

# Sickle Cell Today

## USA Comprehensive Sickle Cell Center

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**SCDAA**  
"Break The Sickle Cycle"®

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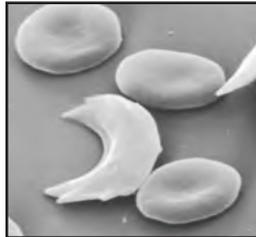
**USA Health System**

## SEPTEMBER IS NATIONAL SICKLE CELL AWARENESS MONTH

### *Iron Overload in Sickle Cell Disease*

#### **Why are blood transfusions needed in sickle cell disease?**

Sickle cell disease (SCD) is an inherited disorder of the hemoglobin molecule, which is a component of red blood cells. Transfusions in SCD raise the oxygen carrying capacity of blood and decrease the proportion of sickle red blood cells. A sudden fall in hemoglobin accompanies many of the manifestations of SCD. Episodic transfusions provide healthy red blood cells and are given to reverse immediate complications such as: aplastic crises, splenic sequestration, acute chest syndrome, and prolonged priapism. Transfusion therapy can be a life-saving therapeutic procedure in such instances. Long-term monthly transfusions are given to prevent future complications such as strokes<sup>1</sup>.



#### **What is iron overload?**

Apart from other risks associated with transfusions, a major long-term problem is the extra iron brought into the body with each transfusion. Animals and humans eliminate excess body iron which deposits into various organs of the body: liver, heart, pancreas, pituitary, and

joints. This condition is called iron overload syndrome or transosis. Iron excess can lead to organ damage in the form of liver fibrosis called cirrhosis, diabetes mellitus, joint disease, impotence in males, conduction abnormalities in the rhythm of the heart, and heart failure<sup>2</sup>.



#### **How is iron overload diagnosed?**

Iron is stored as ferritin in the body. Serum ferritin levels can be measured via a blood specimen obtained by a simple needle stick ordered by your physician. Levels can be unreliable at times because values are altered by inflammation and liver disease. Direct assessment of hepatic iron content by liver biopsy is the best indicator of the total body iron storage and can be safely performed by an experienced physician. As a non-invasive test the superconducting quantum interference device (SQUID) is an acceptable method for measuring liver iron<sup>3-5</sup>. Other methods such as MRI are promising but require further studies before they can be adopted for routine clinic use<sup>5</sup>.

*(continued on page 2)*

## Iron Overload in Sickle Cell Disease

(continued from page 1)

### How is iron overload treated?

Chelators are drugs that help in excretion of a metal ion. Chelation therapy is the only accepted method to manage transfusion-related iron over-load in SCD. Supplementation with vitamin C, may help to increase iron excretion<sup>5</sup>. Two chelating agents are currently available in the United States (US). Desferal (deferroxamine) is commonly given through a small mechanically driven pump which is adjusted to deliver the infusion subcutaneously over 8 to 12 hours, 5-7 nights/week. Desferal is well tolerated; however, compliance with prolonged infusion regimens is difficult and some serious side effects may occur that include increased risk of bacterial infections, anaphylaxis, and hearing and vision impairment<sup>6</sup>.

In November, 2005, the U.S. Food and Drug Administration approved the oral iron chelator Exjade (deferasirox) for the treatment of iron overload due to multiple blood transfusions in patients aged 2 years and older<sup>7</sup>. Exjade is a pill that can be dissolved in water and taken by mouth once a day. A short term study in SCD patients has shown Exjade to be comparable to Desferal in terms of efficacy<sup>8</sup>. Exjade is now commercially available and is preferably prescribed for SCD patients with iron overload. Common side effects of Exjade include nausea and abdominal pain; however, monitoring of laboratory tests measuring kidney and liver function and testing of hearing and vision before and during treatment is recommended. Studies to demonstrate long-term safety and efficacy in SCD patients are underway by the manufacturer.



### When should iron overload be treated?

The best indication to begin chelation therapy is a rise in liver iron stores to 7 mg/g dry weight<sup>5</sup>; however, serum ferritin levels above 1,000 ng/mL are most widely used for beginning and monitoring the therapy.



