Summer Edition: September-November 2011 Volume 1 Issue 4

SCAGO

Sickle Cell Awareness Group of Ontario

Contributors

- Ms.Dotty Nicholas RN- Rouge Valley Health Systems- Centenary site
- Dr. Melanie Kirby-Allen & Ms. Sandra Newton - Hospital for Sick Kids
- NDr. Ivy- A patient with over 20 years usage of Hydroxyurea
- Caroline Dankwah

INSIDE

Message from our Vice President- Ms. Doreen Alexander

SEPTEMBER is Sickle Cell Month ... what are you doing to make it known....
The silence that accompanies this disease because of shame must come to a halt. There is nothing to be ashamed of, The more we make Sickle Cell Disease known, the more we will get help to lobby the government for the help and support that we need.

We must continue to lobby for comprehensive care, for those living with the disease, so that they can have a better quality of life; until a cure is discovered. The time is now, knowing that September is Sickle Cell awareness month. Together we can and we must continue to press on because it is said that united we stand and divided we fall; this is not the time to divide ourselves. There is much to be done so, please be encouraged and get involved in the fight.
The cold weather is fast approaching hence; you must remember to follow the rules that will keep you well. Please remember that the choice is yours so, take ownership for your well-being by making the right choices. Sickle Cell Disease will only control you if you allow it. Therefore, be motivated and continue to do whatever possible to stay healthy. It is mandatory to eat nutritious meals, hydrate yourselves and keep warm to help avoid as much illness and complications as possible. Please remember that Immunization is another vital part that must be carried out to help keep you safe during the winter months ahead.
Keep well and have a safe Autumn Season.

Youth Corner by Caroline Dankwah

The SCAGO/HSK Sickle Cell Youth Network is striving to raise awareness within young adults of the problems that arise with people who have Sickle Cell Disease. Our goal is to give an open forum to young people living with Sickle Cell Disease and have them feel welcomed and inspired.

At the sessions, the group members freely share their concerns and coping strategies. We also encourage and celebrate our union through group activities such as lunch and dinner gatherings, movie nights, inspirational talks, while reaching out to the community.

During our last session, the group talked about ways for other young adults living with Sickle Cell to join and be committed to the cause (especially the kids who are in transition from Sick Kids to the adult hospital).

Since September is Sickle Cell Awareness Month, there will be information about the disease at Sick Kids Hospital and a walk in other to raise more knowledge. Our group suggested a Facebook page where people in and outside of the group can join. This project, along with the walk and the information session is aimed at helping more people understand what Sickle Cell Disease is.

SAVE the DATE

September 17th, 2011
Sickle Cell Awareness Walk
Start location: York Gate Mall (Jane/Finch)
Time: 11am

September 20th
Sickle Cell Awareness Day
Location: Hospital for Sick Kids
Time: 10am-2pm

October:
Comprehensive Pain Management Session
Speaker: Dr. Melanie Kirby-Allen & the Pain Management Team. Hospital for Sick Kids
Time & Location: TBA
Must RSVP to attend session

November
SCAGO'S Annual AGM
Location: TBA

"Sticking With It"

One adult's experience with long-term hydroxyurea treatment for sickle cell disease

By Sandra Newton

When Donnell Ivy, MD, MPH, a National Institutes of Health Medical Officer in Bethesda, Maryland, reflects on the challenges of living with sickle cell disease, he does not consider taking hydroxyurea to be one of them. Now 39 years of age, and in his 15th year of hydroxyurea therapy for treatment of homozygous (Haemoglobin SS) sickle cell disease, he shares his experiences before and after starting treatment with SCAGO newsletter readers.

Dr. Ivy grew up in North Carolina. As a child and adolescent, he often experienced severe pain episodes from sickle cell disease that challenged his ability to cope. "I would have crises very frequently, particularly in the wintertime. When it got cold or when the weather started changing, that's when I would start feeling the symptoms the most," he notes. He recalls becoming ill more easily and more often than his very active brothers, though he did his best to keep up with them at play.

