A Rare Cause of Dyspnea and Arterial Hypoxemia*

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Platypnea-orthodeoxia is a rare pattern of dyspnea with arterial hypoxemia. Platypnea is defined as dyspnea induced by upright posture, and it is relieved by the recumbent position. Orthodeoxia refers to arterial desaturation resulting from assuming an erect or upright position. The case reported involves a 59-year-old man with profound, unexplained dyspnea despite extensive investigation performed at the referring institution. The difficulty in diagnosis persisted until it was recognized that the investigations, in having been performed under “standard” (supine) conditions, were insufficient and therefore misleading. Despite normal supine intracardiac pressures, a patent foramen ovale was shown to give rise to a large orthostatic intracardiac shunt, demonstrated by means of an echocardiogram performed with the patient supine and upright. Surgical closure of the foramen was followed by dramatic clinical improvement. Among dyspeic patients, discernment of a pattern of platypnea and orthodeoxia is key to effective evaluation.

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Key words: anoxemia; atrial septal defects; contrast media; dyspnea; echocardiography; heart catheterization; intracardiac shunting; orthodeoxia; platypnea; posture

The observation of dyspnea induced by upright posture (platypnea), together with orthostatic cyanosis, was reported as early as 1956, in a patient following pneumonectomy.1 The terms “platypnea” and “orthodeoxia” appear to have been combined for the first time in a detailed 1976 report.2 The opportunity to evaluate a patient with this uncommonly recognized syndrome suggested that the caveats learned in the course of his evaluation were worth sharing.

CASE REPORT

A 59-year-old man with a 20 pack-year history of smoking presented with a 6-year history of progressive breathlessness. In the previous 3 years, it had become prominent while sitting or standing and was fully relieved by recumbency. At his regional medical center, arterial blood oxygen tension while breathing room air was 75 mm Hg while seated and 46 mm Hg while standing. While breathing 100% oxygen, PO2 was 354 mm Hg while supine and 200 mm Hg while standing. He had a normal chest x-ray film, ECG, and contrast two-dimensional echocardiogram. A radionuclide lung scan detected evidence of right-to-left shunting and low probability for pulmonary embolism. Pulmonary angiography revealed normal pulmonary vasculature; this finding prompted his physician to refer him for further assessment of the unexplained shunt.

On physical examination, values for blood pressure, pulse, and respiratory rate were within normal ranges. No cyanosis, clubbing, or telangiectasia were found. Heart sounds were of normal intensity with physiologic splitting and without audible murmur. The lung fields were clear. Results of the remainder of the examination were normal.

Oxygen saturation was 92 to 95% with the patient supine and sitting and 85% while standing. Pulmonary studies while the patient was in the sitting position demonstrated normal spirometric results and lung volumes but a reduced diffusing capacity at 49%. A high resolution CT scan of the chest was normal. Catheterization of both left and right sides of the heart was performed and disclosed a patent foramen ovale but could identify no shunting. Pressure readings in the right atrium were 8 and 7 mm Hg during atrial contraction and ventricular contraction, respectively; the mean pressure was 5 mm Hg. In the left atrium, the pressure readings were 10 and 11 mm Hg during atrial contraction and ventricular contraction, respectively; the mean pressure was 7 mm Hg. Pressures in the right ventricle were 22/3 and 7 mm Hg (systolic diastolic and end-diastole), and in the main pulmonary artery, they were 20/9 and 14 mm Hg (systolic diastolic and mean). Blood oxygen saturation was determined in the superior vena cava to be 56%; inferior vena cava, 66%; right atrium, 64%; right ventricle, 65%; main pulmonary artery, 65%; and in each of the major pulmonary veins and left atrium, 100%.

A contrast echocardiogram, performed with the patient in the supine and sitting positions revealed a large right-to-left intrapulmonary shunt solely with the patient in the sitting position. Surgical closure of the patent foramen ovale was performed. Intraoperatively, the patent foramen ovale measured 0.5X0.3 cm. The left lung showed bullae, which suggested emphysema disease, and a biopsy specimen revealed distal acinar irregular emphysema along with interstitial fibrosis and mild nonspecific bronchiolitis.

