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Uncovering the Chronic Impacts of Sickle Cell Disease

Announcer: This is ReachMD. Welcome to this special series, “Changing the Conversation about Sickle Cell Disease,” sponsored by Pfizer. Here is your host Dr. Matt Birnholz

Dr. Birnholz: Welcome to “Changing the Conversation about Sickle Cell Disease.” I’m Dr. Matt Birnholz. Joining me is Dr. Caroline Freiermuth, Associate Professor in the Department of Emergency Medicine at the University of Cincinnati. Dr. Freiermuth and I will be diving into the greater and lesser known chronic impacts of sickle cell disease to get a better sense of how, when, and where we can intervene for these patients. Dr. Freiermuth, welcome to the program.

Dr. Freiermuth: Thank you so much for having me. I’m excited to be a part of it.

Dr. Birnholz: It’s great to have you with us. So, to start, Dr. Freiermuth, can you give our audience a breakdown of the pain symptoms patients with sickle cell disease most commonly present with, if even such a thing as common presentation actually exists?

Dr. Freiermuth: So, I think you need to think back to why patients are having a pain crisis. So, in sickle cell disease, rather than having the nice round red blood cells that the normal person has, their cells take a sickle shape, which is where the name comes from, and then they do not flow through the capillaries very well. So, any time you get down to those very small blood vessels, the cells are misshapen, and they’re actually stickier than the normal person’s as well, and so they stick to each other and cause blockages, and so basically you can get pain anywhere, anywhere where there’s not enough oxygen to those blood vessels, anywhere where there might be some local inflammation, and those cells really start sticking together, and so people may experience pain in their hands and legs, they may experience pain in their large joints. A lot of patients complain about bone pain, but we also have to think about, when people come in with chest pain, is this because their lungs aren’t getting enough blood flow? When people have renal issues, this is also due to these areas of microischemia from their disease process.

Dr. Birnholz: Interesting, and when you talk about experience of pain, I just want to clarify this from the start because the pathophysiological process for these patients in terms of the sickling of their cells, the stickiness that you talked about, that’s going to be a similar process, but does that translate into a similar pain experience? Or do these patients actually have different pain experiences?

Dr. Freiermuth: So, pain is such a unique problem. It’s extremely subjective. None of us have a great way to measure it, and everyone has their own way of dealing with it. So, as we know, in the emergency department, we see patients with all sorts of different pain express themselves differently. Some people may scream out when they have pain, some people might writhe around, some people may rock, you know, some people use a heating pad or an ice pack to try to relieve the pain, some people try to distract themselves, other people, you know, just curl up in a ball and cry, and so for each patient, that manifestation of pain is so different, and they have learned experience throughout life, you know. So, whatever’s worked for them in the past, that’s what they gravitate to in the future, and for some of our patients, they even learn a pseudo-addictive behavior where they may have maladaptive coping mechanisms because they know that it gets more attention, and so they know that if they’re a little bit more expressive about their pain that people might get to them quicker, might treat them faster, or might treat them better, and they really, you know, are more demonstrative than others.

Dr. Birnholz: Right, and on that note, we know that there is a stigma associated with these pain crises for patients in terms of the frequency of the ER visits, the level of pain control that they’re demanding, the type of attention-seeking behaviors that they have to adopt in order to get their pain control under control. Can you elaborate on the perceptions that this creates in the health care settings?

Dr. Freiermuth: Definitely. So, one of the things I like to remind all of my residents is that these patients actually rarely come to the emergency department. Most patients will only have about one crisis per year. Forty percent of patients will have no crises in a year,

and you never see them in the emergency department, but there's that 1-2% of patients who have, you know, six to ten crises a year and really will come to the emergency department quite frequently, and then you have a very few patients who come to the emergency department for things outside of crisis but are also looking to escape whatever it is they're escaping, whether it be, you know, problems at home or chronic pain or psychologic pain, and they also come in and complain of that same pain trying to get that relief, and so what I try to remind people is, we are not seeing a majority of the patients, and even the majority of patients we do see, we're only seeing once or twice a year, but our perceptions really do get colored by those patients who come in very frequently, and so I try to remind people, one, that they're not very frequent, and two, that even for those patients who are here very frequently, we know from research that patients who have more crises per year have greatly increased morbidity and mortality, and so we need to still make sure that each time one of those patients come in, we're screening them for the common complications, we're looking for the things that might be underlying this pain crisis so that we can help expand their lifespan.

