



Sickle Cell Center for Adults at Johns Hopkins

May 2004, Quarterly Newsletter

EDITOR'S NOTE

In this month's newsletter you will find information on erythropoietin, new childcare services for our support group, coumadin, and patient's corner.

On various occasions, I have been asked how do I do it? Nurses, PA's, and physicians have asked me how do I handle managing sickle cell patients. In my eyes, sickle cell patients are no different from any other patient. Sickle cell patients are human beings seeking healthcare. This chronic disease usually requires the use of narcotics for adequate pain control. Healthcare professionals prescribe narcotics to manage patients in sickle cell crisis and are part of the cause and effect equation. Patients may or may not be educated on the side effects of narcotics prior to their treatment. Those individuals who suffer from avascular necrosis (AVN) experience chronic pain requiring narcotics daily. With the use of daily narcotics, patients can have a better quality of life. However, narcotic dependency is a consequence of continued narcotic usage. As a result, our sickle cell team has recruited a drug counselor, Mr. Brian Estes, to help our patients understand, cope, and better manage their narcotic usage.

Dependence relates to the preoccupation with a drug (narcotic pain medication). When the drug is discontinued, your body experiences withdrawal symptoms such as sweats, diarrhea, shakes, and/or back pain. Addiction is an illness in which there is a pattern of misuse of a drug, problems with social life or work, and/or evidence of tolerance or withdrawal. An example of addictive behavior is when a patient changes a prescription to receive a higher amount of narcotics. Only 10 % of our patients are addicted to narcotics; therefore, 90% are not addicted. Yet it is unfortunate that the majority of patients are thought to be drug seeking. We must remember that these individuals did not ask to be born with sickle cell disease, nor did they manipulate the management of the disease making the standard of care narcotics.

I happen to enjoy managing sickle cell patients and I welcome the challenges that arise. What amazes me is the initial astonishment of patients whom I first meet with this chronic illness. They are relieved and overjoyed to know that someone truly cares about them and believes their subjective complaint of pain. Usually physicians see the disease and not the patient. But with sickle cell disease, some only see the patient and forget sicklers have a real disease. Unfortunately, a handful of patients may abuse the system, making management of the majority challenging. In order to treat the patient, we must believe the patient.

By Yvette Roane, PA-C

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UPCOMING EVENTS:

- NATIONAL
SICKLE CELL
DISEASE
ASSOCIATION
ANNUAL
CONVENTION
SEPT 2004
- SICKLE CELL
WALKATHON
SEPT 2004
- PEDIATRIC &
ADULT PICNIC
DATE OPEN

ERYTHROPOIETIN AND ITS USE IN SICKLE CELL

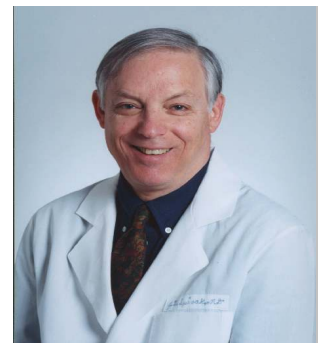
The major function of the red blood cell is to carry oxygen from the lungs to the tissues. Red blood cells are produced in the bone marrow and the production of these cells is controlled by a hormone called erythropoietin. Erythropoietin is produced primarily in the kidneys and its production is controlled by the amount of oxygen reaching the kidney cells that produce it. When there are too few red cells to carry enough oxygen to the kidneys, kidney cells make more erythropoietin, which stimulates the growth of additional red blood cells. When the oxygen supply is restored to normal, the production of erythropoietin by the kidneys is reduced to baseline.

In sickle cell anemia, erythropoietin production is increased because there are too few red blood cells to adequately supply oxygen to the kidneys. Unfortunately, the increased stimulation of red cell production by erythropoietin cannot keep up with the increased rate of death of the sickle red cells. The result of this imbalance is anemia and a continuously elevated erythropoietin level in the blood.

The failure of erythropoietin to produce enough red blood cells in sickle cell anemia patients to compensate for those that are being destroyed is unclear. It may relate in part to the fact there is only so much space in the bone marrow to make red cells and that the demand for it is greater than the space available for its supply. In some sickle cell patients with severe kidney damage, anemia will be more severe because of the inability to produce erythropoietin. In this situation, a genetically manufactured form of erythropoietin (recombinant erythropoietin) has usually been given to replace the erythropoietin that the kidneys can no longer make. Unfortunately, this has not proved to be as successful in patients with sickle cell anemia as it is in patients with severe kidney disease who do not have sickle cell anemia. Furthermore, the manufactured form of erythropoietin is also not that useful for correcting the anemia of sickle cell patients with normal kidneys.

