Internal Medicine
The Japanese Society of Internal Medicine
doi: 10.2169/internalmedicine.9904-17
Intern Med 57: 971-973, 2018
http://internmed.jp

【 CASE REPORT 】

# Platypnea-orthodeoxia Syndrome Induced by Multiple Vertebral Compression Fractures and an Atrial Septal Defect

Ko Harada, Koji Nakagawa, Hiroaki Ohtsuka, Yoichi Takaya, Teiji Akagi, Kazufumi Nakamura, Hiroshi Morita and Hiroshi Ito

**Abstract:**
Platypnea-orthodeoxia syndrome (POS) is a rare condition that is characterized by dyspnea and arterial oxygen desaturation, which worsen on standing and which are relieved by recumbency. We treated an 80-year-old woman with an atrial septal defect (ASD) who demonstrated POS following thoracic and lumbar vertebral compression fractures. The surgical closure of the ASD relieved her symptoms. The etiology might have been multiple compression fractures causing kyphosis and aortic distortion producing right atrial compression and increased right-to-left flow through the ASD. POS should be considered in the differential diagnosis of patients who develop dyspnea after vertebral compression fractures. The careful assessment of the patient's history and clinical condition helps in the diagnosis of POS.

**Key words:** platypnea-orthodeoxia syndrome, atrial septal defect, vertebral fracture

**(Intern Med 57: 971-973, 2018)**
**(DOI: 10.2169/internalmedicine.9904-17)**

## Introduction

Platypnea-orthodeoxia syndrome (POS) is a rare syndrome characterized by dyspnea and hypoxia when a patient is sitting or standing (1). The most common anatomical cause of POS is a right-to-left (RL) shunt through interatrial communications, including an atrial septal defect (ASD) or patent foramen ovale (PFO) (2). We herein report the case of an elderly ASD patient who developed POS following thoracic and lumbar vertebral compression fractures and who was successfully treated with the surgical closure of the ASD.

## Case Report

An 80-year-old Japanese woman presented to our hospital with dyspnea in the sitting and standing positions. She had a history of paroxysmal atrial fibrillation, for which she had been using an oral anticoagulant. Two months prior to her admission to our hospital she had presented to another hospital with multiple vertebral compression fractures. After being discharged from the hospital, she developed dyspnea in the sitting and standing positions. She also reported an episode of syncope while she had been standing at home. The development of POS was highly suspected and she was referred to our hospital for further evaluation and treatment.

A physical examination revealed that her blood pressure was 152/91 mmHg, and her pulse rate was regular at 95 beats/minute. Although the patient's oxygen saturation on room air was 98% in the supine position, it declined to 83% in the sitting position and dropped further to 65% in the standing position. An anteroposterior X-ray of her chest in the supine position revealed no lung abnormalities, and her cardiothoracic ratio was 59% (Fig. 1A). Electrocardiography showed a normal sinus rhythm with a normal atrioventricular conduction. Computed tomography (CT) of the spine showed thoracic 12 and lumbar 1 vertebral fractures (Fig. 1B). Contrast-enhanced CT revealed the elongation of the aorta, which was compressing the right atrium and distorting the position of her heart (Fig. 2). A transthoracic echocardiogram (TTE) revealed a normal ventricular func-

Department of Cardiovascular Medicine, Okayama University Graduate School of Medicine, Dentistry and Pharmaceutical Sciences, Japan
Received: July 30, 2017; Accepted: August 7, 2017; Advance Publication by J-STAGE: December 21, 2017
Correspondence to Dr. Ko Harada, me422084@s.okayama-u.ac.jp

