UNDERSTANDING SICKLE CELL DISEASE

SCD is a blood disorder

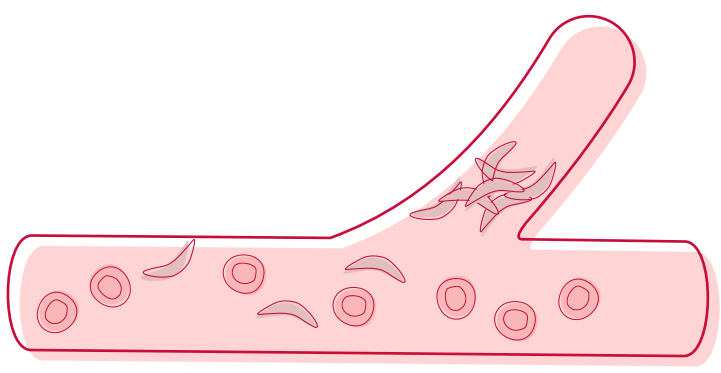
Sickle cell disease (SCD) is an inherited blood disorder that affects red blood cells. Normal red blood cells are round and flexible, which lets them travel through small blood vessels to deliver oxygen to all parts of the body.

It causes misshapen blood cells

SCD causes red blood cells to form into a crescent shape, like a sickle.

And creates painful complications

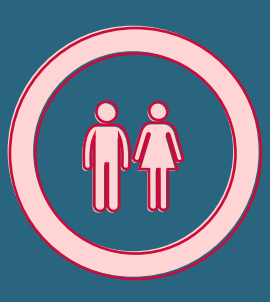
The sickle-shaped red blood cells break apart easily, clump together, and stick to the walls of blood vessels, blocking the flow of blood, which can cause a range of serious health issues.



WHAT CAUSES SCD?

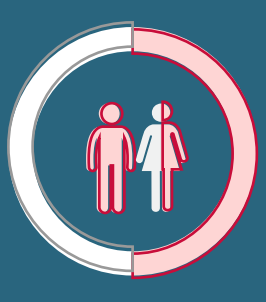
SCD is a genetic condition that is present at birth. It is inherited when a child receives two sickle cell genes—one from each parent. If only one sickle cell gene is inherited, the result is sickle cell trait (SCT). People with SCT can pass on the disease when they have a child.

When both parents have sickle cell disease



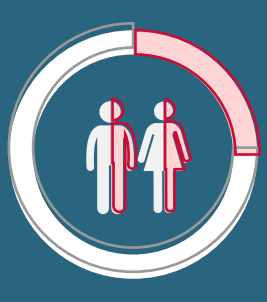
100% chance a child is born with SCD

When one parent has sickle cell disease, one parent has sickle cell trait



50% chance a child is born with SCD

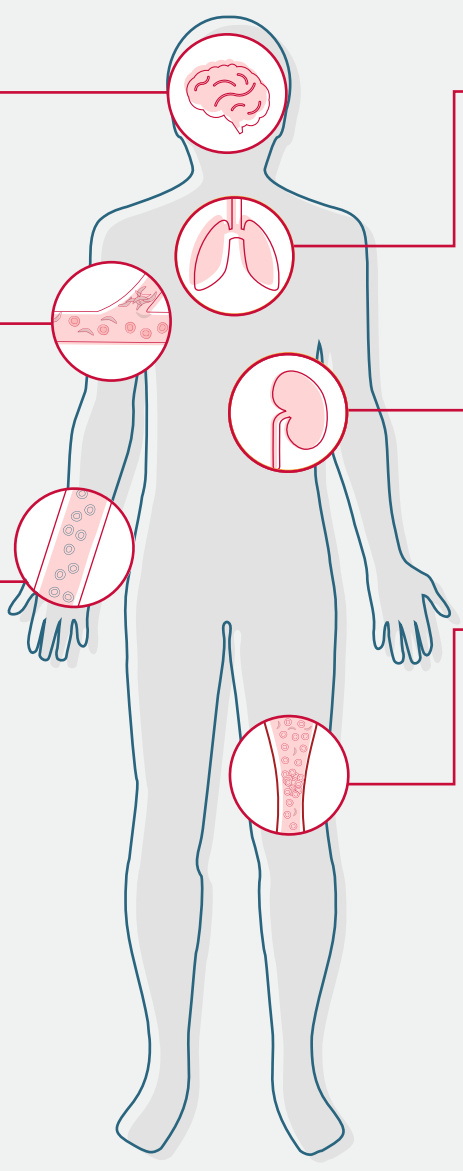
When both parents have sickle cell trait



25% chance a child is born with SCD

COMMON COMPLICATIONS OF SCD

Individuals living with SCD face many challenges and complications, ranging from mild to severe, throughout their life. Common complications include:



STROKE

Sickled cells can clog blood flow to the brain and cause a stroke.

