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How to Improve Sickle Cell Care in Emergency Medicine: A Case Review

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 in the Conversation about Sickle Cell Disease. I am Dr. Matt Birnholz. Joining me is Dr. Bernard Lopez from Thomas Jefferson University Hospital. Dr. Lopez and I will be walking through an updated approach to emergency medical care for patients with sickle cell disease, and we will do that with a representative case review. Dr. Lopez, welcome to the program.

Dr. Lopez: Thanks, it's a pleasure to be able to do this.

Dr. Birnholz: So, Dr. Lopez, to give us some background before we examine this case, can you tell us about the general perception towards sickle cell patients in emergency department settings?

Dr. Lopez: Sickle cell disease is probably not a very common condition that your typical emergency physician will encounter. It is not very common so for that reason emergency physicians are likely to not be very familiar with them, so there is that stigma of a sickle cell patient being that rare disease kind of person. Additionally, for those that are familiar with sickle cell patients, there is really the drug-seeking stigma, that perhaps that patient really does not have pain, or their pain is not that severe but there is that perception that they are presenting simply because they are seeking analgesics, which is typically going to be an opioid.

Dr. Birnholz: Well then why don't we talk about the types of pain affecting patients with this condition. I understand that there is a common perception among clinicians that pain from vaso-occlusive crises and other presenting issues may not actually be real since patients often do not exhibit the clinical signs to match the symptoms of their pain. So, what are your thoughts on that?

Dr. Lopez: Sickle cell patients have been living with their disease really ever since they were born. It is very common that over the course of their lifetime they will have repeated episodes of acute pain. They may have had hundreds of these episodes by the time they present on any given day in the emergency department, so they are, in many ways, used to their pain. When they present to the emergency department, unlike your typical emergency department patient, they may present without any of the typical signs or symptoms of pain. They often will not have tachycardia. They will not have tachypnea. They may not exhibit that typical pain response, so for that reason, there is that perception that, "Well maybe their pain is not real." Maybe they really do not have it. They often require much higher doses of opioids and when they visit the emergency department and that fact, the fact that they needed maybe higher doses, they need it more frequently, often plays a role in how the clinician views that patient and whether they actually

believe that they have real pain.

Dr. Birnholz: Excellent. So, with this understanding in mind, let's review a representative case. To start us off, how does the patient typically present to the emergency department?

Dr. Lopez: The typical sickle cell patient presents with severe pain. Often, they have been living with this pain. They will tell you that they have had this pain for a day, two days, three days, mostly because they are trying to manage that pain at home, so they do not commonly come in saying, "I've had pain for the last day, two or three days." They are also telling you that it is certain areas of their body which they will typically get. It's in their arms and legs and they will tell you that, "This is when I get my typical sickle crisis pain." They can have pain anywhere, but most commonly they will present telling you it is in their extremities, their low back, their abdomen and even their chest, but again, they will tell you, "This is my typical pattern of pain." They may also tell you, "Well this is what happened, this is what's new that may have precipitated this pain," but most commonly it seems as if the pain just comes out of nowhere and you cannot really identify any cause.

Dr. Birnholz: What are your first steps in triage?

Dr. Lopez: First and foremost is assessing the level of pain. Pain is considered an emergency, so the first step is assessing their pain. Commonly that is done by some sort of a pain score. As an example, on a scale of 1-10, "If 10 is the worst pain you have ever had, what number is your pain now?" That is first, assessing their pain. Second and as importantly, is vital sign measurement and looking for any indicators of infection; looking for some sort of an indicator that there is an additional process that is ongoing or that perhaps there is another process that is present that is really not their vaso-occlusive sickle cell crisis. Those are typically the first steps that we will take when assessing one of these patients.

Dr. Birnholz: Let's focus on the diagnostic tests. What tests do you order and what you looking to rule out?

Dr. Lopez: Generally, testing is done based on the patient's presentation, so if there are any signs of an infection; if the disease is presenting differently, I am going to start to look around to see, "Well are there any causes that I might be able to diagnose with testing. I use the patient's clinical presentation to guide any testing, such as a chest x-ray, a urinalysis – that is guided by their presentation. For a sickle cell patient, the most common test that I may order is a complete blood count, a CBC, and the reticulocyte count. The CBC will really give me an indication of the patient's hemoglobin level. Sickle cell anemia is a disease in which the patient has anemia. I am looking to see in the emergency department if they have a much more severe level of anemia, because that is going to guide my therapy and it is not uncommon to see a sickle cell patient with a lower than their usual hemoglobin level. A reticulocyte count simply measures their bone marrow activity. It measures the patient's ability to produce red blood cells. Sickle cell patients are at risk for their bone marrow simply not being able to function at their normal level, and I am looking to make sure that that patient is actually able to produce red blood cells. A sickle cell patient is at risk for something called aplastic anemia, and this is a condition in which the patient is unable to produce the red blood cells. This leads to more severe form on anemia and it can simply worsen the condition of that particular patient.

