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

**Who gets it?**

Sickle cell disease can affect anyone from any race or nationality. It is most common in people of African descent. About 1 in 365 African-American children born in the U.S. has sickle cell disease. About 1 in 12 African-Americans has sickle cell trait.

**What is sickle cell disease?**

Sickle cell disease is a genetic disorder that affects red blood cells. 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. This can lead to swelling, pain, and other health problems.

**What are the symptoms?**

People with sickle cell disease can have complications such as:

- Infections
- Fatigue
- Painful swelling of hands and feet (dactylitis)
- Pain
- Strokes
- Organ damage
- Hemolytic anemia
- Sickle cell crisis
- Vaso-occlusive crises

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**

  1. **Collection**
     - Stem cells are collected from a donor’s bone marrow or blood.
  2. **Processing**
     - Blood or bone marrow is processed in the lab to purify and concentrate the stem cells.
  3. **Chemotherapy**
     - Chemotherapy is given to the patient.
  4. **Infusion**
     - Thawed cells are infused into the patient.

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

**What research is being done to help sickle cell patients?**

- **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.

- **A new study called SCGRIP (Sickle Cell Clinical Research and Intervention Program)**

  - Aims to find long-term outcomes in sickle cell disease.

- **St. Jude doctors are conducting research in the laboratory to develop new treatments for sickle cell disease.**

**Sources**