

# Sickle Cell Today

## USA Comprehensive Sickle Cell Center

Volume 6 Issue 2

September 2009

### Table of Contents

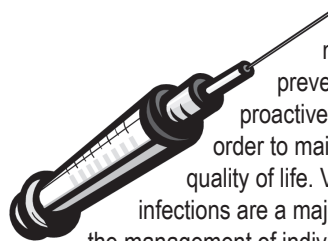
Flu Vaccine	1
Conference Gets Better and Better	2
From the Director's Desk Changing Trend in Health Care	2
Acute Splenic Sequestration Crisis	3
Social Worker's Corner	6
Blood Drive	6

USA Comprehensive Sickle Cell Center  
Main Office (251) 470-5893  
Fax (251) 470-5895  
Clinical Research (251) 471-7703  
Pediatric Clinic (251) 405-5147  
Adult Clinic (251) 470-5890  
Community-based Program (251) 432-0301



## Just Around the Corner, Flu Vaccine

Basic Facts and Recommendations - [www.cdc.gov/flu](http://www.cdc.gov/flu)



Health maintenance and prevention are key proactive steps to take in order to maintain a healthy quality of life. Viral and bacterial infections are a major complication in the management of individuals with sickle cell disease. The influenza infection is a contagious respiratory illness caused by the influenza virus and can cause severe morbidity in individuals with sickle cell disease. Flu viruses spread in respiratory droplets caused by coughing or sneezing and direct or indirect contact. Most healthy adults are contagious and spread the infection to others as early as one day before symptoms develop and up to five days after becoming sick. **The influenza vaccination is recommended yearly for individuals with chronic illness and it's the best way to prevent the flu. Proper hand washing with soap and water is also an effective mechanism to prevent the spread of germs.** The Centers for Disease Control reports on average 5% to 20% of the United States population gets the flu every year, more than 200,000 people are hospitalized from flu complications, and about 36,000 people die from the flu and associated flu complications such as bacterial pneumonia. The flu vaccine promotes immunity to the influenza virus by stimulating specific antibody production. Antibodies are proteins that help the body fight infections. Each year the flu vaccine is reformulated according to the United States Public Health Services standards and must be given every year to prevent the flu. The optimal time to receive the flu vaccine for adults and children is October-November and prior to exposure to influenza. The flu vaccine can be given into the month of December and later as long as the vaccine is available. The flu season may last as late as May. However with the recent flu epidemics and vaccine shortage, it is advantageous to receive your flu vaccine early.

### What are the symptoms of the Flu?

- high fever
- headache
- muscle aches
- extreme tiredness
- dry cough
- sore throat
- runny or stuffy nose
- stomach symptoms (nausea, vomiting, diarrhea)

### Who Should Receive the Flu Vaccine?

- Persons > 65 years of age
- Adults and children with a chronic disorder
- Adults and children who have required medical follow-up or hospitalization because of renal dysfunction, hemoglobinopathies (sickle cell disease), or immunosuppression.
- Adults and children with conditions which may compromise respiratory function
- Women who will be pregnant during the flu season
- Children 6-23 months of age
- Healthy adults 50-64 years of age
- Individuals who are in close contact with children 0-23 months of age
- Healthy persons who may transmit influenza to those at risk
- Persons who smoke

### Who Should Not Receive the Flu Vaccine

- Infant younger than 6 months of age
- Individuals with allergy to the influenza vaccine
- Individuals with allergy to eggs
- Individuals with infection or fever

### Possible Side Effects

- Irritation at site of injection
- Muscle pain

### Reasons to Call Your Healthcare Provider Immediately

- Signs of life-threatening reaction-wheezing, chest tightness, fever, itching, bad cough, blue skin color, swelling of face, lips, tongue, or throat.
- fast heart rate
- seizures
- hives
- rash
- weakness

continued on page 5

## Conference Gets Better and Better Each Year

“Informative, excellent program, well planned, and simply wonderful” are just a few of the 2009 conference attendees comments describing the annual sickle cell conference entitled “Practical Issues IX: Bringing Hope to Healthcare in Sickle Cell Disease”. The speakers’ evaluations were rated as knowledgeable and excellent. The Cecil Parker Distinguished Lectureship featured James R. Eckman, MD, Professor, Department of Internal Medicine, Division of Hematology and Medical Oncology at Emory Winship Cancer Institute and Director of the Georgia Comprehensive Sickle Cell Center, Grady Health System in Atlanta. Dr. Eckman spoke on, “Health Disparities in Sickle Cell Disease”. Other conference speakers included: J. Hoxi Jones, Strategic Partnership Specialist, Texas Health and Human Services Commission-Houston; Kathy Hall, Deputy Commissioner of Program Administration, Alabama Medicaid; and Felicia Wilson, MD (pictured), Associate Professor, Department of Pediatric Medicine, Hematology/ Oncology Division, University of South Alabama College of Medicine. Ms. Jones, a patient advocate, spoke on the value of patient empowerment and the need for the patient to assume a lead role in their care. Ms. Hall updated the audience on the current state of Alabama Medicaid and new programs that are in their early implementation phase. Dr. Wilson addressed the pros and cons of stem cell transplantation as a potential curative treatment in sickle cell disease.



