



Happy New Year!

FROM THE SICKLE CELL CENTER AT
JOHNS HOPKINS

As the year comes to a end, we should reflect on all of the small steps that have been taken to further educate, promote, and inform patients as well as the public on sickle cell disease. In the last few issues we have discussed health maintenance, prescriptions, and advocacy. As this year comes to an end, hopefully all the ideas, advice, and information will play a part in keeping everyone healthy in 2007. Let's strive to put in place what we have learned in 2006 to practice in 2007, for the patients as well as those who support our patients. Please enjoy the last newsletter of this year as we continue to enhance the understanding of sickle cell disease. Thank you.

Mandy David, PA-C

HAPPY BIRTHDAY!!

NOVEMBER:

KIMBERLY, SHIRLEY, BRANDI, QUANTINA, JOHN, ADRIENNE, KEVIN, JOANN, CHRISTINE, LAMART, JOANNE, DAMALI, SHANIKA, DEMON, DEBORAH F, CREIGHTINA, DEBORAH L, FATMATA, MONAE, CARLA, BRITTANY,

DECEMBER:

ARLEEN, CLAUDETTE, LINDA, OMLARA, DION, GLORIA, SHAMIA, CAROLYN, DAVID, FRACIE, JAMES, SEBASTIAN, FRANCINE, MOJIBADE, MARTINA, FLORECITA

JANUARY:

KIMBERLY, THOMASINE, CALVIN, ANTOINETTE, TASHA, AMINA, CEDRIC, CARLTON, ROGER, TIFFANY, FRANCIS, SHANEKIA, AKIN-YEMI

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WHY DO I NEED A CASE MANAGER?.....

CASE MANAGEMENT AND SICKLE CELL DISEASE

As nurse case managers with Johns Hopkins Health Care (JHHC), we are often asked, “What can you do to help me?”

The primary goal of the Johns Hopkins Health Care Hopkins disease/case management programs is to provide members with **care coordination** and health care of the highest possible quality. While participating in one of our **voluntary** case management programs a registered nurse will work closely with your physician to reach your health care goals by providing the services listed below:

- ◆ An initial health assessment to help determine specific needs
- ◆ Periodic telephone check-ups to see how you are doing
- ◆ Prompt responses to your questions
- ◆ Coordination with all the physicians, home care agencies, equipment and other health care professionals that provide you with care
- ◆ Educational materials about your health
- ◆ Assistance in accessing social services and community resources

The relationship formed between a case manager and their patient is often long term and includes the involvement of family members and other support systems.

The JHHC Care Management Department offers services to Priority Partners, Uniformed Services Family Health Plan, and Employee Health Plan members. For other health insurances, please contact your customer service department to see if case management services are available under your plan.

Provided by Case Managers: Joann Rease, RN and Kay Concha, RN



WHAT IS PULMONARY HYPERTENSION?.....

Pulmonary Hypertension in Sickle Cell Disease

Stephen C. Mathai, MD MHS

Lung disease is common in patients with sickle cell disease. In fact, in large, long-term studies of patients with sickle cell disease, one of the most common causes of death was chronic lung disease. Pulmonary hypertension, a form of lung disease, may be responsible for many of the problems associated with the lungs that ultimately cause death.

What is Pulmonary Hypertension?

Pulmonary hypertension is high blood pressure in the lungs. To understand what pulmonary hypertension is, we first have to understand the normal function of the lungs and heart. Normally, the air you breathe travels through your windpipe or trachea into your lungs. Small blood vessels within the lungs carry blood to pick up oxygen. The blood that is now oxygenated, or high in oxygen content, travels to the left ventricle of the heart and is pumped out to the rest of the body to provide oxygen to the tissues. The now de-oxygenated blood is collected in veins and returned to the right atrium of the heart. From there, the blood travels to the right ventricle and is pumped into the blood vessels of the lung. The process of picking up oxygen in the lungs and dropping it off in the tissues then repeats.

