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Falls and Delirium: Platypnea-Orthodeoxia Syndrome

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PRESENTATION

An 85-year-old woman's inability to remain standing was ultimately connected to an undiagnosed congenital defect. The patient was admitted to the geriatric unit of a university hospital for new frequent falls and subacute delirium, both of which began 1 month before presentation. Her medical history included ischemic stroke 10 years earlier, mild neurocognitive impairment, and Von Recklinghausen neurofibromatosis with isolated skin involvement.

ASSESSMENT

On examination the patient's blood pressure was 104/70 mm Hg, heart rate was 100 beats per minute, oxygen saturation was 85%, and body temperature was 98.8°F (37.1°C). She weighed 99.2 lb (45 kg). Her breathing was normal while she was in the dorsal decubitus position. Findings from a lung examination and heart auscultations were within normal limits, but she had a distended jugular vein and peripheral cyanosis.

A neuropsychological examination confirmed that her confusion, marked by anxiety, agitation, daytime sleepiness, and increasing disorientation, was associated with acute psychomotor abnormalities, including backward disequilibrium, reactional hypertonia, alteration of postural reactions, and fear of falling. She had no focal neurologic deficits. It was impossible for her to remain standing. An electrocardiogram showed a sinus rhythm at 90 beats per minute and first-degree atrioventricular block with no other conduction or repolarization abnormalities. Chest radiography revealed no parenchymal or mediastinal disorders.

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Results of a complete blood count and a basic metabolic panel were within normal limits, as were thyroid, renal, and hepatic assays. No inflammatory syndrome was revealed. The initial arterial blood gas analysis showed a severe shunt effect; partial pressure of oxygen was 47.6 mm Hg, partial pressure of carbon dioxide was 25.5 mm Hg, pH was 7.48, and bicarbonate was 22 mmol/L. Because the patient's unexplained desaturation and confusion led us to suspect pulmonary embolism, emergency contrast-enhanced thoracic and cerebral computed tomography scans were performed.

Owing to our concern for thromboembolic disease, she was strictly maintained in the dorsal decubitus position until the imaging results were available. No abnormalities, apart from the sequelae of a left cerebellar lacunar stroke, were found on the computed tomography scan. Her delirium, cyanosis, and oxygen saturation, obtained via transcutaneous oximetry, improved while she was lying on her back (**Table**).

DIAGNOSIS

We believed the patient had platypnea-orthodeoxia syndrome, on the basis of her positional hypoxemia, which occurred when she was upright and resolved when she was recumbent. These findings contrasted sharply with what we expected to discover, because her distended jugular vein first suggested congestive heart failure.

Transthoracic echocardiography was performed in an effort to find a source for the patient's signs and symptoms. Her left and right ventricular ejection fractions were within normal limits, as were her pulmonary arterial pressure and cardiac output. Both atria were clear. A bubble study followed, and transthoracic echocardiography then disclosed an intracardiac right-to-left shunt caused by a patent foramen ovale and multiple defects in the interatrial septum, including an aneurysm (**Figure**).

Three years earlier the patient underwent transthoracic echocardiography to rule out emboligenic heart disease after

Table Arterial Blood Gas Parameters

Parameter	Before Intervention		After Intervention
	Sitting (T0)	Supine (T0 + 20 min)	(Sitting)
O_2 (L/min)	2	2	0
SaO_2 (%)	86.3	92.4	93.0
PaO_2 (mm Hg)	50.2	62.3	64.0
$PaCO_2$ (mm Hg)	33.3	32.5	28.1
HCO_3- (mmol/L)	23.2	23.2	21.6

HCO_3- = bicarbonate; O_2 = oxygen; $paCO_2$ = partial pressure of carbon dioxide; PaO_2 = partial pressure of oxygen; SaO_2 = oxygen saturation.

she developed a sudden decrease in visual acuity. When the images were retrieved and examined the interatrial septum aneurysm was visible but patent foramen ovale and other defects were not. Cerebral nuclear magnetic resonance spectroscopy, performed 8 months earlier during a workup for cognitive dysfunction, confirmed the presence of left cerebellar ischemic damage and highlighted bilateral vascular frontoparietal hyperintensities. A causal relationship between the intracardiac shunt and the cerebrovascular lesions was ruled out because of their old and nonspecific nature and the absence of venous thrombosis.[1]

Our final diagnosis was platypnea-orthodeoxia syndrome, leading to subacute delirium and acute psychomotor disadaptation syndrome, which is characterized by psychomotor difficulties like those experienced by our patient while she was in the sitting and standing positions.[2] These were consequences of acute hypoxemia, caused by an intracardiac shunt from a patent foramen ovale and interatrial septum defects in a frail, debilitated patient.[2] Cardiac abnormalities, especially interatrial septum defects, are frequently noted among patients with neurofibromatosis.[3,4] However, to our knowledge this association has never been reported in elderly persons, because it is unusual for a patient with neurofibromatosis to reach an advanced age before the abnormality is discovered.