Dr. Birnholz: And maybe we can focus in on that idea of moving beyond what is currently on the surface to understand more about what's beneath the surface for the pain that this disease afflicts on the patients, and that's understanding the impacts that are not widely known. What can you tell us about that?

Dr. Freiermuth: So, again, sickle cell affects all organ systems because it does create these little microischemic areas throughout your body, and so we see, you know, most often in kids when they're first presenting, we see dactylitis, which is inflammation of the fingers and toes. They'll come in with big swollen fingers and toes, and you'll think, "Oh my goodness, what has happened to this poor child?" and it's just that there's that venous congestion in those extremities, and it can't go back through. You know, as they reach puberty, we see a lot of children with priapism from sickle cell disease. We know that avascular necrosis is a long-term problem for patients with sickle cell disease. They're not getting the blood flow to their large joints, and so they end up with chronic hip pain, chronic shoulder pain. A lot of these patients need joint replacements if they live to adulthood. We see renal failure as quite common. Median age for renal failure is somewhere in your twenties, and about 30% of patients with sickle cell disease will wind up with renal failure in their forties. Once they get put on dialysis, the outcome is very, very poor. They do not have nearly the lifespan as those other people in America would have once they get put on dialysis. So, if we look, there have been some studies looking at how long these patients actually live, and some data from the nineties shows that the average age of death for a male with sickle cell disease is around the age of 42, and for a female, it's around the age of 48. Some studies from 2006 show that the average age span might actually be somewhere between 39 and 45. So, if you think about the patient population this affects, which is predominantly African-Americans here in America, the average lifespan for those patients is 69 for males and 72 for females, and so this disease really does kill people early, and I think a lot of people forget that.

Dr. Birnholz: Right, that's sobering statistics, and as some of the interviews that we've talked about before having spoken to you, we've gotten a sense of just how widespread the disease affects the body and how that affects people's longevity. I want to then dive into the effects and impacts on the organ systems specifically. Maybe we can start with the cardiopulmonary system, which for many of us immediately brings to mind acute chest syndrome, but are there any other related impacts to this organ system?

Dr. Freiermuth: Sure, acute chest is definitely the thing we drill into our triage nurses. We want them looking for that. It is the number one killer of patients with sickle cell disease. It's the number two reason for admission with pain crisis being the number one admission, and so we want people looking for – Do these patients have cough? Do they have fever? Do they have increased sputum production? Are we noting them to be hypoxic? If any of those are the case, we definitely want them to be brought right back. We want that chest x-ray, we want to get them treated, and even for those patients who do not have acute chest, we like to remind people that the only thing we know of right now that decreases the risk for acute chest during an acute pain crisis is the use of an incentive spirometer. So, we like people to use these machines, take those deep breaths, expand their lungs, and so if your patients are there for just a regular pain crisis, if they're going to be holding in your emergency department, if they're going to be in your obs unit, if you're admitting them to the hospital, please remember to get that machine out and really encourage the use because we see these patients who are in pain crisis that curl up in bed, and they take very shallow breaths because everything hurts, and then that just increases their risk of developing acute chest during a regular crisis. Outside of acute chest syndrome, we also see that many of these patients develop pulmonary hypertension through the years, so about 30-35% of patients with sickle cell disease will end up with pulmonary hypertension, which means that, you know, their hearts are working harder to pump blood into their lungs, they're not getting the oxygen and blood to match up within their lungs, and these patients end up on long-term oxygen, they end up having to be on pumps where they have medications continuously infused to help decrease the strain that pulmonary hypertension causes to the heart.

