image of chest-enhanced computed tomography (a) and three-dimensional reconstruction of the outer view of the heart and right ventricular inflow tract of the patient (b). The patient’s right ventricular inflow tract was severely compressed by the aortic root (yellow circle) compared with two control elderly females (c, d) who were diagnosed as normal on transthoracic echocardiography.

Figure 2. Transthoracic echocardiography findings. Visualization of the atrial septum was difficult due to the severely enlarged and elongated aortic root in both the short axis and apical four chamber views (a, b). After agitated saline infusion, bubbles appeared on the left side of the heart (yellow circle) 1 and 2 beats after they appeared in the right ventricle (yellow arrows) (c, d), indicating the presence of intracardiac shunting. RV: right ventricle, AV: aortic valve, LV: left ventricle, LA: left atrium, Ao: ascending aorta.

augmented by the displaced aortic root in the right lateral decubitus position, which in turn causes compression of the right atrium. According to these proposed mechanisms, the left lateral position for a TTE examination would thus be unsuitable for detecting POS. We recommend contrast TEE with simultaneous SpO2 monitoring as the assessment of
Figure 3. Contrast transesophageal echocardiography obtained in (a) the left lateral decubitus and (b) left lateral sitting positions. In the left atrium, the amount of injected saline significantly increased in the left lateral sitting position. The patent foramen ovale slit, which widened to 5.5 mm (arrow) in the left lateral sitting position, is shown in (c). LA: left atrium, RA: right atrium.

Figure 4. Simultaneous SpO2 monitoring during transesophageal echocardiography under 4 L/min of oxygen inhalation. The arrows show the times when Fig. 3a and 3b were recorded.

choice to identify the underlying cause of POS, as used for the present patient. This approach offers an effective solution to the problems associated with TTE because there is no obstacle between the TEE probe and the atrial septum. Furthermore, clear images of augmented right-to-left shunting can be obtained using TEE with patients in the right lateral sitting position.

In conclusion, although POS is rare, the possibility of a missed diagnosis due to difficulties with TTE imaging cannot be dismissed. The improvement of the diagnostic techniques for POS is clinically relevant, as the risk of aortic root elongation, as well as other risk factors for POS, increases with aging. Patients presenting with hypoxia exacerbation in an upright position should be investigated for potential POS using lung perfusion scintigraphy and contrast TEE, regardless of the results upon TTE. With the recent advances in intravascular therapy for PFO/ASD occlusions (7), the accurate diagnosis of POS will become more important as treatment becomes readily available.

The authors state that they have no Conflict of Interest (COI).

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References


