

SICKLE CELL CENTER FOR ADULTS AT JOHNS HOPKINS

February 2004, Quarterly

EDITOR'S NOTE

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BLACK HISTORY MONTH



In this quarter's newsletter you will find information on Community Health Charities, Sickle Retinopathy, Hematology's new anticoagulation clinic, Why Cells Sickle, Patient's Corner, Support Group, and Black History Awareness Month. February is black history awareness month. Take a moment to reflect on the many accomplishments of African Americans and test your knowledge on page 6. If you would like to contribute something to our Patient's Corner, please contact Yvette Roane at (410) 502-6997 or by email at yroane1@jhmi.edu.

As most of you know, I have been out on maternity leave for the past two months. I am back to work now in my full capacity. I gave birth to a precious baby girl. Her name is Imani. She was 9lbs 8 oz at birth. Imani developed an infection hours after birth and was hospitalized at Johns Hopkins Children Center ICU. It was the scariest time of my life. The optimum treatment methods available at Hopkins nursed Imani back to good health. I am so grateful to Hopkins Children Center for the outstanding care they provided. No one ever wants their child to be a patient, but if it happens, I recommend The Johns Hopkins Children Center. Thank you from me and my family.

By Yvette Roane, PA-C

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](http://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.



## LASER TREATMENT FOR SICKLE RETINOPATHY

When sickle cell disease affects the eye (sickle retinopathy), blindness can result if left untreated. Fortunately, timely laser treatment can halt or prevent vision loss. The retina is a delicate tissue at the back of the eye containing photoreceptors necessary for eyesight. Normally, blood vessels present since birth nourish the retina, but in sickle cell disease, additional vessels develop later in life. These abnormal vessels are fragile and can bleed in front of the retina, blocking vision. After bleeding, vision sometimes returns as the blood disappears. However, scar formation, retinal detachment, and permanent loss of vision can also result.

The risk of sickle retinopathy is highest in individuals with SC disease. Sickle cell carriers are not at risk for sickle retinopathy. With routine eye exams, abnormal blood vessels can be identified when they first appear. You and your eye doctor can decide whether to carefully monitor these vessels or to treat them with laser. Laser retinal photocoagulation is safe and causes very little discomfort. For patients with sickle cell SS or SC disease, regular eye exams are an important way to preserve useful eyesight.



By Geoffrey G. Emerson, MD,

## HEMATOLOGY ANTICOAGULATION CLINIC

Over 250,000 people in the US develop blood clots in their legs (known as “deep venous thrombosis or DVT”) or lungs (known as “pulmonary embolism or PE”) each year. Deep venous thrombosis and pulmonary embolism are treated with anticoagulants (blood thinners) at first heparin and then warfarin, which prevent further clotting and allow the clot to gradually resolve. Since warfarin makes the blood less likely to clot it puts patients taking it at a higher risk of bleeding. Therefore, it is very important for patients taking warfarin to be closely followed. Fortunately, the Hematology division has opened a new Anticoagulation clinic to provide expert assistance for patients taking warfarin. The clinic is located on the 7th floor of the Johns Hopkins Outpatient Center in Suite B and is open from 8 AM to 12PM Monday, Wednesday, Thursday and Friday. The clinic staff, Michael Streiff, MD, Paula Biscup, PharmD and Marla Polo-Zisa, PA-C specialize in the management of patients requiring anticoagulation and look forward to helping any patients with sickle cell anemia on warfarin.

By Michael Streiff, M.D.



## WHY SICKLE CELLS SICKLE?

Since the red blood cell is the most visibly affected cell in the blood of patients with sickle cell anemia, there is a tendency to ignore the other elements of the blood: white blood cells, platelets and plasma. While much of this attention is well deserved due to treatment with hydroxyurea, there is a growing realization that the sickle red cell does not operate in isolation, but rather interactions with other elements of the blood also play a role in causing the symptoms of sickle cell anemia. The red blood cell can be thought of as the tractor-trailer truck of the body. In the lungs it picks up oxygen that it transports to the other cells of the body through the blood vessels. Once the red cell has delivered its payload of oxygen, it picks up carbon dioxide, the waste product of the body's metabolic furnace, and transports it back to the lungs to be disposed of (exhaled).

