Transcript Details

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2022 ESC/ERS Guidelines for the Diagnosis and Treatment of Pulmonary Hypertension: Updates on Diagnosis

Announcer:
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Dr. Elwing:
Welcome. My name is Dr. Jean Elwing and I'm a professor of medicine and the director of the Pulmonary Hypertension Program at the University of Cincinnati. We will be reviewing today the 2022 ESC and ERS Guidelines for the diagnosis and treatment of pulmonary hypertension. We will review the updates and potential implications.

So, let's talk about what's new and what's recommended by these guidelines. One of the most impactful change is the recommendation to change the hemodynamic definition of pulmonary hypertension. This change included a revised pulmonary vascular resistance cutoff with a change from the previous three or more Wood units to a new cutoff of two or more Wood units to meet the criteria for precapillary pulmonary hypertension. Additionally, exercise pulmonary hypertension was revisited with a new definition of exercise-induced pulmonary hypertension included in the recommendations. Additional updates affected classification, as well as the diagnostic algorithm with a goal of achieving earlier detection of pulmonary vascular disease. Risk stratification was updated and modified based on recent publications and included new echocardiographic determinants of risk, as well as MRI prognostic indicators. The ERS/ESC Guidelines recommended a three risk strata to evaluate newly diagnosed pulmonary hypertension patients to determine risk of mortality and severity of illness at diagnosis and then adopted a four-risk strata risk assessment at follow-up evaluation with the continued goal of achieving low-risk status with aggressive medical therapy. Other recommendations were those that were related to chronic thrombolic disease and chronic thrombolic pulmonary hypertension and recommended evaluating for both with persistent symptoms of dyspnea and exercise limitation after pulmonary emboli. They also discussed the role of catheter-based interventions in both groups. Additionally, standards for pulmonary hypertension centers were included in these guidelines. In today's "PH Weekly," I would like to focus on the recommendations of the diagnostic algorithm. The goal of these changes in this guideline was to increase the index of suspicion for patients at risk of pulmonary hypertension and promote earlier detection and diagnosis of this condition.

The updates in the diagnostic algorithm proposed in the ERS/ESC Guidelines focused on two major tasks. The first was raising early suspicion for pulmonary hypertension and ensuring fast tracking of referral to pulmonary hypertension centers in patients that had a high likelihood of pulmonary arterial hypertension, chronic thrombolic pulmonary hypertension, and other forms of severe and significant pulmonary hypertension. The second is to identify underlying diseases, especially in patients at risk of pulmonary hypertension in the setting of left heart disease and lung disease with the goal of correct classification, accurate risk assessment, and optimal treatment strategies. For risk assessment of patients diagnosed with Group 1 pulmonary arterial hypertension, the guidelines proposed some modifications of the previous 3-risk strata model proposed in the 2019 ERS/ESC Guidelines. That was recommended at diagnosis to be used. At follow-up, the current ERS/ESC Guidelines propose using a 4-risk strata model with defined cutoffs for functional class, six-minute walk distance, NT-proBNP and classification is low, intermediate-low, intermediate-high, and high-risk status.

See the updated initial assessment tool proposed by the ERS/ESC Guidelines here. We'll be concentrating on the echocardiographic parameters, cardiac MRI addition, and the hemodynamic criteria for risk assessment.

In the 3-strata risk assessment tool used at the diagnosis of pulmonary arterial hypertension, there were several updates from the 2019 ERS/ESC risk assessment tool that were recommended. When using echocardiography, an RVPA coupling assessment was added with the use of TAPSE over systolic PA pressure. Cardiac MRI parameters were added to the risk assessment tool with RVEF, stroke volume index and RVN systolic volume index included. Additionally, when performing right heart catheterization, stroke volume index was added to the hemodynamic assessment of pulmonary hypertension patients to evaluate risk.

Now let's move on to the recommended diagnostic algorithm for unexplained dyspnea or suspected pulmonary hypertension. This algorithm begins with the general practitioner obtaining a medical history, doing a physical exam, and obtaining routine testing, including an NT-proBNP. At this point, if there's warning signs for rapidly progressive symptoms, severely reduced exercise capacity, presyncopy or syncopy on mild exertion or right heart failure symptoms, the patient would be fast-tracked to a pulmonary hypertension center for further evaluation. If there are no warning signs, the patient would then be referred to a pulmonologist or a cardiologist based on local practice for further testing. After that, cardiac or pulmonary assessment is complete. The probability of pulmonary hypertension would be assessed. If the probability of pulmonary hypertension is low, it is recommended that the patient undergo additional testing for further evaluation of the etiology of symptomatology. If with further testing, the patient is then determined to have newly discovered pulmonary hypertension risk factors, they then would go on to the pulmonary hypertension center. If the patient is found to have an intermediate or high probability of pulmonary hypertension on that evaluation with pulmonary or cardiology, the patient would be referred onto the pulmonary hypertension center.

Now, let's talk a bit about the testing that is used in the diagnostic assessment of patients with suspected pulmonary hypertension. A great deal of weight is placed on echocardiography in this diagnostic algorithm. Echocardiography is recommended as the first line noninvasive diagnostic investigation in these patients. It is used to assign an echocardiographic probability of pulmonary hypertension based on the TR jet velocity and the presence of other echocardiographic features suggestive of pulmonary hypertension, taking into account the right ventricular changes, pulmonary artery findings, as well as changes in the IBC and right atrium that can occur in the setting of elevated pulmonary pressures. Based on the 2022 guidelines, it is recommended to maintain the current threshold of a TR jet velocity of greater than 2.8 meters per second for the echocardiographic probability of pulmonary hypertension.

Additional recommendations in the diagnostic assessment of patients undergoing pulmonary hypertension evaluation include the following: ventilation/perfusion or perfusion lung scanning is recommended in patients undergoing evaluation for unexplained pulmonary hypertension to assess for chronic thrombolic pulmonary hypertension. CT pulmonary angiography is recommended in the workup of patients with suspected CTEPH. Routine biochemistry, hematology, immunology, HIV testing and thyroid function testing is recommended in all patients with PAH to identify associated conditions. Abdominal ultrasound is recommended to screen for portal hypertension. Pulmonary function testing with DLCO is recommended in the initial evaluation of patients with pulmonary hypertension. It is important to note that open or thoracoscopic lung biopsy is not recommended in the evaluation of PAH patients.

Once patients are diagnosed the guidelines do provide recommendations regarding timing of testing and followup as you see here. In a new patient diagnosed with pulmonary hypertension, it is recommended to obtain a full assessment with functional class, walk distance, NT-proBNP, EKG, echocardiogram, or cardiac MRI, ABG or pulse oximetry and a right heart catheterization. Additional testing is recommended at three to six-month intervals or earlier in case of worsening.

In summary, there have been many changes in the 2022 ERS/ESC Guidelines. The main diagnostic algorithm for pulmonary hypertension has been simplified following a three-step approach from suspicion by first-line physicians, detection by echocardiography, and confirmation by right heart catheterization in PH specialty centers. Warning signs associated with worse outcomes have been identified which justify immediate referral and management by those PH specialty centers. The 3-strata risk stratification assessment in PH has been refined after being validated in multiple registries. The MRI and echocardiographic criteria have been added to the ESC/ERS table, refining noninvasive evaluation at diagnosis. A 4-strata risk stratification dividing the large, intermediate-risk group into intermediate-low and intermediate-high risk group has been proposed to be used at follow-up. Thank you so much for joining us for this "PH Weekly." We look forward to you joining us again in the near future.

Announcer:
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