In pulmonary hypertension, small blood vessels in the lungs become narrowed and fill with either debris or blood clots. These narrowed or blocked blood vessels increase the pressure in the lungs. The right ventricle, which is responsible for pumping blood into the lungs, now has to work much harder to get blood into the lungs. Unfortunately, the right ventricle can not adapt very well to higher pressures and eventually begins to fail. Because the right ventricle is not pumping blood forward, blood and fluid begins to back up into the abdomen and legs, causing swelling. Further, less blood is pumped into the lungs to pick up oxygen and less oxygen reaches the tissue. Eventually, the heart can not pump enough to maintain blood pressure or oxygen levels, leading to death.

Symptoms of pulmonary hypertension include shortness of breath, especially with exercise. Chest pains, swelling of the legs or abdomen, or passing out may suggest pulmonary hypertension. However, these symptoms can be related to other lung or heart problems. It is possible to have pulmonary hypertension and not have any symptoms.

Why do people with Sickle Cell Disease get Pulmonary Hypertension?

Although it is not fully understood why patients with sickle cell disease get pulmonary hypertension, it is thought to be related to several problems that regularly occur in sickle cell disease. First, hemolysis, or break down of the red blood cells, releases factors in the blood that cause the vessels to narrow. When this occurs in the lungs, oxygen levels drop, causing the blood vessels to narrow further. Second, hemolysis also increases the tendency of blood to clot. Because these blood vessels in the lung are so narrow, they may be more likely to develop clots. Additionally, hemolysis releases factors that may increase the tendency of other red blood cells to become sickled.

WHAT IS PULMONARY HYPERTENSION?.....

Who is going to get Pulmonary Hypertension?

Several studies have shown that about one-third of patients with sickle cell disease will get pulmonary hypertension.

How is Pulmonary Hypertension diagnosed?

An echocardiogram, or ultrasound of the heart, is the best screening tool for pulmonary hypertension. This test can detect abnormalities of both the left and right side of the heart and importantly, gives an estimate of the pressure inside the lungs. If this estimate of the pulmonary pressures is high, a right heart catheterization is necessary to confirm the pressures. A right heart catheterization is an outpatient procedure that takes about one hour to complete. This procedure directly measures the pressure in the lungs and the amount of blood the heart pumps each minute. This test is the only way to definitively make the diagnosis of pulmonary hypertension.

How do you treat Pulmonary Hypertension?

There are only a few studies that have looked at treatment of pulmonary hypertension in sickle cell disease. However, there are some general recommendations for treatment. For patients with very low hemoglobin levels, transfusions may help reduce pulmonary pressures. Hydroxyurea may also have a role in therapy. Medications that have been used for other forms of pulmonary hypertension have also been used to treat sickle cell disease. These drugs include bosentan or Tracleer®, sildenafil or Revatio®, and epoprostenol or Flolan®. Each of these medications may help reduce the pressure in the lungs and improve shortness of breath. However, these medications can only be started if pulmonary hypertension has been diagnosed by right heart catheterization.

What should I do if I want to be evaluated for Pulmonary Hypertension?

The Pulmonary Hypertension Program at Johns Hopkins Hospital, led by Drs. Paul Hassoun, Reda Girgis, and Ari Zaiman, has expertise in the evaluation and treatment of pulmonary hypertension. Please consult your doctor to see if an evaluation by a pulmonary hypertension specialist is advised.

WHY DO MY EYES HAVE TO BE DILATED EVERY YEAR?



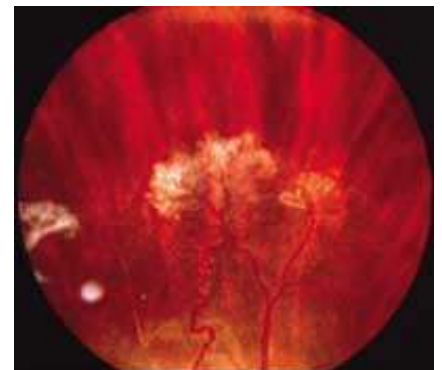
Why do I have to have my eyes dilated every year? I can see very well, why do I have to see the eye doctor now? These are some of the questions that I often hear when it is time to see an eye doctor as part of routine health maintenance. Retinopathy is defined as “any disease of the retina.”