Poverty and family members' mistrust of the health system further complicated Dr. Ivy's condition. He was rarely taken to a doctor and encouraged to endure his pain at home instead. "[It wasn't] a good thing, but what it did teach me is that there would be times that I would just have to figure out how to get things done, even though I was having pain or in crisis," he notes.

Dr. Ivy has experienced significant complications of SCD, such as avascular necrosis (deterioration of the hip joint) and pneumonia from an early age. Nonetheless, he considers himself fortunate and to have done relatively well. He knows, however, that the impact of sickle cell disease is ongoing, even during periods when he feels relatively well. "Just because you're not having an acute event or a crisis, doesn't mean you're not having complications of sickle cell, or that damage is not being done. Because of the way the disease is, you can have damage occur that you're not made aware of until much later [such as silent infarcts or mini strokes]," he stresses.

During his junior year of college, Dr. Ivy experienced a bout of acute chest syndrome, his worst complication of sickle cell to date. He was moved from a hospital in Raleigh to the University of North Carolina Chapel Hill Hospital, where he was intubated to assist with breathing and treated with blood transfusions for the first time. When his condition started to improve, he learned that doctors had feared that he might not survive the episode. "It was a very bad crisis. When I got out, I learned that they had been calling my mother to come see me, because they didn't think I would live to the next hour," he states. While in hospital, Dr. Ivy was introduced to Dr. Eugene Orringer, a physician with a special interest in sickle cell disease and hydroxyurea treatment.

This meeting was a turning point for Dr. Ivy. During their visits, Dr. Orringer recommended treatment with hydroxyurea. Dr. Ivy was a biochemistry and physiology student at the time and understood the research and how hydroxyurea worked; however, he still had some concerns about the treatment. When he mentioned this, Dr. Orringer offered something helpful to think about. "He made me realize that, [while there were risks to taking hydroxyurea], sickle cell is a very severe and dangerous condition also, and it should be taken seriously. I could possibly develop [a complication like cancer] in 15 years because of taking hydroxyurea, or I could not take hydroxyurea and have another acute chest syndrome or crisis in 6 months that [I might not survive]. So that made me realize that I had to weigh my odds and come to grips that this fear of developing cancer several years from now, if it ever happened, was a risk factor, but so was dying from acute chest syndrome and complications of sickle cell [in the nearer future]."

It has now been 15 years since Dr. Ivy made the decision to begin – and stick with – hydroxyurea treatment, and he has not looked back. After a short time on treatment, he noticed increased energy, weight gain, and improved grades due to better health. "I used to have crises two or three times a year that would require hospitalization, but since taking hydroxyurea, I go four or five years without experiencing hospitalization or having a pain crisis. It

helped me to start focusing on school. I managed to make the Dean's List every semester after I started [treatment]." For him, downsides of taking the medication have been minimal, to none. "I've been fortunate, probably more fortunate than others, that I haven't really experienced many side effects from hydroxyurea. There could have been something that I overlooked, but the benefit of hydroxyurea for me has far, far outweighed any risks. It has really helped me to improve my life."

Although pleased to be able to share his story, Dr. Ivy acknowledges that beginning treatment with hydroxyurea, or any therapy, is an individual and personal choice. Each person's situation and experience is unique. "You have to [talk with your doctor] and weigh the risks and the benefits carefully," he states. Dr. Ivy acknowledges that there are still many questions to be answered about the medication. "The [links] to cancer haven't been well demonstrated in humans. Tests in animal models are done a little differently than what happens in real life in humans, so there's more research that needs to be done there." It is also unclear why individuals may respond differently to hydroxyurea. "It would seem from theory that everybody would have a pretty similar course because of the way the medication works, but unfortunately, we do know that not everybody benefits the same from it. Understanding why those differences occur is a big part of it," he notes.

