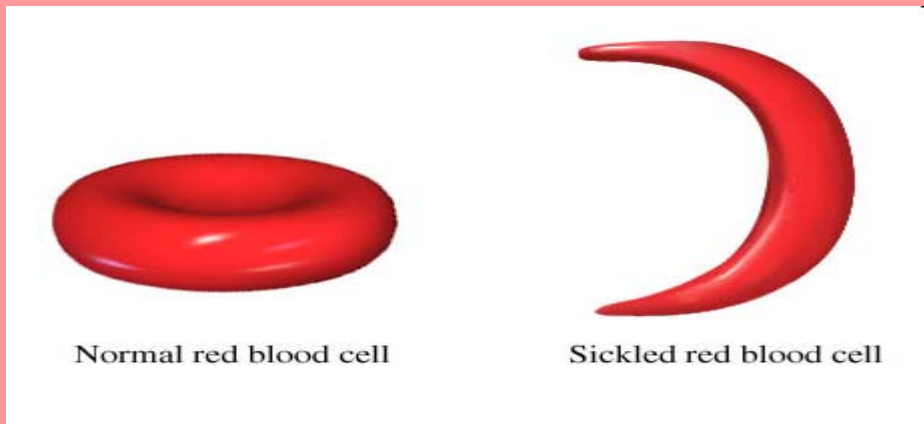
Sickle Cell Anaemia

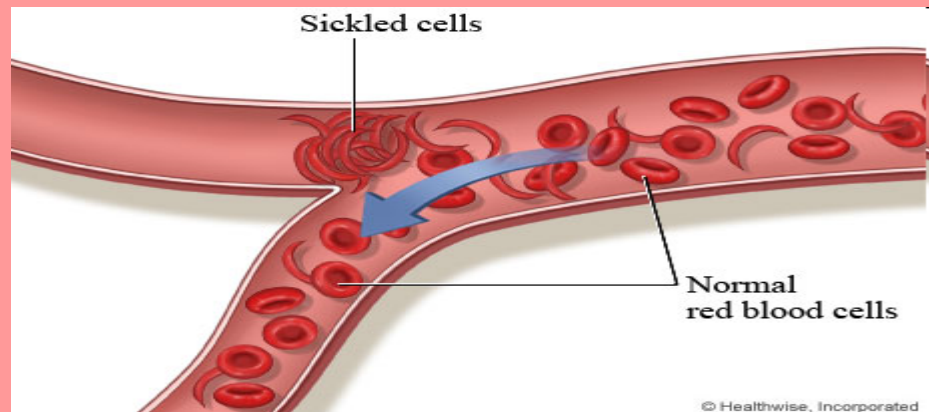
What Is Sickle Cell Anemia?

- Sickle cell is a serious disease in which the body makes sickle-shaped red blood cells. “Sickle-shaped” means that the red blood cells are shaped like a "C."
- Normal red blood cells are disc-shaped and look like doughnuts without holes in the center. They move easily through your blood vessels. Red blood cells contain the protein hemoglobin. This iron-rich protein gives blood its red color and carries oxygen from the lungs to the rest of the body.



What Is Sickle Cell Anemia?

- Sickle cell anemia is one type of anemia. Anemia is a condition in which your blood has a lower than normal number of red blood cells. This condition also can occur if your red blood cells don't have enough hemoglobin.
- Red blood cells are made in the spongy marrow inside the large bones of the body. Bone marrow is always making new red blood cells to replace old ones. Normal red blood cells last about 120 days in the bloodstream and then die. They carry oxygen and remove carbon dioxide from your body.



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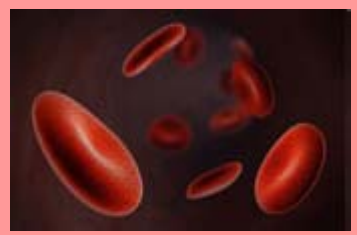
What Is Sickle Cell Anemia?

- In sickle cell anemia, a lower-than-normal number of red blood cells occur because sickle cells don't last very long. Sickle cells usually die after only about 10 to 20 days as compared to normal red blood cells that lasts up to 120 days. The bone marrow can't make new red blood cells fast enough to replace the dying ones.
- Sickle cell anemia is an inherited, lifelong disease. People who have the disease are born with it. They inherit two copies of the sickle cell gene—one from each parent.

What Is Sickle Cell Anemia?

- People who inherit a sickle cell gene from one parent and a normal gene from the other parent have a condition called sickle cell trait. People who have sickle cell trait don't have the disease, but they have one of the genes that cause it. Like people who have sickle cell anemia, people who have sickle cell trait can pass the gene to their children.



Did You Know?

