J Clin Case Rep 2017, 7:9 (Suppl)
DOI: 10.4172/2165-7920-C1-015

Platypnea orthodeoxia syndrome in a patient with pulmonary arterial hypertension

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Platypnea orthodeoxia syndrome (POS) is a rare syndrome, traditionally defined as dyspnea or hypoxemia. It is usually observed when changing from a recumbent to an upright or seated position. A patent foramen ovale (PFO) is often part of the underlying pathophysiology. In the present case, a 70-year-old woman with known PFO presented with new symptoms of a POS. A pulmonary arterial hypertension (PAH) Nizza Group 1 associated with a connective tissue disease was also known and a treatment with ambrisentan had been initiated five years ago. In the absence of an established method to diagnose the platypnea orthodeoxia syndrome we performed continuous measurements of heart rate and oxygen saturation in lying, sitting and standing positions using a standard pulse oximetry. We observed a slow but continuous decrease of the oxygen saturation accompanied by an increase of the heart rate and development of symptoms (dyspnea).In order to clarify the etiology of the new hemodynamic state, we performed a transoesophageal echocardiography(TOE) initially in lying and then in standing position. Despite a documented decrease of the oxygen saturation (from 89% to 81%), the blood flow through the pulmonary veins remained unchanged and the PFO showed no evidence of intracardiac shunting or flow obstruction. A further investigation showed a chronic obstructive pulmonary disease, the treatment of which eradicated the symptoms and eliminated the drop of oxygen saturation in the standing position.This is the first described case of POS in a patient with PAH. While a simple pulse oximetry was successful in identifying the symptoms, a dynamic transoesophageal echocardiography might be needed. Although, PFO is a common cause of POS and a PFO closure is often advised, careful differential diagnosis should be kept in mind.

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