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PH Management in Unique Patients and Conditions

Announcer:
Welcome to CME on ReachMD. This activity, titled “PH Management in Unique Patients and Conditions, “is provided by Total CME, LLC and is supported by Actelion Pharmaceuticals US, Inc., a Janssen Pharmaceutical Company of Johnson & Johnson.

This replay of a live broadcast addresses challenges in the management of pulmonary hypertension in unique patient populations and patients with complex conditions.

Dr. Preston:
The management of pulmonary hypertension in unique patient populations and complex conditions requires addressing certain challenges. Today, we will be going to present the case of a patient with comorbidities, as well as psychosocial challenges like many of the patients that we see in our practices. We hope to provide some guidance and resources to help manage these unique patient situations.

This is CME on ReachMD and I'm Dr. Ioana Preston.

Dr. Elwing:
I'm Dr. Jean Elwing.

Martha Kingman:
And I'm Martha Kingman.

Dr. Preston:
We have a lot to discuss today, so let's begin. I'm going to turn it over to Martha. Martha, would you please present the patient case starting with the diagnostic process and then later we can update the case as situations evolve.

Martha Kingman:
Okay, thank you, I'm happy to. Our case today is a 67-year-old female with scleroderma and also mild hypertension. She's got a 6-month history of shortness of breath on exertion that has been worsening lately.

She went to see her primary care doctor who referred her to a pulmonologist, and the pulmonologist, appropriately, ordered pulmonary function testing and an echocardiogram. The PFTs were normal, unremarkable. The echocardiogram, however, showed some mild right ventricular enlargement and mild right ventricular dysfunction. The RVSP was estimated at 45 mmHg and the LA was mildly enlarged.

The pulmonologist placed a referral to the pulmonary hypertension expert center, who then completed some additional testing. They did a right heart catheterization, which revealed a mean pulmonary artery pressure of 32, a pulmonary capillary wedge pressure of 15, cardiac output of 4 L, cardiac index was 2.0, and her pulmonary vascular resistance calculated out to 4.25 Wood units.

Her Functional Class was II and on REVEAL Lite risk assessment she was low risk. Her NT-proBNP was 600. Her 6-minute walk distance was 370 m. Vital signs: blood pressure was 115/67, her heart rate was 87, her creatinine is 0.6, and her GFR was greater than 60.

Dr. Preston:
So, Martha, tell me, what do you think about her REVEAL score?

Martha Kingman:
So her REVEAL risk score turned out to be 5, as you can see here on the calculator, putting her in the low-risk category.

Dr. Preston:
Jean, can you tell us what do you take into account when deciding on how sick this patient is, and what do you look at?

Dr. Elwing:
Well, I think as Martha presented, she presented a lot of data points to help us understand where the patient was, where they're going, and how they feel right now. So I think we have to take all of that into account, and I'm sure you guys do too when you look at a patient.

And we take everything into account when we assess the severity of illness in our patients. We do look at that risk score because that helps us understand her likelihood of worsening and mortality over the short term in patients like her that we've seen in registries. But we have to look at her individually also. We have to look at the echocardiogram and the impairment of the right heart size and function related to her pulmonary hypertension. And she does have some mild right ventricle enlargement and decreased function, so something we have to be mindful of and watch over time.

And we also want to know about her comorbidities because we have to manage those to help her do as well as possible with our therapies and in light of those other comorbidities we may encounter as we treat her. And of course, we want to get additional testing to see if there's anything else that's contributing to her shortness of breath.

And Martha did a great job telling us a lot about this patient, so it's very helpful to get a picture of how she is doing right now in terms of her functionality and in terms of her hemodynamics also. So we really look at things globally, including all of those parameters. So when I see this patient, I see a lady who has scleroderma, so I know she's at higher risk of worsening than some of our other PAH patients, so I'm going to watch her very carefully.

But I do have, still, some questions about other things that could be contributing to how she's doing and how she's feeling, so I do want to ask Martha about did she get a V/Q? Did she have any additional labs that may help me understand more about how she is doing?

Dr. Preston:
Okay, Martha, what do you tell us?

