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Who is at Risk for Pulmonary Arterial Hypertension (PAH)?

Narrator:

Welcome to **Medical Breakthroughs** from Penn Medicine, advancing medicine through precision diagnostics and novel therapies.

Dr. Caudle:

This is ReachMD and I am your host, Dr. Jennifer Caudle. Joining me today is Dr. Akaya Smith who is the Medical Director for the Pulmonary Hypertension Program and an Assistant Professor of Medicine at the Hospital of the University of Pennsylvania. We will be discussing pulmonary arterial hypertension. Dr. Smith, welcome to the program.

Dr. Smith:

Thanks so much for having me.

Dr. Caudle:

What is pulmonary arterial hypertension and who is at risk for developing it?

Dr. Smith:

Pulmonary arterial hypertension is actually a quite rare disease, occurring in only about 10 in a million people. It's marked by stiffening and narrowing of the pulmonary arteries and because these vascular changes there is a strain on the right heart and ultimately these patients may develop right heart failure as the disease progresses. About half the patients in this country have idiopathic disease and we really just don't know why they get it. Connective tissue disease, especially scleroderma, congenital heart disease, especially ASDs and VSDs, really comprise the next largest group. Finally, HIV and liver disease, as well as some drug use, can increase the risk of pulmonary arterial hypertension. The classic drug associated with pulmonary arterial hypertension is the anorexiant, fen-phen, but that's now off the market and we're now seeing more and more disease related to methamphetamine abuse. The people who get it, much more common in women than in men, and often in the prime of life, occurring in the 30s and the 40s. There is an earlier peak in childhood often related to congenital heart disease and then a peak later in life, often related to liver disease or connective tissue disease.

Dr. Caudle:

Okay great. That's very interesting. What do practitioners need to know about diagnosing pulmonary arterial hypertension?

Dr. Smith:

So, that's a great question. Pulmonary arterial hypertension, PAH for short, actually falls under the larger umbrella disease of pulmonary hypertension. Pulmonary hypertension just means there is high pressure on the right side of the heart, but really doesn't tell you why you have it, and it's most commonly caused by left heart disease. Lung diseases such as COPD and interstitial lung disease are the second leading cause of pulmonary hypertension. Pulmonary arterial hypertension is really rare and one of the rarest causes of pulmonary hypertension, but I consider it a must-not-miss diagnosis. Though it's rare, it's progressive, it's deadly, and therapies can significantly change prognosis. Unfortunately, the symptoms are nonspecific and it really requires an awareness and an index of suspicion when patients complain of progressive shortness of breath. It's especially important to think of this rare disease when they have no other reason to be short of breath, such as lung disease or left heart disease, or if they're in one of the increased risk categories I mentioned before, if it's a patient with HIV or liver disease, and especially scleroderma. Scleroderma patients really require kind of a special mention. Even though PAH is 10 in a million, in patients with scleroderma it's 1 in 10. They have a 10% chance of getting this disease and so we actively screen those patients. The echocardiogram is probably the most important screening test, really focusing on the right ventricular size and function. We do also pay attention to the estimated pulmonary arterial systolic pressure, the PASP, but we look at the whole picture with the RV size and function. The echo is not diagnostic and you really do need a confirmatory right heart catheterization if the echo suggests pulmonary hypertension. Certainly you should never start any medications for pulmonary hypertension without a right heart catheterization. If the echo is truly normal then you really don't need the cath.

Dr. Caudle:

That's very comprehensive and very helpful. So, can you please discuss how patients with pulmonary arterial hypertension are treated and can you also discuss the overall prognosis for these patients?

Dr. Smith:

The treatment has really changed dramatically in the last 20 years. If we go back to 1995, there really was no specific treatment for these patients and median survival was only about 2.8 years. Now we have more than 12 therapies to consider and survival is more than 9 years. The most important part to begin is that we get the diagnosis correctly as PH therapies, pulmonary hypertension therapies, are really only indicated in those patients who have confirmed pulmonary arterial hypertension, and it's important that we find these patients early and that we treat them aggressively, as studies suggest that these patients really don't do as well when we miss the diagnosis or when we under-treat them. The sickest of our patients require IV therapy and registry data suggests that half of these patients aren't actually getting the IV therapy that they need. Ultimately, some patients will need lung transplantation if they have inadequate response to medical therapy.

