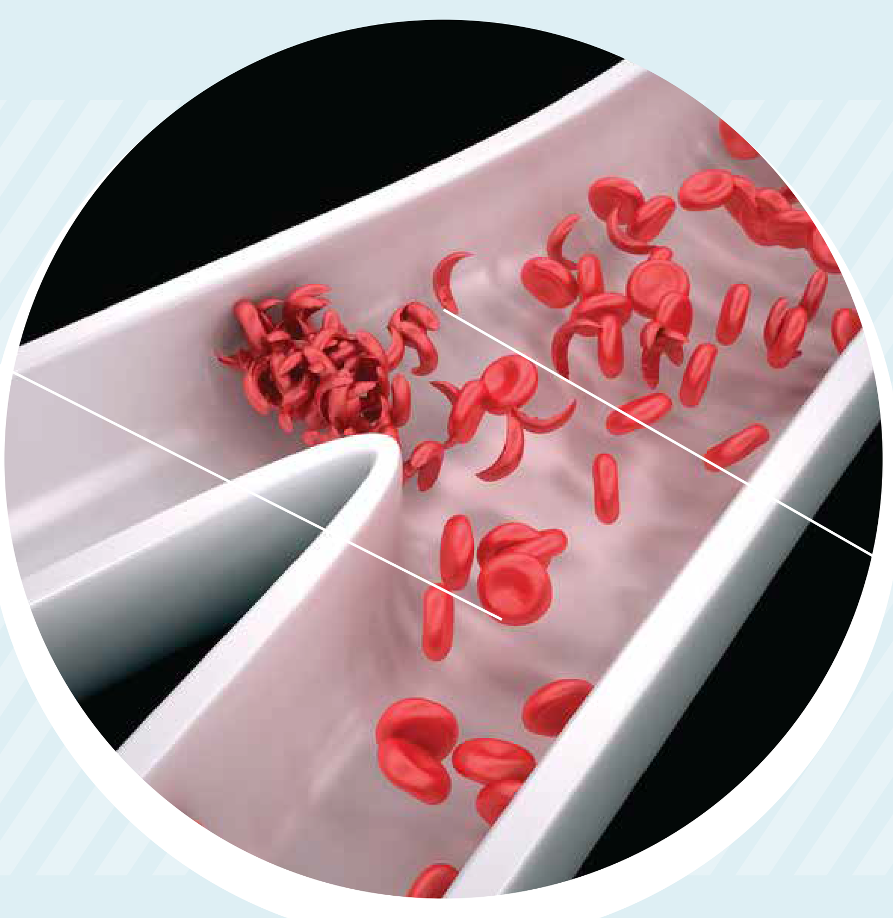
SICKLE CELL DISEASE

What is sickle cell disease?

Sickle cell disease is an inherited blood disorder that affects a person's red blood cells.

All red blood cells contain hemoglobin, which carries oxygen O_2 O_2 from the lungs to all parts of the body.

People **without sickle cell** disease have red blood cells that contain mostly **hemoglobin A.**



People **with sickle cell** disease have red blood cells that contain mostly **hemoglobin S**, which causes the red blood cell to change from a round circle shape to a banana shape.

Who gets it?

Sickle cell disease is an inherited disorder.

If both parents carry the trait for sickle cell

each child will have

- **25%** chance of not carrying the trait at all
- **50%** chance of carrying the trait
- **25%** chance of developing sickle cell disease

90,000 to 100,000 people in the U.S. have sickle cell disease

1 OUT OF 365 African-American children born in the US has sickle cell disease

1 OUT OF 12 African-Americans has sickle cell trait

Any race or nationality can carry the sickle cell trait

What are the symptoms?

People with sickle cell disease can have **complications** such as:

- Infections
- Painful Swelling of hands & feet (dacytlitis)
- Fatigue
- Stroke
- Organ Damage
- Pain

In some cases, people with sickle cell trait can have complications under **certain conditions** such as:

- If they are dehydrated
- If they are in a place of high altitude
- If they are exercising heavily

How do you treat it?

Stem cell transplant is the only approved cure for sickle cell disease, but gene therapy is currently being studied in clinical trials and could offer a cure.

Stem Cell Transplant Process

Collection
Stem cells are collected from a donor's bone marrow or blood.

Processing
Blood or bone marrow is processed in the lab to purify and concentrate the stem cells.

Chemotherapy
Chemotherapy is given to the patient.

Infusion
Thawed cells are infused into the patient.

IN 1983 a St. Jude patient was FIRST IN THE WORLD to be cured of sickle cell disease through a stem cell transplant

Many patients receive treatment with blood transfusions and an oral medication called hydroxyurea.

Transfusions of red blood cells - given every three to four weeks. This is the main treatment for fighting the strokes that can occur in sickle cell disease.

Hydroxyurea - used to help reduce cell sickling, as well as many of the major complications of sickle cell disease.

Other treatments include:

- **Endari (L-glutamine)** – used to reduce pain and fatigue.
- **Adakveo (crizanlizumab)** – used to reduce pain.
- **Oxbryta (voxelotor)** – used to reduce cell sickling.

*The patient had leukemia and sickle cell disease. A St. Jude doctor performed a stem cell transplant to cure her leukemia using bone marrow donated from her brother. Ultimately, **the patient was cured of both leukemia and sickle cell disease.***

What research is being done to help sickle cell patients?

IN PROGRESS

1. **The St. Jude-Methodist Sickle Cell Disease Transition Clinic**
   - Created to help 18 year olds make the leap from St. Jude to adult-care facilities of their choice.
   - Clinicians hope the transition program will become a national model for similar programs that encourage teens with sickle cell disease to continue their treatment as adults.
2. **A new study called SCCRIP (Sickle Cell Clinical Research and Intervention Program) looks at long-term outcomes in sickle cell disease.**
3. **St. Jude doctors are conducting research in the laboratory to develop cures for sickle cell disease.**

To learn more about sickle cell disease, go to **stjude.org/sickle-cell.**

St. Jude Children's Research Hospital
Finding cures. Saving children.

Sources

https://www.stjude.org/disease/sickle-cell-disease.html
https://www.stjude.org/content/dam/en_US/shared/www/patient-support/hematology-literature/fact-sheet-sickle-cell-trait.pdf