

## **WORLD SICKLE CELL DAY**

### **June 19, 2015**

Sickle cell anemia, also called sickle cell disease, is a condition that affects the red blood cells. Red blood cells carry oxygen to organs in the body. Normal red blood cells are round.

When people have sickle cell anemia, some of their red blood cells can have an abnormal shape. They look like an antique tool called a “sickle”. The abnormal shaped red blood cells get stuck in the blood vessels easily, and so they do not carry enough oxygen to the body’s organs. This can cause pain or organ damage.

Sickle cell anemia is a life-long condition that people are born with. It usually affects people of African or Mediterranean descent. It is caused by an abnormal gene. To get the disease, people need to get the abnormal gene from their mother and father. If people get the abnormal gene from only one parent, it is called “sickle cell trait”. People with sickle cell trait do not usually have any symptoms of sickle cell anemia.

What are some of the symptoms of sickle cell anemia? Symptoms usually start after a baby is a few months old. Most commonly, people with sickle cell anemia have episodes of pain. This pain is known by the term “sickle cell crisis”. The pain can be in the bones, chest, back and other parts of the body. The pain can be mild or severe and can last for hours to days. People can have other symptoms with their pain including fever, swelling and trouble breathing.

Most episodes of pain just happen because of the abnormal red blood cells and can’t be predicted. However, sometimes the pain can be related to other things such as:

- Infection – people with sickle cell anemia are more likely than others to get certain infections.
- Dehydration – dehydration is when the body loses too much water.
- Weather or air conditions.
- Travel to high-altitude places – these are places where there is less oxygen in the air, such as mountains. Flying in an airplane might also cause symptoms for people with sickle cell anemia.

Sickle cell anemia can also cause many different health problems including:

- Anemia – anemia is a condition in which the body has too few red blood cells. It makes people tired or weak.
- Getting a lot of infections.
- Lung problems – symptoms can include trouble breathing or chest pain.
- Strokes – a stroke is when part of the brain is damaged due to lack of blood flow.
- Open sores on the skin, usually on the legs.
- An erection in men that last too long and is very painful. This is known as priapism.

Is there a test for sickle cell anemia? Yes. A blood test can show if someone has sickle cell anemia. All newborn babies in the United States are tested for sickle cell anemia before they leave the hospital. Some parents choose to test their unborn baby for sickle cell anemia; this can be done in different ways.

How is sickle cell anemia treated? Sickle cell anemia involves life-long treatment. Some treatments help prevent the different problems sickle cell anemia can cause. Other treatments are aimed at relieving pain for treating other problems like infection. Treatments include:

- Antibiotics – daily antibiotics help prevent infections in young children.
- Vaccines – getting all vaccines helps prevent infections. This is especially important in children and also true for adults.
- Hydroxurea (sample brand names: Droxin, Hydrea) this medicine helps the body make more normal red blood cells.
- Blood transfusions – a blood transfusion is when a person receives blood that was given by another person. People who have a high chance of having a stroke might get a blood transfusion to help prevent strokes. People might also receive blood transfusions to treat anemia.
- Folic acid – a vitamin called folic acid (also called Folate) is needed by people with sickle cell anemia to make red blood cells.
- Pain medicines – there are many types of pain medicines which can be used. Severe pain sometimes requires admission to hospital for intravenous pain medicine.
- Medicines to treat infections – it is important for people with sickle cell to be treated for infections right away.

A procedure called a “bone marrow transplant” or “blood stem cell transplant” can potentially cure sickle cell anemia. This procedure replaces the cells that make blood with healthy cells. This treatment is not done very often for sickle cell anemia because it has many side effects.

When should I call the doctor? You should call your doctor right away if the person with sickle cell anemia has any of the following problems:

- Gets a fever (temperature higher than 101.5°F) or has other signs of infection.
- Has pain that doesn't get better after it is treated at home.
- Has an erection that lasts longer than 2 to 4 hours.
- Has trouble breathing.
- Has signs of a stroke which include weakness of the arms or legs, slurred speech, facial drooping.

What else can people with sickle cell anemia do to stay healthy? They should have regular checkup and follow their doctor's instructions about treatments and tests. They should receive recommended vaccines that could prevent certain serious infections and they should also take good care of themselves which includes regular exercise, staying well hydrated, eating a healthy diet and getting enough sleep.

What if someone with sickle cell anemia wants to get pregnant? You should talk to your doctor and they may recommend that you see a “genetic counselor”. This person can tell you your baby's chances of having sickle cell anemia or sickle cell trait. If you have sickle cell and are pregnant, your doctor will follow you and your baby closely throughout your pregnancy. That is because sickle cell anemia can cause problems for both you and your baby.

Article Submitted By: Guy T. Easterling, DO, FACP  
Chief Medical Officer  
Taylor Health Care Group