Dr. Birnholz: Dr. Lopez, help us understand some of the hidden conditions on the differential diagnosis list that clinicians are at risk for mistaking as just another vaso-occlusive crisis.

Dr. Lopez: I mention infection. I cannot stress that enough. There is a reason I mention it frequently is that a sickle cell patient is simply at high risk for infection. That is one of those "hidden conditions" that a clinician really needs to look for. An important condition that a clinician really needs to look for is acute chest syndrome. Acute chest syndrome is simply tissue ischemia, tissue infarction in the lung, and a sickle cell patient is at high risk for this condition. Acute chest syndrome may not be so obvious. Sometimes it looks like pneumonia and sometimes it looks like a typical sickle cell crisis presentation. Acute chest syndrome is outside of infection and the leading cause mortality in the adult sickle cell patient, so it is really important that the clinician, at the very least, think about this condition and perhaps do testing that looks for the signs, the presentation, so that they can make that diagnosis.

Dr. Birnholz: I think the next natural follow-up question here then is when and how do you address this patient's pain and what factors guide you in a particular pain management direction?

Dr. Lopez: Pain is considered an emergency, so when you treat it, you treat it just like any other emergency, you treat it as quickly as you can. The idea is the sooner you can treat the pain, the better the chance of success that you will have in relieving that patient's pain. We like to try to treat patients as soon as we can. We do not wait for testing. There is actually no testing that will tell me how severe a patient's pain is, so while we are doing testing, if we choose to do so, we try to treat the pain immediately. Studies have shown that the quicker you treat, the more aggressive you are in the treatment of pain in sickle cell crisis, the higher the success rate in treating that pain, the less time it will take for that patient's pain to resolve, so really what guides it, is really making sure that you treat it as quickly as possible and that is really based on how that patient is feeling, the severity of pain that they present with.

Dr. Birnholz: Dr. Lopez, under what circumstances do you admit this patient versus deeming well enough to send home?

Dr. Lopez: The simple yet common answer to that is we decide based on the patient's report of relief or non-relief of pain. As there is no objective measure for pain or objective measure for pain relief, we simply go on the patient's report of did they receive enough relief of their pain in the emergency department to be able to go home and manage it there. If that is the case, we discharge them, and this is assuming that there is no other concomitant condition. If the patient says, "I don't feel well enough, the pain is still too bad," we typically admit them to the hospital.

Dr. Birnholz: Let's consider the care continuum beyond the ER setting. What are your priorities as an emergency medicine physician for ensuring the best continuing management beyond the ER?

Dr. Lopez: The best management outside of the emergency department is ongoing and long-term care, so my job is to assure that the patient has some sort of follow up. Having that continued, ongoing care is the best way to assure that therapies that help to manage the disease on a chronic basis that the patient is receiving any available therapies that might lessen the frequency of their acute crisis, maybe lessen the severity of their disease. There are therapies such as hydroxyurea that have been shown long term that if a patient takes this on an ongoing basis, it lessens the frequency of their sickle cell crisis. This is a complex and complicated disease that really requires probably a team approach to assuring that all aspects are addressed, so as far as the priorities, it is really assuring that they have ongoing, long-term management.

Dr. Birnholz: Dr. Lopez, before we wrap up, is there any additional comments or closing remarks that you would like to impart to our audience?

Dr. Lopez: Well, the big keys to treating a sickle cell patient who presents to the emergency department are first and foremost, they are in pain. Severe pain is an emergency, consider that. Believe the patient, treat the pain. That is consideration number one. Number two – these patients are at risk for serious illness such as infection. Consider infection as a cause of their pain; consider their infection as the reason why they are presenting the way that they are. I had mentioned acute chest syndrome. You need to consider that a patient who presents to the emergency department may have acute chest syndrome, so make sure that you consider that condition. Lastly, this is a different group of patients in the way that they present. These patients typically require high doses of opioids, doses that are far higher than your typical emergency department patients. They typically need far more frequent dosing that is far different than your typical emergency department patient, yet that is the sort of treatment that they need in order to be successful in terms of treating their pain and their condition. And lastly, I'll say this one more time – believe the patient.

Dr. Birnholz: With that, I very much want to thank Dr. Lopez for joining me today in discussing an updated approach to emergency medical care for patients with sickle cell disease. Dr. Lopez, it was great having you on the program today.

Dr. Lopez: Thanks. It is a pleasure.

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