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Case 1 and Panel- Limitations of Risk Assessment

Announcer:
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Dr. McLaughlin:
And we're going to move into some real-world cases now, and again, if you have questions, please feel free to go ahead and speak out. So, I put together a couple of cases and these are not diagnostic dilemmas. I just want to tell the group about two patients and really get their opinions on risk assessment in these patients. And these two patients were chosen to really review, or really highlight the limitations of some of our objective risk stratification tools. Now, don't get me wrong, I think you should do an objective risk stratification tool no matter what you want to use, if you want to use REVEAL, if you want to use 4-Strata, like, you should do that. Like, I really want to emphasize that. But it's not always perfect and sometimes you need to think about other things.

So, this first patient is a patient I've been seeing for about 5 years. She was really sick when we first met her, and she was started on triple therapy. She is on epoprostenol at a pretty decent dose of 44 ng/kg/min, tadalafil, and macitentan. Her last right heart cath was about a year ago, and you can see there she really has severe pulmonary arterial hypertension, a mean pulmonary artery pressure of 81, right atrial pressure of 7, wedge pressure 14, cardiac index of 3.6, and pulmonary vascular resistant of 16 WU. I have seen her more recently than that and she's about the same, but she's functioning well. She's functional class 1, her hall walk is 617 meters, her BNP is 48. You know, she's a young woman, she's otherwise healthy. She's an idiopath, she doesn't have any other comorbidities, so despite the severity of her pulmonary hypertension, she's functioning well. And she technically calculates to low risk using both 4-Strata and REVEAL Lite 2. But I showed you her PVR is 16.

This is her most recent echo. She still has a big right ventricle, it's not dead, but you know, obviously she's got some dysfunction of the right ventricle and you know, that's about as much pressure overload that you're going to see on a short axis. So, she's got a really ugly-looking right ventricle.

So, in this patient, you know, you look at the guidelines and you say she's low-risk, I don't need to do anything else. She's already on triple therapy. The only other else thing to do for her would be transplant and you're not going to do a transplant of lung on someone who's functional class 1 and walks 600 meters. But I don't want to be under any delusion that this patient is low-risk and I'm not worried about this patient. This is the sort of patient that actually keeps me up at night. So, here's an example of, despite being low risk, I'm still worried about this patient.

And they exist in the opposite direction, too.

So, this is another one of my patients. She is 75. I have been following her for over a decade, over a decade. She has scleroderma. We actually caught her early, you know, we have an aggressive screening program. When we first diagnosed her, her mean PA pressure was 33, her wedge pressure was 11, her cardiac index was 3.1, her PVR was 3.2. So, you know, even with the old definition, just barely met criteria. Put her on a PDE5 monotherapy. Her most recent cath was a number of years ago. Her PVR was 2.8. But when I see her in the office, she's functional class 3, you know, she gets short of breath with stairs, she can't do as much as she thinks she should do.

Her hall walk is 244, you know, a pretty poor hall walk. BNP is a little elevated. And she falls into the intermediate high-risk category using the 4 strata technique, and the intermediate risk category using REVEAL. So, gosh, this patient's not at low risk, they're not at goal. Do we need to do something more?

And this is her echo. She got a nice happy right ventricle, right? Her right ventricle is normal in size and function. She's getting a little old, right? Her left atrium is big, her interatrial septum is bowing left to right, and in the short axis, her right ventricle is just this little sliver around her left ventricle, the septum still belongs to the left ventricle. So, even though she falls into intermediate high by 4-Strata, and intermediate by REVEAL, I look at this echo and I'm not worried about pulmonary vascular disease. I feel that her scleroderma, her age, maybe she's starting to develop some diastolic heart failure, deconditioning is why her functional class is poor and her hall walk is poor, and she falls into those risk categories. But if someone were to look at our database they would say, you're not optimally treating this patient. She's not at low risk. But she doesn't worry me. I think I would just cause her more harm than good by starting another therapy.

So, those are the two cases that I wanted to go through, you know, to just give you some examples of the risk stratification tools that we have, and then open it up for discussion from my colleagues here about how they manage this as they see patients in clinic.

Dr. Moles:
So, I'll start first, right. This happens not uncommonly unfortunately, and I think this is a little bit of the reality of what we see when we follow patients long-term. I mean, they may have a diagnosis when we meet them, but then when things progress, they may develop other diagnoses. I mean, the scleroderma population, I think that diastolic dysfunction, heart failure with preserved ejection fraction is a common diagnosis. My suspicion is that if we put that last patient that you talked about on an exercise heart cath, we will see that wedge pressure go up pretty significantly with exercise.

How to manage them is a little bit more difficult because we – and to me it represents a diagnostic dilemma because we're calling them PAH, but at the same time we need to acknowledge that they may have other comorbidities. So, I think this patient would be appropriate to exercise on a right heart cath and with a bike and see what that shows.

The first patient that you showed, I think would be a great patient for a clinical trial.

Dr. McLaughlin:
She overwalks everything. Her hall walk is over 600. She can't. We've tried. She's functional class 1 with a hall walk of over 600, she doesn't qualify for clinical trials. She's considered too well.

Dr. Moles:
That's true, that's true.

Dr. Cuttica:
But in theory I agree with you. That's exactly what I was thinking, and I was like, no, she would never get in. Her walk test is just too good.