The 2009 conference presented the Planning Committee with a record breaking, enthusiastic response with over one hundred registrants, which prompted the relocation of the conference for the first time in conference planning history. Health care professionals, social workers, parents, and clients attended the 2009 half day conference. Conference participants took advantage of the opportunity to freely network and interact with the conference organizers and speakers. The conference offers the latest information on the most innovative treatment options in the management of sickle cell disease and its complications. The conference is held annually during the spring and offers continuing education credits to conference attendees. For additional information regarding future conferences or to be added to the conference mailing list, contact our office at (251) 470-5893.

## FROM THE DIRECTOR'S DESK

### **A Changing Trend in Health Care Delivery for Adults with Sickle Cell Disease: The Need for More Primary Care Physician Involvement**

One of the major goals of Healthy People 2010 is to eliminate health disparities with substantial interest given to disparities associated with race and ethnicity. Sickle cell disease (SCD) is a genetic disorder that is seen in individuals of various ethnic backgrounds, but in the U.S. is most prevalent amongst Americans of African descent. In comparison to whites, minority Americans lag far behind in health care quality measures including effective patient-physician communication, overcoming cultural and linguistic barriers, access to health care and insurance coverage. In SCD, this is compounded by ethnic/racial driven under-treatment of recurring, acute, pain episodes, and chronic pain syndromes (the most common reason medical attention is sought) along with negative attitudes towards effective pain management by nurses and physicians.

In spite of the barriers listed above, which negatively impact the quality of health care, mortality and morbidity for the child, adolescent, and adult with SCD have improved over the last two decades as a result of educational and medical advances. Higher survival rates have increased the numbers of adolescents confronting the issues of transitioning from pediatric to adult services. With improved survival, the lack of access to healthcare providers with expertise in the management of SCD is a growing, national problem,

particularly for adults. In the United States, the number of non-malignant hematologist in training continues to decline. This leaves a greater burden of responsibility on the primary care provider in delivering care for their SCD clients. With increased survival, physicians are also seeing more chronic end organ damage, such as, chronic kidney disease and pulmonary hypertension, not previously experienced. This begs for not only a need for more primary care involvement, but also the involvement of sub-specialists beyond hematologist, i.e., nephrologist, pulmonologist, cardiologist, neurologist, etc.

With this being said, the question has to be raised whether the hematologist alone can carry the brunt of responsibility in the care of the sickle cell client. If not, who other than primary care providers can assume some of this responsibility? In order to have primary care providers who are comfortable and capable of caring for individuals with SCD, academic sickle cell centers must lead by creating educational and clinical opportunities for residents in primary care disciplines (internal medicine, and family medicine) to engage in the general care of adult clients with SCD. This will not obviate the need for hematologist, but provide a wider net of health care access to patients with SCD. With this primary care net in place, centers with hematologist and sickle cell specialist can serve as a resource to underserved communities and referral site when more complex care is needed.

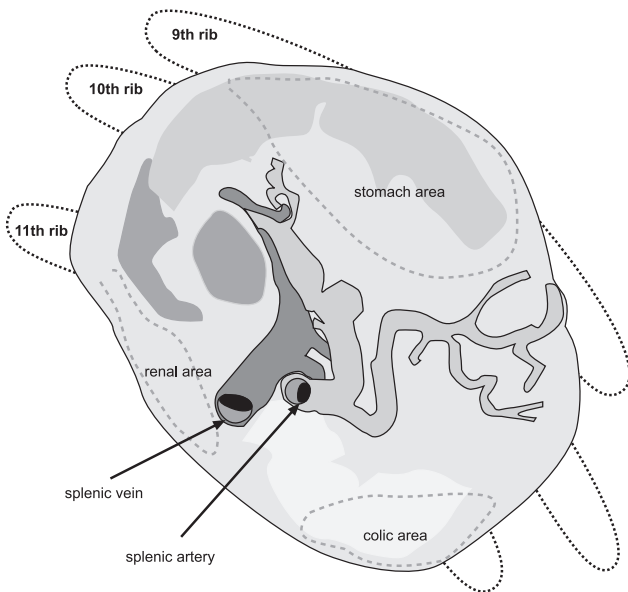
Again, many, many thanks.