Dr. Caudle:

If you're just tuning in, you're listening to Medical Breakthroughs from Penn Medicine on ReachMD. I'm your host, Dr. Jennifer Caudle, and I'm speaking with Dr. Akaya Smith, Medical Director for Pulmonary Hypertension Program and an Assistant Professor of Medicine at the Hospital of the University of Pennsylvania. So, moving forward, I've had an opportunity to speak to a number of your colleagues and I understand that Penn is the Center of Excellence with regards to pulmonary hypertension care. What does this mean and can you talk about this a bit?

Dr. Smith:

This is something we're actually very proud of. As I mentioned, pulmonary arterial hypertension is very rare and so many pulmonologists and cardiologists may only have a handful of patients in their practice, maybe one or two. As a center we care for more than 400 of these patients. Understanding the importance of early and accurate diagnosis, as well as appropriate treatment, the Centers of Excellence Accreditation initiative really sought to objectively evaluate centers with special expertise in the diagnosis and the management of patients with pulmonary arterial hypertension, and really focused on quality of care, comprehensiveness of care, as well as patient outcomes. Penn was named one of the first six centers in the country back in about, in 2014. Even now, there are only two centers in Pennsylvania and we're the only center in all of Philadelphia.

Dr. Caudle:

So, what kind of patients should be cared for in a pulmonary hypertension center and when should patients be referred?

Dr. Smith:

So, we take care of all forms of pulmonary hypertension, those caused by left heart disease and lung disease, as well as the rare form, pulmonary arterial hypertension. Pulmonary arterial hypertension is obviously the more rare and more dangerous disease, and also the one for which we have new therapies. This is what I consider a must-not-miss diagnosis and we'd much rather see a patient who ultimately doesn't have PAH than miss a diagnosis of PAH. So, the short answer is we'll see all patients who have pulmonary hypertension. Certainly if the diagnosis is unclear, if the disease is progressing, or if the patient requires advanced therapies, they're probably best served having their care in collaboration with a highly experienced center for PH. If the diagnosis is unclear, if the disease is progressing, or if the patient requires advanced therapies, they're probably best served having their care in collaboration with a highly experienced center. These patients are complex and it's important that there's an active communication between their center and their PH team and so we really focus on making sure we maintain open communication with our referring physicians.

Dr. Caudle:

Okay, great. And can you tell us a little bit about your background and the training of those in your center who treat pulmonary hypertension?

Dr. Smith:

I really think about pulmonary hypertension as where the heart meets a lung. While it's a pulmonary vascular process, ultimately it's the right heart that fails. For this reason, PH physicians are trained as pulmonologists or cardiologists, but you have to understand quite a bit about both disciplines. I'm trained as a pulmonologist and we have one of the few centers in the country to actually function as a collaboration between cardiologists and pulmonologists. So, we have, we do have two cardiologists and a handful of pulmonologists who work together to take care of these patients.

Dr. Caudle:

Wonderful, and finally, how do you think this field will evolve over time?

Dr. Smith:

I think pulmonary arterial hypertension has actually been a remarkable story in the lifespan of medicine. It's not often that you see a disease go from no therapies to more than a dozen therapies in only just 20 years. The development of these therapies has actually really picked up pace and we've had 3 therapies come out in just the last 2 years, after years and years of nothing. There are active studies on novel agents and novel pathways and new ways to deliver the medication for those patients that require IV therapy. That, I think, is probably the most exciting thing is that those patients who are the sickest have to walk around with pumps all the time for continuous infusions, and we're looking at more ways to deliver these specific medications orally and maybe with an implantable device. Those are on the horizon and not yet available, but are certainly very promising for some of our sickest patients. Certainly the ultimate goal is to have a true understanding of why patients develop this disease and work towards a cure, but we're certainly not there yet.

Dr. Caudle:

Well, that's very exciting. Before we close today, is there anything else that you would like to add?

Dr. Smith:

Thank you for asking that question. Pulmonary arterial hypertension is such an important, but rare, disease. I think that just having an awareness of it and raising awareness of the community, we can keep an eye out for these patients is key. Having a high index of

suspicion when we think about patients and young women, who are short of breath, maybe can help us get to these patients earlier and improve their outcome.

Dr. Caudle:

Wonderful. Well, Dr. Smith, thank you so much for joining us today and sharing your insights.

Dr. Smith:

It was my pleasure.

Dr. Caudle:

I'm your host, Dr. Jennifer Caudle and thank you for listening.

Narrator:

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