Martha Kingman:
Okay. So the story continues at the pulmonary hypertension center. There was a V/Q scan that was done and was negative, so that ruled out CTEPH, so we don't have to worry about CTEPH anymore. She had her overnight oximetry and did not need any oxygen.

We repeated the NT-proBNP and it was 580, so about the same as it was, and the pulmonary hypertension center decided to start her on combination up-front therapy because she did not fall into the high-risk category. So she was started on a PDE5 and an ERA.

The PDE5 was started first and they chose sildenafil, which she tolerated well. And then, she started the ERA and did develop some swelling, some lower extremity edema, a few days after starting it.

Dr. Preston:
And, Martha, swelling after starting an ERA is not rare, and we have to think about how to manage this. Jean, can you tell us your thoughts on managing a lower extremity edema in this setting?

Dr. Elwing:
Well, first I agree with you. It is not rare, and I think, because of that, the first thing I try to do is counsel patients so they're not surprised, they don't get worried about that developing. And I'm sure you guys also do that, warn so that people know that it might happen. And we are very tempted, when we have an adverse symptom, to pull away a medication, but I would avoid doing that in this individual. We know that this combination of PDE5s and ERAs is effective in our patients with PAH and so we don't want to withdraw an effective medication because of this side effect. So I'd rather try to mitigate this by starting some diuretics.

But in doing so, we have to make sure we're watching her labs and watching her electrolytes so we don't get into other issues with that, and then we'll follow that over time. We'll follow her edema, we’ll follow her weight, and we’ll follow her NT-proBNP to make sure that that is all going in the right direction. And we know she might be a little bit more at risk for swelling because she did have some enlargement of her left atrium. And we had some concern for possibly some diastolic dysfunction when we initially evaluated her, looking at her echocardiogram. So this might be someone we could even consider starting a diuretic even before we start the ERA, preemptively. So that's something we could consider.

Dr. Preston:
I agree, Jean. We can usually manage fluid retention, and ERAs are an important component of this patient's plan. This is a great example of using shared decision-making with a patient where we can explain that the benefits likely outweigh the risks of side effects, such as edema, and that we can manage this together.

Martha, any other thoughts about this case?

Martha Kingman:
I would just add a little bit about patient education and would make sure that we talk to the patient about sodium restriction. She does maybe have some diastolic heart failure with her LA enlargement and her history of hypertension, so I'd make sure she's on a sodium-restricted diet; 2 g is generally what we recommend. And then I would talk to her about weighing herself every day, writing it down, doing it in the morning. Use the restroom and then weigh yourself, write it down, and then give her some parameters of when to call the clinic. So that if the swelling does continue to get worse despite our increase in our diuretics, then we would catch it earlier and be able to act on that.

Dr. Preston:
Yes, thank you, Martha. And I think these measures are very important, especially at the start of the new medication. And once we find a normal maintenance dose of a diuretic and the patient starts learning how much sodium she can afford and she can eat, or he, I think it's very important for long-term tolerability and adherence to the drug.

So we agree we can manage this comorbid condition of mild heart failure with preserved ejection fraction in this truly Group 1 PAH patient and continue the endothelin receptor antagonist.

Martha, we have managed her swelling and she comes back for follow-up. Can you present the next part of her case study?

Martha Kingman:
Sure, happy to. So she comes back to clinic after about 3 months, which is pretty typical time frame. And unfortunately, she's not doing as well. She has worsening shortness of breath on exertion. She's noticed when she's climbing up the stairs in her home that she's feeling winded now, and on physical exam she does appear to be euvolemic after her diuretic, so we are not thinking that it's really a fluid retention problem going on.

Dr. Preston:
So, Jean, what would you do to assess this patient for worsening pulmonary hypertension?

Dr. Elwing:
Thank you for that very good question, because I think it's really important to understand what is driving her worsening. Is it pulmonary arterial hypertension or is it something else? So I think we need to go back to the parameters we looked at initially and then compare. And I would start with some basic labs, make sure we're not missing something like anemia. Look at her NT-proBNP. Is it rising? Is it out of proportion to what we expect in this individual? I would repeat her echo. I want to know what her right heart is doing. I want to know if there's any new parameters, like a pericardial effusion. And look at her walk distance. Is it the same? Is it lower? Is she now needing supplemental oxygen? Assess her functionality with functional class. Put all of those parameters into risk assessment, and if we think this is worsening pulmonary arterial hypertension, I would do a right heart cath to really understand what's going on hemodynamically.