The red cell is able to perform its job because it contains hemoglobin, a protein that can bind and release oxygen and carbon dioxide. In patients with sickle cell anemia, the structure of hemoglobin is altered so that when it releases oxygen it becomes sticky and aggregates into large masses which can distort the shape of the red cell such that it can no longer fit through the blood vessels. The result is clumping of cells. Red cells flow into the vessel, but are unable to get through. Therefore, the cells served by these vessels become oxygen-starved. The cellular consequences of a lack of oxygen are the same whether it occurs in the heart (a heart attack) or in the bones of the leg or back (a sickle cell crisis) – PAIN.

This is the concept of a sickle cell crisis that I was taught when I was a medical student (which despite my appearance was not that long ago). Red cells generally do not spend enough time in the small vessels to become distorted and form a blockade. Sickling only happens when red cells are waylaid in these tiny vessels, which is where white blood cells and platelets come in. White blood cells fight off infection and release chemicals that make vessel walls stickier so that blood cells are more likely to stick. This observation may explain why crises seem to be triggered sometimes by infections. Platelets, the blood cells which aid in clotting when we cut ourselves, also seem to be involved. When activated, they release proteins that cause sickle cells to stick to vessel walls. These platelet proteins may slow down sickle red cells enough to "sickle" and cause a crisis. In support of this theory, some sickle cell researchers have found evidence that platelets and clotting proteins in the blood are activated during sickle cell crises. Thus far, however, blood thinners have not proven to be useful for sickle cell disease. A collaborative study at Johns Hopkins, Howard University and the National Institutes of Health is looking at clotting proteins in patients with sickle cell disease and high blood pressure in their lungs.

Fortunately, there are a number of things sickle cell patients can do to prevent crises. Avoid dehydration. The blood gets thicker and flows slower when you are dehydrated. Drinking plenty of water and avoiding alcohol (which can cause dehydration by telling the kidneys to make more urine) are useful preventive medicine. Since sickling is caused by low blood oxygen levels, it is also important not to smoke. Cigarette smoke has carbon monoxide (the same stuff that comes out of the tail pipe of your car), which prevents the hemoglobin in your red cells from binding oxygen. It can trigger sickling so you should avoid smoking. Since infections can trigger crises, it is also important that you make sure you get an influenza (flu) vaccine each year and a pneumococcal vaccine every 5-6 years to protect you against infections. Finally, we are fortunate that hydroxyurea not only changes the red blood cells making them less likely to sickle but also decreases white blood cell and platelet counts making them less likely to trigger sickle cell crises. If you have questions about what you can do to prevent crisis, please contact us.

By Michael Streiff, M.D.

## PATIENT'S CORNER

### Inner Thoughts of Roger Carter

Let me tell you a little about my life  
Both before and after meeting my wife

I was born in Washington, DC  
I went to school and worked in that city

My younger brother Steve and I have Sickle Cell disease  
Both of us were small, fragile, and teased

Our mother was very protective and I'm sure a lot afraid  
Having one child die at a very early age

My mother said I was in and out of the hospital a lot as a very young child  
Thinking about what she must have gone thru just blows my mind

I remember my brother's high school years  
He went thru so much pain, medical treatments, and tears

Steve was so much smaller than any other kid  
He was on so much medication he looked just like Gary Coleman as a kid

Steve and I both graduated  
He in 1981 and me in 1980

Out of school I started hanging out, working and experiencing the world  
Smoking, drinking, clubbing, and girls

