



Sickle Cell Center for Adults at Johns Hopkins

Aug 2004, Quarterly Newsletter

EDITOR'S NOTE

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UPCOMING EVENTS:

- SICKLE CELL
PICNIC AT
PATTERSON
PARK
11 AM-4 PM
SEPT 19TH
- SICKLE CELL
WALKATHON
AT DRUID HILL
PARK FROM
9AM-3PM
SEPT 26th

In this month's newsletter you will find information on pulmonary hypertension, pregnancy outcome in Hb SS patients, substance abuse services, patient's corner, and voting registration.

How can you help? Many of you have voiced your opinions about the wait in the Emergency Department when you arrive in crisis. Some have even asked me "What programs or functions are held in honor of sickle cell patients?" You might have even wondered why there isn't a sickle cell center in Baltimore. These questions are valid and have motivated our sickle cell team to work harder for you. But we can't make the needed improvements without the help of our sickle cell population. One voice may not be heard, but it is hard to ignore hundreds of sickle cell patients who come together.

Did you know that ONE sickle cell patient came to our first walk-a-thon? Did you know that only 4-5 patients attend our support group monthly. We can help you only if you are willing to help yourself. If all of our patients attend the picnic and walk-a-thon, we can go to congress and lobby for a 24 hour sickle cell center.

Why do we need a center dedicated to the care of sickle cell patients? We need a comprehensive 24 hour sickle cell center run by professionals who are: trained in caring for patients with sickle cell disease, compassionate, and culturally sensitive. You deserve the best care. Come and be counted at the picnic, walk-a-thon, support group, and any other future events. Thank you for your time and support.

By Yvette Roane, PA-C

PULMONARY HYPERTENSION AND ACUTE CHEST SYNDROME IN SICKLE CELL DISEASE

Introduction: Sickle cell disease (SCD) is the most common genetic disorder affecting African-Americans. Many patients with SCD suffer from strokes, acute chest syndrome, bone pain and other complications thought to arise from obstruction due to excess red blood cell sickling within the blood vessels. These changes contribute to what has been termed vaso-occlusive events. Lung problems associated with SCD, such as pulmonary hypertension (PH) and the acute chest syndrome (ACS) cause many problems for those with sickle cell. Recurrent episodes of ACS develop in many patients (between 20 and 80%), often progressing to chronic lung disease and PH, although the link between ACS and PH in SCD has recently been questioned. The prevalence of PH in SCD is not known, however, it has recently been suggested, based on echocardiographic screening, that about one third of adult patients with SCD develop PH. This study by Gladwin and colleagues, published this year in the *New England Journal of Medicine*, suggests that SCD patients with PH have a significantly higher risk of death compared to patients without PH. However, since not all deaths in SCD patients with PH are directly attributable to complications of high pulmonary pressure, it remains unclear whether PH is a direct cause of death or a mere marker of disease severity.

Disease Mechanisms: A growing body of knowledge indicates that significant disturbances in the nitric oxide (NO) pathway, which normally helps blood vessel to relax, contributes to the development of sickle cell-induced vaso-occlusive events. Nitric oxide is generated by cells which line the inner part of all blood vessels. This has led us to conclude that vaso-occlusive events, such as ACS, and PH may be related to general blood vessel wall dysfunction and decreased nitric oxide availability.

Potential Therapy for Complications of SCD: Recent promising therapies, such as inhaled nitric oxide or hydroxyurea (a drug used to treat patients with SCD, which prevents RBC sickling and increases hemoglobin F), may act through their effects on nitric oxide. For instance, increased pulmonary artery pressures seen in children with ACS can be lowered by inhalation of nitric oxide. In addition, short-term oral administration of arginine which is needed to make nitric oxide, also leads to improvement in symptoms in patients with sickle cell disease and pulmonary hypertension.

Current Research in SCD: In a collaborative effort between the Pulmonary, Hematology, and Cardiology Divisions at Hopkins and investigators at Howard University, we hope to receive funding through the National Institute of Health to do a study that will help us better understand what happens in the lungs of patients with sickle cell disease.

By Paul Hassoun, M.D.

