Experts Explain: Tools for Determining Prognosis With Myelofibrosis

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Chuck:
Hi, everybody. My name's Chuck.

Andrew Schorr:
And what condition are you dealing with, Chuck?

Chuck:
I'm dealing with primary myelofibrosis. I was diagnosed 18 months ago. I'm grateful to be asymptomatic for the most part and being treated with hydroxyurea (Hydrea). My question is, and it's general—and as I talk to people in the room and talk to patients waiting in the waiting room at the cancer center, how do you determine risk? And you touched on it a moment ago, Dr. Daver. And there are certain risk assessments that I've tried to plow through and have found them very difficult. And even with my oncologist, sometimes getting a direct answer from them is difficult. But I think most people wanna know what's my risk? How long am I gonna live?

Andrew Schorr:
Who wants to start with that one? Do you wanna start?

Lindsey Lyle:
Sure, I'll start. Sure.

Andrew Schorr:
Go ahead.

Lindsey Lyle:
So, it's not uncommon for you to be fumbling through all of these different risk assessment tools because we have a bunch of them, actually. And more and more are being developed the more that we learn about the disease biology, the more that we learn about different mutations. And Dr. Daver alluded to not just the driver mutations but perhaps these other mutations that are present also that can predict a more aggressive course. But at the very basis of things, there's a score called the International Prognostic Scoring System, IPSS, and this was the first one that was used in the clinical trials for the development of ruxolitinib (Jakafi). And this is probably the most basic that may make more sense to you.

So, age over 65. You get points for different things. So, in the IPSS, all of these points are just one point for each one. Or a white blood cell count greater than 25,000, circulating blasts which are baby cells which are not supposed to be in the peripheral blood, symptoms of the disease, a large spleen, weight loss, these sort of things, and anemia with a hemoglobin less than 10. So, generally, those are the things at a very basic level aside from different things that we might see in the bone marrow such as chromosome changes or genetic mutations as well as now, more recently, platelets are also involved in this prognostic scoring system. But is that helpful to you?

Chuck:
Yes.

Andrew Schorr:
Okay. And anything you wanna add, Dr. McMahon?

Dr. McMahon:
Yeah. Exactly what Lindsey said. And I think that it can get really sticky too because as Lindsey said, as we're learning more about this, we're learning that there may be an accumulation of additional mutations that may alter overall prognosis too. And we're looking into that. There's some ongoing validation studies looking into those to…

Andrew Schorr:
Anything you wanna add?

Dr. Daver:
Yeah. So, I think one of the important things is these scoring systems were developed retrospectively on trials that were already done. So, we don't necessarily use them right now to decide treatment. And this is one condition which is very different from other leukemias, acute leukemias, where we use scoring systems or MDS, another disease that I mentioned where you use scoring to treat because here, the Jakafiwas approved for improvement in spleen and symptoms and liver. Later on, we found there's a survival advantage, but the study wasn't part for that, so we don't know the clear benefit of that.

So, these scoring systems, the only reason one may use them would be to consider transplant, no transplant that your doctor will discuss with you. But the Jakafi is really started only if you have big spleen, big liver, lot of symptoms. So, that's something you could discuss with them.