Cardiac Catheterization A Must in PAH Diagnosis

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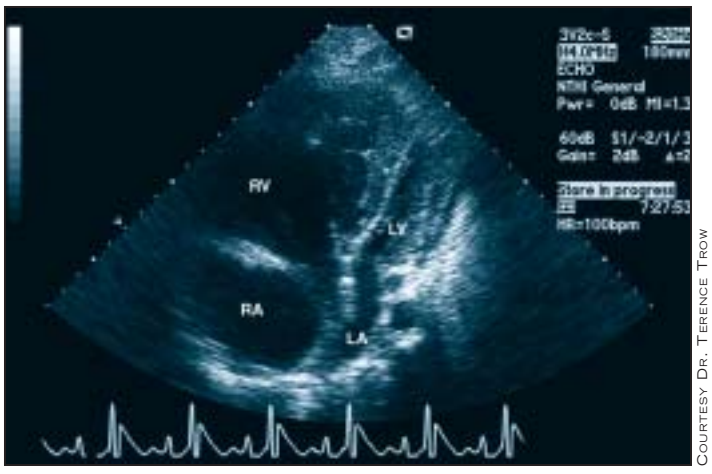
NEW YORK — Any patient with suspected pulmonary hypertension must have a thorough work-up, including right heart catheterization, before initiating treatment, Dr. Roxana Sulica said at a meeting sponsored by the Pulmonary Hypertension Association and the University of Michigan.

Because the typical presenting symptoms of pulmonary arterial hypertension (PAH) are subtle and nonspecific, with dyspnea, fatigue, and syncope or near syncope being the most common, a high index of suspicion is needed or the diagnosis may not be made until the disease is advanced and the prognosis is poor, she said.

Risk factors for PAH include underlying connective tissue disease, especially limited scleroderma and mixed connective tissue disease, a family history of PAH, the presence of congenital heart disease, and environmental factors such as exposure to anorexigens.

Clinical assessment of the patient with possible PAH includes an electrocardiogram, which may show changes in the right ventricle, including right axis deviation, right atrial enlargement, and right ventricular hypertrophy, said Dr. Sulica.



COURTESY DR. TERENCE TROW

An echocardiogram shows right atrial and right ventricular enlargement that is impinging on the left side.

A chest x-ray may reveal prominent proximal pulmonary arteries, peripheral hypovascularity, and reduced retrosternal air space.

An echocardiogram should then be done, and typical—but not diagnostic—findings on the echocardiogram include right atrial and ventricular enlargement, right ventricular dysfunction, and intraventricular septal flattening.

The definitive diagnosis of PAH can only be made by cardiac catheterization, which can exclude congenital heart disease, measure wedge pressure, and establish the degree of hemodynamic impairment, according to Dr. Sulica, who is director, Beth Israel Pulmonary Hypertension Program, Beth Israel Medical Center, New York.

The hemodynamic definition of PAH is a mean pulmonary artery pressure greater than 25 mm Hg, with a pulmonary capillary wedge pressure less than 15 mm Hg and a calculated pulmonary vascular resistance greater than 3 Wood units.

Right heart catheterization also permits a determination of the pulmonary vasodilator reserve through vasodilator testing using inhaled nitric oxide, intravenous epoprostenol, or intravenous adenosine. A positive response is defined as a reduction in mean pulmonary artery pressure of 10 mm Hg or more to a mean of 40 mm Hg or less, with an unchanged or increased cardiac output. Only positive responders should be given treatment with calcium channel blockers, Dr. Sulica cautioned.

Newly released guidelines from the American College of Chest Physicians emphasize the limited role of calcium channel blockers, which have been studied for PAH for 2 decades.

The guidelines point to a study of 557 patients with idiopathic PAH who underwent acute pulmonary vasodilator testing, with the 70 positive responders receiving long-term oral calcium channel blocker monotherapy. By 1 year, only 38 (6.8% of the total group) showed a favorable clinical response (Circulation 2005;111:3105-11).

Another recent study found that inappropriate—and potentially harmful—calcium channel blocker use remains common. In a registry that enrolled 1,360 PAH patients, 31% were on calcium channel blockers at the time of referral to a tertiary care center (Eur. Respir. J. 2007 Sept. 5 [Epub doi:10.1183/09031936.00042107]).

In patients who are not responders to vasodilation, the use of calcium channel blockers can decrease cardiac output and systemic vascular resistance, without improving pulmonary artery pressure or pulmonary vascular resistance. Routine vasodilator testing before treatment initiation could eliminate this inappropriate use of calcium channel blockers, Dr. Sulica said.

The prognosis for PAH is still fairly poor. Two-year survival among patients with scleroderma complicated by PAH is only 40%, compared with 80% among those without pulmonary complications (Respiratory Care 2006;51:368-81).

The prognosis also is grim for patients in advanced functional classes and those with poor exercise endurance, as well as for those whose hemodynamic findings include elevated right atrial pressure and reduced cardiac index. But improved understanding of pathophysiology and recent advances in medical therapy may change this.

"What we have learned so far is that screening patients with scleroderma can lead to an earlier diagnosis of PAH. Soon we may see if early treatment can improve the long-term prognosis," Dr. Sulica said. ■

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