Hydroxyurea is an important drug for the treatment of sickle cell anemia because it increases the production of hemoglobin F. Hemoglobin F, also known as fetal hemoglobin, prevents red blood cells from sickling. Hydroxyurea also reduces the white blood cell count. Since it is thought that white blood cells are also involved in the sickling process, this may be another way in which hydroxyurea helps to prevent red cell sickling. Erythropoietin also increases the production of hemoglobin F and has been used experimentally with hydroxyurea to increase hemoglobin F levels in sickle red cells. Unfortunately, the dose of manufactured erythropoietin required for this was 3-9 times greater than normally used in humans and, of course, the manufactured erythropoietin had to be given frequently and can only be given by injection. Because of these considerations, manufactured erythropoietin is not in routine clinical use in sickle cell anemia.

By Jerry Spivak, M.D.



WELCOME ANITA CARTER: CHILDCARE PROVIDER



Join me in welcoming Anita Carter as our childcare provider for the monthly support group. Anita is an employee at Johns Hopkins in the surgical pathology department. She has worked for Hopkins for seven years as a laboratory supervisor. She was a Sunday school teacher for five years. Anita is presently a Sunday school coordinator at New Antioch Baptist Church in Randallstown, Maryland. For the past two years she has coordinated blood drives at the American Red Cross.

Anita loves art, music, and working with children. She has two years of college education and a desire to volunteer. Her dream is to travel the world and meet new people. She has a twelve year old son, and is married to a wonderful man with sickle cell disease. Thank you Anita for opening our doors to childcare. It has already expanded our support group participation. Call ahead and sign up for daycare.

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.



COUMADIN: WHAT YOU NEED TO KNOW ABOUT BLOODTHINNERS

Coumadin®, known by some as its other name, warfarin, is used for many reasons in patients with sickle cell disease. Some patients may be using it to treat or prevent clots, and prevent strokes.

How It Works:

Coumadin works against clots by lengthening the time it takes for your blood to clot. This also puts you at a slightly higher risk of bleeding, which is the major side effect of taking ‘blood-thinning’ medications. There are many things that influence how Coumadin works in your body, so the dose may need to be changed to make sure it is working just right for you. Regular monitoring of the PT/INR and regular evaluation by providers at clinic appointments every few weeks will help to prevent some of these bleeding complications.

Signs and Symptoms of Bleeding:

The stomach and intestines are the most common site where bleeding can occur. If blood is in your bowel movement or vomit, report this to your provider who manages your Coumadin. You may notice that you bruise easier, and that it takes longer for bleeding to stop when you cut yourself. If you can’t get bleeding to stop after 20-30 minutes of applying pressure to an injury, consider going to the closest emergency room for evaluation. Also, if you fall down or hit your head very hard, get to the closest emergency room so that scans can be done to be sure you aren’t bleeding in your head.

Drug Interactions:

Coumadin interacts with many prescription and over-the-counter medications. It is important to remind all of your health care providers that you are taking Coumadin (warfarin) when they give you a new prescription medication or before you have any procedures, such as dental work, scopes of the colon or throat, or biopsies. Unless your doctor tells you differently, avoid taking aspirin, ibuprofen, Motrin, Advil, or Aleve as these can increase your risk for bleeding.

Food Interactions:

Coumadin interacts with a vitamin in your diet, called vitamin K. Vitamin K is found in green leafy vegetables, vegetable oils, and liver. Be careful about vegetable drinks such as V8 and dietary supplement drinks such as Ensure, Boost, or Slim Fast as these all have vitamin K in them. It is also best to avoid beef or chicken liver, and large amounts of mayonnaise (which contains vegetable oils).

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PATIENT'S CORNER: "I am Not One of You"

I have always associated the phases I go through while dealing with my disease, as with the 5 stages of grief. They say these stages are what a person goes through when they've lost someone or something due to a death, a divorce, and even good health. Therefore, when I began to be aware of multiple crisis, blood transfusions and hospital stays, I felt I had lost my good health. I grieved over 'my health' for many years. The 5 stages of grief are denial, anger, bargaining, depression, and acceptance. I added a few stages of my own and they are, understanding, tolerance, and admittance. These stages came before acceptance for me. Stage number one is where I have been stuck in most of my life. The stage of denial. I believe that you can enter and re-enter the various stages of grief during your personal phases of understanding what it is you are dealing with. It would have been great to go through the five or rather in my case, the eight and be done with it! Voila, you arrive at acceptance and go on with your life. But in my case, it just didn't happen that way. Unfortunately, I struggled with anyone of these stages for over 20 years.

