BLUE CROSS BLUE SHIELD OF MICHIGAN

Sickle Cell Disease Toolkit

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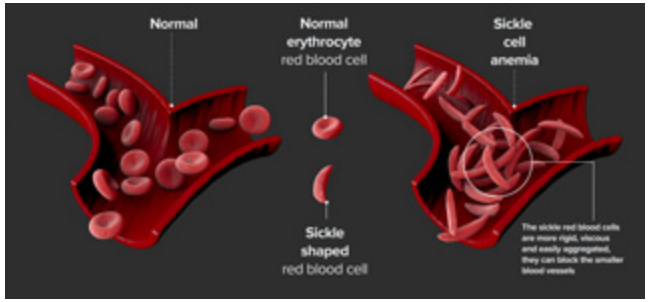
What is Sickle Cell Disease (SCD)?

a. Sickle cell disease (SCD) is a group of inherited red blood cell disorders.[1] Affected individuals have inherited a single sickle cell gene from both parents.[2]

- Healthy red blood cells are round and flexible; they move through the narrow blood vessels and provide oxygen to all parts of the body.
  - In SCD, the red blood cells become hard, sticky and fragile. They are crescent shaped like a farmer's sickle.
  - Sickle cells can get stuck in the blood vessels and block the flow of blood and oxygen to vital organs in the body. This can lead to repeated episodes of severe pain and organ damage.
  - Sickle cells break down early, causing anemia.
- Sickle cell trait vs. sickle cell disease
  - Individuals with SCT have inherited one sickle gene and one normal gene and typically do not have health problems related to SCD.
  - SCT cannot turn into sickle cell disease.
  - Unlike SCD, individuals with SCT only carry one defective gene and typically live normal lives.

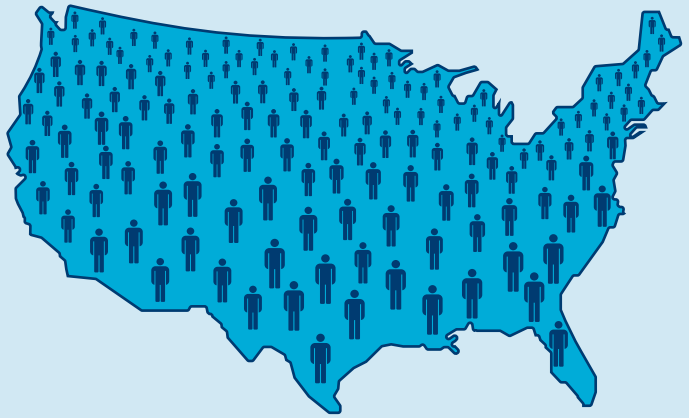


[1] CDC
[2] American Society of Hematology
[3] SCDAA
[4] CDC

b. Who does SCD impact?

According to the Sickle Cell Disease Association of America, it is estimated that

100,000 people in the United States have **SCD**



- Approximately 2,000 babies are born with SCD annually in the U.S.[3]
- In the U.S., cases of sickle cell disease are highest among African Americans, affecting 1 in every 365 births. Additionally, 1 in 13 African American babies are born with the sickle cell trait.[4]
- 1 out of every 16,300 Hispanic American babies are born with sickle cell disease in the U.S.

What is Sickle Cell Disease (SCD)?

c. What are the symptoms of SCD?

- Although quite variable, possible complications of SCD include:
  - Severe and unpredictable episodes of acute pain referred to as a "pain crisis"
  - Anemia
  - Increased susceptibility to infections
  - Swelling in extremities
  - Fatigue
  - Delayed growth
  - Vision problems/blindness
  - Lung and/or Kidney damage
  - Stroke
  - Shortened life expectancy
  - Joint damage
  - Leg ulcers
  - Yellow jaundice
  - Acute chest syndrome (ACS)[5]

What To Know about Sickle Cell Disease

a. *Myth: Those with sickle cell disease seek out pain medicine, even though they don't need it.*

