Peds Pearls

Suffering, Suffocating, and Stroking Sickle Cell Emergencies

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"*Frequent Fliers*"... In the EMS or ER world, that term evokes images of patients seeking seemingly constant attention for physiological or psychological conditions. All too often, those images are of a negative nature. Nevertheless, we have an obligation to provide appropriate medical care to all, regardless of circumstance. Patients living with sickle cell disease may be among those we see as "frequent fliers" and those individuals can be challenging for us. However, the more we know, the better prepared we are. So, with that in mind, let's explore some important factors regarding the nature and treatment of sickle cell disease.

Suffering: One of the most common reasons that we see patients with sickle cell disease is for management of a vaso-occlusive crisis, also known as a "pain crisis." When the abnormal sickled (crescent shaped) RBCs become stuck in the smaller blood vessels and capillaries, it can cause blockages and lead to tissue ischemia which results in severe pain. The pain crisis can come on quite suddenly and some of the more common triggers include dehydration and changes in body temperature. The severe pain can manifest itself in any part of the body and can be severe enough to send the patient (and parents) to the hospital seeking pain relief! Fortunately, after age 10, the rates of painful crises decrease, but the rates of complications increase!

Here are some of factors regarding treatment for those suffering from pain related to sickle cell disease:

- **Oxygen**: If they ask for oxygen, feel free to provide it. If they are short of breath, this treatment seems self-explanatory. What makes the treatment for these patients different is the fact that some experts recommend supplemental oxygen if the oxygen sats are less than 95% (not <90%).
- **Fluids**: If the patients are not dehydrated and able to tolerate PO fluids, PO fluids work just fine. If they are dehydrated or in need of IV fluids for other reasons (antibiotics, analgesia), certainly an IV or IO is appropriate. In some facilities, another option is the administration of subcutaneous (SQ) fluids and medications including saline, morphine, and Dilaudid (hydromorphone). For patients whose veins tend to be almost non-existent on a good day, SQ (or even elective intraosseous with local anesthesia) infusions can come to the rescue!
- **Pain Medication**: Experienced practitioners remember giving Demerol, Demerol, and more Demerol. Recently, however, the American Academy of Pediatrics has developed a formal position statement on the treatment of sickle cell pain. Essentially, it says 1) Demerol (meperidine) is not so good, 2) There are much better pain meds out there, and 3) If a sickle cell patient has never received Demerol, it would be better to avoid it altogether, and provide alternative medications instead. The many problems with Demerol include 1) Outside of anesthesia, very few providers give enough Demerol, 2) If you do give enough Demerol, patients tend to barf all over the place, and 3) The metabolites from boatloads of Demerol in kids can induce nasty seizures. Given that background, it is easy to understand the evolving role of Benadryl (diphenhydramine), NSAIDS (Ibuprofen, Toradol [ketorolac], etc.) and either oral, IV push, or PCA pump morphine or Dilaudid.
- **Protocols:** In terms of sickle cell pain crisis treatment, one of the best things that has come about recently is the development of protocols seen in many ERs. What that means is that when a patient presents in a pain crisis, the "clock" starts immediately and treatments begin. Then, after a specified amount of time, if the patient is still in pain, additional treatments/medications are given. Reassessments and additional orders continue as part of a predetermined treatment plan as time goes on. As the protocol is followed, there is a point at which the decision is made as to admit or discharge home. It makes our job a whole lot easier and provides consistent patient care regardless of the attending provider. Protocols also tend to eliminate the "doctor shopping" that some "frequent fliers" exhibit.
- **Addiction:** Addiction potential can be a real concern for some providers, but when it comes to the management of sickle cell pain crises, one wise pediatric emergency physician suggested the following:

"If I go to work every day and I have 100 patients in pain and if 99 of them are truly in pain and one is addicted, that means I have a 99% chance of making people feel better. If I make 99 children feel better, and feed one person's addiction, I'm OK with those odds."

Suffocating: The number one reason patients with sickle cell disease die is an acute or "clogged up" chest. If the sickle cell disease patient presents with chest pain, shortness of breath, and hypoxia, we have a seriously different situation. If a chest X-ray reveals a big infiltrate, it is probably either pneumonia or an acute chest. So in the ER, we simply treat them for both. The patient gets antibiotics, pain meds, and oxygen, and then once admitted, and with more time and extensive testing, it can be determined which of the two conditions are at play.

Stroking: The triad of really bad things that can come with sickle cell disease includes pain crises, acute or clogged up chest, and in the worst case, sickle cell stroke. As you can imagine, sickle cell stroke is a true emergency and often requires an exchange transfusion. The younger the kid is when experiencing their first sickle cell stroke, the higher the chance is of having another one. Additionally, if a young child's disease is bad enough to actually cause a stroke, then incredibly close follow-up is needed to prevent the devastating sequelae of more sickle cell strokes.



The overwhelming majority of pediatric sicklers hurt. They are praying you will treat their suffering while the parents and the peds ICU pray they don't suffocate or stroke!



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Contact us to have Scott DeBoer or one of our other pediatric experts present at your next conference.

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