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Review article

The multiple dimensions of Platypnea-Orthodeoxia syndrome: A review

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ABSTRACT

Platypnea-Orthodeoxia syndrome (POS) is a rare clinical entity characterized by dyspnea and arterial desaturation while in the upright position. The various pathophysiologic mechanisms leading to POS has puzzled clinicians for years. The hypoxia in POS has been attributed to the mixing of the deoxygenated venous blood with the oxygenated arterial blood via a shunt. The primary mechanisms of POS in these patients can be broadly classified based on intracardiac abnormalities, extracardiac abnormalities and miscellaneous etiologies. A Patent Foramen Ovale (PFO) was the most common reported site of an intracardiac shunt. In addition to PFO, intracardiac shunt leading to POS has been reported from either an Atrial Septal Defect (ASD) or an Atrial Septal Aneurysm (ASA). Most patients with an intracardiac shunt also demonstrated a secondary anatomic or a functional defect. Extracardiac causes of POS included intra-pulmonary arteriovenous malformations and lung parenchymal diseases. A systematic evaluation is necessary to identify the underlying cause and institute an appropriate intervention. We conducted a review of literature and reviewed 239 cases of POS. In this article, we review the etiology and patho-physiology of POS and also summarize the diagnostic algorithms and treatment modalities available for early diagnosis and prompt treatment of patients presenting with symptoms of platypnea and/or orthodeoxia.

Contents

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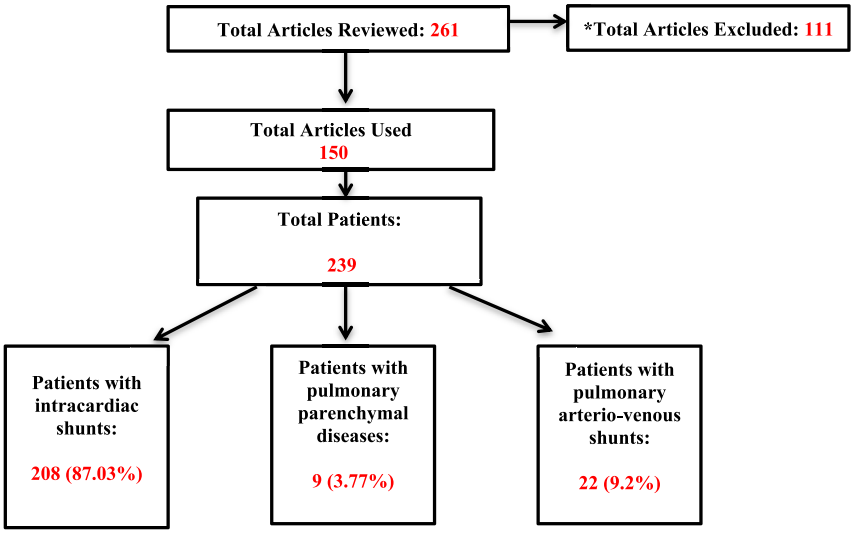
1. Introduction

Platypnea-Orthodeoxia syndrome (POS) is a rare clinical entity that is characterized by positional dyspnea (platypnea) and arterial desaturation (orthodeoxia) while in the upright position. This drop in saturation is defined as a drop in Pao2>4 mmHg or Sao2 >5% from supine to an upright position. In POS, both entities improve with change back to normal from an upright to a recumbent position. The first case of POS was reported by *Burchell at al* in a patient in 1949 with post traumatic intrathoracic arteriovenous shunt [1]. The terms 'platypnea' and 'orthodeoxia' were coined by Altman et al. and Robin et al 1969 and 1978 respectively [2,3]. In these patients POS was a result of hepatic and pulmonary disease respectively. In 1984, POS was first described in a patient with an intracardiac right-to-left shunt with orthostatic accentuation of hypoxemia in the absence of hepato-pulmonary dysfunction or elevated right heart pressures [4]. The pathophysiology of POS has puzzled clinicians for years and in some patients the precise mechanisms remain elusive. This review attempts to describe the mechanisms underlying orthodeoxia and platypnea and formulate a diagnostic and treatment algorithm for POS.

2. Methodology

We conducted a review of literature that included all the articles on PubMed with the keywords 'platypnea' and/or 'orthodeoxia' from January 1949 up to November 2016. The articles were independently examined by two reviewers AA and AP. A total of 261 relevant articles were screened. 111 articles not related to true platypnea or orthodeoxia were excluded (Table 1a). Patients with POS due to complex congenital heart diseases were excluded. Therefore, a total of 150 articles that included 239 patients were analyzed. In addition, another case was reported from our institute [5]. The primary etiologic mechanisms in patients with cardiac POS are reviewed in Table 1b.

Table 1a
Epidemiology of POS.



3. Results

The primary mechanisms of POS in these patients can be broadly classified as intracardiac abnormalities, extracardiac abnormalities and miscellaneous etiologies.

Intracardiac communication between the two atria was the most common cause of POS in 208 of 239 (87%) patients. A Patent Foramen Ovale (PFO) was the most common reported site of an intracardiac shunt. In addition to PFO, intracardiac shunt leading to POS has been reported from either an Atrial Septal Defect (ASD) or an Atrial Septal Aneurysm (ASA).

Apart from an isolated intracardiac defect, most patients with

Table 1b
Primary & secondary anatomic & functional defects in patients with cardiac POS.

Total Patients with Cardiac POS: 208
Primary Anatomic Defect:
Patent Foramen Ovale (PFO) – 139
Atrial Septal Defect (ASD) – 25
Unspecified – 44
Secondary Anatomic or Functional Factors
Aortic dilatation/aneurysm/distortion – 53
Pneumectomy – 29
Diaphragm paralysis - 17
Prominent Eustachian valve - 17
Kyphoscoliosis & Thoracic vertebral fracture – 13
Lipomatous interatrial septum – 4
Abdominal surgery - 6
Pericardial effusion - 3
Chiari network - 2
Right atrial mass – 1
Ventricular dilatation/TAVR – 2
Cardiac Transplant – 2
Pulmonary fibrosis – 1
Single lung transplantation - 1