A reversible cause of refractory hypoxaemia

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A 73 year old man, with a medical history of coronary artery disease, called the ambulance, because of retrosternal chest pain. When the paramedics arrived, his initial vital signs were normal and a pulse oximetry (SpO₂) was 94% on room air as he layed supine in bed. The pain persisted after sublingual nitroglycerin administration, and he was transported to hospital. While he was lying in a semi-recumbent position on the ambulance stretcher, his SpO₂ decreased to 70% and, despite receiving 100% oxygen through a face mask with an oxygen reservoir and oxygen flow of 12 l/min, it only rose to 80%. On arrival at the emergency department, the patient complained of shortness of breath, but his chest pain had subsided. His cardiopulmonary examination was normal. An arterial blood gas analysis confirmed hypoxaemia. There was no sign of alveolar or interstitial processes on his initial chest radiograph. Hypoxaemia, with a clear chest radiograph, points to several possible diagnosis. Pneumothorax and chronic obstructive pulmonary disease were unlikely after physical examination and chest radiography. The probability of a large pulmonary embolism was low as a ventilation/perfusion scan was normal. Finally, the arterial blood gas measurement excluded a hypoventilation syndrome. A clue to the correct diagnosis was the association of the change of position and the decrease in SpO₂. This was confirmed by repeating arterial blood gas measurements in the recumbent and upright positions (see table 1) and led us to diagnose orthodeoxia. Orthodeoxia is defined as an arterial desaturation in the upright compared with a lying position. The platypnea-orthodeoxia syndrome, though uncommon, has been described in disorders frequently encountered in the emergency department: for example, shunts either intracardiac (usually an atrial septal defect), or extracardiac in pulmonary diseases (severe chronic obstructive pulmonary disease), or in liver cirrhosis. Severe refractory hypoxaemia is a critical finding that requires immediate investigation in the emergency department. If the initial evaluation points to a right to left shunting, the platypnea-orthodeoxia syndrome must be part of the differential diagnosis. As in our case, the platypnea (worsening dyspnea when sitting up) can be minimal, and not spontaneously reported by the patient. However, this diagnosis can be suspected if the physician pays attention to the circumstances of the SpO₂ decrease, and when confirmed it can be easily reversed by a simple bedside manoeuvre in the emergency department. Surprisingly, many popular internal or emergency medicine handbooks do not mention this syndrome, and it may be overlooked, delaying therefore the correct diagnosis. Contrast echocardiography is the examination of choice for evaluating a right to left shunt, allowing also for discrimination between intracardiac or extracardiac shunting.

In our patient, contrast echocardiography showed a significant right to left shunt (white arrow) through an atrial septal defect (black arrow), which was considerably increased when the patient passed from a supine (fig 1A arrowhead) to an upright position (fig 1B arrowhead). A right and left heart
catheterisation showed a large atrial septal defect (secundum type without pulmonary hypertension) and severe coronary artery disease. The patient underwent surgical closure of the atrial septal defect and a triple coronary artery bypass. He had an uneventful recovery and the disappearance of the orthodeoxia was confirmed by pulse oxymetry (see table 1).

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