Johnson Haynes, Jr., MD  
Director, USA Comprehensive Sickle Cell Center

# Acute Splenic Sequestration Crisis (ASSC)

by Dr. Arati Rao & Stephanie Durggin RN

**Splenic Sequestration:** It is a complication that can occur in patients with sickle cell disease which can be life threatening. Sequestration is caused by blood being trapped in the spleen. This acutely reduces the red cells in the body circulation and when severe may cause shock and lead to death. Splenic sequestration is often preceded by acute infections such as viral or bacterial infections.



## **What is a spleen and why is it important?**

**Spleen:** An organ on the left side of the body that may be felt below the rib cage.

The spleen has many functions:

- helps in immunity (protection against infection)
- destroys bacteria
- destroys worn out and damaged platelets (graveyard for old cells)
- destroys worn out and damaged red blood cells

## **Why is this important to know?**

In sickle cell disease the spleen usually does not work after 4-6 months of age. It is constantly damaged by the sickled red blood cells (RBC's) and is therefore unable to remove bacteria from the blood. This means that bacteria can grow in the blood and cause septicemia or blood poisoning (infection of the blood).

In Sickle Cell Anemia (SS), this complication can occur as early as 2 months of age. Infants and

young children with SS are at greatest risk of splenic sequestration. By the age of five years, the spleen shrinks and becomes stiff (fibrosed) due to repeat damage from sickled red blood cells and hence it cannot enlarge any more. However in patients with SC disease and Sickle Beta-Plus Thalassemia, spleen damage is less severe and hence the spleen is able to trap and cause sequestration even as an adult.

A study of 14 children who had 20 episodes collectively in which four children died, four had one or more recurrences, four have splenic involution, and two had their spleens removed. The recurrences were within four months of the previous ASSC. An infant or child that has had one episode of splenic sequestration is more likely to have other episodes.

## **What are the symptoms?**

Your child may experience any of the following symptoms:

- Weakness
- Irritability
- Unusual sleepiness
- Paleness
- Big spleen
- Fast heart beat
- Pain in the left side of the abdomen

## **Precautions to be taken:**

- Educate all family members and caretakers about this potential complication
- Practice palpating (feeling) for the spleen on regular basis and when child is ill
- Learn the symptoms that your child may experience with this complication
- Ask to review splenic sequestration video (available during clinic visits)
- Have list of phone numbers for: Hematologist and other emergency contacts- readily available. Provide a list to everyone that cares for the child- grandparents, aunts, daycare, schools, just to name a few.

## **What should you do?**

Have your child seen by a physician immediately.

### ***What treatment should you expect?***

- Immediate blood transfusion is given if the blood count is dangerously low.
- IV Fluids
- Pain medications-when needed
- Antibiotics-when needed
- Blood draws (labs)
- Frequent monitoring of vital signs

If a child experiences several episodes of splenic sequestration, current recommendation is to consider surgery to remove the spleen. However, this should be discussed with your child's hematologist to determine what needs to be done. Sometimes for children under the age of 2 years, surgery is delayed until they reach 2 years, however, the patient will require monthly transfusion until that time. Referral to a surgeon will be done, when the decision for surgery has been determined. All surgeries should be coordinated with the hematologist/ sickle cell physician.

Your child will be required to have the pneumococcal vaccine (PPV-23) and meningococcal vaccine at least 4-6 weeks prior to the date of surgery is to be done.

#### **Important:**

**Prophylactic penicillin will be required to be taken twice-a-day (everyday) after surgery.**

Again, we are available for any questions or concerns you may have about this or other complications that may occur. It is very important to keep regularly scheduled clinic appointments with the hematologist/ Sickle Cell Doctor to discuss potential complications and their management during the visit.

#### References:

Acute splenic sequestration in young children w/ sickle cell disease/Clinical Pediatrics, Vol. 11, No. 12, 701-704 (1972), DOI: 10.1177/000992287201101214

**Sickle Cell Information Center Guidelines:** Edited by James Eckman, M.D. and Allan Platt, PA-C  
The role of the spleen in sickle cell disease by Lewis Hsu, M.D., Ph.D.  
reviewed by Laura Jana, M.D., F.A.A.P.

NEPSCC-New England Pediatric Sickle Cell Consortium  
Project # MCJ-481004 and # 2H46 MC00232-02 from the Maternal and Child Health Bureau (Title V, Social Security Act).Adapted from materials by the Texas Department of Public Health Newborn Screening Program.

*continued from page 1*

Although the flu vaccine is the single most effective way to protect your self against the flu; there are also some other helpful tips:

- avoid close contact with people who are sick
- stay home when you are sick to prevent the spread of germs
- cover your mouth and nose when coughing or sneezing
- wash your hands with soap and water
- avoid touching your eyes, nose or mouth

Contact your health care provider to make an appointment for your flu vaccine. Remember the earlier the better just in case there is a shortage of the vaccine.