Dr. Preston:
Yeah, that's a great reassessment. And what did this repeat evaluation show?

Dr. Elwing:
So unfortunately for her, it showed worsening of her pulmonary hypertension, based on all of those parameters. So when we looked at her data, we found that her NT-proBNP had gone up from 600 at her time of diagnosis to 1,000. Her walk distance had gone down from 370 to 315. She's now Functional Class III. Her blood pressure was a little bit lower, which as I'm sure you guys are feeling also, that's a very worrisome sign for a pulmonary hypertension patient. And then, of course, we did the right heart cath and that tracked with worsening hemodynamics also. The mean pulmonary pressure had increased to 38, the cardiac output had gone down to 3.8, and now the index is less than 2, so a concerning sign. And her pulmonary vascular resistance, 6.8. So all of the things on hemodynamics that would suggest pulmonary vascular disease have progressed. And we also got an echo which now went from mild RV dysfunction to moderate, and our TAPSE is lower and we have some pericardial effusion. So all of the parameters making us concerned that we need to take action.

Dr. Preston:
I was going to add that it's amazing that this worsening happened within 3 months, so this signifies a rapidly progressive disease.

Dr. Elwing:
Very true. And I think that is something we can never get too complacent about when we meet a patient at low risk.

Dr. Preston:
That's why it’s important to follow her very closely. So, Martha, what's her current risk?

Martha Kingman:
Well, this slide shows the 4-strata model from the ERS/ESC guidelines from 2022, and this is recommended to be used at follow-up. Where previously it was 3 stratas, now it's 4, and that is because they cut the intermediate risk into intermediate-low and intermediate-high.

So if you look at the chart here, you can see that now her parameters, where the arrows are, are all falling into the intermediate-high risk. She's Functional Class III, her walk distance fits in that category, as does her NT-ProBNP. So she seems to have moved from a lower-risk patient to now an intermediate-high-risk patient.

Dr. Preston:
So, Martha and Jean, we have a CTD PAH patient with evidence of worsening pulmonary hypertension on dual oral therapy. What additional therapy options may we take for this patient? Martha?

Martha Kingman:
Yes, thank you. There's some options. Generally, when we see someone getting this much worse and they're getting more toward the high-risk category, we definitely would consider parenteral therapy, parenteral prostacyclin, at this point. And we could talk to the patient about IV prostacyclin or subcutaneous prostacyclin. Definitely a conversation with shared decision-making because there are some options here and we have to talk about the risks and benefits of all of them. We can talk about the long-term data and really just see what makes sense in her personal situation.

Dr. Preston:
For those just tuning in, you are listening to CME on ReachMD. I’m Dr. Ioana Preston, and here with me today are Dr. Jean Elwing and Martha Kingman. We are discussing the effective management of pulmonary hypertension in unique patient populations and in those with complex conditions.

So moving more towards an individualized medicine approach. So, Jean, can you walk us quickly through the PAH treatment algorithm for a patient who's already on background therapy, and what are our options?

Dr. Elwing:
Oh, yes, I'd be happy to. So we're going to look at the 7th World Symposium treatment algorithm now. And we know initially she was in the not high-risk category that Martha told us, and she received recommended dual up-front combination therapy. And now we reassessed her and she was at the intermediate-high risk. So we have options of either adding IV or subcutaneous prostanoids, or we could also consider an activin-signaling inhibitor. And so we really have 2 options based on these guidelines, and we really need to counsel her, understand, and take into account how fast her disease is progressing to make the best decision for her.

Dr. Preston:
So, Jean, what about sotatercept, the activin-signaling inhibitor which is one of our 2 options in intermediate-high-risk patients or high-risk patients? Would you consider adding that first or after the prostacyclin or at the same time?

