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JHMI

SICKLE CELL CENTER FOR ADULTS AT JOHNS HOPKINS

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In this quarter's newsletter you will find information on the following: a study involving the long term use of hydroxyurea, dental health in sickle cell patients, and chronic transfusions. Also in this issue we have added a special feature called the Patient's Corner. The Patient's Corner is a section of the newsletter devoted to our patients. Patients can write in and express their thoughts, feelings, and/or experiences. Lloyd Quashie has permitted us to publish his perspective on sickle cell disease and how it has affected his life. As caregivers to patients with Sickle Cell disease, we found his comments to be quite insightful. We hope his article offers hope to those patients who have reached obstacles in their life.

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. We hope you enjoy this quarter's newsletter and have a wonderful summer.

HYDROXYUREA HIGHLIGHTS

- HYDROXYUREA DECREASES THE NUMBER OF PAINFUL CRISIS.
- HYDROXYUREA INCREASES HEMOGLOBIN F
- HEMOGLOBIN F HELPS TO PREVENT RED CELLS FROM SICKLING.

COMMUNITY HEALTH CHARITIES

Community Health Charities of the National Capital Area helps our organization and many others obtain support through workplace giving campaigns. They invited our Adult Sickle Cell Center to participate in the promotion and education of Sickle Cell disease at two of their health fairs. The first fair was held in Silver Spring, Maryland on May 14th, 2003. The second fair was held on June 4th, 2003 in Washington, DC. Members of the community were educated by our Physician Assistant, Yvette Roane, on Sickle Cell disease, its background, statistics, complications, nearby treatment centers, and research advances in the field.

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.



CHRONIC TRANSFUSIONS

For some patients with sickle cell disease, treatment includes the use of chronic transfusion. The goal of monthly blood transfusions is to decrease the amount of sickle hemoglobin. Chronic transfusions help prevent some serious complications of sickle cell disease. The most common reason that patients are placed on a chronic transfusion protocol is because they have had a stroke. There is a lot of data that shows that the use of chronic transfusion decreases the risk of a patient having a second stroke. This treatment for stroke is considered the standard of care. Patients with other complications are also occasionally placed on chronic transfusion therapy. Some patients with chronic leg ulcers, lung problems, or rarely those with very frequent painful crises are placed on transfusion therapy. There are several important risks of chronic blood transfusions that have to be discussed with patients before the decision is made to proceed with this treatment.

The first risk of chronic transfusions is iron overload. Each unit of blood has a large amount of iron. When patients are chronically transfused they become iron overloaded. Iron overload can lead to liver failure and heart failure. To prevent these life-threatening complications, patients on chronic transfusions need to be chelated with a medicine called Desferal. This medicine helps remove iron from the body. Desferal requires nightly infusions typically given under the skin. Another way of preventing iron overload is to use exchange transfusion. Exchange transfusions require a larger number of units of blood to be transfused compared to patients receiving simple transfusions monthly.

The other major risk of chronic transfusion therapy is that patients will develop antibodies to the blood they receive. One quarter of sickle cell patients that are transfused will develop these antibodies. This can make it difficult to find matching blood and can cause patients to have reactions to blood products which can be life threatening.

The final risk, which is most concerning for patients, is the risk of getting an infection from transfused blood. The risk of getting a serious infection such as HIV or Hepatitis is quite low, but is unfortunately not zero.

As health care workers we often weigh the risks versus the benefits of any therapy that we offer. Although there may seem to be many risks associated with the use of chronic transfusions, for many patients it can be a life saving therapy.

Dr. Sophie Lanzkron, M.D.

ANNUAL SICKLE CELL DISEASE CONVENTION

When: September 24-27, 2003

Where: Beverly Hills, California

Please visit www.sicklecelldisease.org for more information.

PATIENT'S CORNER

There ought to be a law, some help, understanding, research, compassion, support, truth, optimism, hope. I was born with Sickle Cell Anemia. It should be called Sickle Cell Full of Lies, Mistreatments, Shame, and Despair. All of my life has been devoted to this thing. Not only do I deal with all of the issues that it brings me, but I have to deal with other people's issues as well. Stupid, isn't it? The biggest part is dealing with the lie that it is your life. There sat a twelve year old boy rejected trying to explain to himself why he was so small. Trying to figure out why everyone else is growing and he is still the size of an eight year old. This prolonged state of physical immaturity causes intense feelings of inadequacies. You will only live to be in your late teens, twenty if you're lucky. I'm not so completely alone though. My big brother, Nana, will always take care of me. He always has and always will. We were born just over one year apart. So close, so in love. I will always take care of him too. He takes such good care of me even though I'm sure he doesn't know it. I need him so badly. We made promises to each other. He was worse off than I. He died one month before twenty-one. I am so grateful to him.

