Detecting pulmonary arterial hypertension (PAH) requires an index of suspicion that is raised by the combination of exertional symptoms and suggestive physical findings such as increased intensity of P2. Cardiac echocardiography is the most useful noninvasive screening test. Diagnosing PAH requires a carefully directed workup that is focused on excluding other causes of pulmonary hypertension. Initially, noninvasive tests are used to confirm the suspicion for pulmonary hypertension and evaluate for secondary contributing factors, and cardiac catheterization is then performed to confirm suspected PAH. Functional testing such as the six-minute walk test and certain blood tests are proving useful in assessing responses to therapy. Additional testing can aid the clinician in determining prognosis and characterizing PAH severity, helping the decision-making process to optimize the choice and timing of treatment.
**Key Words:** pulmonary hypertension diagnosis; echocardiography; exercise testing; pulmonary function; six-minute walk test.

1. **INTRODUCTION**

Diagnosing pulmonary hypertension (PH) and, as much as possible, delineating the underlying pathophysiology are important, because differing pathophysiologies necessitate different treatment approaches. For example, PH associated with left ventricular diastolic dysfunction requires different therapy than idiopathic pulmonary arterial hypertension (IPAH). Thus, correctly detecting and diagnosing PH is absolutely essential in order to formulate a sound treatment plan. Since PAH is mainly a diagnosis made after excluding other, usually more common, diagnoses, the diagnostic workup of PAH requires the utilization of many diagnostic modalities. These include clinical history, physical examination, and noninvasive and invasive testing. The following information represents a synthesis of published data, consensus recommendations (1,2), and clinical expertise used to provide a practical approach to diagnosing PAH.

2. **SUSPICION/HISTORY**

Although PH may be found incidentally, a critical element in the evaluation of PH is the clinical suspicion the diagnostician must have that PH exists, since the symptoms are often nonspecific. This suspicion may be triggered by the patient’s history or physical findings, or it may emerge from one or more abnormal laboratory test results.

Dyspnea on exertion is the most common symptom of PH (3), but is, of course, nonspecific. Also, other nonspecific symptoms may predominate, such as fatigue, lack of energy, and/or syncope, and patients may not readily volunteer that they are dyspneic, leading to delays in detection. In one study (3), the average time from symptom onset to diagnosis was approximately two years. This likely reflects not only the reluctance on the part of patients to attribute significance to their symptoms, but also the clinician’s unwillingness to associate common symptoms with a rare disease such as PAH. Another impediment to the early diagnosis of PAH lies in the fact that PAH symptoms do not typically manifest themselves until pulmonary vascular resistance (PVR) becomes significantly elevated, perhaps years after the onset of pulmonary vascular disease (4).

A focused history facilitates the consideration of important differential diagnoses in the workup of PAH. Dyspnea and/or fatigue