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Platypnea orthodeoxia syndrome: focus on predisposing anatomical factors

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Platypnea orthodeoxia syndrome (POS) is a rare situation with hypoxia and breathlessness in the upright position recovering in the recumbent position. A mechanical inter-atrial septum distortion, causing redirection of flow from the right to the left atrium through a patent foramen ovale (PFO), despite normal pulmonary pressure, is suggested to explain POS. Prevalence of predisposing anatomical factors remain little known

Methods: All patients who underwent a PFO closure for a POS were retrospectively included from 2 CHU. Predisposing anatomical factors were investigated.

Results: 67 patients (Median age 72 y.o., interquartile range 61-80; 58.2% men) were included. All patients had dyspnea (76.2% NYHA III or IV, 53.7% under oxygen-therapy). The remaining patients had a refractory hypoxemia (38.2%) without POS. Most frequent predisposing anatomical factor was an enlarged or unwound aorta (n=29, 43.3% 95CI 31.2-56.0) with an aortic aneurysm in 25 patients (37.3%, 95CI 25.8-50.0). Other factors identified were pneumonectomy (n=8, 11.9% CI95 5.3-22.2), a history of cardiac surgery (n=7, 10.5%, 95CI 4.3-20.3), mechanical ventilation (n=6, 9.0% 95CI 3.4-18.5), kyphoscoliosis (n=4, 6.0% 95CI 1.7-14.6), hepatomegaly (n=4, 6.0% 95CI 1.7-14.6, 2 patients with hepato-renal polycystic disease, one hemochromatosis and one cirrhosis), right ventricle failure (n=2,3.0% 95CI 0.4-10.4), pericardial effusions (n=2,3.0% 95CI 0.4-10.4), right ventricle arrhythmogenic dysplasia (n=2,3.0% 95CI 0.4-10.4), diaphragmatic paralysis (n=1, 1.5% 95CI 0.1-8.0), carcinoid syndrome with tricuspid regurgitation (n=1, 1.5% 95CI 0.1-8.0), a right atrium pace-maker lead (n=1, 1.5% 95CI 0.1-8.0) and a tako-tsubo syndrome (n=1, 1.5% 95CI 0.1-8.0).

Conclusion: Aortic aneurysm and pneumonectomy are the most frequent situation leading to a POS. Other causes were observed such as hepato-renal polycystic kidney, or atrial pacemaker probe that may be underdiagnosed in clinical practice.