Ardie Pack-Mabien, CRNP  
Clinical Nurse Practitioner

## Swine Flu vs Seasonal Influenza; What's the difference?

[www.cdc.gov/h1n1flu](http://www.cdc.gov/h1n1flu)

Swine Flu (novel H1N1) is a new influenza virus causing illness in people. In the United States, this new virus was first detected in humans in April 2009. The virus spreads from person to person in much the same way as the seasonal influenza viruses. High risk individuals include people 65 years and older, children younger than five years of age, pregnant women, and people of any age with certain chronic medical conditions such as sickle cell disease and asthma.

### **What's the difference between swine flu and seasonal flu?**

Seasonal flu seasons vary in terms of timing, duration, and severity which differ from swine flu. The largest number of swine flu confirmed and probable cases have occurred in people between the ages of 5 and 24 years old.

### **What are the signs and symptoms of Swine Flu?**

Signs and symptoms are similar to seasonal influenza and include:

- fever
- cough
- sore throat
- runny or stuffy nose
- body aches
- headache
- chills
- fatigue

\*diarrhea and vomiting has been reported in a significant number of people who have been infected with Swine Flu.

### **How does swine flu virus spread?**

The virus can spread from person to person through coughing or sneezing by people infected with the virus. Sometimes people may become infected by touching something with the virus and then touching their mouth or nose. Center for Disease reports studies showing the influenza virus can survive on environmental surfaces and can infect a person for up to 2-8 hours after being deposited on the surface.

### **What can we do to protect ourselves?**

- Cover mouth and nose with a tissue when you cough or sneeze; throw the tissue away after you use it.
- **Wash your hands often with soap and water**; alcohol base hand cleansers are also effective.
- Try to avoid close contact with sick people.
- **Stay at home if you are sick until you have been free of symptoms for 24 hours.**

### **What should I do if I think I might have Seasonal flu or Swine Flu?**

- Contact your primary care provider or family physician for medical evaluation and management.

Ardie Pack-Mabien, MSN, CRNP  
Clinical Nurse Practitioner



University of South Alabama  
 Comprehensive Sickle Cell Center  
 2451 Fillingim Street, MCSB 1530  
 Mobile, AL 36617-2293

Non-Profit  
 U.S. Postage  
 PAID  
 Mobile, AL  
 Permit No. 506

## FROM THE SOCIAL WORKER'S CORNER

The community-based Sickle Cell Disease Association of America-Mobile Chapter (SCDAA-MC) continues to engage in efforts that will further enhance the quality of client services. Although many of our local clients can access services and programs with virtual ease, the task for providing comparable services to clients residing in the rural extremes can be a bit more challenging. The SCDAA-MC services nine counties. In addition to Mobile, these include Baldwin, Choctaw, Clarke, Conecuh, Covington, Escambia, Monroe and Washington. Much of our time and effort are spent coordinating services with agencies and medical providers located outside the parameters of our local community. And while many of us are aware that clients residing in out lying areas experience a great deal of difficulty in accessing medical care, others may be surprised to learn that the guidance or direction needed to access many community resources is also often lacking. It is important to note that clients residing outside the Mobile community are entitled to the same services that clients have within the community. The only exception is that transportation provided by SCDAA-MC is typically restricted to the inner city limits. Many clients residing in the rural extremes often use lack of transportation to justify noncompliance with respect to missed clinic appointments, laboratory tests and procedures. What many clients may not realize however is that transportation for these medically related commutes can be coordinated by joint efforts facilitated by the SCDAA-MC and Rural County Transportation Provider(s). If the client is a recipient of Medicaid, the provider is typically reimbursed for their travel by the Alabama Medicaid Non Emergency Transportation Program.

If you would like any additional information regarding this or any other program service, please feel free to contact me at (251)432-0301. Until next time, so long from the Social Worker's Corner.

Adrienne Petite, LBSW  
 SCDAA-MC



## Every Three Seconds, Someone Needs Blood

Remember September is "National Sickle Cell Awareness Month". The 2009 community blood drive has been scheduled for Saturday, September 26, 2009 at Franklin Primary Health Center at 1303 MLK Drive from 10am until 1pm. The 2008 blood drive yielded 67 individuals presenting as potential donors. Twenty-one donors were first time donors at the 2008 blood drive. A total of 54 productive units were obtained. Each unit donated in 2008 was separated into red cells and plasma touching the lives of 108 individuals, their families, and friends in the surrounding community. Please come out and participate in the blood drive by "Giving the Gift of Life through blood donation".