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Don't Wait to Escalate—Getting Aggressive in PH Management

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

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Dr. Elwing:
Welcome to CME on ReachMD. My name is Dr. Jean Elwing,

So we know that pulmonary hypertension is quite common. It affects about 1% of the global population. But when we talk about pulmonary arterial hypertension, we're talking about a rare disease. This condition requires an extensive evaluation and hemodynamics to diagnose it. We go through an extensive evaluation that includes medical history, physical exam, EKGs, and when we think that it is likely a patient could be affected by pulmonary hypertension, we then go on to assess with hemodynamics, and that confirms the diagnosis. And once we get to the point of PAH, we have to stop and reassess. We have to know how sick our patient is. And in my practice, at that point, we use a risk assessment tool in the medical record, and we track it each visit.

But when we looked at the number of individuals that care for PAH patients who are actually using risk assessment tools, we found in a survey that was performed by the CHEST Pulmonary Vascular Network that only about 60% of providers were using a tool routinely. And that included many of our options in REVEAL, COMPERA, the French registry, both invasive and noninvasive, and the Swedish data. So many individuals are not using risk assessment tools.

But you say maybe we could just use our gestalt. Well, Dr. Sandeep Sahay looked at that with phone interviews, reaching out to PAH providers and giving them case scenarios. And he found that the gestalt did not match the answers that were achieved when patients were formally assessed with COMPERA, REVEAL, or the noninvasive French. So we were not consistent with any of those risk tools when we used our expert opinion.

So in the most recent ERS/ESC guidelines, they took a step back and said, "What can we do better?" And they added more robust echocardiographic and cardiac MRI prognostic indicators. And they said, "When you meet a new patient, every patient should be risk assessed in an expert center." And they took a 3 strata model when the patients were initially assessed, and then a shorter 4 strata model on follow-up.

So looking at the risk assessment based on ERS/ESC, you can see here there's 3 categories; the green, which is always the goal, the go, low risk. We want patients to meet criteria of low-risk status. And what does that tell us? That the likelihood they will be surviving that first year after diagnosis is the highest we can achieve. We try to keep patients out of that yellow risk, which portends a poorer prognosis with the risk of mortality of 5% to 20%. And we really want to capture anyone in the high-risk group, a greater than 20% mortality in that first year, and change their outcome by adding medications, augmenting therapy to get them to a lower risk status. And what does this look at? Multiple variables. It looks at heart failure, progression, symptoms, functionality, cardiopulmonary exercise testing if they have completed that, biomarkers with BNP or NT-proBNP, echocardiographic features, cardiac MRI, and hemodynamics. And you say, "Oh,

I'm not doing all of those things." Well, you have to give the patient that full assessment.

But you say, "Well, I don't use that risk tool." Well, some people use REVEAL. And just giving you an example of REVEAL 2.0, and that's what I use in my medical record, and I track patients over time, and I put this in a flowsheet. And this gives you multiple variables, both modifiable and non-modifiable variables that then you get a point score for, and then you can put them in that low-, intermediate-, or high-risk zone. Again, guiding therapy.

And how does that play into our next steps in evaluating patients? Well, you saw your patient, you diagnosed them, you confirmed the diagnosis with your full, robust evaluation, and then you risk assessed. And you decided, is this a low- or intermediate-risk patient? And those patients would be treated with dual up-front combination oral therapy. Or are they high risk? And we're going to go and we're going to be the most aggressive we can be with those individuals, because they need the greatest help quickest.

But we don't stop there. The guidelines and our clinical practice suggests we need to reevaluate and reevaluate in a standardized fashion. And medical reassessment, functional class, and testing is recommended multiple times throughout the patient's course, every 3 to 6 months. And we need to reassess with that additional testing with echocardiograms, right heart catheterization, maybe cardiopulmonary exercise testing, cardiac MRI, when the patient deems a need. So this is something that doesn't stop at diagnosis. The only way we get patients to their goal is to reassess, reevaluate, and redetermine the best strategies for the patients in terms of medications.

So when we see patients in follow-up at that 3- to 6-month mark, we would then go ahead and reevaluate their risk status. And based on the ERS/ESC guidelines, they recommended a 4 strata model. And you look at very easy, quickly obtainable information, their functional class, their walk distance, and their BNP. And you get a score for each one of those, and you divide the number by the number of variables you assessed, and that's where you land in terms of your risk.

