Platypnea-Orthodeoxia Syndrome (POS) Is A Rare but Dramatic Cause Of Respiratory failure

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Case

73 year old female who was sent from Primary provider office for:

Evaluation of hypoxia on room air.

Burning chest sensation.
PMH

- SCD s/p AICD
- MR (mild-mod)
- PFO (with moderate-sized shunt)
- HTN.
- CLL (currently on chemotherapy session 3).
- GERD
PE

Vitals: Temp 97.9     HR 65     BP 151/72
SpO2 81-86% on room air, 98% on BIPAP 50% FiO2
Height 5'4"     Weight 139     BMI:
CV: 2/6 Holosystolic murmur at the apex, split S2.
EKG
UP RIGHT POSITION

SUPINE POSITION
## Labs

### Blood Gas

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<thead>
<tr>
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<th>Value</th>
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<td>PCO2 arterial</td>
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<tr>
<td>PO2</td>
<td>54</td>
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<tr>
<td>HCO3</td>
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<tr>
<td>Base Excess</td>
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<tr>
<td>O2 sat arterial</td>
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### Chemistry

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<td>CO2</td>
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<td>AG</td>
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<td>Glu</td>
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<table>
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<td>ALT</td>
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<tr>
<td>AST</td>
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<tr>
<td>BUN</td>
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Echo

TTE: EF 58%.

TEE:

- Large right to left shunt at atrial level, associated with PFO.
- Aneurysmal interatrial septum and a prominent Eustachian valve directing blood towards the PFO
- The degree of shunt is compatible with the hypoxia.
Hospital course:

- Successful percutaneous closure of interatrial septal defect with large shunt using a Gore device size 30 mm under fluoroscopy guidance.

- She had immediate and complete resolution of her symptoms.
Platypnea-Orthodeoxia

- A rare disorder characterized by both dyspnea (platypnea) and arterial desaturation (orthodeoxia) in the upright position with improvement in the supine position.
Pathophysiology

Two conditions must coexist to cause POS:

• *Anatomical component*: interatrial communication.

• *Functional component*: that produces a deformity in the atrial septum and results in a redirection of shunt flow with the assumption of an upright posture.
CAUSES

What Causes Water to Flow Uphill?
CAUSES

• Atrial septal defects.
• Pericardial effusion.
• Constrictive pericarditis.
• Emphysema.
• Amiodarone pulmonary toxicity.
• Pneumonectomy.
• Cirrhosis.
• Aortic elongation.
Atrial Septal Defect (ASD)

- 13% of congenital heart disease.
- 2 per 1000 live births.
- 30-40% of CHD in adults.
- 65—70% : II ASD.
- 50% of Primmum ASD and 40—50% SV ASD are seen in female.
Anatomy

- **PFO:** 25-30% adult
- **Secundum:** 70% of ASD.
- **Primum ASD:** A cleft in the AML and conduction abnormality.
- **SV ASD:** 4-11% of all ASD; 80-85% at SVC junction.

Redrawn from original supplied courtesy of Dr. William D. Edwards, Department of Laboratory Medicine and Pathology, Mayo Clinic, Rochester, MN.
**TEE**

- **Apical four-chamber view**
  - Ostium primum ASD in this view.
  - May lead to false positives.

- **Subcostal view**
  - Often more reliable - can visualize entire atrial septum.
Color Doppler

- Confirm the presence of the ASD.
- Estimate the defect size.
- Evaluate the efficacy of surgery.
Cardiac MR

- Shunt flow.
- Defect size.
- Pulmonary venous return.
Cardiac Catheterization

- Calculate the pulmonary–to–systemic blood flow ratio (Qp:Qs).
- Recommended in ASD and PAH to aid in determining whether ASD closure is indicated.
Treatment

- Symptoms.
- Right-sided cardiac chamber enlargement.
- Age.
- Defect size and location.
- Associated abnormalities such as right heart enlargement, moderate (or less) hypertension, tricuspid regurgitation.
ACC/AHA Guideline

ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease: Executive Summary

A Report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines for the Management of Adults With Congenital Heart Disease)

Developed in Collaboration With the American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons
2. Closure of an ASD, either percutaneously or surgically, is reasonable in the presence of:
   a. Paradoxical embolism. *(Level of Evidence: C)*
   b. Documented orthodeoxia-platypnea. *(Level of Evidence: B)*
Percutaneous Closure

- An alternative to surgical closure for secundum ASDs with appropriate anatomic characteristics.
- Defect < 30mm diameter.
Amplatzer Occlusion Device

- Approved for percutaneous ASD closure in 2001 by F.D.A.

- Consists of two round disks made of Nitinol (nickel + titanium) wire mesh linked together by a short connecting waist.
Case Report

Platypnea-orthodeoxia syndrome in a patient with a pre-existing patent foramen ovale successfully treated with an atrial septal occluder

Ting-Ting ZHANG, Ge-Sheng CHENG, Jun WANG, Xing-Ye WANG, Xue-Gang XIE, Ya-Juan DU, Yu-Shun ZHANG

The Second Department of Cardiology, First Hospital Affiliated to Xi’an Jiaotong University, Xi’an, Shaanxi Province, China
REFERENCE


https://mksap17.acponline.org/app/groups/cv/topics/mk17_a_cv_s9/sections/mk17_a_cv_s9_3_1.

AHA/ACC 2008 guideline.
THANKS