



Understanding MPNs: Experts Explain Essential Thrombocythemia, Myelofibrosis and Polycythemia Vera

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Andrew Schorr:

Lindsey, let's just start with you for a second. We have on our slide there ET, MF, PV. How are these related? What are they? Is it something in your bone marrow? Where does it come from? What is it?

Lindsey Lyle:

Great question. So, MPN, or myeloproliferative neoplasm is sort of an umbrella term for basically a process in the bone marrow is going out of control, essentially. So, it can come in different flavors is the way I like to talk about it. And so, essential thrombocythemia is a problem where the bone marrow, there's a clonal process in the bone marrow meaning that it's not reactive but actually, there's a mutation causing platelets to grow without control. And so, there are too many platelets being made in the bone marrow. And then those go out into the peripheral blood. And that's why we can see elevated platelets on a blood draw. So, that's the main feature of the disease of essential thrombocythemia.

Myelofibrosis is the most progressive type of myeloproliferative neoplasm. And just to be said, these are not necessarily a continuum. ET does not always progress to myelofibrosis. But it can. It can over time. But myelofibrosis is a bone marrow disorder in which too many white blood cells are usually being produced. And there's fibrosis or scarring in the bone marrow which makes it more difficult for the bone marrow to make healthy cells such as red blood cells or platelets. So, those are oftentimes low. And then because of this hyper proliferation or these cells that are growing without control, sometimes patients have very large spleens in myelofibrosis.

This is a disease that, like Andrew said, can be categorized as lower risk or higher risk depending on different features of the disease, which we'll get into in a little bit. But lastly, within this group of MPNs, polycythemia Vera can be manifested in elevation of all three cell lines. So, you have too many white blood cells, too many red blood cells, and too many platelets. But the difference is that polycythemia Vera is characterized by elevated red blood cell counts. So, the other two, essential thrombocythemia and myelofibrosis don't have an elevated red blood cell count above normal. But polycythemia vera does. So, that's the distinguishing feature of that process.

Andrew Schorr:

Okay. Thank you. So, Brandon, I'm gonna try to be a student. And you tell me if I get a good grade, okay? Because I wanna make sure we understand this. So, platelets, that's what helps—if you cut yourself, that's what helps you heal, right? It's stickiness in the blood.

Dr. McMahon:

Yup.

Andrew Schorr:

So, these platelets that—what would be the normal—if you had a blood test, what would be the normal range of platelets?

Dr. McMahon:

There's a little bit of variability, but it's a very broad range. About 150 to about 400 thousand roughly.

Andrew Schorr:

Okay. But if somebody had ET, how high could their platelet number go?

Dr. McMahon:

Sometimes, depending on the person, it could be in the million range. So, the number is reported as 150 to 400, but it's 150 thousand to 400 thousand. So, it could be up as high as a million, a million and a half, and sometimes even higher than that.

Andrew Schorr:

Okay. And polycythemia vera, what would be indications of that in the blood count?

Dr. McMahon:

So, the big key that people will notice is their hemoglobin and hematocrit are going to be elevated. And as Lindsey mentioned, oftentimes the way blood count is high, and the platelet count is high with polycythemia Vera, but not always. But oftentimes the key feature of p. vera is that the hemoglobin and the hematocrit are elevated.

Andrew Schorr:

Okay. And in myelofibrosis, we talked about scarring in the bone marrow. And I think—tell me if I'm right—the bone marrow in your long bones and in your hips, that's where we make our blood. That's the blood factory, right?

Dr. McMahon:

Correct.

Andrew Schorr:

Okay. So, if we get scarring, that can gum everything up.

Dr. McMahon:

It can. And it can also, especially in the case of myelofibrosis, even though the blood cells are predominantly made in the bone marrow, if the bone marrow's less effective, then organs that used to make blood cells when you were a fetus, the spleen being one of them, can get bigger and bigger and bigger, because that tries to take over and tries to make red blood cells and other cells in place of the bone marrow.

Andrew Schorr:

In some people who develop a large spleen, it can go down to your hip.

Dr. McMahon:

Very large. Yes. Can be very painful or cause a lot of symptoms with that too.

Andrew Schorr:

But treatment has made a big difference.

Dr. McMahon:

Big difference. Yes.

Andrew Schorr:

Okay. And the idea of having to remove the spleen is very uncommon now.

Dr. McMahon:

Very rarely do we recommend a splenectomy. And it would typically only be in very specific situations for patients, because there can be complications with the surgery. And now as you mentioned, since we do have a lot more availability with treatments that are not surgical, people can have improvement with their spleen.

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