Dr. Birnholz: These are excellent reminders that many people don't really think about, and especially, as you said, about spirometry having a very big impact on patient outcomes through their hospital stays. Why don't we then turn to the neurological system – how are sickle cell patients affected here?

Dr. Freiermuth: Patients with sickle cell disease have 300 times the risk of stroke than the average American, so that's huge, and most

of those will happen between the ages of two and nine. So, with the kids, we really want to make sure that we're not blowing off any of these neurologic symptoms. When they come in and they're not using, you know, one of their arms or legs, or they're complaining about headaches or their speech just isn't quite right, which is sometimes hard to figure out for a two-year-old, we really want to make sure that we're taking a close look and we're not missing stroke. About 11% of patients with sickle cell disease will have a stroke by the age of 20, and about 45% of them will have a stroke by the age of 45, and so that's a really high number. Even those who don't have obvious strokes where they don't come in with stroke-like symptoms, they will have these micro strokes throughout their lives and have little ischemic areas in their brain. Many of these patients, later in their life when they have an MRI, we can see areas where they have had prior ischemic events that have just never been detected, and so this also affects their cognitive abilities throughout life.

Dr. Birnholz: Right, and between cardiopulmonary and neurological impacts alone, we're getting a very clear picture of just how deeply this disease impacts the morbidity and mortality for these patients. It's staggering, actually. I want to then turn to the immune system impacts as well. We obviously know that there's an increased susceptibility to certain infections conferred by the sickle cell gene, but I imagine that there is a broader cascade effect over time from this kind of vulnerability, similar to what you laid out for the spectrum of a person's life with neurological risk and ministrokes. What can you tell us about the immune system impacts?

Dr. Freiermuth: So, we know that most patients with sickle cell disease will be functionally asplenic by the time they hit their teens. That means that your spleen is then not there to help you clear any infections, especially the encapsulated organisms that it usually helps to catch. So, there were big strides made, you know, decades ago when vaccines really came into play. We saw the death rate for kids with sickle cell disease drop dramatically because all of a sudden, they were being vaccinated against some of these deadly diseases. Then they started prophylactic penicillin for children under the age of five with sickle cell disease, and that again made a great impact, but for some of our adults, we still need to make sure that we take very seriously when they come in with fever. You know, even if it is just a virus, we need to make really sure that we're screening and it's not something more serious. A lot of the kids get blood cultures right away. The aim at a lot of hospital systems is that they get antibiotics within an hour so that we're not missing the window for a serious bacterial infection.

Dr. Birnholz: Excellent, and Dr. Freiermuth, I don't want to leave anything out of the mix here. Are there other downstream impacts that we haven't covered here that may also not be on everyone's radar?

Dr. Freiermuth: So, I think one of the biggest places that gets ignored is the psychiatric effects. So, patients with sickle cell disease, we've talked about, they have pain crises, they can't keep up with their friends as kids because getting dehydrated or getting low on oxygen can trigger these crises, and so they learn how to cope where they don't run around on the soccer field with their friends, and then later in life, you know, they can't hold that part-time job because if you call out sick too many times you get fired, or they don't have the cognitive ability to hold that job, and it really has a long-term effect on the psyche when you can't do the things that everyone else around you is doing. That makes it very hard. We know that 30% of patients with sickle cell disease have some form of depression/anxiety, and yet they're not freely telling us that, and we're not always screening for it, and so it's one of the things, especially when you see increased visits to the emergency department, we really need to be asking those questions and seeing, you know, is this an acute pain crisis? Is it their chronic pain? Or is it pain from some other source, and they don't know how to tell us that?

Dr. Birnholz: Well, these are definitely words to live by from a practice standpoint for these patients, and what I love the most about this is that it helps us question what a typical presentation actually is and what we should be on the lookout for sickle cell disease patients. So, Dr. Freiermuth, I really want to thank you for your time and joining us today, bringing your insights to the table. It was really great having you on the program.

Dr. Freiermuth: Well, thank you again.

Announcer: The preceding program was sponsored by Pfizer. To revisit any part of this discussion, and to access other episodes in this series, visit ReachMD.com/SickCellConversations. Thank you for listening.