ACUTE CHEST SYNDROME

Blockage of blood flow to the lungs can cause acute chest syndrome, a life-threatening condition like pneumonia, which can lead to symptoms such as chest pain, coughing, difficulty breathing, and fever. Abnormal lung function may also occur over time from chronic sickling in the lungs.

PAIN

Sickled cells often get stuck and clog blood flow through small blood vessels, causing chronic or episodic pain that can start suddenly, be mild to severe, and last for a varied length of time.

KIDNEY DISEASE

Sickling of red blood cells in the small blood vessels can cause a variety of kidney complications.

ANEMIA

Sickling causes red blood cells to die early, resulting in anemia. This can cause tiredness, dizziness, difficulty breathing, and pale skin color. Blood transfusions are sometimes needed to treat severe anemia.

VTE
(VENOUS THROMBOEMBOLISM)

Sickling of red blood cells can increase the risk of developing blood clots in the deep veins (deep-vein thrombosis, DVT), or in the lungs (pulmonary embolism, PE). DVT and PE can cause serious illness, disability, and, in some cases, death.

SCD IN THE U.S.

Who it Affects

While SCD can affect anyone, it is most often found in descendents of:

HISPANIC COUNTRIES

AFRICA

THE MIDDLE EAST

INDIA

Prevalence

SCD affects
~100,000 individuals

SCD occurs in
1/365 African American births

~3,000,000 people
have sickle cell trait

Treatment

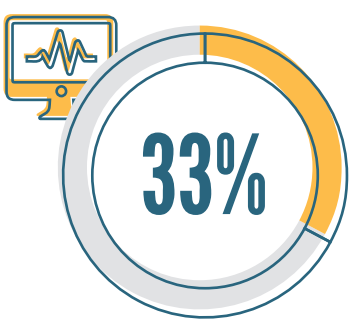
Only 4 medicines:
Hydroxyurea, L-glutamine, crizanlizumab-tmca, and voxelotor are currently the only FDA-approved medicines used to treat SCD.

Stem cell transplantation
has shown success in curing some individuals with SCD, but it is not widely available.

Transfusion
may help deliver oxygen to the body and unblock blood vessels.

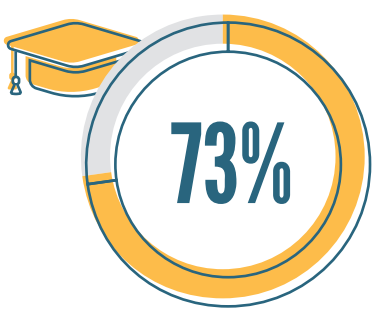
Access to Care

of adults with SCD
and frequent pain crises fail to get hydroxyurea, the recommended treatment.



of children with SCD
receive regular screening for stroke by age 2.

Training & Education



of family physicians
believe that more education and support tools would help avoid complications in managing SCD.

2019–2020 ASH CLINICAL PRACTICE GUIDELINES ON SCD

The American Society of Hematology (ASH) believes it is essential to provide updated treatment guidelines that reflect the newest evidence about SCD, ensuring the medical community can better treat the disease and people with SCD can make the best decisions for their care.

ASH partnered with the Evidence-Based Practice Research Program at Mayo Clinic, one of only a handful of evidence-based practice centers in the U.S., and more than 70 experts including hematologists, clinicians, specialists, and patient representatives to synthesize the research and develop new clinical practice guidelines for SCD.

For more information on these new guidelines, visit:
www.hematology.org/SCDguidelines

The American Society of Hematology (ASH) (www.hematology.org) is the world's largest professional society of hematologists dedicated to furthering the understanding, diagnosis, treatment, and prevention of disorders affecting the blood. For more than 60 years, the Society has led the development of hematology as a discipline by promoting research, patient care, education, training, and advocacy in hematology.

REFERENCES

1. www.cdc.gov/ncbddd/sicklecell/treatments.html
2. www.cdc.gov/ncbddd/sicklecell/documents/SCD-factsheet_What-is-SCD.pdf
3. www.nhlbi.nih.gov/health-topics/sickle-cell-disease