Dr. Elwing:
So I don't think there's a right answer here, but in her case, she is really worrisome to me because she's progressing so quickly. She worsened in almost every parameter that we looked at on her follow-up, so I would opt for the parenteral prostacyclin if she's agreeable, and I would do that based on the data we've had over the last 2 decades, that we know our sickest patients are oftentimes best responsive to that parenteral prostacyclin. And then I would consider adding on the sotatercept after we’ve completed the up-titration.

So let's talk about treatment, and we're going to look in depth at the 7th World Symposium treatment algorithm. The whole thing relies on patients being assessed thoroughly and being risk assessed. And when you meet someone, you have to evaluate, are they at high risk or are they not at high risk? So it's a little bit simpler than it was in the past, just either high risk or not, and our patient, of course, was not high risk. And then based on that, we choose treatment. And most of our patients who are in that not high-risk group are going to be treated with an endothelin receptor antagonist and a PDE5. And hopefully, most tolerate that combination, and then we go on and reevaluate.

But for those patients who are at high risk on initial assessment, they need a combination of 3 medications. They need that PDE5, the ERA, but also, we recommend a prostacyclin because those patients are our sickest patients that we meet. We evaluate and we find them at high risk, we want to treat them as aggressively as possible.

And one caveat is that when you look at the treatment algorithm key points from the 7th World Symposium, they really tell you that this is based on our previous studies looking at patients who have a mean pulmonary artery pressure of 25 or greater and a PVR of greater than 3. But you say, hey, why? You just told me this person had pulmonary arterial hypertension because the mean was greater than 20, but that's because all of our studies have been done, to date, based on those parameters. So just a little bit of some of the things I want to make you aware of.

The other thing I think is really important is 3 to 4 months, or earlier if your patient is having problems, you want to reassess your patient. And it's not just saying, hi, how are you doing? Are you tolerating your meds? It's really doing a risk assessment. And then we're going to try to sort out, are they really at low risk or are they at intermediate-low, intermediate-high, or high risk? And what you do will depend on if they are in that low-risk group because we can continue our therapy and keep watching them if they're truly staying at that low-risk status. But if they're anything else, we need to do more. This is not a disease that remains stable. It will get worse if we don't try to control it as well as possible.

So if you're at intermediate-low risk, you might look pretty good, but you are not where we want you to be, so then we'd offer additional therapy or change in therapy. We could add an activin-signaling inhibitor like our newly approved drug, sotatercept, or we could continue what we're on and consider a switch of the PDE5 to a guanylate cyclase stimulator. So we have some options.

Or we could consider the addition of an oral or inhaled prostacyclin. So that population has a lot of options, and I think, really, we have to use our shared decision-making, try to sort out do they want another drug or would they rather switch, or would they like to add a pill or an inhaler? All of our patients have different experiences in their lives and they may appreciate a different option than another, so we really have to ask important questions when choosing our therapies.

Now, if you're in the intermediate-high or high risk, that’s when we’re thinking about our prostacyclins and, also, we're thinking about our activin-signaling inhibitors. And for our high-risk patients, the answer always is offer prostacyclins, the parenteral prostacyclins. But we also have to consider that not every patient will accept that, and so we do now have something to consider for them, which is sotatercept.

So just giving you this global view of our options for treatment. It is a little complicated; I understand that. However, if you look at this, the backbone is, see the patient, treat the patient, reassess risk, and then add therapy or change therapy based on what you find. And in those patients who stay high risk, either intermediate-high or high risk, we need to think about maximizing our therapies, evaluating for lung transplant, or even thinking about palliative care if we are not going in a positive direction or if the patient does not want to proceed with aggressive measures. So something to chew on a little bit and think about when you're seeing your patients and following them over time.

Dr. Preston:
And how do you talk to patients about prostacyclin side effects, especially in the up-titration period, and the management of these?

