Sickle Cell Crisis and You: A How-to Guide

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ANNALS CASE

A 19-year-old man with sickle cell anemia presented to the emergency department (ED) with a 1-day history of severe bilateral arm pain and swelling. Vitals signs showed a blood pressure of 117/82 mm Hg, a pulse rate of 76 beats/min, and an oral temperature of 36.8°C (98.2°F). Physical examination was remarkable for tense swelling, warmth, and tenderness to palpation of both triceps muscles. Laboratory tests showed an elevated creatine kinase level of 20,734 IU/L. Point-of-care ultrasonography of the triceps showed diffuse muscle thickening with loss of the normal fascicular architecture compared with the unaffected biceps. Diagnosis: sickle cell–induced myonecrosis.[1]

MY, OH, MY…MYONECROSIS!

Myonecrosis is a pretty rare complication of sickle cell disease (SCD).[2] In fact, there are mostly case reports in the literature! But there is a host of other more common, can't-miss complications that can develop as a result of these crescent-shaped deformities of our circulating minilife-givers. Keeping all these straight can sometimes be overwhelming, so we'll run through some of the critical complications of SCD and how to have a sensible approach to these patients.

APPROACH TO SCD FOR DUMMIES

Over the years, there have been several guidelines about the management of acute SCD crises,[3-5] some of which include treatment algorithms. But when it comes to a basic *diagnostic* approach to the sickle cell patient in the ED, we just hear crickets chirping in the midst of awkward silence. Sure, you might have a set of shiny tools, but if you can't even figure out where to use them, they're pretty useless. So here's a simple algorithm (Figure) that can help you keep things straight when all cognitive systems fail (it goes without saying that every patient encounter should be individualized).

The important branch point of this algorithm centers around the point-of-care hemoglobin level, if your facility has the capability. At the heart of SCD is a combination of vaso-occlusion and hemolysis resulting in any number of secondary complications (because of the valine substitute at position…zzz…). Despite this, in uncomplicated sickle cell presentations, a full rainbow set of tubes is not usually needed.[6-8] The approach to SCD in the ED relies on a thorough history and physical examination. Before we get ahead of ourselves, though, let's break down the different pathways of this algorithm; it's so easy you can even do it on your drive home post–night shift.

SEQUESTRATION AND HEMOLYSIS: A PROBLEM OF CONSUMPTION

If the hemoglobin of a sickle cell patient is outside the range of error (a decrease of more than 2 g/dL from his or her baseline)[5], more evaluation is usually warranted to figure out why. The next most important laboratory test to determine why the hemoglobin level decreased is the reticulocyte count, which gives you a snapshot of how well the bone marrow is replenishing the depleted peripheral RBCs. If the retic count is appropriately high (ie, the bone marrow factory is firing on all cylinders to meet market demands), then the worsening anemia is primarily a problem of peripheral consumption of the RBCs. The differential in this case is either sequestration (in the liver or spleen) or hemolytic crisis.

When the Spleen Becomes a Sponge

Question: What's worse than a crashing sickle cell patient? Answer: a crashing *pediatric* sickle cell patient.

