



Platypnea-Orthodeoxia syndrome (POS): An Overlooked Cause of Hypoxemia

A 65 y male with history of partial right pneumonectomy for non-small cell carcinoma (stage I) of lung 2 months ago presented with progressive dyspnea since 1 month. His vital signs were normal except for his room air oxygen saturation of 85% in the OPD. Routine blood investigations were normal. Chest X-ray was unremarkable and ECG and cardiac 2D echo revealed no abnormality. HRCT of chest and pulmonary CT angiogram revealed no significant abnormality. He was hospitalised for further evaluation. A junior resident noticed that his oxygen saturation consistently improved to 95 to 96 % every time he assumed supine position. Patient also insisted on sleeping with a back rest at night. This raised a strong suspicion of Platypnea-orthodeoxia syndrome (POS). Agitated saline contrast echo was done to look for right to left shunting through a PFO. It revealed immediate (within first two beats) appearance of contrast in left atrium and left ventricle (Video 1). Presence of PFO was then confirmed on TEE (Fig 1 & 2). Agitated contrast injection during TEE confirmed the findings seen on trans-thoracic echo (Video 2). The PFO was closed with a 24 mm Amplatzer device (Video 3) with remarkable immediate improvement in symptoms with room air oxygen saturation in sitting position in 98-99% range. This case was done in 2017 and he was doing very well until his last follow up in 2024.

The removal of the lung (particularly right) causes mediastinal shift and diaphragmatic elevation, which anatomically distort the heart, forcing blood from the inferior vena cava directly into the PFO in upright or standing position(1).

Discussion

POS is a rare condition characterized by:
Platypnea – dyspnea that worsens in the

upright (sitting or standing) position and improves on lying down.

Orthodeoxia – a fall in arterial oxygen saturation ($\geq 5\%$) or PaO_2 (≥ 4 mmHg) on assuming the upright posture, with improvement in the supine position.

Pathophysiology

POS requires two components:

1. Anatomical substrate for right-to-left shunt or V/Q mismatch
 - Intracardiac shunt: Patent foramen ovale (PFO), atrial septal defect (ASD)
 - Intrapulmonary shunt: Pulmonary AV malformations, hepatopulmonary syndrome
 - Severe ventilation-perfusion mismatch
2. Functional/postural factor
 - Upright posture alters cardiac or pulmonary geometry causing increased shunting:
 - Redirection of inferior vena cava flow toward interatrial septum
 - Aortic root dilation or elongation
 - Kyphoscoliosis, post-pneumonectomy state

Importantly, POS occurs without pulmonary hypertension.

Clinical Features

- Dyspnea and cyanosis in sitting or standing position
- Relief on lying supine
- Oxygen saturation drops significantly on standing
- Often no signs of heart failure or lung congestion

Etiology

Intracardiac causes

- PFO (most common)
- ASD
- Atrial septal aneurysm

Intrapulmonary causes

- Hepatopulmonary syndrome
- Pulmonary arteriovenous malformations

Other causes

- Post-lung surgery
- Severe kyphoscoliosis
- Aortic root dilation

Diagnosis

- Pulse oximetry: Supine vs upright saturation difference
- Contrast (bubble) echocardiography: Immediate appearance of bubbles in the left atrium suggests intracardiac shunt
- TEE often required to define anatomy
- CT chest or pulmonary angiography if intrapulmonary shunt suspected
- ABG confirms orthodeoxia

Management

- Treat the underlying cause:
- Percutaneous closure of PFO/ASD

(most effective). Use of intra-cardiac echo (ICE) has obviated the need for intra-procedural endotracheal

Intubation for TEE

- Treatment of liver disease in hepatopulmonary syndrome
- Embolization of pulmonary AV malformations
- Supplemental oxygen as supportive therapy

Prognosis

- Excellent when intracardiac shunt is closed
- Dramatic and immediate improvement in symptoms and oxygenation is typical

Reference 1.

Shah AH et al, Percutaneous Intervention to Treat Platypnea-Orthodeoxia Syndrome: The Toronto Experience. JACC Volume 9, issue 18, 26 September 2016, Pages 1928-1938

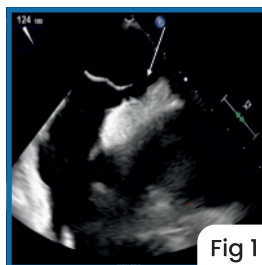


Fig 1

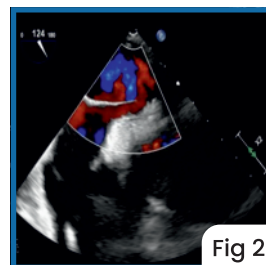


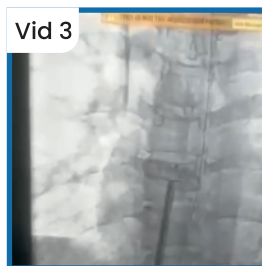
Fig 2



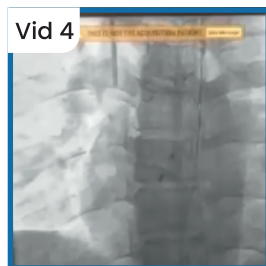
Vid 1



Vid 2



Vid 3



Vid 4



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CARDIOLOGISTS

Dr. Ajit Bhagwat | Dr. Sachin Mukhedkar | Dr. Ranjit Palkar | Dr. Mahendrasingh Parihar

Gut No. 43, Bajaj Marg, Satara Parisar, Beed Bypass Road, Chhatrapati Sambhajinagar. Tel.: +91 240-6632111