

# “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.*

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