

The Future of Sickle Cell Disease: Highlights From the Herrick Symposium

Overview

One hundred years ago, physician James Herrick discovered something unusual in a sample of blood taken from one of his patients, Walter Clement Noel. While healthy red blood cells look like doughnuts without a hole, Mr. Noel's cells were "peculiar elongated and sickle-shaped."

The observation led to Western medicine's first published description of what would come to be known as sickle cell disease. To mark the 100th anniversary of Dr. Herrick's paper, several hundred scientists, clinicians, and members of the public gathered on the National Institutes of Health campus in November for the *James B. Herrick Symposium—Sickle Cell Disease Care and Research: Past, Present, and Future*.

Sickle cell disease is an inherited, lifelong condition. People who have the disease inherit a copy of the gene that causes sickle cell from each parent. The gene codes for production of abnormal hemoglobin, which causes red blood cells to form a sickle or crescent shape. The irregular cells are stiff and sticky, and tend to form clumps and get stuck in the blood vessels. The disease affects between 70,000 and 100,000 people in the United States, the majority of whom are of African or Hispanic descent.

100 years of progress in sickle cell research and care

"Sickle cell disease represents one of the great triumphs of medical research. In my career as a clinician, and as a researcher, there's been an absolutely major shift in the survival rate for sickle cell disease and in the level of complications which are seen. What this reflects is the research base in which we understand both the nature of the disease and the complications which develop. This has enabled us to do early diagnosis and preventative therapies, which have made it so that it is uncommon for children to die of sickle cell disease in this country now, and that the average lifespan is well into middle age," said Dr. Susan Shurin, acting director of the National Heart, Lung, and Blood Institute (NHLBI). Dr. Shurin is a pediatric hematologist who has treated numerous individuals with sickle cell disease over the years. Throughout the symposium, she shared her experiences and looked to the future. She also said:

"Among the really exciting opportunities in sickle cell disease research are some cellular therapies, particularly bone marrow transplant and gene therapies. Bone marrow transplant will cure sickle cell anemia, and we've known that for quite some time. The application of bone marrow transplant is limited by the fact that you have to have a donor who matches you and who does not have the disease.

"Gene therapy is increasingly promising as a potential cure for sickle cell anemia. This is a genetic disease, we can replace the gene, and some of the recent advances in this field lead us to think that this is much closer to clinical application than it has been in the past.

"The NHLBI has a long-standing and permanent commitment to research in sickle cell anemia. We will be continuing to support basic science studies, and we will be continuing to support clinical trials. We're also working with the community to ensure that community input is taken into the design of trials. We expect to be increasing the number of trials that are open over the next several years as we have some very exciting new treatments to test."

[Read more from Dr. Shurin in the Director's Corner](#)

Sickle cell disease clinical practice guidelines

The NHLBI has launched an initiative to develop evidence-based, clinical practice guidelines to manage sickle cell disease across the lifespan. The goal is to develop guidelines that address key aspects of

care in clinical settings, focus on primary care, and identify evidence-based best practices. The guidelines will cover health maintenance, acute and chronic care, hydroxyurea usage, and transfusion therapy. They are expected to be released in the fall of 2011.

The draft chapter "Recommendations on the Use of Hydroxyurea Therapy in Sickle Cell Disease" was released during the symposium and is now available online. The recommendations are intended to assist primary care physicians, patients, and families in assessing whether hydroxyurea might be a beneficial therapy for an individual patient. The draft is available for review and comment.

[Learn more about the guidelines and access the draft chapter](#)

The patient-provider relationship

Do you listen to your patients? Do your patients believe that you are listening to them? Carlton Haywood, Jr., Ph.D., M.A., from the Johns Hopkins University School of Medicine, addressed challenges in sickle cell patient-provider communication in a symposium presentation. As both a researcher and an individual living with sickle cell anemia, Dr. Haywood is uniquely qualified to address patient-centered care.

Research on patient experiences reveals several themes that resonate among individuals with sickle cell disease and among parents of children with sickle cell disease, according to Dr. Haywood. These include:

- Not being treated with respect
- Not having enough done to control pain
- Perhaps being stigmatized as "drug addicted"
- Not being involved enough in decisionmaking and the actual health care process

Health care providers can better recognize the experiences of the individual and the family who are seeking care and better use their experiences to help in the care process.

Patient-centered behaviors that health care providers can foster include:

- Listen carefully.
- Explain things clearly.
- Provide information.
- Show respect for the patient's views.
- Demonstrate empathy.

While these may seem like basic skills, Dr. Haywood emphasized that they can go a long way toward demonstrating a patient-centered care approach, and they can ultimately lead to more beneficial outcomes.

Resources

- [A Century of Progress: Milestones in Sickle Cell Disease Research and Care brochure, 2010](#) (6.9 MB PDF)
- [Sickle Cell Disease Awareness and Education Strategy Development Workshop Report, 2010](#) (2 MB PDF)
- Sickle cell anemia [information](#), [quiz](#), and [widget](#)
- [Patient Voice: Tiffany McCoy Talks About Living With Sickle Cell Disease \(short video\)](#)
- [Sickle Cell Disease Information Center](#)

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<http://hp2010.nhlbihin.net/joinhin/news/professional/HINSCDHrckSymp.asp>