I am so scared now. What life is there for me? I must accomplish it all, everything, now. I was told that I should be classified as disabled. After all I am twenty-one with Sickle Cell. I should give up any and all dreams of life and be realistic. The sooner I accept it, the better for me, and especially everyone else. I should not worry about children, why would I put them through this and all that it would mean for them? Besides, what woman would agree to be my mate? I should not worry about working. I don't need that much money when I'm not going to be around to achieve any goals anyway. I won't need a car, house, vacation, or nice things. I could get a nice room somewhere and do my best to be happy and wait. I challenge it all! I will die trying! It is mine to live, push on, just like everyone else. No, I can't get it through my thick head! Not yet! At least not now.

I work and get sick. I recover again. The drugs earn you a reputation as an addict trying to score. They would rather just deny you treatment. It is safer for all involved, except you. But you don't matter enough, you were just trying to pull one over on them again and they are sick of it. Did you know that all of us are one shade of black or another? So I work and get sick. I recover again

So I was able to start life anew. Never telling anyone the whole truth. I would work, get sick and I would recover. I even started to live a little. Then it hit me! I can live but for how long? If I was to have any life at all I must have it now. I had to pack it all into now! Lie: You can't do the things that other people do; you can't have the normal things. Why are you even dreaming about a family and a house with a picket fence? I don't have long so I've got to hit these goals as soon as possible. If I get married, I want one child or maybe four. If I have children, I must buy them a home to live in. If I have a child, I must prepare for their college, their future. If I own a home, I must make provisions to maintain it, even when I am gone. I've got a lot to do and I've got a deadline.

I went places around the world and gathered experiences. I had fun and learned quickly. I had to. I fell in love and lost it. I did everything that everyone else was doing. You couldn't tell the difference, till I got sick. I was driven and she wouldn't have to put up with me for long. Death would us do part. I did have a child. A beautiful, healthy baby girl. She gave me hope. I had new purpose. I had new goals and I must meet them now. I was getting weaker and weaker. I could tell. They were probably right about living

PATIENT'S CORNER CONTINUED

to thirty. I was trying to buy a house for my little girl. I had learned that you could do so much once you owned a house. That would be her future. I would do more. Her success depended on me. I loved being a father. I hated being so close to thirty. Life is too precious. I could not afford to fake one more minute. Lie: Get used to being alone.

They are almost right about that one. Almost. She was wonderful and that was scary. You are trained to disbelieve that people could be genuine. I'll be myself, and I can accept that. I want to believe her. Finally, I do. She is still so beautiful. She brings tears to my eyes. I am in love. I treasure her. She has decided to stick it out with me. She made me older than thirty! I have another beautiful baby girl! Despite all of the barbaric warnings and offers for "help", she is so lovely. Beautiful and healthy again. Could you imagine if we would have listened to them? She would not be here and your world would be that much worse off. I got sick and worked harder. And I had to do it against huge negative forces. Don't forget that I also dealt with life, as you know it. Life is full of ups and downs and everything that means. I had a lot on my plate.

I was getting sicker and weaker. I had ulcers, viruses, and the pain was amazing. We know that no one makes it to forty. Forty is the most you will live. I tried all of the experiments, procedures, and prayers. I volunteered and tested. I tried everything until my mind turned to mush. Finally, I accepted it. I was beaten into submission. After fighting so hard for so long, this was a surreal feeling. I accepted my fate. They were right this time. Just let me make it to forty.

I accepted that I would be with Nana again. He had watched over me all of this time. He would be there for me again. Nicole didn't. She prayed and kept hope. Thank God for her belief enough for two. Thank God for her selfish hold on me. I thank God for her. There was an ad placed by NIH for a new protocol. I applied and she believed. I got better and stronger. That is how we bought our house. That is how I made it to forty! I have a new dilemma now. What do I do with all this extra time? How much time do I have left? You try living past your deadline and see what happens. Only my God and my family hold it together for me. Now it is you. It is all of you that this might help. You are not alone. Others have gone before you and paved the way. Reach out for each other, help each other, and support each other. You need not be ashamed.

