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Released: 04/29/2022
Valid until: 04/29/2023
Time needed to complete: 1h 59m

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2022 ESC/ERS Guidelines for the Diagnosis and Treatment of Pulmonary Hypertension: A Look At Group I Treatment Guideline Updates

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. Today, we'll be reviewing the 2022 ERS and ESC Guidelines for the diagnosis and treatment of pulmonary hypertension, and we'll be focusing on the updates in the treatment guidelines. So, let's talk about what's new. The 2022 ERS and ESC Guidelines made several updated recommendations and include proposed changes in pulmonary hypertension definitions, including a revised criteria for pulmonary vascular resistance, as well as criteria for the diagnosis of exercise-associated pulmonary hypertension. The guidelines included recommendations regarding updates in the classification, changes to the diagnostic algorithm with modifications on vasoreactivity, and spent time focusing on early detection of pulmonary hypertension. Risk stratification was revisited and included new echocardiographic and cardiac MRI prognostic indicators and changes to the risk table. The new guidelines recommended using a three-risk strata at diagnosis and a four-risk strata for follow-up assessment of pulmonary hypertension patients. There was also a revision of the risk table, as well as modifications to the treatment algorithm to incorporate cardiopulmonary comorbidities, risk assessment, and combination therapy. Furthermore, the guidelines addressed chronic thromboembolic disease and set forth standards for pulmonary hypertension centers. In this installment, we are going to focus on the treatment algorithm and proposed incorporation of cardiopulmonary comorbidities, risk assessment, and combination therapy recommendations.

The updated ERS/ESC Guidelines continue to stress our current focus on pulmonary hypertension risk assessment and treatment to low-risk status. The updated guidelines recommend two possible approaches to newly diagnosed intermediate-risk patients with an initial treatment strategy of either maximal medical therapy with triple combination therapy including a parenteral prostacyclin, or an aggressive titration strategy starting with dual oral therapy. The guidelines discuss the importance of a selection of a treatment strategy utilizing shared decision-making and an awareness of the risks, uncertainties, and potential benefits of maximal medical therapy with upfront triple combination therapy including a parenteral prostacyclin. And if the aggressive titration strategy starting with dual combination therapy is chosen, there should be a plan for rapid reassessment of response within three months of diagnosis.

The guidelines were focused on questions with direct consequences for clinical practitioners regarding the pulmonary hypertension subgroups and included initial treatment strategies for group 1 pulmonary hypertension, the use of oral phosphodiesterase 5 inhibitors for the treatment of group 2 and group 3 pulmonary hypertension, and the use of pulmonary hypertension medications prior to balloon angioplasty for the treatment of group 4 pulmonary hypertension. These questions were considered to be important because most contemporary pulmonary hypertension registries describe variable use of initial monotherapy and combination therapy for pulmonary arterial hypertension patients. And large case series showed widespread use of PDE5 therapy in group 2 and group 3 pulmonary

hypertension. And there currently are no clear guidelines for medical therapy in patients with inoperable chronic thromboembolic pulmonary hypertension prior to balloon angioplasty.

Now, I would like to direct your attention to the updated treatment algorithm for idiopathic, heritable, drug-associated, and connective tissue disease-associated pulmonary arterial hypertension. The 2022 guidelines recommend a pulmonary hypertension center referral for evaluation and confirmation of the diagnosis of PAH. Evaluation of vasoreactivity is included in the diagnostic algorithm and the importance of general measures is stressed throughout the course of disease. The updated algorithm proposes that the non-vasoreactive patient undergo risk assessment and treatment based on severity of disease with newly diagnosed low and intermediate-risk patients via three-risk data be treated with dual combination therapy and those high-risk patients with triple therapy including a parenteral prostacyclin. Patients should then be regularly followed and reevaluated with a four-risk strata assessment and medical therapy be uptitrated if they are not reaching their treatment goals of low-risk status. This algorithm did include taking into account those patients with significant cardiopulmonary comorbidities and modifying medical therapy based on those factors with ongoing reassessment after initial medical therapy management.

