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[1] described this rare syndrome over half a century ago, no more than 50 cases have been reported in the literature.[2]

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.[2]

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?[2] A persistent Eustachian valve can cause interatrial right-to-left shunting with a normal right atrial pressure.[3] 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.[3] 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.[2]

The elegant echocardiographic demonstration by Medina et al[4] 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.[2] 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,[5] as was done in the case of Medina et al.[4]

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4. Medina A, Suarez de Lezo J, Caballero E, et al. Platypnea-orthodeoxia due to aortic elongation. *Circulation*. 2001;104:741.
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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.[1]

From our point of view, Dr Cheng's articles[2–4] help explain the underlying mechanisms of this condition and offer current recommendations for its treatment.

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1. Medina A, Suarez de Lezo J, Caballero E, et al. Platypnea-orthodeoxia due to aortic elongation. *Circulation*. 2001;104:741.
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