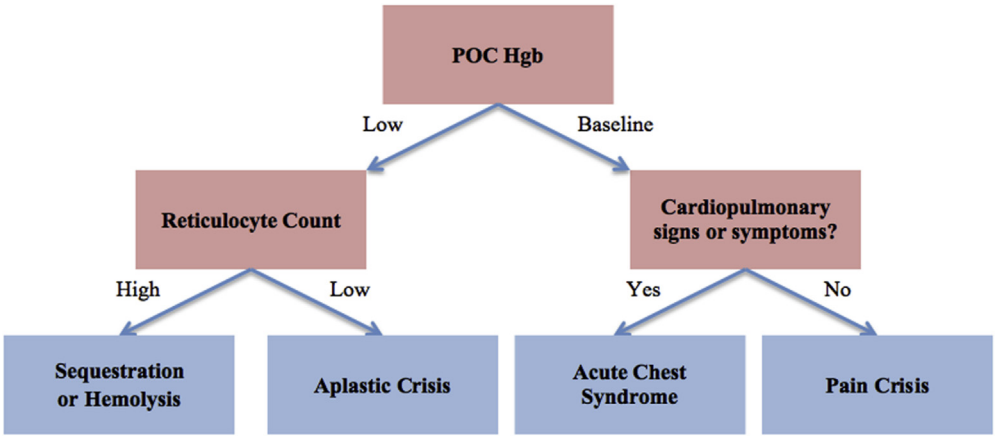

Figure. Algorithmic approach to sickle cell crisis.

Splenic sequestration usually occurs in children, peaking at aged 1 to 4 years[8,9] (although it can occur in adults as well).[10] These patients are usually very ill appearing, presenting in extremis and shock owing to their spleens having greedily hoarded a large amount of peripheral RBCs. Tip-offs to diagnosis, in addition to an acute decrease in hemoglobin level, are clinical: circulatory shock and a very large spleen on ultrasonography or examination. Aggressively treat these patients with fluid resuscitation until you can get the more definitive blood transfusion started, with a goal hemoglobin level between 6 and 8 g/dL. Just to make things even more complicated, the spleen will often release a large amount of peripheral RBCs after transfusion, causing you to overshoot your goal hemoglobin level. This puts the patient at risk of hyperviscosity and subsequent strokes. One recommended transfusion approach is to transfuse the equivalent milliliters per kilogram of blood as the observed hemoglobin concentration in grams per deciliter. For example, if the hemoglobin is 5 g/dL, then transfuse 5 mL/kg at a time.[11] Hepatic sequestration presents in the same way as splenic sequestration (except with an enlarged liver) and is managed similarly…one less thing to think about!

When Red Cells Explode, Measure the Guts

So how do you differentiate a hemolytic crisis from splenic sequestration if both are characterized by a decrease in hemoglobin level and an elevated reticulocyte count? The answer lies in the indirect bilirubin, aspartate and alanine transaminases, and lactate dehydrogenase. When RBCs are destroyed in an acute hemolytic crisis, an increase in lactate dehydrogenase, bilirubin, and aspartate and alanine transaminase levels occurs. This won't be the case in a sequestration crisis. Hemolytic crises are usually self-limited, but patients might need a blood transfusion if they have severe anemia. And as a word of precaution, don't forget the "county handshake" (rectal examination) for patients with an acute decrease in hemoglobin level. Sickle cell patients are prone to gastrointestinal bleeding events the same as everyone else; they just don't tolerate it as well because of their baseline anemia.

APLASTIC CRISIS: A PROBLEM OF PRODUCTION

On the other hand, if the reticulocyte count is low (usually <2%), this suggests that the blood factory workers in the bone marrow aren't working hard enough to meet the peripheral demands of the chronic hemolysis that characterizes SCD. Somewhere out there, Upton Sinclair is rolling in his grave. But really, it's not the bone marrow's fault. Up to 80% of the time, an aplastic crisis in sickle cell patients is caused by parvovirus B19 infection.[12] Infection usually lasts approximately 7 to 10 days and is self-limited. In addition to transfusing these patients for symptomatic anemia and thrombocytopenia, don't forget to put them in isolation so pregnant patients don't develop fetal hydrops.[13]

ACUTE CHEST SYNDROME: THE OTHER ACS

If all is good in the land of enchanted hemoglobins, your job is to ensure that the patient doesn't have acute chest syndrome (ACS). To be clear, when we mention "ACS" in this section, we're not talking about acute coronary syndrome (although technically you could have both ACS and ACS at the same time; how horrible would that be?). So what is this other ACS that we speak of? ACS is defined as some combination of fever, chest pain, hypoxia, respiratory symptoms (cough, wheezing, rales, or tachypnea), and a new pulmonary infiltrate on chest radiograph.[5,7,8] Up to 80% of patients will have isolated