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2022 ERS/ESC PAH Guidelines on Treatment

Announcer:

Welcome to CME on ReachMD. This episode is part of our MinuteCE curriculum.

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Dr. Elwing:

So 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. And I'm going to be talking with you today about essential aspects of the new 2022 ERS/ESC PH Guidelines on Treatment, top-line takeaways.

So, all PAH patients are treated with general measures. And the new guidelines list these in detail and adds some other aspects we should consider. So first, general measures, they recommended supervised exercise, psychosocial support, immunizations, diuretics, long-term oxygen when appropriate, and assessing anemia and treating it if it's present, and looking for iron deficiency and replacing it when indicated. They recommended anticoagulation not be generally used except in certain circumstances, and avoiding left heart failure therapies like ACE inhibitors and beta blockers unless there's another indication. They mentioned special circumstances like use of supplemental oxygen with air travel in individuals who have a PO₂ of 60 or less at sea level. And they talked about working together for individuals who need interventions requiring anesthesia and multidisciplinary consultation, and doing those interventions in a center that has a PAH program.

They talked about pregnancy and individuals who have childbearing potential, and to counsel these individuals early on at the time of diagnosis about risks and certain uncertainties of pregnancies in pulmonary hypertension. They recommended to provide women of childbearing potential with clear contraceptive advice and guidance. They recommended that PAH patients who consider pregnancy or become pregnant receive prompt counseling and guidance. For those individuals considering termination if they become pregnant, it should be recommended that it be performed in a center that has a PAH program to support that individual. And those PAH patients who are looking to have children, adoption and surrogacy with preconception counseling should be considered. And lastly, those individuals who do become pregnant should avoid the use of endothelin receptor antagonists and guanylate cyclase stimulators like riociguat, because these drugs can have teratogenic effects.

So let's talk a little bit about individual groups of pulmonary arterial hypertension patients. First, the vasoreactive patients that have idiopathic heritable or drug and toxin-induced disease, we see, assess, undergo right heart catheterization, and test vasoreactivity. They recommend the use of nitric oxide or inhaled iloprost for testing, and epoprostenol I.V. if that is not available. And then if a patient has a positive response with a drop in their mean PA pressure by 10 or more, reaching 40 or less with no decrease in their cardiac output, then to use a calcium channel blocker and up-titrate to high levels, as tolerated.

So for the non-vasoreactive patients, this is where the algorithm really took a change. They recommend to assess the patient, diagnose, undergo right heart catheterization, and then divide the groups of these non-vasoreactive PAH patients into those with no comorbidities and those with cardiopulmonary comorbidities, and assess them and manage them differently. And this is very important because it is a change from previous guidelines. Monotherapy was recommended in those patients with comorbidities in the setting of non-

vasoreactive pulmonary arterial hypertension. And we'll talk a little bit more about that later.

So why would they recommend that we consider monotherapy in the non-vasoreactive PAH patients? Well, patients with cardiopulmonary comorbidities were underrepresented in many of our trials, and registry data suggests that many of these patients are oftentimes treated with PDE5 therapies as initial monotherapy. Endothelin receptor antagonists and the combination of PDE5 and ERAs are occasionally used, but drug discontinuation is higher than classic PAH. In a subgroup analysis in AMBITION, of these patients with left heart phenotype, showed less clinical improvement and higher rates of discontinuation of drugs because of adverse events. And in the ASPIRE registry, it demonstrated that patients with this cardiopulmonary phenotype had less improvement in exercise capacity as compared to classic idiopathic PAH patients. And we also saw in other studies that ERAs triggered fluid retention in those patients with left heart phenotype, and there was more decline in peripheral oxygen saturations at times in these patients. And there's little published on the experience of use of prostacyclins or prostacyclin receptor agonists in this group. And there is a lack of evidence using these PAH therapies in our elderly patients with comorbidities. So this is what prompted that change.

So for the patients without vasoreactivity, without comorbidities, they recommend to go through our routine evaluation with risk assessment. And initial risk assessment is a 3-strata model. And those individuals who are at low or intermediate risk would go on to combination PAH therapy with a PDE5 and an ERA. Those who are at high risk, receive the oral therapy plus a parenteral prostacyclin.

So when we're looking at drugs, and we're talking about ERAs and PDE5's, and our parenteral prostacyclins, we know there's other therapies in addition to those available and sometimes we do have to change, modify based on side effects and tolerance. But based on guidelines, this is where we're starting in our PAH patients.

But we don't end there. We reassess, and we determine if our medications had the response we want from these individuals. So we look at their functional class, their walk distance and their NT-proBNP and we use this 4-strata model. And we look at the grade of each individual parameter, and we divide that by the number assessed, and we find risk in these individuals. And then this prompts us to change medication if individuals are not at low risk. If they're intermediate risk, we can look at our patients who are on PDE5s and ERAs and we could consider changing that PDE5 to a guanylate cyclase stimulator. Or we could add a prostacyclin receptor agonist.

And our patients that don't meet these low-intermediate risk, and they're in the intermediate or high-risk category, we should consider parenteral prostacyclins as the next step in their therapy, and also consider lung transplant referral.

So in summary, general measures should be implemented to optimize health status in all of our PAH patients. Pregnancy should be avoided in PAH due to risk of cardiopulmonary compromise. Vasoreactivity testing should be followed for our idiopathic heritable and drug and toxin patients associated with PAH. Calcium channel blocker therapy should be considered in these individuals who have a positive challenge. Monotherapy could be considered in those non-vasoreactive patients with significant comorbidities but assessed on an individual basis. PAH combination therapy should be used in our patients with non-vasoreactive PAH without comorbidities and medication choices will be driven by risk assessment.

So thank you so much for joining me. I hope you enjoyed this program.

Announcer:

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