

## Paradoxical Orthodeoxia in a Patient with a Large Thoracic Aortic Aneurysm

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### Abstract

**Introduction:** Orthodeoxia is a rare clinical syndrome characterised by dyspnoea and arterial deoxygenation that accompanies a change from a supine to erect position. **Clinical Picture:** We describe an unusual case of “paradoxical orthodeoxia” in a 70-year-old man with a thoracic aortic aneurysm: arterial desaturation when supine that improved when erect. **Treatment and Outcome:** Non-invasive imaging revealed compression of the left pulmonary artery by the aneurysm (thoracic computed tomography) and patent foramen ovale (transesophageal echocardiography). Nuclear studies show decreased relative left lung perfusion attributable to the former, and right-to-left atrial shunt attributable to the latter. The degree of right-to-left shunt increases in the supine position: nuclear pulmonary shunt study shows shunt extent of 21% when supine versus 10% erect. **Conclusion:** A physioanatomical explanation is proposed.

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**Key words:** Desaturation, Patent foramen ovale, Recumbent posture

### Introduction

We report an unusual case of “paradoxical orthodeoxia” in a man with a large asymptomatic thoracic aneurysm of the ascending aorta, arch and proximal descending aorta. He was found to have arterial deoxygenation when supine, that improved in the erect position.

### Case Report

A large asymptomatic thoracic aortic aneurysm was found in a 70-year-old male chronic hypertensive and ex-smoker on routine chest roentgenogram pre-cataract surgery (Fig. 1). Aortic dimensions on computed tomography (CT) were: ascending aorta 4.5 cm, arch 4.5 cm and descending thoracic aorta 5.5 cm. Two years later, surveillance CT revealed stable ascending aortic dimensions but progressive expansion of the proximal descending thoracic aorta (diameter 6.5 cm). The latter compressed the left pulmonary branch artery against the left main bronchus; reducing relative left lung perfusion, as evident on nuclear lung perfusion imaging (Fig. 1).

The patient was noted to have an unusual pattern of arterial deoxygenation: worse in the supine compared to the erect position (Table 1). As the relative left lung

perfusion deficit on nuclear imaging was almost similar in both positions, pulmonary ventilation-perfusion mismatch alone cannot explain these postural changes in blood oxygenation. Transesophageal echocardiography (TEE) was performed in the recumbent position. This unveiled right atrial compression by the thoracic aneurysm, resulting in atrial septal stretch with widening of the patent foramen ovale (PFO), and persistent right-to-left shunt (Fig. 2). The pulmonary artery systolic pressure was not evaluable as tricuspid regurgitation was absent. There was however no morphological feature to indicate right ventricular pressure overload. Nuclear pulmonary shunt study in both the erect and supine positions demonstrated less pulmonary shunting with the patient erect, compared to when he was supine. This finding is consistent with the observed postural changes in patient’s blood oxygenation (Table 1).

Despite these abnormal findings, patient remained asymptomatic. He declined prophylactic surgical intervention, and was thus managed conservatively.

### Discussion

Platypnea-orthodeoxia is a rare clinical syndrome characterised by dyspnoea and arterial deoxygenation that

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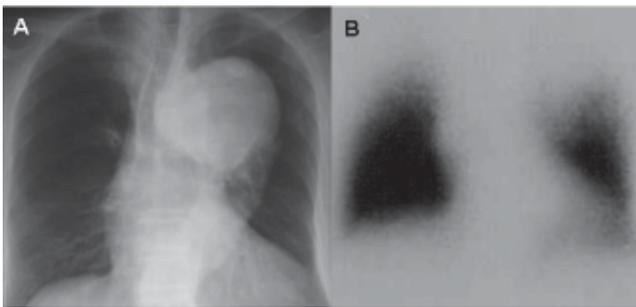


Fig. 1. The chest roentgenogram shows a large thoracic aortic aneurysm (panel A). There was left pulmonary arterial flow embarrassment, evidenced by reduced left upper lung perfusion on lung perfusion imaging with technetium-99m-labelled macroaggregated albumin (panel B).

Table 1. Effect of Posture on Blood Oxygenation, Lung Perfusion and Pulmonary Shunt

	Supine	Erect
Partial pressure of oxygen (mm Hg)	45.0	68.8
Oxygen saturation (%)	84.1	95.0
Relative left lung perfusion* (%)	33.9	35.6
Right-to-left shunt extent† (%)	20.8	9.8

\* Technetium-99m-labelled macroaggregated albumin lung perfusion scan. Left lung perfusion is expressed as a percentage of total bilateral lung perfusion.