**PATIENT'S CORNER CONT'D:**

You see, Sickle Cell was not a concern for me. I was caught up with doing things that I thought was fun. I remember going into a crisis and having pneumonia, laying in the emergency room watching doctors work on my body as I floated above. I've probably had pneumonia about 5 times and many crisis. I don't remember how many times I've been hospitalized. I met Anita, my wife, in November 1999. I told her that I had Sickle Cell. Sickle cell was not a big concern for me, but I told her anyway. It was the right thing to do. Pain every-now-and-then, take some pills, not a big deal too me. Until I went into a crisis for the first time in her presence. She had no idea what to do. She was scared, I was in pain, I was being a pain because I was determined not to go to the hospital. But I was convinced to go to the hospital. That was the start of me, with the love and concern of my wife to be concerned and serious about my disease. We've been married since July 2001 and I had lived past the "expected" life expectancy of a man with sickle cell. I think it was Mother's day weekend 2003, I had run out of pain medication and having a minor crisis, however, my doctor didn't want to give me a prescription for medication. He suggests that I go to the emergency room and get treated. I'm sure some of you can relate to this blow-off from a doctor. We just want pills to get high, not because we need pills to get thru life in as less pain as possible. Anyway, I admitted myself into a Randallstown hospital. My blood count was low and I was given fluids and blood transfusions. I don't remember too much after the 2<sup>nd</sup> day in the hospital. My kidneys and liver all failed at the same time. My wife and my family and my friends are told that I may have only 48 hours to live and they have to decide on what to do. Forty eight hours to live and I don't know what is happening because I'm not conscious. God answers my wife's and I'm sure a lot of other's prayers and Anita moves me to Johns Hopkins Hospital and the doctors their do a remarkable job on me. My wife gives up her life and is with me from day to night, finding doctors and people to pray and help. My family, church, friends all in their own way contribute. I don't remember too much about what happened. I was blessed by God to be able to type this for your reading pleasure. I am continually blessed and I need to thank God right here, right now. The hematology people at Johns Hopkins, thank you, my wife Anita, thank you. I just celebrated my 42<sup>nd</sup> birthday on January 2...I take better care of myself, I try to keep all of my doctor's appointments, I take my hydroxyurea, I drink my Tahitian Noni Juice, I say hello to people, and I'm starting my training to volunteer at a local hospital. Everybody, thank you.

By Roger Carter



We are on the web!  
sicklecellcenter.org

## JHMI

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*S I C K L E   C E L L   T E A M*  
*S O P H I E   L A N Z K R O N ,   M . D .*  
*Y V E T T E   R O A N E ,   P A - C*

## UPCOMING SUPPORT GROUPS

- WHAT:        SICKLE CELL SUPPORT GROUP
- WHERE:      JOHNS HOPKINS OUTPATIENT CENTER, CONFERENCE ROOM 2140
- WHEN:      6:30PM-8PM, 2ND THURSDAY OF THE MONTH
- DATES:      FEB12th, MAR 11th, APR 8th, MAY 13th, JUNE 10th, JULY 8th,  
              AUG 12th, SEPT 9th, OCT 14th, NOV 11th, DEC 9th

## BLACK HISTORY AWARENESS MONTH:

February is black history awareness month. Test your knowledge on black history.

1. Which scientist started the world's first blood bank? \_        A. John Standard
2. Which doctor performed the first open heart surgery? \_        B. Garrett A. Morgan
3. Who invented the lawn mower? \_        C. Charles Drew
4. Who invented the traffic light? \_        D. Brown vs. Board of Education
5. Who invented the refrigerator? \_        E. John Burr
6. First major victory of the modern civil rights movement? \_        F. Daniel Hale Williams
7. First self-made women millionaire from hair products? \_        G. Lewis Howard
8. Who invented the filament within the light bulb? \_        H. Madam C.J. Walker

Answers:

1. C    2. F    3. E    4. B    5. A    6. D    7. H    8. G

