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Primary Biliary Cholangitis: A Natural History Lesson

Announcer:

Welcome to CME on ReachMD. This episode is part of our MinuteCE curriculum.

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Dr. Mayo:

This is CME on ReachMD, and I'm Dr. Marlyn Mayo. And here with me today is Dr. Kris Kowdley.

What can you tell us, Dr. Kowdley, as providers about the progression of PBC?

Dr. Kowdley:

Yes, so it's important to understand that PBC is a progressive disease, but the rate of progression of the disease is variable. The disease typically has a long course. The reason it was called primary biliary cirrhosis when it was first identified as a condition decades ago was because most patients presented with end-stage liver disease and complications of cirrhosis. But we now know that most patients are diagnosed through routine biochemical testing with or without symptoms, and so the patient may have PBC for decades.

But there are a few important caveats for our listeners to be aware of. First of all, PBC being an autoimmune disease and a progressive disease does not have a cure as of yet. But we do have a number of therapies that we can utilize to slow down the rate of progression of this disease.

So the best example is with ursodeoxycholic acid, which, as you know, was approved in 1997 to treat PBC. And the data that has accumulated over the decades since urso has been approved show clearly that the overall number of patients requiring liver transplantation has decreased. We also have identified that the majority of patients who respond to ursodeoxycholic acid do not progress and can achieve and maintain a biochemical remission in many cases. But we know that there is a subset of patients who have rapidly progressive disease and may need liver transplantation sooner rather than later, but these are typically patients who are diagnosed at younger age and also have more severe cholestasis, and sometimes they may develop ductopenia, where the bile ducts in the liver disappear even before the development of cirrhosis.

So we know now that PBC is a chronic progressive disease, but we can manage it medically, and the data with urso has shown that we can actually slow down the progression of the disease and many patients can do very well for many years.

Dr. Mayo:

Right. So the key takeaway, then, would be that this is a progressive disease, although it progresses at different rates in different people. And the therapies that we have may slow that progression but are not a cure. And also, this is not a disease that's going to spontaneously cure itself or go away.

So thank you, Dr. Kowdley, for that little history lesson and thank you to our listeners for tuning in.

Announcer:

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