The doctors and the rest of society must bare the shame now. They must apologize for doing all of those terrible things to us. They made it this way. We have a problem that we can live with. You must understand that when someone else does not understand something, it is easier and better for them to make it and you go away. They must reconcile within themselves that no one is at fault. Do not hide in fear anymore. There is more to life than waiting for death. It will come soon enough. I ask you to start living like everyone else. If you look back on your life, I know that you will be amazed of how far you have come and what you have been able to accomplish in your state of being. You will be amazed at your successes against all of your odds. Let that be the undeniable proof that you need. Be your own inspiration! Don't accept the lies, mistreatments, shame, or despair anymore. Life is what you make it, even for us. Make your own truths and LIVE!

By Mr. Lloyd Quashie

UPDATE ON HYDROXYUREA

In 1995 the results of the Multicenter Study of Hydroxyurea (MSH) were published. [This study showed that subjects treated with hydroxyurea had fewer painful crises, fewer episodes of acute chest syndrome, and required fewer blood transfusions than those subjects that received placebo (sugar pill).] MSH led to the approval by the FDA of hydroxyurea use in individuals with sickle cell disease. Although MSH demonstrated the short term benefit of hydroxyurea (subjects were treated for 2.5 years on average) there was concern over the long term effects of hydroxyurea. Of most concern was the possibility that patients on hydroxyurea for a long period of time might be at risk for the development of cancer.

A recent publication in the Journal of American Medical Association (JAMA 2003;289(13) 1645) addresses concerns over the long term use of hydroxyurea. After the completion of MSH, subjects were given the opportunity to enroll in an observational follow-up study. Along with their physicians, subjects either decided to start taking, continue taking or stop taking hydroxyurea. Two hundred ninety-nine adult patients enrolled in this follow-up study. Subjects had follow-up for as long as nine years. Those subjects that took hydroxyurea and had the expected benefits of therapy (ie elevations in hemoglobin F levels or fewer painful crises) survived longer than those individuals who did not take hydroxyurea. Taking hydroxyurea did not increase the risk of getting cancer.

This study is important as it is the first to address the issue of long term use of hydroxyurea. It is not a perfect study. The study does not compare subjects in a random fashion, but rather it was the subject and the doctor that decided on the use of hydroxyurea. This may have an influence on the outcome of the study. It may be that those subjects that decided not to take hydroxyurea did not receive the same quality of medical care as those who chose to take hydroxyurea. This study is however, the first step toward decreasing our concerns over the long term use of hydroxyurea.



“Hydroxyurea can help me live longer.”

We are on the web!
sicklecellcenter.org

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SICKLE CELL TEAM, Editors
DR. LANZKRON, Director (LEFT)
YVETTE ROANE, PA-C, (Right)

TOOTH LOSS AND TOOTH DECAY

I was glad when Yvette Roane asked me if I wanted to contribute to the newsletter. A lot of the newsletters' readers may be familiar with me from my presence at the outpatient clinic for the months of November 2002 through May 2003. During that time I asked patients to participate in a study on sickle cell disease and oral health, particularly dental decay and tooth loss. I felt this would be a good opportunity to let the newsletter readers know a little more about what we are doing.

Some previous studies, including one that we conducted, suggest that individuals with sickle cell disease may be at increased risk of tooth loss or tooth decay. We wanted to help answer that question more definitively by comparing participants with sickle cell disease to those without the disease. We compared tooth loss, tooth decay and a few other oral health outcomes. We are conducting this study in Baltimore and Washington, DC. This is the largest study of sickle cell disease and dental health so far and while it is too early to talk about any of our findings, the level of enthusiasm shown by all of those involved, including the staff and the patients, made the time I spent in the clinic a very rewarding experience. Hopefully, we will have some interesting findings to share in the next few months.

Happy Birthday Wishes for July:

Crystal, Beverly, Synithia, Allen,
Kenneth, Shelton, Audrey, Nirvana,
Alexander, Antonio, Bryant,
Paulette, Teeda

Upcoming Support Groups

Outpatient Ctr.

July 10th

August 7th

September 4th

October 2nd

November 2nd

December 4th

“Some previous studies suggest that individuals with sickle cell disease may be at risk of tooth loss or tooth decay.”

By Dr. Brian Laurence, DDS