486 A blue man standing

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An 85-year-old was admitted in Internal Medic Ward for recurrent episodes of acute respiratory failure. His electrocardiogram showed a new onset of atrial flutter with spontaneous reversal to sinus rhythm (narrow QRS complex and normal atrioventricular conduction). The dyspnoea was worse with the patient sitting and was better when lying supine. Also, he report back pain associated to his major kyphoscoliosis. Results of chest radiography and blood test were normal. The arterial blood gas test revealed a hypocapnic hypoxemic respiratory failure, therefore the patient oxygen supplementation with noninvasive ventilation (NIV) was started, but peripheral oxygen saturation was persistently below 88% despite the high flow oxygenation. The patient underwent nasopharyngeal (NP) swab (ruling out SARS-CoV-2), high-resolution computed tomography (which was not pathologic), and computed tomography angiography (excluding pulmonary embolism). A transthoracic echocardiography was performed showing concentric hypertrophy, left atriomegaly and severe aortic ectasia of the root and of his ascending part, normal pulmonary pression and an aneurism of the interatrial septum with the presence of right-to-left shunt after using agitate saline contrast with Valsalva maneuver. Transesophageal echocardiography (TTE) confirmed the presence of severe structural interatrial septal abnormality with wide left convex aneurysm and an atrial septum defect (ASD) as ostium secundum, causing severe bidirectional shunt. Therefore, the patient underwent a successful transcatheter closure of secundum ASD with device Amplatzer Septal Occluder 018, while monitored with cardiac catheterization and transesophageal echocardiography. After few days, the patient was discharged at home without oxygen therapy. At the 3-month follow-up visit he reported no respiratory symptoms. Platypnea-orthodeoxia syndrome (POS) is an uncommon disorder characterized by dyspnoea and hypoxemia that occurs when the patient is sitting or standing and disappear quickly when

recumbent. POS is characterized by both an anatomic and a functional component. The anatomic element is typically an interatrial communication such as ASD or Patent Foramen Ovale, which cause blood shunt left-to-right due to a higher pressure in left atrium and a greater compliance of the right ventricle, in the presence of normal pulmonary artery pressure. The syndrome occurs when a functional element, such as an increased atrial pressure or a decreased compliance of the right ventricle, reverses the flow. The shunts are often small and could remain asymptomatic: the high left atrial pressure let the defect close until there is a reverse pression which stretched atrial septum in particular in the upright position. In fact, when the patient is standing, the inferior vena cava comes in line with the defect increasing the right-to-left flow, sparking the respiratory symptoms. This could be linked to a cardiac or an extracardiac condition; in this case the presence of kyphoscoliosis and severe aortic ectasia of the root and of his ascending part, played an important role. The diagnosis is mainly made by echocardiography and cardiac catheterization to verify the mismatch in oxygen saturation between the pulmonary vein and the aorta. The diagnosis of POS is challenging, often considered only after other possible diagnosis is excluded. When POS is triggered by an interatrial defect in the absence of severe pulmonary hypertension, the usual treatment is a percutaneous or surgical closure.