- People with sickle cell disease start to have symptoms during the first year of life, usually around 5 months of age. Symptoms and complications of sickle cell disease are different for each person and can range from mild to severe.
- The reason that babies don't show symptoms of sickle cell disease at birth is because baby hemoglobin protects the red blood cells from sickling. Around 4 to 5 months old, the baby hemoglobin is replaced by adult hemoglobin and the cells begin to sickle.

Symptoms of Sickle Cell Anemia

- Symptoms may include bacterial infections, painful swelling of the hands and feet, fever, arthritis, leg ulcers, fatigue, anemia, eye damage.

  Listed below are some frequently experienced symptoms:

Hand-foot syndrome

This is usually the first symptom of sickle cell disease. Swelling in the hands and feet, often along with a fever, is caused by the sickle cells getting stuck in the blood vessels and blocking the flow of blood in and out of the hands and feet.

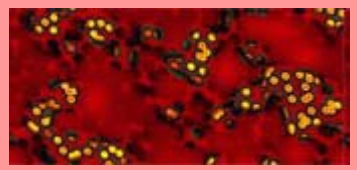
Symptoms of Sickle Cell Anemia

- **Pain Episode or Crisis**

This is the most common complication, and the top reason that people with sickle cell disease go to the emergency room or hospital. When sickle cells travel through small blood vessels, they can get stuck and clog the blood flow. This causes pain that can start suddenly, be mild to severe, and can last for any length of time.

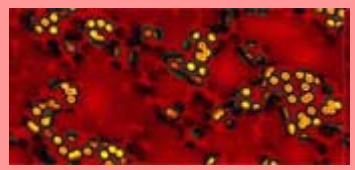
Symptoms of Sickle Cell Anemia

- **Anemia**
  This is a very common complication. With sickle cell disease, the red blood cells die early. This means there are not enough healthy red blood cells to carry oxygen throughout the body. When this happens, a person might have:
- Tiredness
- Irritability
- Dizziness and lightheadedness



Symptoms of Sickle Cell Anemia

- Fast heart rate
- Difficulty breathing
- Pale skin color
- Jaundice (yellow color to the skin and whites of the eyes)
- Slow growth
- Delayed puberty



Symptoms of Sickle Cell Anemia

Acute chest syndrome

This can be life-threatening and should be treated in a hospital. It is similar to pneumonia and symptoms include chest pain, coughing, difficulty breathing, and fever.

Prevention: Adults with severe sickle cell disease can take a medicine called hydroxyurea to help prevent acute chest syndrome. People taking hydroxyurea must be watched closely because the medicine can cause serious side effects, including an increased risk of dangerous infections.

Symptoms of Sickle Cell Anemia

A person who is on bed rest or has recently had surgery can use an incentive Spiro meter, also called blow bottle, to help prevent acute chest syndrome.

Acute chest syndrome

- **Treatment**: Depending on the cause, treatment might include oxygen, medicine to treat an infection, medicine to open up blood vessels to improve blood flow, and blood transfusions.

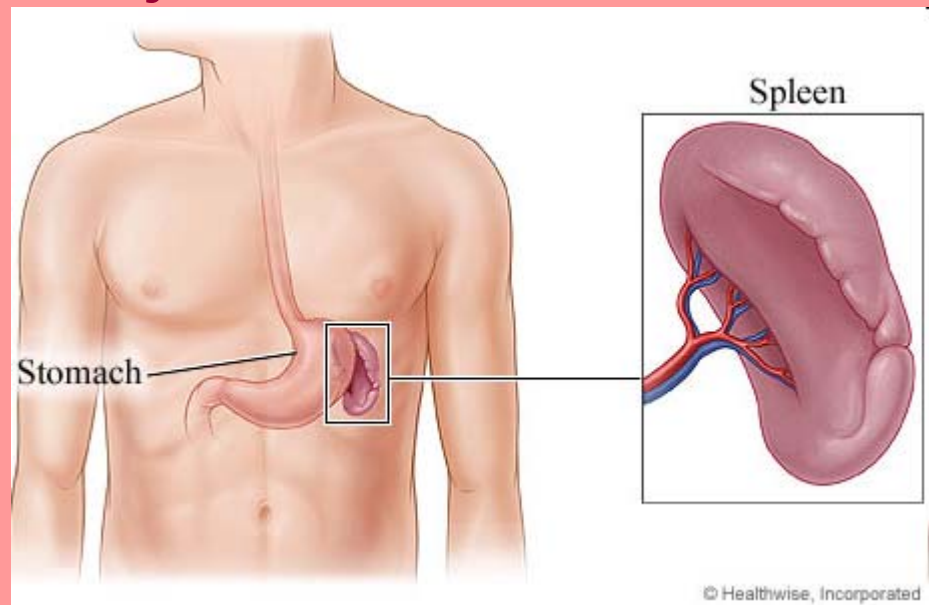
Symptoms of Sickle Cell Anemia

Infection

- People with sickle cell disease, especially infants and children, are more at risk for harmful infections. Pneumonia is a leading cause of death in infants and young children with sickle cell disease.