A sickled cell results when there is an alteration in the structure of hemoglobin and therefore the sickled red cells are rigid, and restrict blood flow. This can occur in the part of the eye known as the retina. Over time when a person has a sickle event in the retina this can lead to a proliferation (growth), of new blood vessels and these new blood vessels can bleed within the retina and cause vision loss.

The severity of sickle retinopathy can depend on the type of sickle cell disease you have, SS, SC or SBTal, those with SC disease are at the most risk. In all types of sickle cell disease, symptoms such as reduced vision, a change in vision, floaters, flashes, dark areas in the field of vision, blurry vision, double vision, peripheral vision loss, and sudden vision loss could suggest the possibility of a retinal bleed. Occasionally there may be no symptoms present, which is why yearly screening is important.

Our goal in the management of sickle cell retinopathy is to screen sickle patients by referring them to ophthalmologists annually. The ophthalmologist will look and see if there is evidence of new blood vessel formation. If there are new vessels, these can lead to retinal bleed and catastrophic vision loss. The ophthalmologist can treat these areas to prevent bleeding with a laser procedure. The goal of the ophthalmologist is to decrease the risk of retinal neovascularization with yearly ocular examinations, and dilated retinal evaluations. With regularly scheduled appointments the damage from retinal disease can be prevented. If you have the sudden onset of floaters or any other visual change you should seek care immediately. The Wilmer Eye Institute has a 24 hour Emergency department and can be contacted by calling (410)-955-5347, for regular follow up appointments please call (410)-955-5080.

Written by: Mandy David, PA-C



PATIENTS CORNER.....

My Life with Sickle Cell

I entered the world, kicking and screaming on November 15, 1978. My parents loved music and I was told that while my Mom was in labor, the sound of Marvin Gaye and the Bee Gees filled the room. My parents knew before my first birthday that I had SC Sickle Cell disease. They were told that their first born would not live past the age of 21. Somehow, they knew that this wasn't true.

Throughout early childhood, my life was filled with trips to the ER and long hospital stays because of ear infections, fevers and painful Sickle Cell crisis. My grandma had a garden with a huge magnolia tree. I remember her picking the magnolia seeds in the fall to make a liniment solution to rub on my aching joints. During the cold days of winter, it was hard to watch the other kids play in the snow for hours when I could only go out for 15 minutes; sometimes not at all. My Dad would always try to make me stay inside, but my grandma would tell him to let me enjoy being a child. In her thick southern accent, she would say, "Never let that chile' think that she can't do anything because of her illness. Always encourage her to do her best." Putting on my coat and hat, I would overhear the conversations that she had with my parents. She birthed 10 children and was very wise. They always welcomed her advice.

Some days I used to think, why would God punish a child to feel such harsh pain? During a crisis, I would feel so overwhelmed with pain. I would cry, moan, and rock back and forth; nothing helped. It felt like the end of the world each time I experienced a crisis. My parents looked helpless as they held me and tried to take my mind off of the pain. They wanted to do everything that they could at home because they knew that the trip to the ER would lead to being admitted. Sometimes I went home after 1 day, other times I was doomed to stay for two weeks; depending on the other symptoms that accompanied the pain. I was in the hospital one Christmas. I was so out of it from the pain meds; I didn't even realize that it was Christmas until I woke up late in the afternoon to a hospital room filled with gifts. It was one of the best holidays I've ever had.

My Mom was always very strong. She took life one day at a time and worked very hard to provide for us. As I became older, she taught me how to notice when I'm receiving proper medical care and when I'm not. If my doctors seemed like they knew very little about Sickle Cell, she would seek another. She never backed down from the long medical terms that were used to deliver my diagnosis. "Always ask questions", she would say. And to this day, I have no problem asking questions.