### Who amongst the SCD patients is at risk of iron overload?

Iron overload in SCD patients could remain undetected. Each milliliter of blood roughly provides 1 milligram of elemental iron. Iron overload, thus, may occur after 10 or more transfusions. Patients who have a history of chronic transfusion therapy or received multiple transfusions in the past should have a baseline screening with serum ferritin level. Please ask your hematologist or sickle cell physician about iron overload syndrome if you currently receive or have received transfusion therapy in the past.

by Hamayun Imran, M.D.  
Assistant Professor of Pediatrics

#### References

1. Ohene-Frempong K. Indications for red cell transfusion in sickle cell disease. *Semin Hematol*. Jan 2001;38(1 Suppl 1):5-13.
2. Kushner JP, Porter JP, Olivieri NF. Secondary iron overload. *Hematology Am Soc Hematol Educ Program*. 2001:47-61.
3. Brittenham GM, Cohen AR, McLaren CE, et al. Hepatic iron stores and plasma ferritin concentration in patients with sickle cell anemia and thalassemia major. *Am J Hematol*. Jan 1993;42(1):81-85.
4. Fischer R, Longo F, Nielsen P, Engelhardt R, Hider RC, Piga A. Monitoring long-term efficacy of iron chelation therapy by deferiprone and desferrioxamine in patients with beta-thalassaemia major: application of SQUID biomagnetic liver susceptometry. *Br J Haematol*. Jun 2003;121(6):938-948.
5. US Department of Health and Human Services. National Institutes of Health. National Heart, Lung, and Blood Institute. *The Management of Sickle Cell Disease*. 4th ed, rev June 2002. Publication 02-2117. Available at: [http://www.nhlbi.nih.gov/health/prof/blood/sickle/sc\\_mngt.pdf](http://www.nhlbi.nih.gov/health/prof/blood/sickle/sc_mngt.pdf).
6. Olivieri NF, Buncic JR, Chew E, et al. Visual and auditory neurotoxicity in patients receiving subcutaneous deferroxamine infusions. *N Engl J Med*. Apr 3 1986;314(14):869-873.
7. Oral drug for iron overload. *FDA Consum*. Jan-Feb 2006;40(1):5.
8. Vichinsky E, Onyekwere O, Porter J, et al. A randomised comparison of deferasirox versus deferroxamine for the treatment of transfusional iron overload in sickle cell disease. *Br J Haematol*. Feb 2007;136(3):501-508.

# Putting Things in Perspective

by Johnson Haynes, Jr., M.D., Director USA Comprehensive Sickle Cell Center

Access to health insurance is vital to the well-being of individuals with sickle cell disease. While known, but commonly ignored, individuals with genetic and chronic medical conditions have difficulties obtaining health insurance. Results of a large-scale survey published in the American Journal of Medical Genetics Part A (Vol. 143A(7):707-717, (2007), addressed experiences and attitudes encountered by individuals with genetic and non-genetic medical conditions when seeking health insurance. In this study 100 adults or parents of children with either sickle cell disease, cystic fibrosis, diabetes, and HIV (human immunodeficiency virus) and 200 adults with or at risk for breast or colon cancer experiences were compared. Of the 597 individuals

who responded to the survey, 27% had either been denied health insurance or offered insurance at a higher cost. Individuals with sickle cell disease and cystic fibrosis were twice as likely to report this bias in coverage when compared to individuals at risk for breast or colon cancer. The authors report that current laws which limit genetic discrimination in insurance are designed to address genetic risk or traits rather than actual disease. Stated differently, current legislation does not address the challenges faced by individuals with genetic conditions such as sickle cell disease or individuals at risk for conditions such as breast or colon cancer who try to maintain access to health insurance. Over one-third of all individuals responding to the survey, thought

there was a high chance that they would be denied health insurance in the future or their insurance would become unaffordable. Independent of the health condition previously described, all expressed concerns regarding the ability to obtain future health insurance. In my opinion, this study raises a very interesting question, "Should health insurance in America be an inherited right, or a privilege?" From the director's desk, remember the USA Comprehensive Sickle Cell Center wants to participate in your care.



