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PH Management Into the Future: Understanding the Next Generation of Treatments and Mechanisms

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

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Dr. Moles:
Welcome to our chapter exploring challenges and barriers to risk stratification in pulmonary hypertension.

This is CME on ReachMD, and I am Dr. Victor Moles.

What I wanted to discuss with you today was how risk stratification directs treatment plan in pulmonary arterial hypertension. So as you know, we have different risk stratification models. Two of the main ones include the European guidelines or 3 strata risk score, and also the REVEAL score. Both of them use multiple data and information from the clinical data that we get in clinic, functional capacity, imaging, and even hemodynamics to come up with 3 strata, meaning low risk, intermediate risk, or high risk. And these 2 models are very comprehensive and probably better used when patients are initially diagnosed with their condition.

For patients who we see routinely in clinic after they've been diagnosed and they are on medical therapy, I think that the 4 strata risk score or the REVEAL Lite 2 score are much easier to implement in the routine clinical practice. The 4 strata risk score uses functional class, 6-minute walk distance, and BNP, and categorizes patients into low, intermediate-low, intermediate-high, and high risk. And the REVEAL Lite 2 score uses comorbidities such as chronic kidney disease, functional class, blood pressure, heart rate, 6-minute walk distance, and BNP to categorize patients into low, intermediate, or high risk.

So the important thing about risk stratification is that it helps guide medical treatment in PAH. The goal is always to achieve a low-risk status. And I really enjoy this diagram from the 6th World Symposium in Pulmonary Hypertension, which walks you through how to use risk stratification in patients with PAH.

So once a patient has been confirmed to have PAH, and I'm talking about those who do not have a positive vasoreactivity testing, the way we want to use risk assessment is to define patients into 2 distinct categories; those with a high risk will benefit from initial combination therapy that includes a parenteral prostacyclin and early consideration of lung transplantation in their care. Patients who have low or intermediate risk would benefit, most of them, from an initial oral combination therapy.

Now, things don't stop there, and when we see patients in clinic, we want to keep using that risk stratification. And again, the goal is to achieve a low-risk status. If we achieve that, we will keep following the patient without changing their medical therapy. But those patients who continue to be on an intermediate- or high-risk status, we want to escalate or add more PAH-specific medical therapy or to increase the dosing, if we're talking about a parenteral prostacyclin. For those patients who cannot achieve a low-risk status, we want to consider lung transplantation in their care at that time.

Now, the important thing about achieving a low-risk status is that it forecasts an excellent long-term prognosis. What you can see here

in this graph is the survival of patients with PAH according to their REVEAL score. And the line that you're seeing on the top, this red line, are those patients who did achieve a low-risk status. And you can see their 5-year survival is excellent comparable to the general population. Now as that REVEAL score increases, the survival is less optimal.

Now I do recognize that risk stratification is not always simple. And what I mean by that is that risk stratification score may not always correlate with PAH disease severity. Here are some examples. Musculoskeletal issues may decrease a 6-minute walk distance and may overestimate the risk. Young and fit patients may show a higher 6-minute walk distance and that may underestimate a risk. Elderly patients with comorbidities such as COPD, CAD, and atrial fibrillation may have a decreased 6-minute walk distance and higher NT-proBNP, which would overestimate the risk. The treatment of essential hypertension may lower blood pressure, overestimating the risk. And arrhythmias may increase heart rate, which may overestimate the PAH disease severity.

So I want to present you with 2 different patients, both of them have in common that they have an intermediate risk by the REVEAL Lite 2 score. Patient A has this echocardiogram. As you can see, the right ventricle is severely dilated and dysfunctional. The right atrium is very dilated. And when we look at the hemodynamics, the right atrial pressure is 12, the mean PA pressure is 60, the cardiac index is 1.9, the stroke volume index is 19, and the pulmonary vascular resistance 10 Wood units.

On the other hand, we have Patient B, who has this echocardiogram. As you can see, the right ventricle, at the most, is mildly dilated. The function is very normal, and the right atrium is very close to being normal in size. The Patient B hemodynamics are a right atrial pressure of 7 and mean pulmonary artery pressure of 38, a cardiac index of 2.6, a stroke volume index of 40, and a pulmonary vascular resistance of 2.5.

As you can tell, both patients have an intermediate risk score by the REVEAL Lite score. But these 2 patients are very, very different, and this is why we need to better understand how to get more granularity on risk stratification.

What I'm showing you here is data from the REVEAL registry, which a few years ago came up with the REVEAL-ECHO score. And what the REVEAL-ECHO score does is takes information from RV enlargement, RV dysfunction, pericardial effusion, TR severity, and PAH subtype, and that helps categorize patients into low, intermediate, and high risk. Now if you take the REVEAL-ECHO score and you merge it with the REVEAL Lite 2 score, you can get a lot more granularity on the patient's risk, and you can come up with 4 different strata: patients who are in low risk, intermediate-low risk, intermediate-high risk, and high risk. And as you can see in the image in the bottom right, there are some patients who have low risk by the REVEAL Lite score, that if they have a high echo score, their risk may be more than low. And also patients in the intermediate REVEAL Lite 2 category may be further differentiated into intermediate-low and intermediate-high.