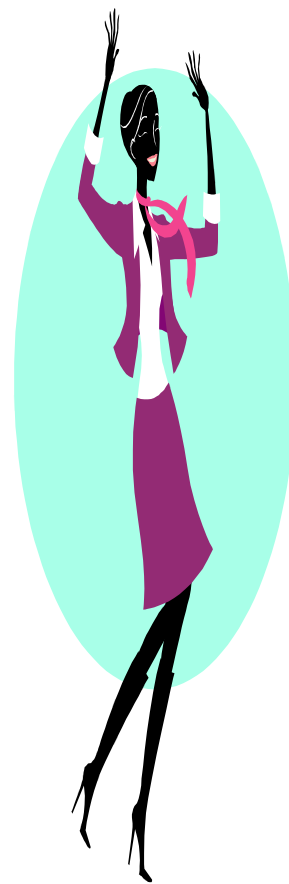
PATIENT CORNER CONTINUED.....

Shortly after the start of my 10th grade year in high school, my Grandmother passed away, and three months later, my mother passed away. I was totally devastated. They were all that I had left. It was such a stressful situation to go from having a sound family unit and support system to having nothing. My sister and I went to live with my paternal grandmother in NW, DC. I didn't have to change high schools, but it still was not the same. To make matters worse, after my 17th birthday, I was told that I could no longer come back to Children's Hospital. I couldn't help but think that all of the wonderful doctors and nurses that my parents and I worked so hard to develop a positive relationship with would also abandoned me.

Transitioning into adult care was horrible. The wait at the ER was three times as long. I didn't know anyone there. The hospital rooms were dark and dreary. And the doctors...most of them were literally looking at books and electronic publications to find out more about the disease and how it should be treated. In my case, I would always have either type of extremist. There was the doctor who just asked, "What are you taking?", and pumped me full of whatever would take the pain away. They usually didn't perform any blood tests or complete a full medical history work-up. I would be blitzed out of my mind within 3 hours and barfing up everything. Then I had the thorough doctor who completed grand rounds on a hematology related topic and was fully equipped to handle a "sickler". I would have an oxygen mask, meds for pain, nausea and itching, fluids, full CBC and urine culture within 1 hour of being seen. Their bedside manner usually wasn't the best and they always knew how I was going to answer every question before I finished my statement, but at least I was being cared for. I wanted more. I wanted the total package. It was like dating almost, trying to find a good doctor. The internet brought on a new playing field because I could conduct some preliminary research on the doctor before I decided on them.

During college, I entered into the workforce. I never gave it a second thought. No one was going to dictate how I spent the rest of my life and what my limitations would be. My family members would say that I was crazy for working full-time and going to school full-time. For me, there was no choice. It was something that I had to do. I would get sick a few times each year, but I always returned to work and school with a positive outlook and eager to move forward. I was learning to cope with the changes of adulthood and the demands of corporate America. I was even dating someone who could help me manage the complexities of my life.

I was still in school when I found out that I was pregnant with my first child. My hematologist at the time told me that I shouldn't keep the baby. The pregnancy would be too complicated. Although I thought about the outcome of either decision constantly, I decided to find a doctor who would be more optimistic about my child's outcome. How could I not give this life a chance?



PATIENT CORNER CONTINUED.....

My first son was born, so perfect and beautiful. For the first time I felt an instant, overwhelming love and connection with another soul. It was truly a magical experience. Two years later, I married my husband. I never thought that I would actually be someone's wife and mother because I spent so much time focusing on my healthcare and making sure that I'm managing a balanced lifestyle. Aside from the birth of my son, it was the best day of my life. I didn't want that day to ever end. Of course the fact that neither my Mom nor Dad was there to share in joy of this day weighed on my mind. But part of me felt their presence. It was as if they were taking each step with me, hand-in-hand.

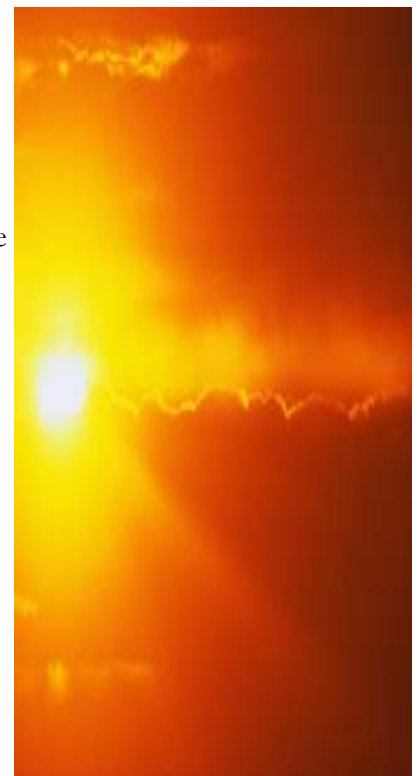
A few years later I started to be admitted into the hospital frequently for various reasons. I now had to deal with being away from my family and not being a Mom and wife for days while I'm in the hospital. My husband and mother-in-law would make sure that everything was taken care of so that I didn't worry. Having that support system again was a great relief.