Two months after surgery, the patient was free of dyspnea, both while standing and while walking long distances. A subsequent pulmonary function test at his regional medical center at 4 months showed mild obstruction and a diffusing capacity of 61%. At 6 months, his PO2 while standing and breathing room air was 64 mm Hg and the saturation value was 92%.

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DISCUSSION

Basic causes for platypnea-orthodeoxia have consisted of intrapulmonary and intracardiac shunting.1-3 Most cases of right-to-left shunting through a patent foramen ovale are attributed to right atrial pressure exceeding that of the left, forcing open a potentially patent foramen ovale, which is typical of chronic cyanotic congenital heart disease.4 The fact that this patient’s supine atrial pressures were lower on the right led to an initial incorrect view that the patent foramen ovale was extraneous to the problem. Subsequent reflection and contrast echocardiography with the patient in the seated position provided the diagnosis, but given the normal supine pressures, one had to reconsider the possible pathophysiologic basis for the shunt.

Seward et al5 described seven patients whose case histories were collected over several years at the Mayo Clinic. In the one patient in whom cardiac catheterization results were reported for both the supine and sitting positions, there was no position dependence of the intraatrial chamber pressures, and therefore, any demonstrable pressure gradient to explain a right-to-left shunt was absent.

Potential explanations for shunting in the supine position include transient right-to-left pressure gradients, altered compliance, preferential flow from the inferior vena cava, and Valsalva maneuver or respiratory factors,5 but to date, the mechanisms for orthodeoxia have not been satisfactorily delineated. Shunting across an atrial septal defect has been shown to occur from the less compliant (ie, “stiffer”) to the more compliant chamber.6 In assuming the erect posture, the combined drop in right ventricular pressure and filling may keep right ventricular compliance constant, whereas compliance of the left ventricle will increase.7 During early diastole, while the relatively more compliant left ventricle more readily accepts left atrial filling, the right ventricle, being relatively less compliant, might offer greater resistance to flow from the right atrium. This might facilitate shunting across the foramen ovale, possibly without a significant right-to-left gradient in atrial pressures. Postural effects, or lung disease of which this patient had some evidence, might have altered the configuration of the atrial structures or the direction of blood flow from the superior vena cava. In an effort to assess these possibilities, transeophageal echocardiography with the patient seated was performed, but technically adequate imaging could not be achieved.

In summary, an infrequently recognized cause of dyspnea has been described. This case suggests that patients with unexplained dyspnea, when accompanied by arterial desaturation out of proportion to their cardiopulmonary disease, should have orthodeoxia easily and inexpensively excluded by measurement of arterial blood gases while they are in the supine and standing positions. Orthostatic desaturation should prompt further investigation. A definitive diagnosis of a posturally related right-to-left shunt can most easily be obtained by contrast echocardiography, which must be performed with the patient both supine and as erect as possible. If this test is negative for abnormalities, the possibility of true intrapulmonary shunts within the lung bases should be considered.5

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REFERENCES

Platypnea-Orthodeoxia Related to Aortic Elongation*

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An 80-year-old woman presented with progressive shortness of breath. There was no history of pulmonary or cardiac disease. Results of a physical examination were normal. She had significant oxygen desaturation while she was in an upright position. Admission to the hospital for workup followed, and evaluation included tilt-table transesophageal echocardiogram and cardiac catheterization. A massive right-to-left shunt through a patent foramen ovale was detected, and surgical intervention resulted in dramatic improvement of symptoms. In this patient, it seems that the syndrome of platypnea-orthodeoxia was related to aortic elongation, allowing significant right-to-left shunt.

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Most cases of significant right-to-left interatrial cardiac shunting are associated with elevation of right-sided cardiac pressures. Platypnea-orthodeoxia is an uncommon

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