This patient might have been predisposed to delirium by her pre-existing neurocognitive dysfunction, whereas hypoxemia has been described as a common precipitating factor for delirium; indeed, hypoxemia is one of the main components in the differential diagnosis when assessing delirious

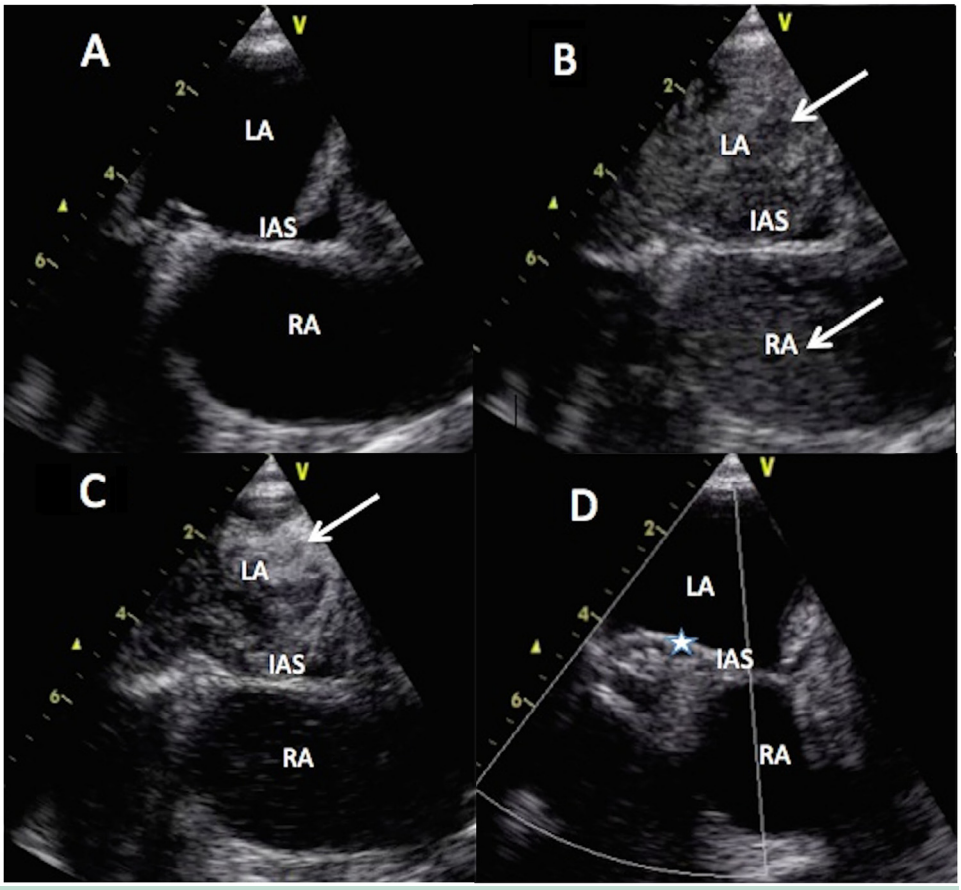

Figure A bubble study was performed with transesophageal echocardiography. **(A)** This image was obtained before the bubble study. **(B)** In the early phase of the bubble study, bubbles (arrow) moved from the right atrium (RA) to the left atrium (LA) through the interatrial septum (IAS). **(C)** In the late phase of the bubble study, bubbles (arrows) were located in the right atrium. **(D)** This image was captured after percutaneous closure of the interatrial septum (star).

patients.[5] It has been suggested that reactive oxygen species induce injury in the hypoxic brain, though the precise mechanisms involved in hypoxemia-induced delirium are not fully understood.[6]

MANAGEMENT

Our patient initially received nasal oxygen therapy, physiotherapy, and psychological and nutritional support. After a multidisciplinary discussion, which included the patient's daughter, the patient was scheduled for percutaneous atrial septal defect closure. Despite her general frailty, we believed that severe hypoxemia significantly impaired her functional prognosis and quality of life.[7] Moreover, the procedure has been reported to be safe and effective in the elderly.[8]

Percutaneous patent foramen ovale and interatrial defect closure was achieved with a 30-mm Cardioform Septal Occluder (W.L. Gore and Associates, Newark, Del) under transesophageal echocardiography and fluoroscopic guidance (**Figure**). The specific roles of the patent foramen ovale and interatrial septum defects in hypoxemia were not known, because the intervention corrected both at the same time. After surgery, hypoxemia ceased immediately. The patient's oxygen saturation in the sitting position increased from 86% to 93%, similar to the level recorded when the patient was in the supine position (**Table**). No residual shunt was seen on an echocardiogram.

Gradually the patient's delirium regressed. Multidisciplinary care for psychomotor disadaptation syndrome was provided in the hospital's rehabilitation center, where daily physiotherapy was continued and supplemented by psychological and nutritional support. She experienced progressive improvement in standing and gait and was discharged from the hospital with an antiplatelet regimen 1 month after the cardiac intervention.[1]

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