† Nuclear pulmonary shunt study. Shunt extent more than 5% denotes significant shunting.

accompanies a change from the supine to the erect position. Two conditions must coexist to produce this phenomenon: interatrial communication, and a functional component that deforms the atrial septum, resulting in redirection of shunt flow with the assumption of an erect posture. The latter comprises diverse entities: pericardial effusion, arteriovenous malformation, liver cirrhosis, aortic aneurysm and aortic elongation.<sup>1,2</sup> In the absence of overt pulmonary hypertension, right-to-left interatrial shunting associated with the platypnea-orthodeoxia syndrome occurs due to positional modification of the atrial septum.<sup>3</sup> Atrial septal distortion attributable to elongation of the aneurysmal aorta has been reported by various authors.<sup>4-6</sup> Faller et al<sup>5</sup> reviewed 6 cases of aortic aneurysms, where the elongated aortic aneurysms caused stretching of the interatrial communication in the erect position. They postulated that an enlarged ascending aorta rotates the heart counterclockwise, resulting in atrial septal shift and the redirection of venous return through the interatrial communication.

Our patient had a large thoracic aneurysm. There were relative left lung perfusion deficit (from extrinsic left pulmonary artery compression) and right-to-left interatrial shunt (via distortion of the atrial septum, resulting in redirection of venous return directly across the stretched

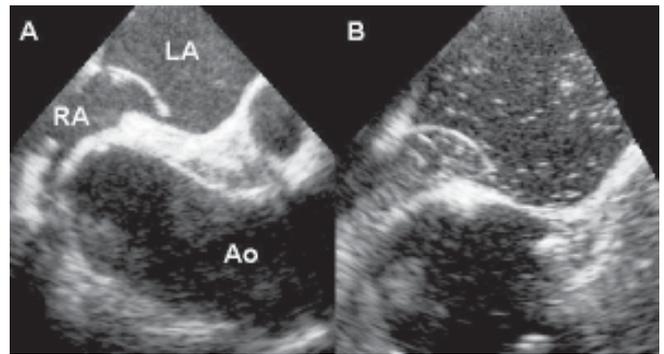


Fig. 2. The transesophageal echocardiogram shows compression of the right atrium (RA) septum by the enlarged aortic root (Ao), resulting in a distorted atrial septum and stretched patent foramen ovale (panel A). Passage of bubbles from the right to the left atrium (LA) was seen after rapid bolus injection of agitated saline contrast (panel B). During the procedure, the stream of bubbles can be seen to preferentially flow through the PFO upon entry of the RA.

PFO). The former appeared to be posture-independent: there were similar reductions in relative left lung perfusion in the supine and erect positions on nuclear lung perfusion imaging. In contrast, there was more severe right-to-left shunting in the supine versus erect position. Consequently, blood oxygenation became worse when patient was supine compared to when he was erect (Table 1). To our knowledge, this unusual pattern of “paradoxical orthodeoxia” has not been previously reported.

In our patient, initial investigation had been directed at the lung, having excluded pulmonary arterio-venous malformations and emboli on serial thoracic CT scans, and postural abnormality on spirometry. Unconvinced that the left pulmonary artery compression was the sole explanation, we searched for an intracardiac shunt. Trans-thoracic echocardiography had failed to show it, but strong suspicion following review of the literature led to the elucidation of the shunt through a stretched PFO on transoesophageal echocardiography.

The pulmonary artery systolic pressure was not evaluable as tricuspid regurgitation was absent, and there was no morphological feature to indicate right ventricular pressure overload. LV size and contractility was normal on both echocardiography done by separate consultants. Mild concentric LVH with some abnormal diastolic LV function was present but we feel would not entirely account for the desaturation. Gated SPECT showed normal LV size and function (LVEF 66%). Coronary angiogram showed only minor disease with LVEF 60% (LVEDP 21 mm Hg). Ideally tilt table cardiac catheterisation study, or evidence of improvement upon surgical repair of the aortic aneurysm would conclusively proof our suspicion. However, we were limited by the patient’s wish which was not to have further invasive procedures. Nevertheless, we feel that

there is enough evidence to propose a physioanatomical hypothesis to explain this phenomenon.

### **Conclusion**

This case emphasises the importance of considering intracardiac shunts in patients with oxygen desaturation. In the presence of a thoracic aortic aneurysm, depending on its anatomy and relation to the atrial septum, we believe that maximal distortion of the atrial septum (and right-to-left shunting) may occur preferentially in positions other than the erect, thereby producing variant patterns of deoxygenation different from that described for platypnea-orthodeoxia.

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