Now hemodynamic values can also help stratify patients. This is data from the French Pulmonary Hypertension Network, and what they have recently showed is that the use of some hemodynamic variables can help in those patients who are at intermediate risk. Patients who are at low risk by the 4 strata criteria or high risk, they don't benefit much from additional hemodynamic information. But patients who have an intermediate-low or intermediate-high risk may be further divided using 2 variables, the stroke volume index or the pulmonary artery saturation. And what this can help create, instead of a 4 strata risk score, is a 6 strata risk score. And patients in the intermediate-low or intermediate-high risk category can further be divided into having a better or worse prognosis if they have the presence of a stroke volume index above 37 mL/m^2 or a pulmonary artery saturation above 65.

So understanding risk stratification is important, but what is, I think, more important, is to translate that into the daily clinical practice. This is what we have done at our institution, at the University of Michigan, we have embedded the risk prediction models into the electronic medical records. And what we have done is created a flowsheet in Epic so every time the patient comes, they have blood work and a 6-minute walk test, we assess the functional capacity, we measure their vital signs, and we can come up with a 4 strata risk score and a REVEAL Lite 2 score. And moreover, what we do is we created a SmartPhrase which translate this information into our clinic note, so we can follow their risk over time and understand when we need to escalate their medical therapy or if we have achieved the goal that we set for each patient.

Now I want to talk a little bit about social determinants of health and disparities. Social determinants of health are the conditions in the environment where people are born, learn, work, play, worship, and age that affect a wide range of health, functioning, and quality of life outcomes and risks. Providers' knowledge of social determinants of health is vital to address individual social needs.

The research has also suggested that proper intervention to address social determinants of health can result in positive outcomes. There was a survey done in 2022 of pulmonary hypertension specialists, and what we found is that clinicians from pulmonary hypertension care centers across the US were highly aware of social determinants of health. The issue was that most centers did not have formal screening methods for social determinants of health.

And what I want to share with you is that there is a simple tool called the 10-Item Social Determinants of Health Screener, where you can ask patients during each clinic visit, or at least once a year, 10 simple questions that can help screen for social determinants of health that may need to be addressed. And I will leave a link to the description, to the link in CMS, where you can find this information from.

Delays in the diagnosis and initiation of PAH treatment are common. Data from multiple registries have revealed a mean time from symptom onset to PAH diagnosis ranging from 1 to 4 years, with most of the delay occurring after the patient's first contact with the medical system.

There was a study called the RePHerral study in 2013 that assessed pulmonary hypertension referrals for 3 major pulmonary hypertension centers in the United States. And what it showed is that 61% of the patients had functional class III or IV at the time of their referral to a tertiary center, 33% of the patients received a misdiagnosis before referral, and 42% of the patients had a right heart catheterization for the first time at the referral center.

Interestingly, there was another study in 2022 from the Veterans Affair Administration that showed that the median time from the diagnosis of PAH to receive an appropriate therapy was 16 weeks. Lower household income was associated with longer delays in times. And you need to take into consideration that the VA substantially subsidizes medications cost, so cost-related medications might not have been the main reason for this delay, but maybe potentially traveling or going to the center. Race/ethnicity were not associated with the time of PAH diagnosis to treatment.

So I wanted to share with you a little bit of what we do in the state of Michigan. We have the Michigan CTEPH Multidisciplinary Conference. As you may know, CTEPH is a rare disease that requires very specialized, multidisciplinary care. And what we do is a monthly virtual conference with PH specialists across the state. These are specialists from PHA-accredited and also nonaccredited centers, and we share information. Our goals are to discuss a diagnosis and treatment options for CTEPH patients. We want to early diagnose CTEPH. We want to create a network of specialists with interest in this condition. And we also discuss morbidity and mortality and also teaching cases. And I think this is a very helpful way to collaborate within the state, but also to provide patients with timely diagnosis and access to PAH treatment.

I also want to point out that patients have to travel sometimes a very long distance to get to a pulmonary hypertension referral center. What you're seeing here is a map of the United States with all the PHA-accredited centers across the country. You will see that the East has more density of centers, with the West having less. And there are some areas in the center of the country where states don't have a PHA-accredited center. I'll also share the map of the state of Michigan, and you can see that patients sometimes have to travel a long, long distance to get to see us.

So as we think about how we can better serve patients, you may want to think how you want to use virtual care or telehealth. And I think that something that has worked for us quite a bit is considering a virtual initial visit as a way of talking to patients, understanding how symptomatic or not they are, reviewing their diagnostic testing, explaining them the importance of diagnosis and treatment, but also planning ahead and doing most of that diagnostic testing in one day, if possible, when they come to the referral center.

So in conclusion, risk stratification models accurately predict outcomes in PAH and help guide medical therapy. Echocardiography and hemodynamics can also assist in giving more granularity to traditional risk models. Integration of risk models to daily clinical practice is key.

Providers need to understand patient social determinants of health to provide an integrated and effective care. Multicenter collaboration can have many benefits, including timely access to care. Then think about what role can virtual care have in your practice.

That's all the time that we have today. Thanks for listening.

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