From a public health standpoint, Dr. Ivy believes that more can be done to help individuals and families understand the medication better. "We need to figure out how to give the individuals the knowledge they need to make an informed decision," he says. There is also a problem of underutilization of hydroxyurea treatment by healthcare providers, a key finding of a 2008 Consensus Conference on Sickle Cell Treatment conducted by the National Institutes of Health. "In the US, [it is mainly] hematologists (blood specialists) using hydroxyurea for sickle cell disease treatment, and unfortunately, not everybody has equal access to hematologists. How do we go about getting primary health care providers, [like] pediatricians and family medicine doctors to use hydroxyurea so that individuals with sickle cell have better access?"

Once the risks and benefits have been weighed and a commitment made to hydroxyurea treatment, Dr. Ivy stresses that patients play a key role in maintaining and maximizing the benefits of treatment. Taking medication regularly and attending medical appointments consistently for close monitoring of hydroxyurea therapy is very important. Children and adolescents taking hydroxyurea should also be taught that these behaviours are part of therapy from the start. "[Taking hydroxyurea] every day, and not just when you're sick, is the way to really get the benefit. You have to use it the way it's meant to be used," he says. Dr. Ivy admits that sometimes sticking with treatment can be difficult. "I had to learn to take my medication every day, and there are some times that I've had to struggle with that," he says. "[At one time], I might have gone too long without refilling the medication, missed a day, or forgotten because I was busy. [Now], I try to keep those days to a minimum."

Generic medications are now available, reducing the cost of hydroxyurea therapy to some extent. However, the cost may remain too high for some. In these situations, individuals are strongly encouraged to speak with their healthcare team about possible solutions so that their outcomes are optimized.

Note: This article, excerpted from SCAGO's July 2011 Learning for Life session, is not intended as a source of medical advice for readers. The viewpoints presented are those of the participants, and not any medical or academic institutions or agencies listed. Please consult your physician or healthcare provider with questions regarding your particular health needs.

Dr. Donnell Ivy is a Medical Officer of Health with the National Institutes of Health, National Heart, Lung, and Blood Institute in Bethesda, Maryland. He completed a medical degree at East Carolina University School of Medicine, a residency in Pediatrics in Savannah, Georgia and a Master of Public Health degree at The University of North Carolina. He is currently involved in the development of the National Institutes of Health Guidelines for Management of Sickle Cell Disease, anticipated in Spring 2012.

Sandra Newton, M.A., is a doctoral candidate in child and adolescent clinical psychology at The University of Toronto. Her dissertation research involves measurement of health-related quality of life in children and adolescents with sickle cell disease.

SICKLE CELL DISEASE AND NUTRITION

A Holistic Approach

Presented by: Dotty I. Nicholas Registered Nurse Clinician

As the life expectancy for individuals with sickle cell disease increases, health care providers will be required give education on life skills on how to effectively manage their disease. Nutrition is generally overlooked and not given the same attention as other therapy. A balanced nutritious diet plays an important role in the health status of those affected with Sickle Cell.

Introducing a diet rich in all the essential minerals and vitamins should begin as early as possible to allow taste and texture to be acquired.

A diet rich in folate for red cell repair and growth also blood production should always be emphasized. Because of the control of folate over Homocysteine an amino acid produced by the body, it gives some protection against heart disease.

Prepare food just before serving. Valuable nutrients are lost when food is allowed to stand.

Meals should be eaten slowly and at regular intervals. Enjoy your meals in a happy and relaxing atmosphere.

FOODS TO INCLUDE IN YOUR DIET

Meats 2-3 servings	Vegetables 3-5 servings	Starches 6-10 Servings
Eggs Chicken Turkey Fish Pork Beef Tofu Seafood Lamb	Green Peas/Beans Carrots Broccoli Red/green peppers Potatoes Spinach Lettuce Cucumbers Collard Greens Okra	Bread Rice Pasta Cereal Crackers Corn Bread Peas /Beans
Fruits 2-4 servings	**Desserts**	**Milk and Milk Products**
Apples Berries Grapes Tomatoes Oranges Pineapples Peaches Pears	Cakes Cookies Pies Tart Ice Cream Yogurt Chocolate Nuts of all kinds Fats and sugars should be used sparingly Liquids 8-10 glasses of water per day	Whole milk 2% Milk Cream Butter Milk Cheese Ice Cream Yogurts