If you choose to use REVEAL, it would be either the full REVEAL 2.0, or you could use REVEAL Lite 2, which looks at fewer variables and is able to discriminate risk very well. You get a low- or intermediate- or high-risk zone for those patients, and then you know what's next for the patient.

So one thing that may be part of our future risk assessment is cardiac imaging and hemodynamics. Recently published, they looked at REVEAL plus echo data, looking at RV size, RV function, tricuspid regurgitant jet severity, and pericardial effusion. And they saw that those parameters helped improve the ability for REVEAL to discriminate those patients from low to high risk. So it helped really tease out the differences in the subgroups and help us to know which patients need more aggressive therapy.

The French group looked at patients in the French registry from 2009 to 2020. And they found that when they looked at hemodynamics and added that to the French risk assessment score, it helped improve the ability to know what to do for those intermediate-risk patients. Those are our biggest group, and they're the hardest to manage because we have to figure out, do we go at a more aggressive pace, or do we change therapy? What is the best option for that individual to achieve low risk with the most tolerable and easy-to-use therapy? So this can be added to our current risk assessment. Looking at data, we already get the right heart catheterization, which I usually repeat a right heart catheterization at that point where I feel like they've reached a reasonable amount of prostacyclin or they have not reached low-risk status by my noninvasive tools. And then I could use that data to say, "Hey, have I, with hemodynamics and maybe the echo in addition to that, really gotten this patient to the lowest risk status I can with my current medications?"

So we get that information, and we go back to our treatment guidelines, and we say, "Yes, I reassessed my patient and they are low risk. That means I can stay on my current therapies and hold tight." I'm not going to forget about the patient. Of course, I'm going to see them, reassess, and reevaluate at each visit.

But if they're in that intermediate zone, I've got to decide, do I need to change therapy, or do I need to add therapy? And if they're at high risk, I need to reevaluate what I'm doing and think about parenteral prostacyclins. And that is one of our most effective but most challenging medications for our patients. So I really need to know that this is going to change how the patient is feeling, how they're doing, how they're maintaining their functionality. And I want to be able to talk with them about that so I can counsel them on the importance of that addition.

So what do I tell patients when we talk about adding therapy or changing therapy? They may be very satisfied with their current overall status, but I want to tell them that, "I want more for you. I want you to be as well as you can." And what has been shown, looking at data from the French registry, is that for patients that are at low risk that stay at low risk, they do very well, and their outcomes can be very similar to another patient without pulmonary hypertension if we remain at that level.

And I want you to see here, this dotted green line, those are patients that were at higher risk and then became low risk. They also have a very good outcome. The solid red line are those individuals that were at high risk and stayed at high risk, and the dotted red line are

those who worsened to high risk, and those patients have a very poor prognosis. They are very affected by their pulmonary hypertension, and it has caused increased risk of morbidity and mortality. So those are patients we really want to target and do everything we can to improve their risk status.

And you see here these 4 panels. Panel A are all comers with pulmonary arterial hypertension; B are idiopaths, heritable, and drug- and toxin-exposed patients; C are the connective tissue disease patients; and D are those with congenital heart disease. So all patients are following that same pattern and should be approached in a similar fashion.

So what about those patients that come to you that are so very sick and you start on triple-combination therapy? Well, I was delighted to see the information from the French registry that was published in 2021 looking at those triple-combination therapy patients. 46% of them had no low-risk features in terms of their French risk assessment with the invasive data. And then at first follow-up, 39% of them had 4 low-risk criteria. That's first follow-up. So we can aggressively manage patients and quickly turn around the processes that are occurring in their bodies and in their cardiopulmonary status to change the risk status in that first follow-up.

And when we look at overall survival with transplant and without transplant, we can see that there's a dramatic difference when we treat patients with aggressive interventions for PAH. And I want to point your attention here to this first graph here, and it tells you overall survival of pulmonary hypertension patients. And then when you look at the lower graph, you'll see the red, the triple-combination therapy patients. Those are our sickest patients. And if you see here at 10 years, we have more than 80% survival overall. And if you look at transplant-free survival, still in the 70% range. So a dramatic difference than when we started caring for patients with pulmonary arterial hypertension many years ago.

So what does the future hold for risk assessment? Well, first, we've got to use it. We've got to increase awareness. We've got to be consistent. We've got to track it. It only works if we use it the way it's supposed to be used. And we have to use standardized tools; we can't use our gestalt. And we should use the information we have. We should add cardiac imaging and add hemodynamics to be able to understand our patients' status the best we can.

So with that, I want to tell you thank you for joining me and learning about risk assessment and the future.

Announcer:
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