Dr. Moles:
Yeah, but apart from – you know, I agree with you, I would be very concerned about that patient apart from it I'd up-titrate a little bit epoprostenol, which you are going to be limited by the cardiac index being in the high end of normal. There's not much that we can do at this point. So, I agree with the dilemma and I'm not sure that I would do anything differently than you're doing.

Dr. Cuttica:
I think this, to me, you look at this and it makes me think of two things, one, I think the risk stratification is a tool that informs the discussion with the patient, it doesn't make the decision about what to do with the patient. Right? Because you can't completely replace Val McLaughlin's clinical acumen in making decisions about how to manage patients, but I think the power of the risk stratification tools is to make you think about these things in the clinic that you take all of the parameters together and say, do I really think this person is low, intermediate, or high risk based on my clinical experience.

And I think for that first case, what it really makes me think of, is stability is a relative concept, right? There is no really, as a pulmonary vascular person, there is no such thing as really a stable PAH patient, right? These are walking critically ill patients and you have to be very careful with them all of the time. And the disconnect between her echocardiogram and her functional status is really striking in that. And I agree, I'd be super worried about that lady because ultimately, we all talk to our patients, and when we talk to trainees, right, ultimately the goal of treating a PAH patient, all of our parameters, everything we measure, walk test, serologic studies, they're all about improving the function of the right side of the heart. And you look at that lady and you say, she's in terrible shape, we are not accomplishing what we need. But clinically she's doing okay. So, I agree, I'd be very worried about her. It wouldn't seem like it would take much to tip her over the edge and have her fall off the cliff, but what to do next is hard. Yeah, try and go up a little bit on the Flolan,

maybe switch around some of the oral agents, probably with minimal benefit to that kind approach. But I'd be worried about her, and I'd want to keep a close eye on her.

Dr. McLaughlin:

Yeah, so, you know, with a PVR of 16 on a pump, even though she is functioning well, I would strongly advise against that. You know, we have gotten a handful of patients off pumps. You know, I think that these days it's harder to get people off pumps because most patients these days go on pumps after they have failed oral therapy. So, to think that after they failed oral therapy and you put them on a pump and they're better, that they're going to still be fine on oral therapy, I think is unlikely. You know, over the years we've had a handful of patients that have come to us critically ill that we've started on upfront triple therapy, including a parenteral prostacyclin, and essentially normalized their right ventricle. And sometimes we discuss withdrawal of the pump in those patients, and I have a couple we've done that on. I have a couple – like I can see her right now. She came to us postpartum, like, she was practically dead. Like, you know, and she has the most normal right ventricle now and whenever I talk to her about it, she's like, nope, nope, I'm just going to stay on this. I'm winning the PAH ballgame. I'm just going to stay on this.

So, like, that lady with that heart, I don't think it's going to happen. I think we're going to have to face this question a lot once sotatercept becomes available. You know, Victor and I have put a lot of patients on sotatercept in the clinical trials. We've seen a fair number that have had pretty nice improvements in their pulmonary vascular resistance. And if you look at some of the papers that have been published, like the most recent paper that was published in the ERJ that really was a deep dive into the hemodynamics and echo changes in the STELLAR trial, you'll see a handful of super responders who almost normalized their PVR and, you know, maybe some of those patients we can talk about going down on their pumps. I would never make a promise about it, but there may be a handful of those. But this lady, with where she is now, I don't even bring it up.

Dr. Patel:

I will say as someone who reads a lot of Mike's patient's echos and Yasmin's patient's echos, my heart always skips a beat when I see, Cuttica is the referring, or Raza is the referring, because we see this echos like case 1 a lot, and you just don't know when that RV is at a point where it is going to fall off a cliff, and it was still kind of bringing it in the echo that you showed, and the base is still moving towards the septum. Parameters might be kind of within the limits of normal, but it's extremely hard to know the trajectory of, you know, clinical RV failure in these patients, and we just don't have great noninvasive techniques that can help us predict when a patient is going to fall off of a cliff. Now, there are some newer techniques available, but whether they are truly predictive is less clear. But I agree, I read a lot of these echos on a daily basis and I, you know, worry about that RV. You know, one minute they're, you know, kind of ok. And it's a syndrome in isolation here, as opposed to the second patient, which is kind of almost death by a thousand cuts as one ages. You know, it sounds like the first patient is, this is her problem and really that's the primary problem, whereas in your second patient, there is the issue of aging, comorbidity accrual, kind of death by a thousand cuts. And so, things are going to add up. That first patient will eventually get there, it's just not there now and when that is, is hard to know in the longitudinal. Obviously, care is going to be extraordinarily important to keep a close eye.

Dr. McLaughlin:

Yeah, I think these are all good points. And, you know, my main reason for showing you these two cases is to, you know, as much as risk stratification is important and we should all do it, risk stratification is just part of the puzzle in taking care of these patients. And sometimes I refer to it as the blender technique that you've got to get a lot of things, and risk stratification is one of them. But sometimes echo is another, and sometimes right heart cath is another, and sometimes all these comorbidities is another, and sometimes it's patient goals and quality of life is another. You've got to get all those things and put them in the blender and then come up with a treatment plan using shared decision-making for the patient.

So, I think that was a great discussion. Thank you, guys.

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