Dr. Elwing:
All very good questions because I think that that may be the most critical thing, is that we warn people, we walk through the whole process with them, and we're by their side. As Martha mentioned earlier about salt and fluid, really guiding them through how to manage these adverse effects we create with our medications that are so necessary to be on for their pulmonary hypertension. So I really try to talk about the different things that may or may not happen to them. But I tell them, you may get all or none of these side effects. And then have a toolbox for them to address them as they start experiencing them. Have on hand some acetaminophen for leg aches and some medication like ondansetron or another antiemetic for nausea and vomiting. And of course, have some loperamide available for the loose stools they may experience. And make sure they understand that these are things that can happen at any time we're up-titrating and that we will go back down on their drug, slow down the titration, so they can tolerate it.

One thing I think we have to always mention is the jaw pain they may experience on first bite of food. We don't usually give them medications for that, but I always warn them because otherwise they think something very strange is going on or they're experiencing very odd side effects. So really warning them, trying to help them understand this. And letting them know their skin may change; they may flush and it may not be an even flush. They may be kind of blotchy or patchy, so they don't think there's something odd going on or they have an allergic reaction, because that's a normal side effect of the parenteral prostacyclins.

Dr. Preston:
Yes. And these are all signs that are more prominent as we up-titrate the dose, and then, during this time, the patients learn the presentation of these side signs of prostacyclin treatment and they become more confident as the time goes.

So, Martha, what are some additional considerations when talking to patients about IV therapy or parenteral therapy, in general?

Martha Kingman:
Yes, there's so many things to educate the patient about when they're going to start on parenteral therapy, whether that's IV or subcutaneous. Now, this patient has connective tissue disease, and sometimes those patients can have dexterity issues because they have sclerodactyly and can have trouble manipulating the pump. So that's one thing to kind of take into account and assess if they're going to be able to do that or if they have a family member who will be able to help them do that. I always like to get the family members involved, the caregivers, and make sure that they understand what this is going to look like and how they can be helpful.

I would want to know if there's any insurance or payment barriers that they may have. This is a big lifestyle change for patients, so they go through a lot emotionally, as well. I like to send patients or at least recommend that they go to the support group because many patients can benefit from just being around other people that have been on IV or subcutaneous therapy. It seems like it really helps that they're not alone in this and they have someone that they can talk to.

And last point I would make is this is a good time, if you haven't already, to talk about palliative care so we could get them on board to help with some symptom management and some planning for the future decisions that they want to make going forward.

Dr. Preston:
That's great, Martha.

So let's check on our patient. How are things going at her next follow-up visit?

Martha Kingman:
Well, she comes back to clinic, and as we very often see, there's some challenges with taking her medications correctly and appropriately. She admitted to us that she was not always getting her third dose of sildenafil every day, and this is a really important point to make is to not assume that patients are taking their medications appropriately or regularly. You have to ask them. What I found, if you ask patients in a very nonthreatening way, they’re going to be honest with you. And I will just kind of easily say, or quickly say, all right, if you're on sildenafil 3 times a day, how many pills do you miss in a typical week? And we often hear 5, 7, 10. So clearly that's a problem. And I'll say, I really appreciate you being honest about it. Let's see what we can do.

And then dig in a little more into why is that happening. Are they missing the dose because they just forget about it, they're going on with their day? Usually, people are good about taking it in the morning and at bedtime, but in the middle of the day they get busy and forget about it is what usually happens. In that case, if the patient's going to continue on sildenafil, I would say to set a reminder on your phone, and it's just super important to do that because if you're not taking it 3 times a day, you're not really getting the full benefit that we want you to get. Sometimes I'll suggest to patients that they take an extra bottle of pills with them in their purse so if they're out and about all day, the alarm goes off and they've got it with them. Just simple things like that.

Now, if the patient tells me that, “Oh, the sildenafil, yeah, I quit that a few months ago or a couple months ago” – we've all seen that, right? They show up and they're not on all their medications, then you got to kind of find out why. Is that because there's a cost issue? Is their copayment too high? There are, if patients are commercially insured, there's copay assistance programs that limit the cost. So if they're not already taking advantage of that, we can try to help them with that. Maybe they can talk to the pharmacist or the social worker, bring some additional team members in, try to get to some solutions if it's a cost issue.

