UNDERSTANDING
SICKLE CELL DISEASE

National Sickle Cell Awareness Month at UR Medicine







WHAT IS SICKLE CELL DISEASE?

Sickle Cell Disease (SCD or sickle cell) is the most common **inherited blood disorder** in the US. Sickled red blood cells change from a flexible disc to a fragile "C" or sickled shape, which blocks blood flow and breaks down more easily.

Poor blood flow causes tissue damage and leads to episodes of **severe pain and eventual permanent organ damage**. Red blood cell breakdown leads to anemia and yellowing of eyes and skin (jaundice).

WHO IS AFFECTED BY IT?

In the US, sickle cell affects primarily people who identify as Black and African American – however, it is not a "black disease," an outdated myth about this chronic illness.

SCD developed in regions close to the equator, and is therefore most common in people with African, Caribbean, South Asian, Middle Eastern, Mediterranean and Latino/a & Hispanic descent. The demographics of SCD in the US invites reflection on the historical context of slavery and the influx of African and Caribbean people into North America.

HOW DOES IT IMPACT PEOPLE?

In addition to medical complications, people with SCD face implicit bias and **stigma** from the medical system, due to structural racism and frequent need for opioid treatment. Missing school and work isolates individuals and creates barriers to academic and job achievement. People with SCD may experience recurrent complex trauma from multiple sources: the disease, family stress, academic & financial struggles, and biased, or even hostile treatment from the medical system, all of which contribute to poor physical and mental health.

HOW IS SICKLE CELL DISEASE TREATED ?

Symptom management starts at home with increased hydration, heat, and oral pain medicine. If the pain does not respond, patients are advised to seek care in the hospital.
IV opioid pain medications are the primary treatment for acute SCD pain in the hospital.

Blood transfusions can be life-saving, and are critical to treat or prevent SCD complications like stroke. Donated blood from people of color is often the best match for SCD patients, but donation rates are low: more donations from people of color are needed to lower the risk of transfusion complication.

Hydroxyurea is a medication that increases the production of healthier red blood cells and has the best, longest track record for disease modification. It reduces pain, protects organ health, prolongs life, and improves quality of life. Only three other FDA-approved medications are available, but they are not as effective and more options are needed.

Bone marrow transplantation (BMT) is the only known cure for SCD. The best results come from having a well-matched donor, like a full sibling, but not every patient will have this option. BMT requires chemotherapy and can be associated with high costs and serious risks.

On the horizon: Gene therapy for SCD was recently approved by the FDA and can be transformative for people with sickle cell. Gene therapy involves manipulation of a target gene in the patient, with no donor needed. Chemotherapy is required as part of gene therapy and long-term outcomes are still not clear. UR Medicine sickle cell treatment teams hope to be able to offer this exciting new option soon.

"WILL THEY SAY I'M DRUG-SEEKING?"

Finding stigma when looking for care

One of the most damaging stigmas towards people with sickle cell is the label of "drug seeking". Patients have **limited options** for treating severe SCD pain episodes. The need to utilize opioids reflects these limitations.

The need for opioids to treat pain can mean patients with SCD face assumptions that they have substance use disorder. Stigma against anyone who utilizes opioids for pain management combines with other types of bias people with SCD often experience related to their race, class, and chronic condition.

This **further damages** the livelihood of people with sickle cell because they may delay needed treatment and avoid going to the hospital to escape the discrimination that tends to happen in health care environments.

Sickle Cell Treatment @ UR Medicine

At UR Medicine, we provide evidence-based SCD care, guided by the **biopsychosocial model** from birth through adulthood. The pediatric SCD team offers a full spectrum of preventive, acute, and curative therapy at Golisano Children's Hospital. Teens and young adults are supported in their transition to the only adult SCD program in Upstate New York. The adult program provides comprehensive primary, specialty, and inpatient care, with attention to the specific needs of patients with SCD. The pediatric and adult teams work hand-in-hand with patients and the whole sickle cell community to continue to address barriers to quality care.

AWARENESS INTO ACTION

Education and language awareness can go a long way in supporting positive health care relationships.

Seek education: professional development and presentations (including from in-house experts!), documentaries & media like *Supacell (Netflix)*, community events, and making connections with people with SCD. Visit the QR code to learn more at www.sicklecelldisease.org.

Blood donation, particularly by people of color. The American Red Cross hosts frequent blood drives. For URMC staff, check the intranet for regular hospital drives (staff time for donation is protected: see HR policy 172).

Bone marrow donation: join the national donor registry with a simple cheek swab - learn more at www.bethematch.org.

LANGUAGE TO AVOID

Sicklers
Replace with *"people with sickle cell"*

Frequent Flyer
This phrase is dehumanizing and can indicate compassion fatigue - the need for frequent hospitalization is out of patient control.

"Your labs look fine" (pain assessment)
Labs and vital signs don't accurately communicate pain and distress levels - only the patient can do that!

Non Compliant
Try a less stigmatizing phrase, like *"not adhering to medication plans/protocols"*

"Did you take your home meds?"
While not inherently stigmatizing, this can be asked on repeat during a pain crisis - replace with *"What have you tried for the pain so far?"*

"How long have you had sickle cell?"
Because SCD is inherited, a better question is *"when were you diagnosed?"*

UR MEDICINE

*This micro-learning tool was developed as an initiative of the UR Medicine Interdisciplinary Sickle Cell Disease Workgroup to support SCD awareness and education during **National Sickle Cell Awareness Month**. This workgroup includes members of clinical and non-clinical teams, including pediatric & adult health care staff & faculty, patient navigators, the Nursing Office of Diversity, Equity, & Inclusion, members of the UR Office of Equity and Inclusion, and other passionate leaders in this field throughout the UR medicine system.*

UR MEDICINE RECOGNIZES SICKLE CELL AWARENESS MONTH