Sickle cell disease pain can occur daily and is much more severe than previously thought

January 14 2008

A new prospective study of pain in adults with sickle cell disease finds that daily pain is far more prevalent and severe than previous large studies have indicated.

Authors of the study gave diaries to 232 sickle cell disease patients to record daily pain and indicate whether they used hospital emergency or unscheduled ambulatory care for their pain.

Previous estimates about sickle cell pain assumed that if patients didn't go to the hospital or seek medical care for pain, they didn't have pain. Previous sickle cell treatments also were based on the number of visits to hospitals to relieve pain.

"The major finding of our study," said Wally R. Smith, MD, who directed the new study, "was that pain in sickle cell disease is a daily phenomenon and that patients are at home mostly struggling with their pain rather than coming into the hospital or emergency department."

The study, "A Prospective Study of Daily Pain in Adults with Sickle Cell Disease," is published in the Jan. 15, 2008, issue of *Annals of Internal Medicine*.

Sickle cell disease is a group of hereditary red blood cell disorders. In the United States, sickle cell syndromes are present in 1 in 400 African Americans. The disease is also found in high frequency in individuals from certain areas of the Mediterranean basin, the Middle East, and

India.

Sickle cell disease is caused by a mutation in a red blood cell gene that changes smooth, round blood cells into a sickle-shaped or C-shaped cells that are stiff and sticky and tend to clot in blood vessels. When they get stuck in small blood vessels, the sickle cells block blood flow to the limbs and organs and can cause pain, serious infections, and organ damage, especially in the lungs, kidneys, spleen and brain. Because of the potential for damage to organs, people with sickle cell disease have shorter-than-average life expectancy.

Hydroxyurea is the only FDA-approved treatment specifically for the disease. Other drugs can help symptoms and complications of the disease. Bone marrow transplantation can be curative.

Sickle cell disease causes both acute (rapid, severe, short-term) and chronic (long-lasting) pain. In sickle cell disease, the acute pain episodes are called crises. Previous studies of sickle cell pain have focused on crises.

In the current study, over half of the sickle cell disease patients completing up to six months of pain diaries reported having pain on a majority of days. Almost one-third had pain nearly every day.

“I believe that this study could change the way people view the pain of the disease. It is a chronic pain syndrome,” said Dr. Smith. “And the study results have implications for medical care, and research. We need more drugs to prevent the underlying processes that cause pain in this disease. And we need better treatments to reduce the chronic pain and suffering that these patients go through.”

Source: American College of Physicians