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Recognizing Pulmonary Manifestations of Chronic Liver Disease

Dr. Nandi:
So how much thought have you given to understanding the pulmonary manifestations and complications of chronic liver disease? On today's program, we are joined by an expert in the field: Dr. Karen Krok of Penn State Health Milton S. Hershey Medical Center. Dr. Krok is a Professor of Medicine and the Associate Director of the GI Fellowship Program. She is also the Vice-Chair of Clinical Affairs and the Medical Director of the Liver Transplant Program at Penn State-Hershey.

Dr. Krok, welcome to the program.

Dr. Krok:
Thank you very much for having me. It's a pleasure.

Dr. Nandi:
Absolutely. So I'm an IBD-ologist, and we do often see liver disease, but I'm blessed that I have some great liver colleagues here to help me with the pulmonary complications, but I do want to learn how I can better recognize these pulmonary complications in my chronic liver disease patients. So first off, can you give us an overview? What are the pulmonary manifestations of chronic liver disease?

Dr. Krok:
Absolutely, and I'm really thrilled that you want to learn more about this as well. So there are really 3 main pulmonary complications of liver disease that we should be thinking about. So the first is hepatohydrothorax, which is a pleural effusion that tends to be on the right side, although doesn't have to be, and it's really basically ascites that has gone through the diaphragm and caused a pleural effusion. The second would be portopulmonary hypertension, so high blood pressure in the pulmonary vasculature that is associated with portal hypertension. And the last, and the one that I'd like to maybe spend the most time talking about today, is hepatopulmonary syndrome. And hepatopulmonary syndrome is when a patient develops tiny, small, microscopic, dilated blood vessels in the pulmonary vasculature that is leading to poor aeration, and patients will end up developing hypoxemia associated with that.

Dr. Nandi:
So these are probably, in my opinion, underdiagnosed. With my basic exam skills and clinical history-taking, I hope that I can detect if a patient is having hepatohydrothorax. But tell us: what are the clinical pearls? How can clinicians recognize these types of manifestations earlier?

Dr. Krok:
I really wish it was something simple that we could recognize. Just like you said, hepatohydrothorax is probably going to be the easiest one to recognize because you'll put your stethoscope on their lung and you're not going to hear any breath sounds going through there, or my favorite is the tactile fremitus and you don't feel the vibrations in your hand over that pleural effusion, but the portopulmonary hypertension, you might not recognize that. They may just be developing edema or more dyspnea on exertion. And the same with

hepatopulmonary syndrome; it may also be very difficult to recognize. But some of the clues that we should make sure that we bring up: So, number 1 is clubbing. So clubbing is when there's the distension of the nail bends on your fingers. So if you see clubbing, that could be a sign that someone is developing hepatopulmonary syndrome or portopulmonary hypertension. Someone with a lot of spider angiomas, so those little, dilated blood vessels that tend to be on the upper chest wall that if you press on them they blanch and then fill again, that is another sign that they might be more likely to have hepatopulmonary syndrome. And then my favorite is orthodeoxia and platypnea. Do you remember those words, Dr. Nandi?

Dr. Nandi:
They sound really familiar, but I cannot tell you exactly what they all mean.

Dr. Krok:
Excellent. Well, I like to think of them as sort of the opposite of what someone with heart failure would feel. So platypnea is when a person feels more short of breath when they are going from a reclined position to an upright position, meaning that it's the opposite of heart failure. So in heart failure you feel worse when you lie down. People will sleep when they are sitting up. But in platypnea, our patients prefer being in the recumbent position instead of sitting up. And orthodeoxia actually refers to the fact that when they do sit up, their oxygen level decreases compared to when they are lying down, and so, if a patient tells you they just get so much more short of breath when they are sitting up compared to when they are lying down, ding-ding-ding, you should be thinking about hepatopulmonary syndrome in those patients.

Dr. Nandi:
Okay, well that's fantastic.

For those just tuning in, you're listening to *GI Insights* on ReachMD, and I've been speaking with Dr. Karen Krok on the pulmonary manifestations of cirrhosis and its impact on the transplant outcomes.

Now let me ask you, Karen, when people develop these types of pulmonary complications, how does it impact their ability to tolerate and undergo a transplant or their transplant candidacy and their outcomes? That's a big question.