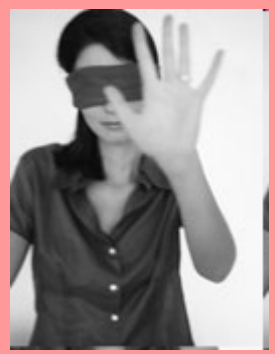
Splenic sequestration

This can be life-threatening and should be treated in a hospital. It happens when a large number of sickle cells get trapped in the spleen and cause it to suddenly get large. Symptoms include sudden weakness, pale lips, fast breathing, extreme thirst, abdominal (belly) pain on the left side of body, and fast heart beat.



Vision loss

- Vision loss, including blindness, can occur when blood vessels in the eye become blocked with sickle cells and the retina (the thin layer of tissue inside the back of the eye) gets damaged. **Prevention**: People with sickle cell disease should have their eyes checked every year to look for damage to the retina. If possible, this should be done by an eye doctor who specializes in diseases of the retina.



Leg ulcers

- This usually occurs on the lower part of the leg. They happen more often in males than in females and usually appear from 10 through 50 years of age. The cause of leg ulcers is unclear.

Stroke

- A stroke can happen if sickle cells get stuck in a blood vessel and clog blood flow to the brain. About 10% of children with sickle cell disease will have a stroke. Stroke can cause lifelong disabilities and learning problems.

Other Possible Complications

- Damage to body organs, tissues, or bones because not enough blood is flowing to the affected area(s).
- Gallstones.

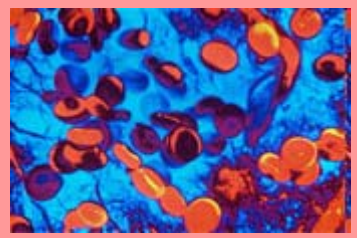
Fast Fact

- Taking iron supplements will not help people with sickle cell disease. This type of anemia is not caused by too little iron in the blood; it's caused by not having enough red blood cells.

  In fact, taking iron supplements could harm a person with sickle cell disease because the extra iron builds up in the body and can cause damage to the organs.

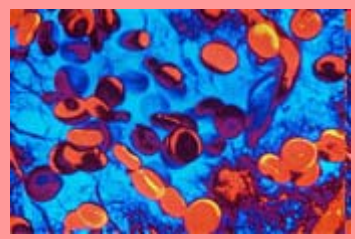
Treatment for Sickle Cell Anemia

- Aims to manage and prevent the worst manifestations of the disease and focuses on therapies that block red blood cells from stacking together, which can lead to tissue and organ damage and pain.



Prevention:

- There are simple steps that people with sickle cell disease can take to help prevent and reduce the number of pain crises:
  - Drink plenty of water.
  - Try not to get too hot or too cold.
  - Try to avoid places with high altitudes (flying, mountain climbing, or cities with a high altitude).
- Try to avoid places or situations with low oxygen e.g. (mountains)



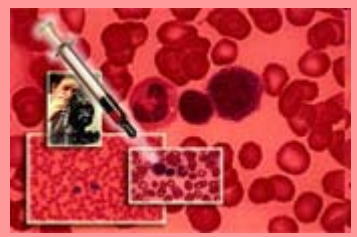
Prevention:

- Adults with severe sickle cell disease can take a medicine called hydroxyurea to help reduce the number of pain crises.

- People taking hydroxyurea must be checked often by a doctor because the medicine can cause serious side effects, including an increased risk of dangerous infections.

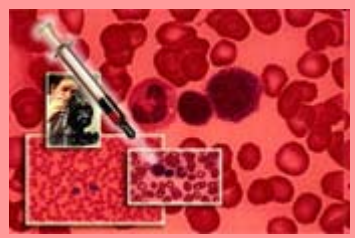
Prevention:

