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ReachMD
www.reachmd.com
info@reachmd.com
(866) 423-7849

Assessing Risk: What Needs To Be Measured at Every PH Visit?

Dr. McLaughlin:
So let's talk about assessing risk. What needs to be done at every PH visit? And I think this is really easy. It's very, very straightforward.

I love this particular graph that really highlights the importance of a multiparametric risk assessment, and we do this in clinic. So the clinical assessment, you talk to a patient, you assess their functional class, you assess how rapidly they've been progressing, you ask about syncope, you examine them, you look for signs of right heart failure. That's all very, very important as we manage our PH patients. Exercise tests are important, and we do a six-minute hall walk in just about every patient at just about every clinic visit. Some centers use more sophisticated exercise testing, such as cardiopulmonary exercise testing. We measure biochemical markers. We draw their blood at every clinic visit. We're checking for volume status or CBC, lights. We're assessing some of the potential side effects of therapies like LFTs. So we do a BNP or NT-proBNP with their blood draw at every clinic visit. And then we do imaging. We don't necessarily do an echo every visit, but we do it relatively frequently, and we look at some of the signs listed there, right atrial area and pericardial effusion. Those are the things in big databases that have been associated with prognosis, but in reality, I think this is one of the limitations of our current system. We don't have an imaging assessment of RV function, and when I go look at my own echoes, I'm looking at that RV function, and that is something that I think is very important in terms of managing a patient. And then of course, we don't do chemodynamics at every clinic visit, but occasionally we repeat chemodynamics, and we look at those very important markers of RV function on hemodynamics, including right atrial pressure, cardiac index, and mixed venous saturation.

So those are the multi-parameter variables. And while we may have many different risk assessment tools, many of them look at the same important variables. REVEAL 2.0 has 14 variables that include both modifiable and non-modifiable variables. We've talked a little bit about the variables included in the French Pulmonary Hypertension Registry and in the COMPERA registry. And here's a list of the ones that overlap in all these registries and are very important in predicting prognosis: functional class, hall walk, biomarkers, cardiac index, right atrial pressure, and SvO2, which tracks very closely with cardiac index. So those are all important markers. Again, those first three, we do it every single clinic visit and the predictive consistency of these variables makes them indispensable in the accurate prediction, no matter which treatment or which risk score, which algorithm you use.

So here's a little comparison of some of the algorithms. Again, REVEAL is more complex, and some of them have as few as three and as many as six variables, and most of these registries were very big. Most of them included different types of pulmonary hypertension. Although the French Registry really was limited to idiopathic anorexigen and, or I should say drug and toxin and heritable PAH. They've all defined low risk in different ways, but they're all very consistent. If you are at low risk, your one-year mortality is very, very low.

Now, as we talk about scoring and the like, we should really focus on some patients in whom screening is appropriate as well. And this was a topic of conversation at the 6th World Symposium. It was also a topic of conversation in the ERS/ESC Guidelines. There are certain patients, certain diseases or diagnoses that predispose to pulmonary hypertension. PH due to interstitial lung disease is something that we look for quite aggressively now, particularly given the availability of treatment for those patients. The most common cause of pulmonary hypertension in general is left ventricular systolic or diastolic dysfunction. Of course, patients with connective tissue diseases should be screened. And of course, patients who've had acute PEs may go on to develop chronic thromboembolic pulmonary hypertension. I think the most clear cut group to think about screening are those patients with the connective tissue diseases, given the

high prevalence of PAH in the scleroderma population. There's some different methods for screening. At Michigan, we use the detect equation, but there are also options for looking at echo, DL, biomarkers and the like, but it's important to screen those patients. These are the patients that I see that have the least advanced disease, and it's really very rewarding and great for the patients.