Dr. Johnson Haynes, Jr.

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## Health Maintenance and Prevention

Gardasil, the first vaccine for prevention of cervical cancer, was approved by the United States Food and Drug Administration in 2006. This vaccine combats the common types of human papillomavirus (types 6, 11, 16, and 18) responsible for cervical cancer, vulvar and vaginal precancers, genital warts, and low grade cervical lesions. Genital human papillomavirus (HPV) is the most common sexually transmitted infection in the United States. It has been reported an estimated 6.2 million persons are newly infected every year with HPV.

The vaccine is approved for preadolescent girls age 9 years, adolescent girls, and women 26 years and younger. The vaccine should ideally be administered before potential exposure through sexual contact. The vaccine is not recommended during pregnancy. The vaccine is given intramuscularly in three separate injections (initial dose, second dose two months after the first dose, and third dose six months after the initial dose).

Gardasil is not a replacement for regular, routine cervical cancer screening annually by your Gynecologist. Gardasil is not a treatment for active genital warts or cervical cancer. For more information on the Gardasil vaccine contact your primary care provider or Gynecologist.

### References:

HYPERLINK "<http://www.uptodate.com>" [www.uptodate.com](http://www.uptodate.com)

HYPERLINK "<http://www.cdc.gov/mmwr>" [www.cdc.gov/mmwr](http://www.cdc.gov/mmwr)

Ardie Pack-Mabien, MSN, CRNP  
Clinical Nurse Practitioner

## Learn to Talk the Talk

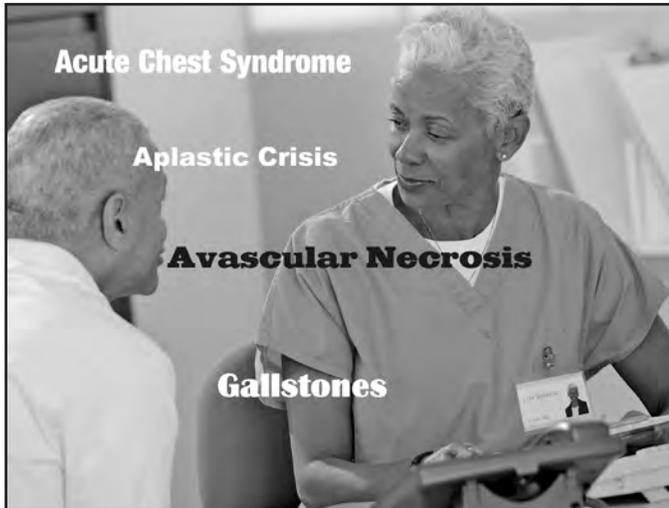
### Terms You Need To Know



Stephanie Durggin, RN  
Pediatric Sickle Cell Nurse Coordinator

#### **ACUTE CHEST SYNDROME:**

Individuals with sickle cell disease who have a new abnormality on chest X-ray with associated chest pain, coughing, difficulty breathing, and fever. The cause may be due to infection, inflammation or trapped red blood cells in the lungs.



#### **APLASTIC CRISIS:**

An episode when the bone marrow stops making blood cells. The blood counts may fall much lower than usual. If it happens, it is usually with a fever or infection.

#### **AVASCULAR NECROSIS:**

A condition in which poor blood supply to an area of bone leads to bone death. The head of the femur (hip) and shoulder are the most common sites of involvement, also called aseptic necrosis and osteonecrosis.

#### **GALLSTONES (CHOLELITHIASIS):**

Small pebble-like substances that develop in the gallbladder. Gallstones form in sickle cell disease from a substance called bilirubin.

## *Quick Reminder! Flu season is just around the corner.*

Adults and children who have a chronic disorder, requiring medical follow-up or hospitalization due to kidney disease, hemoglobinopathies (sickle cell disease), or conditions that compromise lung function should receive the flu vaccine annually. The flu vaccine promotes immunity to the influenza virus by stimulating specific antibody production.