In the updated guidelines, vasoreactivity testing is recommended in idiopathic, heritable, and drug and toxin-associated pulmonary arterial hypertension patients. The subgroups of non-responders at vasoreactivity testing and acute responders at vasoreactivity testing have been included in the guidelines to help drive treatment decisions. Calcium channel blocker therapy is recommended in selected vasoreactive patients and continuing high-dose calcium channel blockers is recommended in patients with idiopathic, heritable, or drug and toxin-associated pulmonary arterial hypertension who achieve or maintain functional class I or II status and have marked hemodynamic improvement with this therapy. In patients with a positive vasoreactivity test but insufficient long-term response to calcium channel blockers who require additional PAH therapy, continuation of those calcium channel blockers should be considered.

Now let's focus on the non-vasoreactive patients with idiopathic, heritable, or drug and toxin-associated pulmonary arterial hypertension without significant cardiopulmonary comorbidities and see how they track through this algorithm. In patients who meet these criteria who present at the time of diagnosis with high-risk status, initial combination therapy with a phosphodiesterase 5 inhibitor, an endothelial receptor antagonist, and a parenteral prostacyclin should be considered. In patients who are presenting at follow-up who do not achieve low-risk status, who are maintaining an intermediate low-risk status while receiving an endothelin receptor antagonist and a phosphodiesterase inhibitor, the addition of a prostacyclin receptor agonist should be considered. In patients presenting again at follow-up not achieving low-risk status and remaining at intermediate or high-risk status while receiving an endothelin receptor antagonist and a phosphodiesterase 5 inhibitor should be offered the addition of a parenteral prostacyclin either intravenously or subcutaneously and should also be considered for referral for lung transplant evaluation.

Now let's drill down on some specific medication combinations that were discussed in the ERS/ESC Guidelines for initial combination therapy for group 1 non-vasoreactive patients without significant cardiopulmonary comorbidities. The initial combination therapy with ambrisentan and tadalafil was recommended with a class I and level of evidence B recommendation. Initial combination therapy with macitentan and tadalafil was also recommended with the class I and level of evidence B recommendation. Initial combination therapy with other endothelin receptor antagonists and phosphodiesterase 5 inhibitors should be considered with the class IIA and level of evidence B recommendation. The initial combination therapy with macitentan, tadalafil, and selexipag was not recommended with a class III and level of evidence B recommendation.

Sequential drug combination therapy for group 1 pulmonary arterial hypertension was also reviewed in the guidelines. It is recommended to base treatment escalations on risk assessment. The addition of macitentan to a phosphodiesterase inhibitor or an oral or inhaled prostacyclin analog is recommended to reduce the risk of morbidity and mortality events with a class I level of evidence B recommendation. The addition of oral treprostinil to an endothelin receptor antagonist or a phosphodiesterase 5 inhibitor or riociguat monotherapy is recommended to reduce the risk of morbidity and mortality events with a class I and level of evidence B recommendation. The addition of riociguat to bosentan should be considered to improve exercise capacity with a class IIA and level of evidence B recommendation. The addition of bosentan to sildenafil is not recommended to reduce the risk of morbidity and mortality with a class III and level of evidence B recommendation.

As mentioned earlier, a modified approach to the non-vasoreactive group 1 idiopathic, heritable, or drug and toxin-associated pulmonary arterial hypertension patients with significant cardiopulmonary comorbidities was proposed and the guidelines recommend initial monotherapy with phosphodiesterase 5 inhibition or endothelin receptor antagonists, and patients should be followed closely. If individuals remain at intermediate or high risk while receiving monotherapy PDE5i or ERA therapy, additional pulmonary arterial hypertension medication could be considered on an individual basis.

In summary, key takeaways and remaining gaps in knowledge were addressed in the updated ERS/ESC Pulmonary Hypertension Guidelines. The treatment algorithm for PAH has been simplified with a clear focus on risk assessment, cardiopulmonary comorbidities,

and treatment goals. Initial combination therapy and treatment escalation at follow-up when appropriate are current standards. The four-strata risk assessment, dividing that large intermediate group into intermediate-low and intermediate-high risk is proposed at follow-up. The efficacy and safety of PAH therapies in group 1 patients with mean pulmonary pressures between 21 and 24, pulmonary vascular resistance between two and three, and exercise-associated pulmonary hypertension has yet to be established. The role of pulmonary arterial hypertension therapies in different PAH subgroups including schistosomiasis-associated PAH needs to be further explored. And finally, the impact of PAH therapies and treatment strategies on survival needs to be further assessed. With that, I want to say thank you so much for joining me to review the 2022 ERS/ESC Guidelines for the diagnosis and treatment of pulmonary hypertension, focusing today on treatment updates for group 1 patients.

Announcer:
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