PREGNANCY OUTCOME IN HEMOGLOBIN SS PATIENTS

A study involving 94 pregnancies in 52 subjects and 157 pregnancies in 68 controls was conducted in Jamaica (Obstet Gynecol 2004;103:1278-85 by the American College of Obstetricians and Gynecologists) to determine if sickle cell patients with Hb SS disease were more likely to have pregnancy related complications. Subjects were pregnant Hb SS patients, while the controls consisted of pregnant females without sickle cell disease. The study participants were screened from birth to pregnancy, and followed during pregnancy. Outcome measures included the age at menstruation, years to pregnancy, result of pregnancy, and complications during pregnancy. Study participants were followed for 3-6 months and motivated to return for care when ill. They received a thorough examination and supportive care during pregnancy. Blood work was taken at 6 month intervals on well subjects and controls.

Prophylactic transfusions, a preventive measure, was not an option in Jamaica for those subjects not displaying symptoms. Subjects with symptoms were transfused during 10 of the pregnancies (4 acute chest syndrome, 3 postpartum hemorrhage, 2 anemia, 1 multiple complications) and 4 controls (2 postpartum hemorrhage, 2 anemia). Transfusion therapy decreased pain crisis during pregnancies and episodes of acute chest syndrome, but had no effect on birth weight. Gestational hypertension was more common in Hb SS patients, but there were no increased instance of pregnancy induced hypertension or pre-eclampsia.

The average age at first menstruation was 15 years old in subjects and 13 years old in the control group. First pregnancy occurred earlier in controls at age 20 compared to age 23 year olds in Hb SS patients. The difference between the two groups was very small. Subjects had more spontaneous abortions, fewer live deliveries, more low birth weight babies, and more premature infants. Spontaneous abortion was defined as an abortion occurring before 24 weeks. It was found to be the major culprit to fetal loss. Pre-maturity was defined as pregnancy at 37 weeks, and low birth weight as birth weight below 2500 grams. Successful pregnancies occurred in 57% of subjects, compared to 89% successful pregnancies in the control group. The main reason for fetal loss was attributed to spontaneous abortions.

The previous assumption of infertility or lack of sexual response in Hb SS patients was not a finding in this study. One new finding showed that a retained placenta was more common in Hb SS patients. Subjects usually gave birth on average at 37 weeks, while the average gestational age for the control group was 38 weeks. Total hemoglobin and fetal hemoglobin were factors influencing clinical complications. Two subjects died from pregnancy related complications, which is between 100-500 times greater than controls. The outcome of Hb SS pregnant patients remains variable and unpredictable. A better understanding of the mechanisms involved could improve our success rates for Hb SS patients.

COMMUNITY HEALTH CHARITIES

Community Health Charities of the National Capital Area helps our organization and many others obtain financial support. If you would like more information on Community Health Charities, visit their website at www.healthcharities-nca.org. If you are interested in donating to the Sickle Cell Center for Adults at Johns Hopkins, please contact Yvette Roane at (410) 502-6997. For federal government employees, our donation code is #6017 through your workplace giving campaigns.



SUBSTANCE ABUSE SERVICES AVAILABLE

My name is Brian Estes and I am the substance abuse counselor for the Sickle Cell Center for Adults at Johns Hopkins. This is a brief introduction to a new service that is available for patients in the sickle cell clinic. I have joined the sickle cell team along with Dr. Sophie Lanzkron and Physician Assistant Yvette Roane. My clinic day is Friday morning. I evaluate, educate, and counsel patients in clinic who are on narcotics. My goal is to provide a support system and educate individuals on the side effects of narcotics. The initial interview involves a brief history, discussion of coping skills, and completion of a questionnaire on depression. The effective management of your pain is vastly important in your total health care. What is important to you is important to me.

Addiction is a concept that is often misunderstood and not mentioned often in connection with sickle cell disease. Addiction and dependence are terms commonly used together, often confused, but are very different.. Addiction is an illness often involving negative behaviors leading to misuse of a drug, problems with social life or work, and/or evidence of tolerance or withdrawal. An example of addiction would be if you made attempts to alter your prescription, or buying narcotics on the street illegally. The term dependence involves physical dependence and actual withdrawal symptoms which are experienced when the medication is stopped abruptly. An example of withdrawal symptoms might include stomach pain, backache, diarrhea, tremor, and/or running nose. A small percentage of our patients are addicted to narcotics. Many of our patients are dependent and we anticipate that this will happen when we start patients on narcotics. The goal is to make sure patients don't become addicted.