After the birth of my second son, about 1 year ago, I was surfing the internet and found out about the Johns Hopkins Sickle Cell Center for Adults. There was an e-mail address for Dr. Lanzkron. I could not believe it. I have an e-mail address for a doctor and I can freely ask questions! Do I finally have my total package?

After the first visit, I knew that she was THE ONE :0) I could not believe how much she cared about me as a patient and the dedication that she displayed for the cause was unbelievable. She as well as her awesome staff has helped me with so much personally and professionally. Most of all, she has ignited my drive to fight for better medical care. The world needs more doctors with as much passion for ensuring that patients have great health care.

I know that sometimes it's hard to see the light at the end of the tunnel and the pain that no one else can seem to understand is so hard to bare. But remember that there are people out there that feel the same pain and understand what you are going through. I am now 28 years old, 7 years past the death sentence that my parents were given when I entered this world. It's ok to wait on the world to change, but it's so much more rewarding to help it along and fight to see change come sooner.

Written by: Lashanta Whisenton



MEET THE SICKLE CELL TEAM.....



Sophie Lanzkron, M.D.



Mandy David, PA-C

Contact Info:

Phone: 410-502-6997

Fax: 410-614-8601

Email: info@sicklecenter.org

Clinic: Tuesday 9:00am-12pm

Thursday 1:10pm-3pm

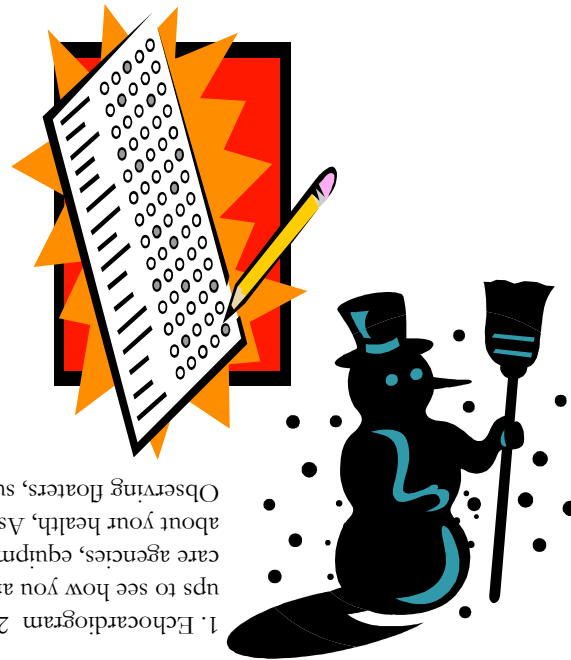


Dawn Hatcher, Resource Coordinator

If you aren't receiving the quarterly newsletter or support group flyers in the mail, please contact our office so that we can add your name to our mailing list. We wouldn't want anyone to miss out on the informative news we have to share.

QUIZ TIME!!

1. According to Dr. Mathai , what is the best screening tool for Pulmonary Hypertension?
2. Name one important role a nurse case manager can offer you.
3. How often do you need to see an ophthalmologist if you have sickle cell disease?
4. Name one symptom related to your vision for which you should seek emergency room care.



1. Echocardiogram 2. An initial health assessment to help determine specific needs, Periodic telephone check-ups to see how you are doing, Prompt responses to your questions, Coordination with all the physicians, home care agencies, equipment and other health care professionals that provide you with care, Educational materials about your health, Assistance in accessing social services and community resources 3. Annually (once a year), 4. Observing floaters, sudden loss of vision