Dr. Krok:
So in the liver transplant world, obviously we know that there's the MELD score. The MELD score tells you where you're going to fall on the transplant wait list. But we know that there are patients that are "sicker" than this MELD score and that there are exception points for portopulmonary hypertension as long as certain criteria are met—if their mean arterial pressure is at certain levels and their pulmonary vascular resistance is at certain levels— but if you're unable to get a patient to that with medication, those patients are 100% not transplant candidates. Their mortality in the operating room is very high, particularly when they do reperfusion of the liver, because that pulmonary vasculature is so tight it's unable to handle the intense blood flow that would occur when the new liver is put in, so recognizing that early, getting patients started on treatment early, can make a big difference.

When it comes to hepatopulmonary syndrome, there are also very validated and approved exception points as well. A patient needs to have a PaO2 of less than 60 to get exception points for hepatopulmonary syndrome. Some studies show as little as 4% of patients have hepatopulmonary syndrome, but most studies are showing that it's as high as 25–50% of patients with portal hypertension and cirrhosis will have hepatopulmonary syndrome. So what that implies to me is that any center that has fewer than that is underdiagnosing it and that's because the symptoms early on are very, very subtle. How it is diagnosed though is with a contrast-enhanced transthoracic echocardiogram. And so what they do is they agitate saline, and they will inject the saline into an IV, and then they'll look to see when those bubbles go from the right side of the heart to the left side of the heart. If the bubbles go between 3 and 7 or 8 beats, that's more consistent with hepatopulmonary syndrome, meaning that there is a connection or vascular malformations occurring in the pulmonary vasculature. And if the bubbles are present after 8 heartbeats, that's normal. There is sort of a magic window, because if the patient has too low of a PaO2, then the transplant is a higher risk. In the past it was felt that the PaO2 needed to be above 50, but there's been some good data coming out of some groups showing that perhaps going down to as low as 45 or 40 might still be adequate for patients to get a liver transplant as well.

Dr. Nandi:
Now Karen, we've just spent a lot of time speaking about portopulmonary hypertension. I kind of want to regroup here and have our clinicians who are listening to the podcast hear from you. How can they work best with a transplant center in identifying these outcomes, and what can they do to start the process of evaluating the patient for portopulmonary hypertension?

Dr. Krok:
Absolutely. So if you have a patient that seems to have more dyspnea on exertion than what you might expect or more shortness of breath or worsening lower extremity edema compared to ascites, you should be thinking about portopulmonary hypertension and order an echocardiogram. A very simple echocardiogram can diagnose that and get those patients evaluated by pulmonary as quickly as possible to get them started on treatments.

As far as hepatopulmonary syndrome goes, that's the dilated blood vessels more so seated with hypoxemia. Those patients, if you start having that platypnea or orthodeoxia, a lot of spider angiomas, order that transthoracic echocardiogram—but not just the standard transthoracic. Order it with contrast so we can get the bubble study, and that will help diagnose it. If it's diagnosed, that would 100% be an indication to refer that patient for a transplant evaluation—not saying that they're going to automatically qualify for exception points, but knowing that that has a poorer outcome in those patients and it can progress pretty rapidly, and so you want to refer those patients for transplant evaluation even if their MELD score is only 10 or 11 or 12 and you wouldn't typically refer them. If you do diagnose hepatopulmonary syndrome, 100% refer that patient for a transplant evaluation, or at least discuss it with a transplant hepatologist.

Dr. Nandi:
Karen, this has been fantastic. And folks, you heard it right here. There is a role for each and every one of us as GI clinicians to further enhance the management strategy and transplant candidacy for our patients who have advanced liver disease, and if you have a patient who has respiratory symptoms, now you know that you have the right questions to ask about orthostatic changes in their breathing and what tests you can order to kind of accelerate the management process in getting the patient diagnosed with one of these pulmonary complications and working hand-in-hand with your local liver transplant center.

Karen, thank you so much for joining us today and sharing your GI insights with our greater GI community.

Dr. Krok:
Thank you so much for inviting me. It's been an absolute pleasure.

Dr. Nandi:
Thank you, Karen. For ReachMD, I'm Dr. Neil Nandi. To access this episode and others from *GI Insights*, please visit ReachMD.com/GIInsights where you can Be Part of the Knowledge. As always, thanks for listening.