Many sickle cell patients often are introduced at an early age to pain medications. Problems arise when the medication becomes more important than the sickle cell crisis and its complications. The importance of your living situation, the stress associated with having a chronic illness, and your family support system are a few factors influencing your life. The ability to understand and cope with your pain is enormously important in managing your disease. The ability to manage your pain medications in an appropriate and effective manner is important, and some people need the assistance of a skilled professional.

My services are available to help you manage your narcotic usage. The balance between pain management and safe narcotic use can be maintained through education and support.

By Brian Estes

PATIENT'S CORNER:**A LITTLE COMPASSION GOES A LONG WAY**

Sickle cell disease or sickle cell anemia, call it what you want. It's what I have, not who I am.

I feel so fortunate to be here in this world. I have been blessed with my husband of 14 years of marriage and a beautiful 5 year old son. I try to live my life simple and in peace. Like many of you, I am too a wife, mother, daughter, sister, employer, teacher, friend, and yes— even a patient.

A sickle cell crisis is very, very painful. The pain can attack any part of my body from head to toe. The pain is sooo deep inside your body. I know doctors try to help us by asking us what's our pain level on a scale from 1 to 10, with 10 being to worst pain ever- so that they can try to make us as comfortable as they possibly can. The way I describe my pain is like this: it exerts power, and is sharper than any two edge sword (saw), and pierces even to the dividing of my soul, joints, and marrow.

For me there is only one thing that's worst than having sickle cell disease or a sickle cell crisis: a doctor who shows no compassion. I feel bad already for having to leave my family at home and come to the hospital in great pain. How dare they come to me and say "You are still in a lot of pain? I gave you a lot of pain medication." Or they may just give me a look like 'you can't be in that much pain, I just gave you some more drugs.'

Please try to remember it's not always what you say, but how you say it. When we, the patients, have to come to the emergency room in great pain, you don't have to embrace us or cherish us. No, that is not why we come to the hospital.

I just want you to know you can give me all the pain medication in the world. There is nothing that would ease my (heart) pain more than if you showed me a little COMPASSION.

The next time a sickle cell patient have to come to the emergency room, please try your best to treat us like you would want your IMMEDIATE FAMILY MEMBER.

And always remember: A LITTLE COMPASSION GOES A LONG WAY.

By Shellie Dejesus

P.S. Thank you so much Yvette. You are the best, and I adore you.

We are on the web!
sicklecellcenter.org

JHMI

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at Johns Hopkins

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Clinic: Tuesday 8a-12p, Friday 9a-12p



SICKLE CELL TEAM
SOPHIE LANZKRON, M.D.
YVETTE ROANE, PA-C

UPCOMING SUPPORT GROUPS:

WHAT: SICKLE CELL SUPPORT GROUP- CHILDCARE AVAILABLE
WHERE: JOHNS HOPKINS OUTPATIENT CENTER, CONFERENCE ROOM 2140
WHEN: 6:30PM-8PM, 2ND THURSDAY OF THE MONTH
DATES: SEPT 9th, OCT 14th, NOV 11th, DEC 9th

IMPORTANCE OF VOTING

I would like to urge you to go out to the polls this November to vote. It is very important to make your vote count. Many people think their vote doesn't matter. Well I disagree. One vote can and has made a difference. Voting makes your citizenship count. You have an active voice in the decisions that shape the world. Your vote will help decide the following: who will lead our country, how the national budget will be spent, the fate of our soldiers, and the appointment of 180 federal judges.

Why is voting such a milestone for African Americans? Well before the civil war, qualifications for voting were not a concern for the Constitution or the federal laws. At that time, a few northern states allowed a small percentage of free black men to register and vote. Slavery and restrictive state laws prevented black people from voting. The 14th Amendment gave citizenship to all persons born in the United States when it was approved in 1868. In 1870, the passing of the 15th Amendment stated that the right to vote shall not be denied or reduced on the basis of race, color, or slavery. The Enforcement Act of 1870 held criminal penalties for interference with the right to vote. As a result, recently freed slaves registered to vote. Black candidates began for the first time to be elected to state, local, and federal offices and play a meaningful role in their governments. Think of the future of yourself, your children, and of those ancestors who fought to give us a voice. You can register to vote by calling 1-866-316-VOTE or log onto yourvotematters.com. See you at the polls.

MARK YOUR CALENDARS: NOVEMBER 2ND, ELECTION DAY.

By Yvette Roane, PA-C