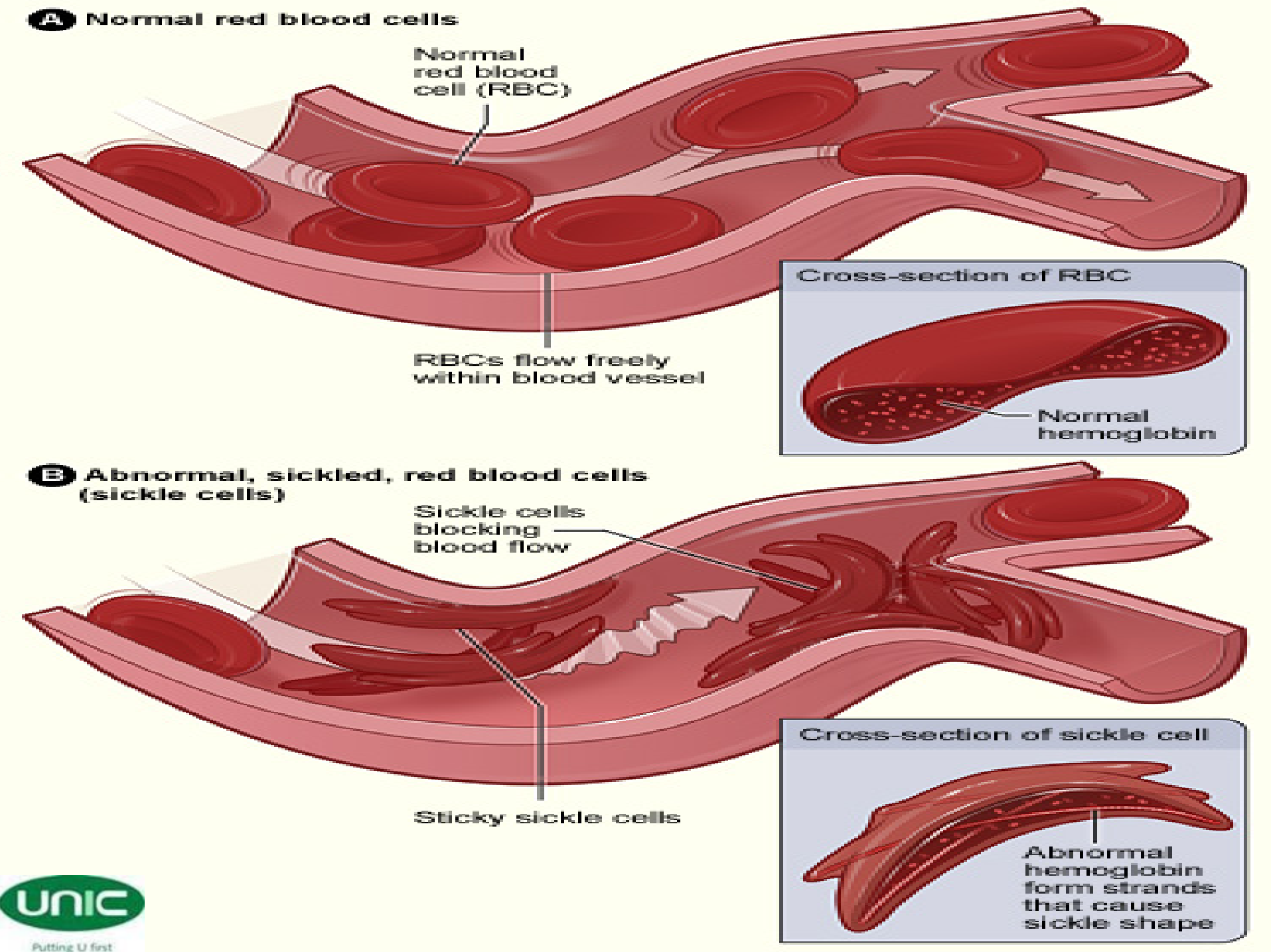
- : Vaccinations can protect against harmful infections. Babies and children with sickle cell disease should have all of the regular childhood vaccinations, plus a few extra. The extra ones are:
  - Flu vaccine (influenza vaccine) every year after 6 months of age.
  - A special pneumococcal vaccine (called 23-valent pneumococcal vaccine) at 2 and 5 years of age.
  - Meningococcal vaccine, if recommended by a doctor.



Prevention:

- In addition, children with sickle cell disease should receive a daily dose of penicillin, an antibiotic medicine, to help prevent infections. This can begin at 2 months of age and continue until the child is at least 5 years of age.





Living Well With Sickle Cell Disease

- Get regular checkups. Regular health checkups with a primary care doctor can help prevent some serious problems.
- Babies from birth to 1 year of age should see a doctor every 2 to 3 months.
- Children from 1 to 2 years of age should see a doctor at least every 3 months.
- Children and adults from 2 years of age or older should see a doctor at least once every year.

Living Well With Sickle Cell Disease

- **Prevent infections.** Common illnesses, like the flu, can quickly become dangerous for a child with sickle cell disease. The best defense is to take simple steps to help prevent infections.
- **Learn healthy habits.** People with sickle cell disease should drink 8 to 10 glasses of water every day and eat healthy food. Try not to get too hot, too cold, or too tired.
- Children can, and should, participate in physical activity to help stay healthy. However, it's important that they don't overdo it, rest when tired, and drink plenty of water.

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