Helping Patients Understand the Complex Pulmonary Hypertension Workup

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When patients are being evaluated for pulmonary hypertension (PH), they are often asked to undergo numerous diagnostic tests in order to reach a definitive diagnosis. An important role of the PH nurse coordinator is educating the patients regarding the purpose and importance of the various procedures.

The classification of PH has gone through a series of changes since the first classification in 1973.1 The current guidelines recommend certain diagnostic testing be performed on patients suspected of having PH in order to provide diagnostic clarity. The American College of Cardiology Foundation (ACCF)/American Heart Association (AHA) 2009 Expert Consensus Document attests to the complexity of diagnosing and caring for patients with PH (Figure).² One important recommendation is that pulmonary arterial hypertension (PAH) patients be evaluated and treated in conjunction with a PH center of excellence.² The following are some of the recommendations for the PH workup.

History and Physical Examination: Patients need to understand the value of an accurate history and thorough physical examination. Inviting a family member who is familiar with the patient's course of illness can be extremely helpful, particularly if the patient has difficulty providing an accurate history.

Fatigue and dyspnea are the symptoms patients most often report.³ Thoughtful questioning of the patient and family may reveal the presence of chest pain, shortness of breath, palpitations, presyncope, or syncope. Patients are often asked if

they have taken diet pills or used methamphetamine and should be counseled regarding the importance of divulging this relevant information.

The physical examination is one of the most important aspects of the initial PH evaluation. The patient can expect a focused cardiac examination that will provide information about the functioning of the right side of the heart. The patient should know that this will be a part of every subsequent examination.

Electrocardiogram (ECG): Patients may ask why an ECG is being performed. This is a quick, noninvasive way to assess electromechanical changes occurring in the right heart. Their first ECG will serve as a baseline for comparison with future ECGs.

Transthoracic Echocardiogram (TTE): The patient needs to know that the diagnosis of PH cannot be made on the basis of an echocardiogram alone.⁴ This noninvasive test can, however, provide important information about the size and function of the right ventricle, as well as evaluate for congenital heart disease or left sided heart failure. Using a heart model can be an excellent visual way of illustrating changes that can occur to the right side of the heart.

The TTE can also allow for specific measurements, such as tricuspid annular plane systolic excursion (TAPSE), which can help predict survival in these patients.⁵ Patients should know that a TTE will be repeated at certain intervals based on clinical presentation, as well as the individual PH center's routine for repeating TTEs.

Pulmonary Function Tests (PFTs): The results of PFTs will provide important information on ventilatory function as well as gas exchange. They can also be helpful in

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the evaluation of unexplained shortness of breath. The results may reveal obstructive and/or restrictive disease, which will allow placement of the patient in the proper PH classification and also guide treatment.⁶

Overnight Oximetry: This relatively simple test can be done in the patient's home. A local oxygen supply company can provide the necessary (and minimal) equipment. Patients are asked to wear a pulse oximeter while sleeping for one night. Results can identify the degree and frequency oxygen levels drop during sleep. Based on results, the patient may need to wear nocturnal oxygen or have a formal sleep study to evaluate for obstructive sleep apnea.

Laboratory Tests: The patient will require very specific laboratory work when seen at the dedicated PH center. Included in a wide variety of labs will be brain natriuretic peptide (BNP) or NT-pro-BNP. Elevations of either can alert clinicians to the presence of right ventricular strain and possible right heart failure.⁷ The patient should know that following trends could provide valuable information regarding effectiveness of PHspecific therapies. Additionally, laboratory tests may be ordered to evaluate for the presence of connective tissue disease, as well as HIV and hepatitis B and C.

6-Minute Walk Test (6MWT): The patient should know this is a simple test requiring a walk on a flat, hard surface for 6 minutes.⁸ Speed, distance, and oxygenation levels will be measured. Results provide a good sense of functionality and oxygen requirements with exertion. Subsequent walk test results can help in the assessment of disease progression as well as response to therapy.

Right Heart Catheterization (RHC): The patient should know that a RHC will be required to establish the diagnosis of PAH.⁹ They should be educated on how this inva-



Figure: Diagnostic approach to PAH.² General guidelines for the evaluation of pulmonary hypertension. Since the suspicion of PH may arise in various ways, the sequence of tests may vary. However, the diagnosis of PAH requires that certain data support a specific diagnosis. In addition, the diagnosis of idiopathic pulmonary arterial hypertension is one of excluding all other reasonable possibilities. Pivotal tests are those that are essential to establishing a diagnosis of any type of PAH either by identification of criteria of associated disease or exclusion of diagnoses other than IPAH. All pivotal tests are required for a definitive diagnosis and baseline characterization. An abnormality of one assessment (such as obstructive pulmonary disease on PFTs), does not preclude that another abnormality (chronic thromboembolic disease on VQ scan and pulmonary angiogram) is contributing or predominant. Contingent tests are recommended to elucidate or confirm results of the pivotal tests, and need only be performed in the appropriate clinical context. The combination of pivotal and appropriate contingent tests contribute to assessment of the differential diagnoses in the right-hand column. It should be recognized that definitive diagnosis may require additional specific evaluations not necessarily included in this general guideline. 6MWT indicates 6-minute walk test; ABGs, arterial blood gases; ANA, antinuclear antibody serology; CHD, congenital heart disease; CPET, cardiopulmonary exercise test; CT, computerized tomography; CTD, connective tissue disease; CXR, chest X-ray; ECG, electrocardiogram; HIV, human immunodeficiency virus screening; Htn, hypertension; LFT, liver function test; PE, pulmonary embolism; PFT, pulmonary function test; PH, pulmonary hypertension; RA, rheumatoid arthritis; RAE, right atrial enlargement; RH Cath, right heart ostheterization; RVE, right ventricular enlargement; RVSP, right ventricular systolic pressure; SLE, systemic lupus erythematosus; TEE, transesophageal echocardiography; VHD, valvular heart disease; and VQ Scan, ventilation-perfusion scintigram. Reprinted with permission.

sive test will be performed and by whom. Upon completion of the RHC, the patient and family will need a careful explanation of the findings and how they will guide the treatment plan.

The list of tests above is not exhaustive and varies depending on the protocols of individual PH-treating facilities, but this does highlight some of the pivotal tests that will be done. The patient may need additional testing such as an exercise RHC, ventilation-perfusion (V/Q) scan, cardiopulmonary exercise testing, cardiac MRI, highresolution computerized tomography (CT), chest CT, or pulmonary angiography.

Most importantly, patients need to know that we are here to educate them, advocate for them, and certainly support them as they deal with the chronicity and progressive nature of PH.

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