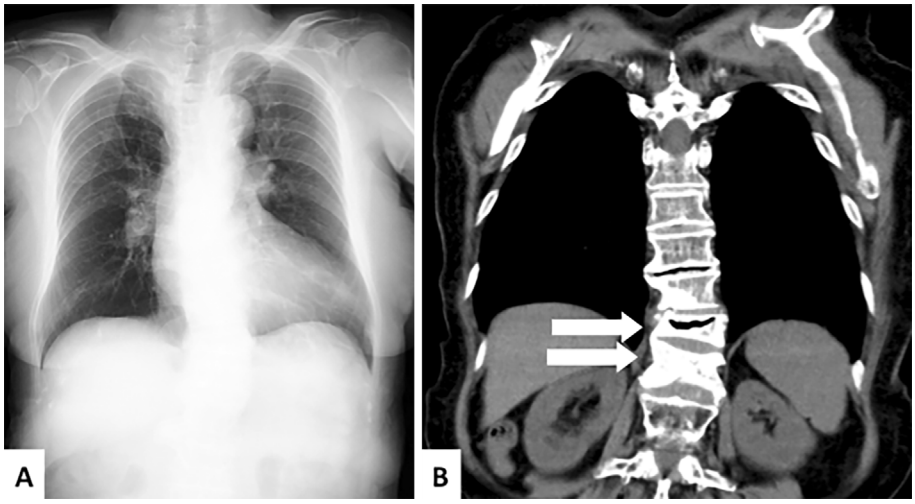


**Figure 1.** **An anteroposterior chest X-ray in the supine position revealed no lung abnormalities and showed a cardiothoracic ratio of 59% (A). Computed tomography (CT) of the spine showed thoracic 12 and lumbar 1 vertebral fractures (B arrows).**

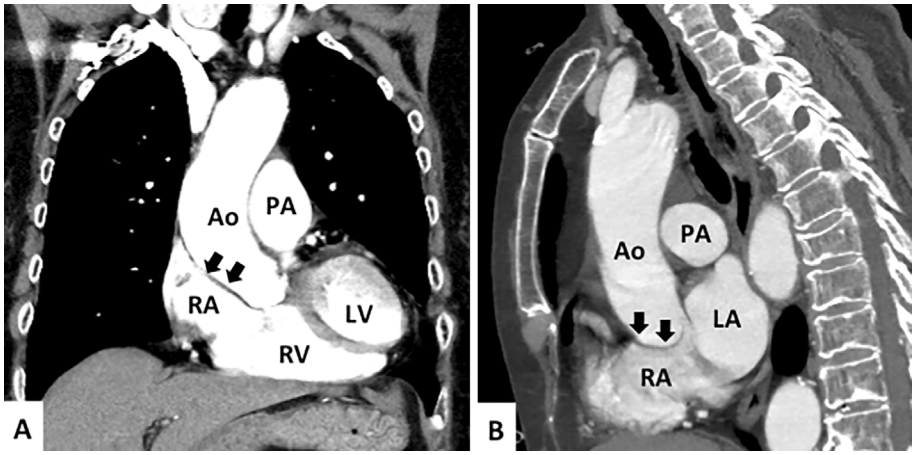


**Figure 2.** **Contrast-enhanced chest CT revealed aortic elongation compressing the right atrium and distorting the heart position, which was evident on the coronal (A: arrows), and sagittal (B: arrows) views. Ao: ascending aorta, RA: right atrium, RV: right ventricle, LV: left ventricle, PA: pulmonary artery, LA: left atrium**

tion without pulmonary hypertension, and a secundum ASD was detected. A transesophageal echocardiogram (TEE) showed an oval-shaped ASD measuring 14 mm × 7 mm in size with a deficient aortic rim (Fig. 3). We did not observe a eustachian valve or Chiari network. A TTE bubble study was performed in both the left lateral decubitus and sitting positions using intravenously administered agitated saline contrast solution. The amount of micro-bubbles appearing on the left side of the heart was increased by changing her position from the left lateral decubitus to the sitting position, indicating the presence of an RL shunt through the ASD, which was exacerbated while sitting.

The percutaneous transcatheter closure of the ASD was attempted on the 3rd day after admission; however, because the ascending aorta was highly deviated toward the right atrium, this procedure was unsuccessful. The surgical closure of the ASD was performed on the 10th day after admission. Following surgery, her interatrial RL shunt was found to have completely resolved, and her oxygen saturation in the sitting and standing positions recovered to 98% on room air. She was discharged on the 23rd day after the surgery without any complaints of platypnea. The patient was followed up at the outpatient clinic, and no further complications were observed.

## Discussion

We herein describe a case of POS, which developed in an elderly woman following multiple vertebral compression fractures. POS is a rare but clinically important syndrome characterized by dyspnea and arterial oxygen desaturation, which worsen on standing and which are relieved by recumbency (3). POS is caused by the coexistence of anatomical abnormalities, including PFO and ASD, and functional com-

