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Why Is Formal Risk Assessment Critical to Effective Treatment of PH?

Dr. McLaughlin:
Hello, and welcome to this program on "Maximizing PH Care Using Risk Assessments to Improve Outcomes". I'm Valerie McLaughlin, Director of the Pulmonary Hypertension Program at the University of Michigan. And I'm so glad you can join me for this.

So why is formal risk assessment critical to effective treatment of pulmonary hypertension? Well, we've learned so much about risk assessment over the past decade or so. We know it's really critical in many chronic diseases and through many databases, we've learned that comprehensive patient assessment in PAH allows us to determine a patient's prognosis, monitor their disease progression, and their response to therapy. It really helps us make treatment decisions. It's become a critical part of the ESCERS guidelines for the care of patients with pulmonary hypertension. And those guidelines recommend that risk assessment be conducted regularly every three to six months in stable patients. And it's really critical that the risk assessment include multiple parameters to evaluate disease progression and the patient response to therapy. So risk assessment should include a range of clinical hemodynamic and exercise parameters. And we really look at all of these together. There's no one variable that we rely on to provide definitive prognostic information.

So we know that pulmonary hypertension is a complex disease and often actually the patients have really advanced symptoms by the time they present to us. There are a number of factors that influence the development of pulmonary hypertension, including genetic and genomic factors, but also environmental factors as well. And we know early in the course of disease, the pulmonary pressures go up and the patient may not be very symptomatic. And because the right ventricle is coping with those pressures, but at some point in time, the right ventricle becomes stressed by that high pulmonary vascular resistance. And it doesn't function as well. And the pulmonary pressures and pulmonary vascular resistance go up and ultimately the right ventricle becomes compromised, and it may even fail. So by the time the patient becomes symptomatic, the pulmonary vascular disease is really quite advanced. And the thing that causes patients to get sicker is not that the pulmonary pressures keep going up and up and up, it's that the right ventricle has trouble coping with that high resistance and the right ventricle fails. In fact, as the patient gets sicker, you might even see the pulmonary artery pressure decline because the right ventricle just can't generate enough output. We'd see this in left heart failure as well.

Now as I said, patients are often very advanced by the time they get diagnosed, even current day, most of the patients I see have functional class three symptoms at the time of diagnosis which is unfortunate because we know that the sicker they are when they get to us, the worst their outcomes are. So on this slide, our data from three different registries looking at baseline risk assessment and survival. The Swedish PAH register on the left and COMPERA on the far right use essentially the same methodology. They look at the variables from the ERSESP guidelines and really assign a score of one if it's low, two it's intermediate and three if it's high and then divide by the number of variables and round to the nearest integer to get low intermediate and high risk. And you can see in both of those registries, the patients at high risk depicted in red at the time of diagnosis have a much poorer prognosis than those at intermediate risk depicted in yellow or those at low risk depicted in green. Very, very similar results in those two databases. Now if you look in the middle, there's the French registry and they do things a little bit differently. They basically said our goals of being low risk include four parameters, one, to have functional class one or two symptoms, two, to have a six minute haul walk of greater than 440, three, to have a right pressure of less than eight, and four, to have a cardiac index of greater than 2.5. And if you meet all four of those variables, you can see in green, the prognosis is very good. That means you're less ill at the time of diagnosis. The blue is three

variables, the red is two, the blue hashed is one and the gray is zero. And you can see that the prognosis gets worse the fewer variables you have or said another way, the prognosis gets worse, the higher risk that you are. So this is one of the reasons it's important to emphasize awareness, emphasize early diagnosis, and an early referral to a pH center, because we know that the more advanced you are at the time of diagnosis, the worse your prognosis is.

However, the good thing is that even if you have advanced symptoms or advanced risk at the time of diagnosis, you still have the opportunity to respond to medical therapy. And if you respond to medical therapy and if we can improve your risk status, then your prognosis improves. On this slide, you see those same three registries. And what we're looking at here is the patient's risk assessment at the time of their first follow up. So they were diagnosed, their risk was assessed, they were started on therapy, and now we're risk assessing them again on therapy. And the good thing is, is that if we can improve their risk on therapy, their prognosis is much better and it's consistent across all of these databases, but let's focus on the far left, on the Swedish PAH register. Cause I think this demonstrates it very, very nicely and is a little more simple to look at. So on the left, you see the same sort of risk assessment looking at the variables from the ERSESC and kind of adding their scores dividing by the number of variables measured and assessing whether or not they hit low risk status. And what you can see in the green line are the patients who presented at low risk and are still at low risk, stayed at low risk at the time of their follow up. And their subsequent prognosis is very good. But I call your attention to that lighter blue line on the top. These are patients who were not at low risk at the time of diagnosis, they were at intermediate or high risk at the time of diagnosis. And then with therapy, they improved to low risk. You can see that their subsequent five year survival is essentially the same as the patients who were at low risk and stayed at low risk. This is really critical, and this is one of the reasons our treatment algorithms are based on achieving low risk, because we know that no matter where you start, if we can get you to low risk, your prognosis is good. On the other hand, in the darker blue line, you see the patients who were at intermediate or high risk and stayed there. So the prognosis is not so good. And even worse are the patients who were at a better risk level and did response therapy and progressed and worsened to intermediate or high risk. And you can see their survival is very poor, 20% die within the subsequent year and more than half of them die within five years. And we won't go through the other two registries, but they're really very much the same. If we can improve to low risk, the prognosis is better.

So I hope this helps convince you that risk assessment to monitor prognosis and PAH is an important part of care. And included in this risk assessment are clinical functional exercise, non-invasive and sometimes invasive therapies, or invasive variables that all go into our risk calculators. Now there are a number of risk calculators available. There are different models, the French pulmonary hypertension registry uses those four variables or there is a less invasive way that swaps out NT-proBNP for the hemodynamics of right atrial pressure and cardiac index. There's pH connection, there's COMPERA and Swedish which we've talked a little bit about, which are really based on the ERSESC guidelines. There's a Scottish score. And then of course there's the reveal risk equations, reveal is a very large US based registry that has created a calculator and a number of equations and including more recently, a less complex equation that only has six variables reveal like two.