I define my chronic illness as an internal and external war with self. I pretended that it just did not exist in my world. Therefore, I'll do whatever I want. I'm just as normal as the next person. Don't I look it? Don't I act it? On many occasions this attitude worked, on many others, it did not. For years I could not even say the words Sickle Cell Anemia, let alone the word disability or God forbid 'handicap.' Oh, not me! I don't have those silly cells. I was informed however, by the California Vocational Rehabilitation Department, that yes I was indeed disabled according to the federal government and I qualified for handicap services. I shut those people out as fast as I could. Except, those two words rang in my ears for days. DISABLED! HANDICAPPED! It took me 20 years and a baby before I applied for my handicap parking card. The words are no longer a threat to me, in fact as a woman with a lot of goals and big dreams, I am finally trying to find out just what those two little words can get me! Silly cells - silly me. Denial is just another word for fear in my opinion. Quietly creeping behind fear is disrespect. Fear and disrespect ruled my life for two decades. Fear of the unknown, fear of others knowing, fear of my future, fear of my past, fear of my present, and the ultimate fear of death. Disrespect of self, of life, and of God. Most devastating of all was the disrespect I showed my blood disease called Sickle Cell Anemia, so much so, I couldn't even call it's name. So I spent 20 years running away from it all. I went from pain to pain medicine. Pain medicine to drugs, and finally from drugs to God. All in the name of seeking peace, seeking revenge, seeking destruction, seeking surrender, but never seeking help. Finally and ultimately I sought God and began to achieve an understanding and some peace of mind. Now, I am trying to untangle the past 20 years and process the real from my own illusions or disillusion, in hopes of arriving at a balanced state of harmony between my mind, my body, and my spirit.

"Moving towards balance is a life long journey" – Yoga Magazine

Proverb 4:7 says, "Wisdom is the principal thing; therefore get wisdom: and with all thy getting, get understanding." This scripture changed my life, recently as a matter of fact, and I decided to attend a Sickle Cell Support Group for the first time in my adult years. I am 44 years old. How can I change the course of my life if I don't ever accept a very special part of it? I received a card on Mother's Day that said, "Believe in the special person you are, and the rest of the world will follow." Denial is nothing but fear. And fear is no longer an option for me. It has already wasted so much time. I possess a great will, much strength and determination. I even tell my 7 year old 'son-flower', it is with the help of those three traits, that he came to be. On May 13th, 2004 I took another step in owning my power. Sickle Cell Anemia is a disease that deserves respect along with proper care and treatment. Perhaps with that state of mind, balance and harmony can be achieved. Have I reached Nirvana or Peace? Not quite. But a little more understanding and a lot more acceptance goes a long way. Was the meeting well worth it? Absolutely! I discovered as I sat around this group of amazing survivors, I am one of you and you are just like me! I leave this quote as a gift to you. It's just a thought. Perhaps a step, or maybe a beginning. Begin within, seeking wisdom, letting go and letting God in. The path to acceptance is through understanding, reading, learning and studying. Who are you? Where are you in the stages towards acceptance? Why are you here? Become who you want to be. **"ACTION IS A PHYSICAL MANIFESTATION OF WILL POWER."** (I can't remember where I heard this, but it's great isn't it?) I'm on a mission...how about you? Won't you join me? Tracy@healthgoddess.com! Namaste! It means, "The light within me, honors the light within you."

—Tracy

We are on the web!
sicklecellcenter.org

JHMI

Sickle Cell Center for Adults
at Johns Hopkins

Phone: 410-502-6997

Fax: 410-614-8601

Email: sicklecellcenter@jhmi.edu

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



SICKLE CELL TEAM
SOPHIE LANZKRON, M.D.
YVETTE ROANE, P.A.-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: JUNE 10th, JUL 8th, AUG 12th, SEPT 9th, OCT 14th, NOV 11th, DEC 9th

COUMADIN (WARFARIN): CONTINUED FROM PAGE 4

Green Leafy Foods To Avoid:

Asparagus, Endive, mustard greens, avocado, green scallion, peas, broccoli, kale, spinach, brussels sprouts, lettuce, turnip greens, cabbage (green & red), liver, watercress, collard greens, mayonnaise, olean (wow chips).

Take your Coumadin at the same time every day, usually in the evening. If you forget to take Coumadin at the usual time, and you remember within about 4 hours, go ahead and take your usual dose. If you remember much later than 4 hours, just skip that dose for the day. DO NOT take extra Coumadin the next day. Remember to tell your health care provider that you missed a dose. This will help to interpret the PT/INR blood test correctly.

Coumadin is a safe and effective medicine and it is important to have it monitored closely to avoid further clots and bleeding events. You need to do your part to attend clinic appointments, report events as they occur, and stay current on your Coumadin refills. If you have any questions or concerns about what you've read about Coumadin, contact your health care professional.

By Paula Biscup, Pharm.D.