Commentary

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Dr Soto's case presentation and discussion elegantly highlight an increasingly common challenge that we are faced with in practice, the diagnosis and treatment of left heart etiologies of pulmonary hypertension. As the population ages, and as the prevalence of other comorbid diseases such as diabetes, systemic hypertension, obesity, and obstructive sleep apnea increase, we are going to be faced with this dilemma with greater frequency. While well summarized in Dr Soto's case and discussion, I would like to highlight a few pearls at different points in the evaluation of such patients.

History

The clinician's index of suspicion for left heart disease as the etiology of the pulmonary hypertension should be increased in patients who have a past medical history of systemic hypertension, particularly if it has not been optimally controlled; diabetes; obesity; coronary artery disease; and obstructive sleep apnea; or symptoms of paroxysmal nocturnal dyspnea or orthopnea. Although paroxysmal nocturnal dyspnea and orthopnea can occur in very late stages of PAH, that diagnosis is generally clear cut at this stage.

Noninvasive Testing

Atrial fibrillation is uncommon in PAH and its presence should sway one toward a left heart etiology of pulmonary hypertension. If electrocardiography does not show right axis deviation, left heart disease also moves up on the differential.

There are many echocardiographic features that should raise the index of suspicion of left heart disease as the etiology of the pulmonary hypertension. These include absence of right heart chamber enlargement; left atrial enlargement; left ventricular hypertrophy; impaired diastolic relaxation by Doppler indices; and modest elevation of pulmonary arterial pressures (ie, 50's rather than 90's).

If the index of suspicion is high after the initial evaluation and noninvasive testing, it might be appropriate to optimize medical therapies and repeat an echocardiogram prior to moving on to invasive testing. In some situations, with optimal blood pressure and fluid management, symptoms and evidence of pulmonary hypertension on echocardiography will improve and this might obviate the need to proceed to cardiac catheterization.

Cardiac Catheterization

The most important points here were emphasized in Dr Soto's case and discussion, but I will briefly reiterate.

It is critical to obtain an accurate measurement of left heart filling pressure. In many instances, it is difficult to obtain a perfect wedge pressure tracing in these patients. Over or underinflating the balloon even just a bit can dramatically alter the wedge pressure tracing. Like Dr Soto, we generally obtain a direct measurement of left ventricular end diastolic pressure in patients in whom we suspect pulmonary venous hypertension. In the absence of mitral stenosis (which should be apparent on echocardiography) the left ventricular end diastolic pressure is an accurate reflection of left heart filling pressure, and of the pulmonary venous pressure.

Often the left heart filling pressure is substantially elevated in such patients, and the transpulmonary gradient is normal or just mildly elevated, and the diagnosis is clear. Nothing further may be required in these cases to confirm the diagnosis. However, it is not uncommon to see an upper normal or just mildly elevated left heart filling pressure, particularly in patients who has been well diuresed. In these instances, either a fluid challenge or exercise will help clinch the diagnosis. Dr Soto used exercise in his case. By increasing heart rate one reduces the diastolic filling time, which in a patient with impaired diastolic function can substantially increase the filling pressures. A volume load in the catheterization laboratory challenges the impaired left ventricle in a similar fashion. An important cautionary note to remember here is that although exercise echocardiography may demonstrate an increase in pulmonary arterial pressures, it is often difficult to delineate how much of this increase is a result of elevated left heart filling pressures given the increased heart rate and reduced diastolic filling time.

Dr Soto also nicely demonstrated the reduction in pulmonary artery pressures that occurred as the result of the reduction in systemic arterial pressures in this case with nitroprusside. This is also my drug of choice to delineate the physiology in a patient with severe systemic hypertension in the catheterization laboratory. Typical agents used to test for vasoreactivity, epoprostenol, adenosine, and nitric oxide, may increase pulmonary blood flow and result in increased filling pressures in those with a left ventricle that is unable to accommodate that increased venous return.

Treatment

As in Dr Soto's case, treatment should be directed toward the underlying pathology, which at times is difficult. Optimal blood pressure control, sodium restriction, and volume management are key in most patients. Treatment of atrial fibrillation may be of benefit (if possible) as restoring atrial kick is crucial in those with diastolic dysfunction. If this is not possible, rate control should be emphasized, to allow adequate time for diastolic filling. Weight loss should also be encouraged in the obese patient, and compliance with treatment of other comorbid diseases, such as sleep apnea, is crucial. Dr Soto's patient had a very nice response to this approach, but sadly, this is not always the case. Often patients are still symptomatic despite this approach, or are not compliant with these therapies. There is very little evidence on how to approach treatment of pulmonary hypertension in such a patient. As Dr Soto highlights, there is no evidence that any PAH-specific therapy is helpful in such a patient, and there is the theoretic risk of increasing pulmonary blood flow and inducing pulmonary edema. It would be interesting to see a controlled clinical trial in this population. ■