What's the Biggest Problem Sickle Cell Patients Face?

For many sickle cell sufferers, severe and constant pain is part of everyday life.

October 30, 2015 By Jeanette L. Pinnace

Last year, the Food and Drug Administration published the results of a public meeting held in early 2014 to hear from sickle cell disease (SCD) patients about the most significant effects of their illness and the therapies available to them for treatment. In general, the adults who participated in this forum said acute pain crises and chronic pain were among the most significant symptoms of this inherited red blood cell disorder.

According to one participant, "The pain has complete control, to the point that three times in my life I prayed to God to let me die because I didn't think I could take any more."

But when acute pain crises drive SCD patients to emergency rooms for treatment, many face suspicion about their motives for requesting the high doses of strong pain meds, often opioids, they need to get relief.

Real Health spoke with W. Keith Hoots, MD, director of the Division of Blood Diseases and Resources at the National Heart, Lung, and Blood Institute (NHLBI) about this problem.

What are some of the most common misconceptions clinicians have about sickle cell disease, and how are they being educated about these misconceptions?

For patients with sickle cell disease, particularly the ones who have the most severe manifestations of sickle cell disease and recurrent pain, this pain increases with age and it increases with organ dysfunction. In some cases, this is pain they have to live with almost daily, and because of that they require a lot of intervention to help them manage the pain. Otherwise, their lives are almost intolerable as far as getting through the day because the pain is so severe. They're on drugs that are designed to treat pain, many of which are opioids. I think the most common misconception in emergency centers and by, perhaps, some clinicians who don't regularly take care of sickle cell disease, is that these patients come in on a lot of pain medication yet they're still complaining of pain. [Some clinicians may wonder], are these patients abusing the medications in the way that a drug abuser would be abusing them?

Overwhelmingly, SCD patients are just trying to manage their pain. The one true fact of the opioids is that the more you take, the more tolerant you become to them, so even though a sickle cell patient may be receiving baseline pain medicines that include opioids, such as the ones that are mostly in oral form, the pain still breaks through. Therefore, these patients still have a lot of pain. When they come to the emergency center, some people not sophisticated in managing sickle cell pain may say, “Well, they’re just drug-seeking.” That can result in a very bad outcome for the individual. This suspicion doesn’t only delay SCD patients’ access to enhanced pain medication, it also creates a distrust between the health professional and the patient that originates within the system. This means that as the patient goes forward in the management of the disease, it’s less likely that care providers will believe them when they say they’re getting better or worse. That’s a bad paradigm for clinical management. Of all the misconceptions, this suspicion of sickle cell disease patients and their legitimate need for opioid medications is at the top of the list.

Are there any other problems that concern you about the interaction between health care providers and adult sickle cell patients?

I think there are also health care providers who probably underestimate just how much organ dysfunction could be ongoing particularly as children age into adulthood. A lot of times in our country the system set up for children with sickle cell are multidisciplinary. They do extraordinarily good things for the children and their family. But when children with sickle cell disease reach a certain age, when the child is now a young adult going off on their own, the system doesn’t replicate those support services as well for them. Adult sickle cell patients are left to manage everything alone, and this is a hard disease to manage without a lot of support.

What do you think clinical guidelines from health care agencies, such as NHLBI, can do to educate health care providers about the pain treatment needs of adult patients with sickle cell disease?

NHLBI clinical guidelines were developed primarily for primary practitioners—not necessarily hematologists (blood specialists), but doctors such as family practitioners, internists, pediatricians, to give them the scope of what is established treatment based on either scientific research or, at least in a few cases, a consensus among experts. The guidelines cover the full gamut of the clinical manifestations of sickle cell and how they should be managed. That includes issues such as ways to deal with central nervous system sickling that can lead to stroke, how to deal with potential renal (kidney) disease, how to deal with potential iron overload, and a whole cadre of potential clinical manifestations. In addition, guidelines cover how practitioners who don’t necessarily treat a lot of patients with sickle cell disease, and therefore would benefit most from these kinds of guidelines, should proceed with the management of SCD.

In certain emergency centers health care providers have made great progress and they’ve done it by, first of all, knowing the patient over time. They’ve created a system that is responsive to the patient. When the same patient with sickle cell comes back again and again for care, care providers get to know them. In addition, even if there’s new staff, the sickle cell patient’s chart reflects the fact that this person has a consistent spectrum of pain because of sickle cell disease.

Therefore, when that patient says they’re having severe pain, they show a track record of having very difficult pain to manage. This immediately removes some of the distrust, because the care provider already knows the subject.

When the care provider is used to managing patients that way, and the person isn’t a new patient, the care provider doesn’t see the SCD patient as someone who is on a lot of medications and trying to get more drugs. The care provider starts with what’s going on with the individual right now that is making the pain break through their usual management, and that’s a whole different paradigm. I think that it’s a system-building project. We’re trying to work on the research side to look at those system changes that some people have implemented and test them to see how effective they are. After that, we have to ask if there are other systemic changes that could be translated to other places where those health care providers may not be as far along in managing a large population of SCD patients.
