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Beyond the Data: Improving the Lives of People with Sickle Cell Disease

Thorpe:

Welcome to Beyond the Data. I'm Dr. Phoebe Thorpe here to talk about sickle cell disease with Dr. Hassell. Thank you very much for joining us.

Hassell:

Thanks for having me.

Thorpe:

Dr. Hassell is a hematologist who specializes in sickle-cell disease. Tell me what is sickle-cell disease?

Hassell:

Sickle-cell disease is a disorder of the blood where the hemoglobin, the parts in the red blood cells that carry oxygen, is by an inherited basis is deformed. And so one has to have two genes, one from mom, one from dad, to be abnormal in a way that can create sickling inside the red blood cell. Sickle-cell disease is really interesting though because we think about as a red cell disease because it uses abnormal genes inside the red cell, but actually what happens with sickle-cell disease is that the red cells can circulate through the body and can break apart and cause damage from the contents that they spill into the organs and cause damage, but also can block the flow through the blood vessels. And in many ways the blood vessels change and create misshapen blood vessels and other harm so that people with sickle-cell disease are actually in some ways like people with diabetes because blood sugar goes everywhere and can damage many different organs and circulate through the blood. And for people with sickle-cell disease the red blood cells actually behave like a too high a blood sugar and cause harm to various organs. Most devastatingly, however, if those blood cells get stuck in the blood vessels they actually can cause very severe pain when they all of a sudden get blocked in one particular area and the blood supply is cut off to an area the body, creating what's called sickle-cell crisis or excruciating painful events that hit all of a sudden and very hard.

Thorpe:

And they can affect, they also, you can get that blockage in the brain and in the spleen and the liver, it's, it's anywhere blood flows.

Hassell:

Yeah, it actually, and it's, as like diabetes, I often have people and providers think about it that way, because when a person with diabetes comes in, they don't just think about what is the blood sugar and when people with sickle-cell come in I have them think about not just how anemic is the person, which is a consequence of having misshapen red blood cells, but also to consider where have the blood cells been stuck creating damage and injury. So it's really a whole body disease and a change in the organs in the body system

not just a disease of the blood.

Thorpe:

And, so, some things, like diabetes, there's prevention and there's treatment. What, what, what are they doing in healthcare now for people with sickle-cell disease?

Hassell:

So, in the medical system, sort of on the medical side there are ways to change what hemoglobin is made inside the red blood cells. There's a treatment called hydroxyurea which is an oral chemotherapy, although it's not the kind make your hair fall out or make you sick, but it prompts the red cells to make the fetal or the baby hemoglobin instead of the sickle hemoglobin that you make after you're born if you have the sickle-cell disease. And so of those cells don't sickle is much they aren't as toxic or damaging to the rest of the body and the anemia is less severe. And of course if it's a problem with blood, one can simply give normal blood cells in place of the sickle red blood cells and so transfusion treatments are used to prevent complications like stroke, which can devastate up to 10% of children with sickle-cell anemia. And/or other acute complications of a person severely ill, transfusion therapy, putting in good healthy red blood cells can really make a difference, so that's the way we impact what happens with sickle-cell disease. In healthcare more broadly that's a trickier question because we may have treatments or even cures coming, but people need to have access to the healthcare system and to people who know about the disease in order to really benefit from these therapies.

Thorpe:

That was going to be my next question, is where else is there room for improvement in, in, in providing care for them? But not just care but also improving their lives?

Hassell:

Yeah, so I think, one of the things that people are often aware of from high school genetics classes is that sickle-cell disease often affects persons at least in North America who have African descent, of persons of Hispanic descent are affected, persons who are Caucasian, who are from the Middle East, so a variety of communities affected but predominantly in North America communities who are disadvantaged. Socioeconomically, they're sometimes disenfranchised in the healthcare system. They have less access to resources for healthcare. And so even as we advance what's taking place in healthcare and the opportunities for treatment of sickle-cell disease we need to find a better way to get these people into the healthcare system. Affordable healthcare, access not just to any health care, but to healthcare providers who understand what the system's about, and understand the sickle-cell disease and how to render the cures. And beyond that I think we have to recognize that often people affected by this disease because they come from disadvantaged communities are making choices between their chemotherapy pills and whether or not the kids have school supplies or they have transportation to come in for the clinic visit that's necessary to help monitor these therapies or even just general healthcare assessments. We need to take a broader perspective on how healthcare and health interplays with your overall life circumstances and how we can improve all of those aspects, and all that matters for quality of life.

Thorpe:

One of the speakers mentioned that the, let's make sure I can get it right, in 1973 the average lifespan was 14 and today while 2008 it's vastly improved by getting to 42 but when you think of the average life equivalency of 42 it's jaw-dropping just to think of that here in the United States and it shows in 14 to 42 shows you how far we've come, but also how much more we have to do. And what else would

you like to see us do to make those outcomes beyond just healthcare are there more supports for community and things like that?

Hassell:

So there's a real movement of what I think in this country, especially the last 10 to 15 years. There is a national support organization called the sickle cells disease Association of America or SCDAA, and they work with local community support groups to enhance the support services that literally support groups for people to come and talk about their sickle-cell disease, disease, to share resources. And, the other thing that the federal government has done is sought not just to empower researchers with grant monies, there's never enough, but, you know, at least they're giving some toward sickle-cell disease. But the NIH for example, the most recent project is called the sickle-cell disease implementation projects. So it's not new discovery in the scientific lab, but it's actually figuring out how to get these advances into the hands of providers and knowledgeable healthcare systems so they can actually render them and support the individuals and their families who are seeking care from the system. And that HERSA the federal agency has a lot of projects going that look at whether you're modifying the healthcare system and the providers are enhancing the patient supports with things like community health workers. So there's a lot of things afoot to try and connect patients and their families with all of their needs not just literally the healthcare but integrating the whole picture.

Thorpe:

And then the, the part, that hopefully, public health can play and that is the collecting information. Our strength is surveillance and helping people figure out what works.

Hassell:

Absolutely, and so I think that the tie that binds really is, we need to understand where people currently are at, both in terms of services they may need, what their healthcare is like, and what are the health outcomes. And then where are the gaps, where are the opportunities to fill those gaps and honestly public health plays a key role in population-based assessments of where the needs are really how can we bridge the gaps that we can identify. Each of us works in our own sector, our research lab or our clinic-based or community organization and those data, the information that we seek to glean as sponsored by CDC and other entities really is gonna be the glue that pulls it all together.

Thorpe:

I hope, I really hope, I know that the, that's the sickle-cell data collection, I hope that that really comes together in a way that provides this information that you all are seeking to, to really improve the outcomes in the lives of people with sickle-cell.

Hassell:

Well, I think it's you know, we've seen today with the presentation, there is such a clear opportunity here and we're really supportive of get moving forward.

Thorpe:

Thank you so much for joining us and for your work. I appreciate it very much.

Hassell:

Thank you so much for having me today.

Thorpe:

And thank you for joining us see you next time on Beyond the Data.