Pulmonary Hypertension Prevalence in Sickle Cell Disease Varies With Diagnostic Method

In a study to determine the prevalence of pulmonary hypertension (PH) in patients with sickle cell disease, Florence Parent, MD, from the University of Paris-South, and colleagues found that 6% had confirmed PH on right-heart catheterization. In contrast, initial echocardiographic evaluation had a low positive predictive value for PH; the negative predictive power could not be determined. "The optimal approach for screening such patients for right heart catheterization remains uncertain," the investigators said in the July 7 New England Journal of Medicine.

A total of 398 adult outpatients with sickle cell disease (mean age, 34) recruited from three referral centers in France were included in the analysis. Participants were in stable condition and had no severe renal insufficiency, severe liver disease, or chronic restrictive lung disease. The researchers performed transthoracic echocardiography (continuous-wave Doppler sampling) to screen patients for PH. In patients with suspected PH (ie, a tricuspid regurgitant jet velocity of at least 2.5 m per second), the investigators also performed right heart catheterization. Based on these tests, patients were classified as follows:

- Group 1 - a tricuspid regurgitant jet velocity < 2.5 m per second or that could not be measured, without other echocardiographic signs of PH or a decrease in the diffusion capacity of carbon monoxide corrected for alveolar volume.
- Group 2 - a tricuspid regurgitant jet velocity of ≥ 2.5 m per second without confirmed PH on right-heart catheterization.
- Group 3 - a tricuspid regurgitant jet velocity of ≥ 2.5 m per second with confirmed PH on right-heart catheterization (ie, mean pulmonary arterial pressure ≥ 25 mm Hg). Group 3 patients were further classified as having either precapillary or postcapillary PH (ie, pulmonary-capillary wedge pressure ≤ 15 mm Hg or > 15 mm Hg, respectively).

Prospective follow-up evaluations were conducted for three years.

While the prevalence of tricuspid regurgitant jet velocity of at least 2.5 m per second was 27%, the prevalence of PH as confirmed on catheterization was 6%—making the positive predictive value of echocardiography 25%. Of the 24 Group 3 patients, 11 had precapillary PH. Group 3 patients were older and had poorer functional capacity and higher levels of N-terminal pro-brain natriuretic peptide than patients in the other groups. Patients in Groups 1 and 2 had similar characteristics.

Shortly after catheterization, three patients experienced an episode of vaso-occlusive crisis that necessitated a brief hospitalization but no permanent sequelae related to the event.

If data had been used from the 47 patients who were excluded from the analysis due to severe end-organ damage or clinical instability, the prevalence of PH might have been higher, the researchers said. "Given the high prevalence of sickle cell disease in many regions, this complication may affect a large worldwide population," they contended.

—Adriene Marshall

Suggested Reading

Analysis of Load Cell Data May Be a Useful Sleep Apnea Screening Method

MINNEAPOLIS—Through the use of load cells, physicians may have an unobtrusive way to diagnose and monitor sleep apnea, according to data presented at the 25th Anniversary Meeting of the Associated Professional Sleep Societies, LLC. A load cell is a transducer that converts force under the supports of a bed to a measurable electrical output.

"Although sleep apnea is typically diagnosed using polysomnography (PSG), PSG is performed over a single night in a laboratory and cannot efficiently monitor patients over extended periods of time," explained Zachary Beattie from the Department of Biomedical Engineering at Oregon Health and Science University, Portland, and the Oregon Center for Aging and Technology, and colleagues.

"We are investigating the use of load cells placed under the supports of a bed to unobtrusively monitor patient respiration and automatically screen for sleep apnea in the home."

Load cell data were collected simultaneously with overnight diagnostic PSG in Oregon Health and Science University patients. The PSG data were scored based on American Academy of Sleep Medicine standards; all episodes of disordered breathing (ie, central apneas, obstructive apneas, hypopneas) and 20 periods of breathing (20 seconds in duration) were identified from the PSG of each patient.

A computer algorithm also was developed to classify the corresponding load cell data segments as normal breathing or disordered breathing. Load cells placed under the supports of one bed identified slight changes in pressure at each support. The small displacement of mass caused by the patients' breathing was detected by the load cells as a quasi-sinusoidal signal.

Fifteen patients (mean age, 50; mean BMI, 36; mean apnea-hypopnea index, 46.5) were included in the analysis. When classifying data from all patients collectively, the researchers found that the sensitivity of the computer algorithm to detect disordered breathing segments was 0.75 and the specificity was 0.82.

However, when classifying each subject separately, the sensitivity of the computer algorithm was better able to discriminate normal from disordered breathing (sensitivity, 0.86; specificity, 0.79). Age, sex, BMI, weight, and apnea-hypopnea index severity did not affect the sensitivity or specificity of the classifications, Beattie and colleagues noted.

"Load cells are capable of differentiating disordered breathing events from normal respiration without direct patient contact," the investigators concluded.

KEY POINT

The prevalence of pulmonary hypertension was 6% in adults with sickle cell disease as confirmed by right-heart catheterization, according to the results of a recent study.

"Further work is aimed at detecting events within a continuous collection. This technology could ultimately provide low-cost, contact-free monitoring for respiratory events over many nights in the comfort of a patient's own bed.

—Frederique H. Theuvenin

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