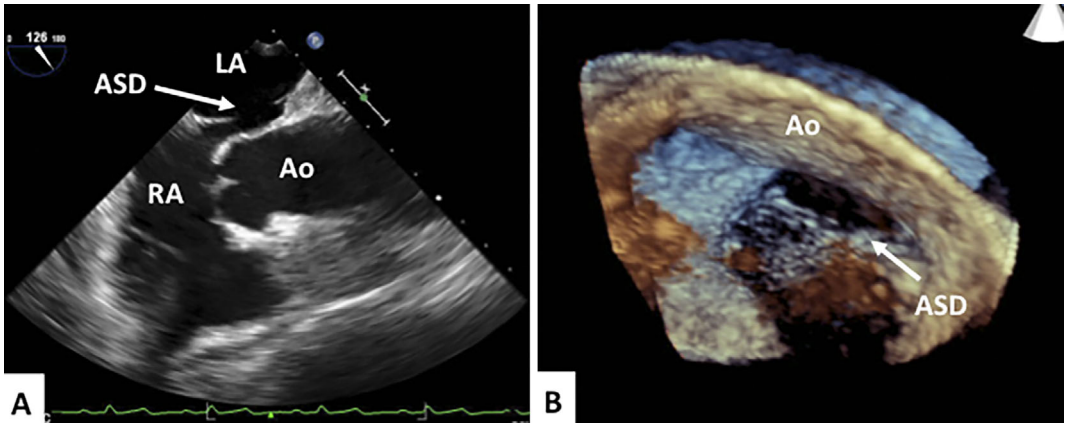


**Figure 3.** **Two-dimensional (A) and three-dimensional (B) transesophageal echocardiography (TEE). An oval-shaped ASD measuring 14 mm ×7 mm in size with a deficient aortic rim (arrows) was observed. LA: left atrium, ASD: atrial septal defect, RA: right atrium, Ao: ascending aorta**

ponents, which accelerate RL shunting, such as pulmonary emphysema, pericardial disease, or aortic aneurysms (3). Aortic elongation another possible aggravating factor that exaggerates RL shunting; this occurs because venous flow can be directed from the inferior *vena cava* into the left atrium through the anomalous interatrial communication when the ascending aorta compresses the right atrium and distorts the atrial septum (4, 5).

In our case, based on the patient's clinical course, her multiple vertebral compression fractures were suspected to have triggered the exacerbation of the RL shunting. To the best of our knowledge, only three cases of POS induced by vertebral fractures have been reported in the English literature (4, 6, 7). A possible etiological mechanism to explain the induction of POS by vertebral fractures is as follows: the multiple vertebral compression fractures caused kyphosis and the subsequent distortion of the aorta resulting in right atrial compression and increased RL flow through the ASD.

Rodrigues et al. reported that among 188 cases of POS, 33 cases showed anatomical changes of the aorta, including aortic dilatation, aneurysm, or distortion (1). However, they did not mention vertebral fractures as an associated anatomical change that could exacerbate RL shunting in POS cases. Based on the fact that PFO is present in approximately 15-35% of the normal population (8), and that the incidence of vertebral compression fractures has been increasing in our aging society, POS should be considered among the differential diagnoses when a patient develops dyspnea following vertebral compression fracture.

POS is recognized as a relatively uncommon condition and might be overlooked when establishing a diagnosis in patients presenting with dyspnea following vertebral compression fracture. The careful assessment of the patient's history and clinical condition are essential for the accurate diagnosis of this rare disorder.

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

## References

1. Rodrigues P, Palma P, Sousa-Pereira L. Platypnea-orthodeoxia syndrome in review: defining a new disease? Cardiology **123**: 15-23, 2012.
2. Cheng TO. Platypnea-orthodeoxia syndrome: etiology, differential diagnosis, and management. Catheter Cardiovasc Interv **47**: 64-66, 1999.
3. Knapper JT, Schultz J, Das G, Sperling LS. Cardiac platypnea-orthodeoxia syndrome: an often unrecognized malady. Clin Cardiol **37**: 645-649, 2014.
4. Takaya Y, Akagi T, Kijima Y, et al. Transcatheter closure of right-to-left atrial shunt in patients with platypnea-orthodeoxia syndrome associated with aortic elongation. Cardiovasc Interv Ther **29**: 221-225, 2014.
5. Shiraishi Y, Hakuno D, Isoda K, Miyazaki K, Adachi T. Platypnea-orthodeoxia syndrome due to PFO and aortic dilation. JACC Cardiovasc Imaging **5**: 570-571, 2012.
6. Fukuoka R, Kawasaki T, Koga H, et al. Platypnea-orthodeoxia syndrome associated with a thoracic vertebral fracture following a car accident. Intern Med **53**: 35-38, 2014.
7. Diaconu C, Abdou V, Safar B, Monsel F, Amara W. Platypnea-orthodeoxia syndrome: a case of hypoxemia associated with a patent foramen ovale. Ann Cardiol Angeiol (Paris) **64**: 406-409, 2015 (in French, Abstract in English).
8. Sun YP, Homma S. Patent foramen ovale and stroke. Circ J **80**: 1665-1673, 2016.