Sometimes people will quit taking their medications because they have a side effect. It just makes them feel bad, so they quit taking it. Maybe, I don't know, one of the ERAs causes some nasal congestion sometimes and people say, “Oh, yeah, I couldn't do it. I couldn't stand the nasal congestion, so I stopped taking that a month ago.” So we can look at other options, what we can do about nasal congestion. And it's a good time to remind the patient we understand sometimes things come up; it's difficult. Let us know right away because we don't want you to be off your medication for 1 or 2 months because during that time, the pulmonary hypertension can get worse, and we don't want that to happen when we have some things that we could do to help you and get you back on a therapy that you can tolerate.

Sometimes patients are completely uninsured, and that's back to the cost part of this, and there are patient assistance programs for patients who aren't insured. So if they're not insured or they lose their insurance, then we need to make sure we get them connected to the pharmaceutical company's patient assistance programs.

Dr. Preston:
Okay. All right. So we have some adherence problems here. So taking a medication 3 times a day is challenging for most people.

Jean, what would you recommend?

Dr. Elwing:
Oh, I would assess if there's just forgetfulness or is it a cost issue, or is it that she really has some adverse effect of the sildenafil? Is she getting acid reflux, or is she getting nasal stuffiness that she's really trying to avoid it because of that? And if she's tolerating it and doing well with the medication, it's just forgetfulness on that midday dose because she's busy and there's a lot of things going on, then I would really talk to her about the possibility of making a switch to a simpler medication for her. And we learned recently with the A-DUE study that there is a combination therapy that is effective and can be used in our PAH patients, our first combination therapy. So they studied, in the A-DUE study, macitentan versus tadalafil versus the combination. And they saw in the short term, 16 weeks, looking at PVR, which can have a significant reduction in PVR when we use the fixed-dose combination as compared to the single monotherapy drugs. And this was recently brought to market and now is available to our patients and has allowed them an easier way to take that ERA and PDE5. So I would offer her that, and if insurance allows, that might be a nice way to reduce her pill burden and hopefully improve adherence.

Dr. Preston:
So, Martha, what may be some benefits regarding the combination tablet?

Martha Kingman:
I think there's several. Really, in this patient's situation, it would reduce her pill burden from 4 down to 1, so that's really nice for the patient. It gets rid of that whole 3-times-a-day dosing that is often difficult to comply with. It's one less copay for the patient. So now instead of an ERA copay and a PDE5, they have one copay so that can save them some money. And then, it's also one less drug that the patient has to remember to get refilled. So they have to know they get their medications refilled, so that's one less thing to worry about. And then from the nursing standpoint, it's one less prior authorization for the nurses to have to complete. So kind of a win-win for everybody really.

Dr. Preston:
Absolutely, absolutely. And especially for the patient.

So before we wrap up, can you tell us what a typical shared decision-making discussion would be like when adding sotatercept, Jean?

Dr. Elwing:
Oh, thank you, very good question. And I think that we have to balance what we know about sotatercept and its outcomes in terms of clinical trials, what the patient wants, and what the risks of the drugs are. And I think we have to be really up front about the fact that the patient has to do labs regularly because we're watching for changes in hemoglobin, changes in platelets that we know will likely occur for that patient. And we also have to counsel them about the real risk of bleeding, especially epistaxis, and make sure they understand that those things may happen and we will watch them very closely, but they have to be on board with all that it takes to safely use this medication, especially those labs that they'll repeat every 3 weeks before their dose for at least 5 or 6 times. And then we can potentially stretch that out and space them to less frequently over time once they become stable. But I think we really have to go into it explaining what the benefits of the medications are but also what are the key things they need to do to use it safely.

Dr. Preston:
And lastly, I think we all can tap into the palliative care programs, which doesn't mean that we're giving up on our patients, but they're a wonderful resource that are nurse-staffed that are sometimes the most amazing nurses in palliative care.

They can help patients with advanced diseases, such as advanced pulmonary hypertension, into dealing with the everyday activities and hurdles that these patients encounter.

Martha Kingman:
I agree. Absolutely.

Dr. Elwing:
I agree, too. I do have a question for both of you. This is so stressful for patients. How do you integrate mental health care in our expanse of things we have to do to maintain patients' health in terms of their pulmonary hypertension? Do you talk about it at each visit? How do you do it?