- Fact: According to the CDC, the most common symptom of Sickle Cell Disease is severe pain. Often, this pain doesn't respond to over-the-counter medicines, so people may need opioids to give relief.
- Over time, an individual develops a tolerance to opioids. This leads to people needing higher doses to manage pain, leading to the perception of people with SCD as "drug seekers."[6]

b. *Myth: Individuals with SCD don't live past the age of 21.*

- Fact: More than 90% of those with sickle cell disease live well into adulthood.[7]

c. *Myth: Sickle cell trait is a mild form of sickle cell disease.*

- Fact: Sickle cell trait is not a disease – people with SCT usually do not have any of the symptoms associated with SCD and live a normal life.[8]

d. *Myth: Only people of African ancestry can have sickle cell disease.*

- Fact: SCD occurs more often in people whose ancestors lived in a region affected by malaria.[9]

e. *Myth: Bone marrow (stem cell) transplant is a universal cure.*

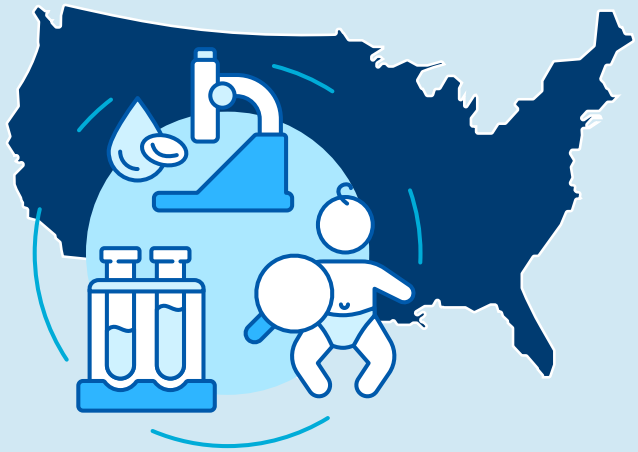
- The CDC reports that bone marrow or stem cell transplants have associated risks including mortality. Usually, the best donor is a sibling, as the bone marrow must be a close match for the transplant to work.
- These treatment options are most common in severe cases among children who have minimal organ damage from the disease.

[5] CDC
[6] Sickle Cell Disease Coalition
[7] American Society of Hematology
[8] CDC
[9] CDC

SCD Diagnosis and Treatment

a. Diagnosis:

In the U.S.,
every baby
is tested for
SCD
at birth.



- This testing also identifies infants with SCT.
- Newborn screening for SCD allows for the initiation of life-saving penicillin prophylaxis and early family education.
- If an adult is unaware of their sickle cell trait status, a test can be administered by their doctor. This allows individuals to make informed decisions about childbearing with respect to SCD.

b. Treatment:

- Care options could include:
  - Antibiotics for infection
  - Pain medication, including opioids
  - Disease modifying therapies
  - Blood transfusions
- Currently available disease modifying medications include:
  - Hydroxyurea
  - Voxelotor (Oxbryta)
  - Crizanlizumab (Adakveo)
  - L-glutamine (Endari)

c. Prevention:

- Living a healthy lifestyle by staying hydrated (at least 8 to 10 glasses of water) and eating a balanced diet of folic acid-rich foods are a few ways to combat complications of sickle cell.[10]
- Getting regular checkups with a primary care doctor can help prevent serious problems.

Talk with primary care doctor, go to local pharmacy or health department.

[10] Mayo Clinic

Available Resources

a. CDC Sickle Cell Disease (SCD)

b. American Society of Hematology (ASH)

c. Sickle Cell Disease Association of America

d. Sickle Cell Disease Association Michigan Chapter

e. MDHHS Newborn Screening Program

f. MI Blues Perspectives – Sickle Cell Disease and its Disproportionate Effect on African Americans in the U.S.

g. A Healthier Michigan

h. Sickle Cell Disease | CS Mott Children's Hospital | Michigan Medicine

i. Sickle Cell Lifespan Clinic | MSU Health Care | Michigan State University

j. What Is Sickle Cell Disease? | Henry Ford Health – Detroit, MI