A Chronologic Perspective on Sickle Cell Disease

Joseph R. Hageman, MD

When I finished my first year of medical school at the University of Illinois in Chicago in 1974, I thought often about aspects of sickle cell disease (SCD) that I had learned about and wanted to know more. It was great to know that, as long as each infant with SCD had enough fetal hemoglobin, they were protected from developing vaso-occlusive crisis.\(^1,2\) However, at approximately age 6 months, when the production of sickle hemoglobin increased and fetal hemoglobin decreased, children with SCD begin to develop dactylitis.\(^1\) I also remember reading about two adults with SCD and how hydroxyurea had induced increased fetal hemoglobin production in both of them.\(^3\)

Flash forward to my time as a pediatric resident at Children’s Memorial Hospital in Chicago (1977-1980) and I had the opportunity to care for many patients with SCD who presented with complications including vaso-occlusive crisis, acute chest syndrome, pneumococcal bacteremia, *Salmonella* osteomyelitis, and stroke. A young patient with Italian ethnicity who presented with SCD crises periodically during my time at Children’s Memorial Hospital taught me how the disease can affect children of Mediterranean descent.\(^1,2\)

Presently, as a pediatric hospitalist and a pediatric intensive care unit (PICU) physician at Evanston Hospital, some of my former patients with SCD do come to mind. One was an infant who presented to the emergency department with splenic sequestration, but passed away in the emergency department after resuscitation efforts; and the other was an adolescent who presented many times with acute chest syndrome, and I would perform partial exchange transfusions to reduce his sickle hemoglobin. Subsequently, he became one of the earliest patients to undergo a hematopoietic stem cell transplant at Children’s Memorial Hospital and was “cured” of his disease.\(^4\) Now married and doing well, he expressed gratitude for the previous transfusions I had performed when I saw him again recently.

One of my roles at The University of Chicago Comer Children’s Hospital is to help the residents with research projects and papers. During the past 7 years, one of the residents had a patient with SCD who presented with acute chest syndrome and, unfortunately, developed acute hepatic sequestration in the PICU; the patient’s hemoglobin dropped to 3 g/dL and her liver was palpable 17 cm below the right costal margin. Fortunately, the patient recovered.\(^5\) Each resident class is required to do a quality improvement project and one class studied the incidence of positive blood cultures in 390 children with SCD presenting with fever. Follow-up care is on an outpatient basis if patients are clinically stable and pending a negative blood culture for 72 hours. In this particular study, there was only one true positive (at 48 hours) *Streptococcus pneumoniae* blood culture; the patient was hospitalized with acute chest syndrome. After the review was completed, it was decided that because of the high rate of compliance with acute care visits and early time to positivity, treatment of these children could be accomplished on an outpatient basis.\(^6\) Although lead toxicity in pediatric patients with SCD is uncommon, clinicians should be knowledgeable as the clinical presentation of crisis in SCD can be similar to lead toxicity and the two examples in the Jung and Peddinti\(^7\) article illustrate that point.

Finally, I want to relay a couple of other observations related to SCD. The first is that hydroxyurea is commonly used clinically to decrease the occurrence of vaso-occlusive crisis and acute chest syndrome in patients with SCD, and it works by increasing production of fetal hemoglobin\(^3\) (this concept is what brought me back to reading about hydroxyurea and fetal hemoglobin as
a new attending physician). The second observation is that hematopoietic stem cell transplantation is now more commonly used to “cure” children with SCD who fulfill specific clinical criteria, including chronic transfusion therapy, multiple vaso-occlusive crises, acute chest syndrome, and stroke.4,8

REFERENCES