Mechanisms of Platypnea-Orthodeoxia: What Causes Water to Flow Uphill?

To the Editor:

Platypnea-orthodeoxia is a relatively uncommon but striking clinical syndrome characterized by dyspnea and deoxygenation accompanying a change to a sitting or standing from a recumbent position. Since Burchell et al described this rare syndrome over half a century ago, no more than 50 cases have been reported in the literature.

Two conditions must coexist to cause platypnea-orthodeoxia: an anatomical component in the form of an interatrial communication and a functional component that produces a deformity in the atrial septum and results in a redirection of shunt flow with the assumption of an upright posture. The former may be an atrial septal defect, a patent foramen ovale, or a fenestrated atrial septal aneurysm. The latter may be cardiac, such as pericardial effusion or constrictive pericarditis; pulmonary, such as emphysema, arteriovenous malformation, pneumonectomy, or amiodarone toxicity; abdominal, such as cirrhosis of the liver or ileus; or vascular, such as aortic aneurysm or elongation.

Under normal conditions an interatrial communication allows blood to shunt from left to right due to a higher pressure in left atrium than right atrium and a greater compliance of the right ventricle than the left ventricle. Right-to-left interatrial shunting is usually associated with spontaneous or induced pulmonary hypertension and, therefore, in the absence of a right-to-left pressure gradient, what is the mechanism for a right-to-left shunt? Or put in another way, what causes water to flow uphill?

A persistent Eustachian valve can cause interatrial right-to-left shunting with a normal right atrial pressure. Platypnea-orthodeoxia could be explained on the basis of positional modification of abnormal shunting. Standing upright could stretch the interatrial communication, be it a patent foramen ovale, an atrial septal defect, or a fenestrated atrial septal aneurysm, thus allowing more streaming of venous blood from inferior vena cava through the defect, whether or not a persistent Eustachian valve coexists. This redirection of flow caused by an anatomic distortion of the right atrium or the atrial septum also might occur from a loculated pericardial effusion, an aortic aneurysm, or aortic elongation.

The elegant echocardiographic demonstration by Medina et al is an example of the last mentioned situation. The readers are referred to my recent editorial for a more detailed discussion of the etiology, differential diagnosis, and management of platypnea-orthodeoxia. Suffice it to say that the definitive treatment for platypnea-orthodeoxia is closure of the interatrial communication, which can be carried out nowadays by transcatheter techniques, as was done in the case of Medina et al.

Response

We appreciate very much Dr Cheng’s comments regarding the definition, pathophysiology, differential diagnosis, and management of platypnea-orthodeoxia syndrome, and we completely agree with them.

From our point of view, Dr Cheng’s articles help explain the underlying mechanisms of this condition and offer current recommendations for its treatment.