The optimal time to receive the flu vaccine for adults and children is October-November and prior to exposure to the influenza virus. The flu vaccine can be given through the month of December and later as long as the vaccine is available. The flu season may last as late as May. There have been shortages of the flu vaccine in the past and it is strongly recommended you do not delay obtaining your flu vaccine.

Contact your primary care provider or local health department for the availability of the flu vaccine and an appointment.

## *9th Annual Blood Drive*

The 9<sup>th</sup> Annual Sickle Cell Center Blood Drive will be held on Saturday, September 22, 2007, at Franklin Primary Health Center at 1303 MLK Drive from 10 am until 2 pm. Forty-two productive blood donors presented last year (120% of the target goal). Eighty-four patients, their families, and friends were touched as a result of the generosity of donors. We thank all who came to show their support last year and look forward to this year's event. This year's blood drive will again be sponsored by the Alpha Phi Alpha Fraternity, Inc, the USA Comprehensive Sickle Cell Center, Sickle Cell Disease Association of America, Mobile Chapter, and Franklin Primary Health Center.

## ***From the Social Worker's Corner***

Many of the adult sickle cell patients we follow in the Comprehensive Sickle Cell Center are pursuing their vocational goals by attending college or working full or part-time jobs. Of course, there are always an industrious few that manage to accomplish both. However when a pain crisis or other sickle cell related complication occurs, the end result often leads to a disruption in their academic curriculum or excessive absences from work. When absences from work or school become extreme, the employer or class instructor(s) will typically request that some form of medical verification be presented that supports the patient's reason for being absent. Many universities and other institutions of higher learning currently have programs in place that provide assistance to students that have a documented disability. We encourage patients to consult with their individual institutions regarding program availability. An enormous benefit of this program is arranging for a specific class lecture(s) to be transcribed or recorded when a student is unable to attend class due to illness. Upon their return however, they may be requested to submit the appropriate documentation such as a hospital discharge summary or emergency room release orders. When a patient's illness results in an absence from work and the cause is justified, we will provide some means of medical verification to the employer if requested to do so.

Hopefully, many of you will find the above information beneficial and should you require any additional direction regarding this issue or any other issues perceived as socially challenging, please contact me at (251) 432-0301. Until next time, so long from the Social Worker's Corner.

*Adrienne Petite, LBSW*

## ***Welcome...***

*The University of South Alabama Comprehensive Sickle Cell Center welcomes Ms. Marilyn Chancellor to the staff. Ms. Chancellor joined the Center February 2007 and will serve in the capacity of Secretary V. She has been employed by the University of South Alabama, College of Medicine for seventeen years. She comes to the Comprehensive Sickle Cell Center with a wealth of experience, knowledge, and skills.*



*Ms. Marilyn Chancellor*

## ***Scholarship Opportunity***

### **Tylenol Scholarship Available**

Award Amount: \$1,000 - \$5,000

Total number of awards available 150.

Two-year and Four-year college student majoring in a health related field.

Application available on the following website: <http://scholarship.tylenol.com>

## CLINICAL TRIALS

### You can help doctors better understand sickle cell disease!

*Are you a male with sickle cell disease?*

*Do you have time to answer a few questions about sickle cell disease?*

If so, you may be eligible to take part in a survey study.

*What would I have to do?*

Give informed consent

Answer a few questions about your sickle cell disease

You do not have to answer any questions you do not want to answer

*Is that All?*

Our research staff will also record information from your medical record about other health problems you have had with sickle cell disease.

*How will this information be used?*

To gain a better understanding of complications of sickle cell disease

To improve treatment of certain complications of sickle cell disease

AS A THANK YOU FOR YOUR TIME, YOU WILL RECEIVE COMPENSATION FOR PARTICIPATING IN THE STUDY

For more information, please contact: University of South Alabama Comprehensive Sickle Cell Center at (251) 470-5893 or 470-5889.

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