Martha Kingman:

I have to admit that I think I have fallen short in serving the patients really well here, because like you mentioned, you have 20 or 30 minutes and we're trying to talk about the disease process and the medications and the side effects and all of these things and go over the echocardiogram that sometimes, unfortunately, it doesn't rise up to the top of the list of our priorities. I'm sure it's a patient's priority. I do, again, not often as I should, but I generally find that if you ask the patients if they're depressed, they will tell you if they're depressed or not. And I will often preface it with, the studies in pulmonary hypertension have shown that up to 50% of patients with PAH understandably suffer from depression or anxiety. Is any of that going on with you? Because if so, I want to make sure we can get you some help.

Dr. Preston:
And what I find is that the family support is the most important tool to anchor these patients and help them in the long run. But families sometimes are difficult, right? So they may not be there for our patients all the time. So going back to the primary care physician and discussing pharmacological options for clinically depressed patients is very important. And then, of course, referring them. Although, there is a shortage of counselors and psychiatrists around the country, so we have to take that into account. But working closely with the primary provider to screen for depression and treat depression when it's there is very important, Jean, absolutely.

Dr. Elwing:
Yeah, I find it's a comorbidity, because we're talking about comorbidities, that oftentimes just doesn't come to the forefront, and I myself have to work harder on doing that also.

Dr. Preston:
As we all are.

Martha Kingman::

Again, I think that getting the patients, or at least referring them, into the patient support groups can be very helpful as an adjunct to other antidepressant medications or therapy that they may be getting.

Dr. Preston:
Yeah.

Dr. Elwing:
Yeah, definitely.

Dr. Preston:
So this case illustrates some of the key challenges that patients with pulmonary hypertension can face. Key learnings and takeaways include the importance of a complete diagnostic workup, including the use of risk assessment to guide therapy decisions. Patients need to be followed closely and therapy escalated when there is worsening pulmonary hypertension. A new combination of an ERA/PDE5 inhibitor tablet is now available, which may offer some benefits, including improved adherence. A proactive side effect management plan is important when studying prostacyclin therapy and, in reality, any advanced therapy in pulmonary hypertension.

Well, this has certainly been a fascinating conversation, and before we wrap up, Jean and Martha, what are your final take-home messages? Jean?

Dr. Elwing:
Okay. I think my take-home message is Martha presented a very common patient for us to see. Someone with a little bit of mixed-up disease at the beginning.

Do they have diastolic dysfunction? How severe is it? But true pulmonary arterial hypertension that needs management. But we can't be complacent because they're low risk; we have to watch them carefully. And we have to react if they're getting worse because if we would have waited 6 or 9 months on Martha's patient, that patient may have been in the ICU in severe right heart failure before we figured out what was going on. So I think this just is a great case to tell us that these patients require very close monitoring, and despite doing everything right, patients may get worse and then we need to aggressively manage the worsening.

So thank you, Martha, for giving us that case.

Dr. Preston:
Martha?

Martha Kingman:
Yes, I agree with everything Jean said. All great take-home messages. I would just add a couple of things. One is that we do have an evidence-based guideline from the 7th World Symposium available, so it's important that that's followed, as it was in this case. And then

what we talked about early on, the patient is put on an ERA and gets swelling, some fluid retention. We see that all the time and definitely our first thought is not to stop the ERA. We want to work through it and try to keep the patient on that really important category of medications and usually can do that by adjusting diuretics.

And then, as Jean talked about, again, that being proactive with the side effect management plan for the patients that are going to go on a parenteral prostacyclin so they can kind of be armed with their tool kit at home and be able to manage some of these side effects before they become a problem.

Dr. Preston:
So that's all the time we have today, and I want to thank our audience for listening in. And I also want to thank Dr. Jean Elwing and Martha Kingman for joining me and for sharing all of your valuable insights and expertise. It was great speaking with you today.

Dr. Elwing:
Ioana, thank you for having us. This was so much fun, and I was so glad we could discuss this case.

Martha Kingman:
I agree. Thank you so much for allowing me to be part of this. And good-bye.

Announcer:
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