Module 4: Complications of SCD

Contents

- **Infections**
- **Aseptic Necrosis (Bone Changes)**
- **Gallstones**
- **Problems with Kidneys**
- **Problems with Bladder**
- **Strokes**
- **Other Brain Problems**
- **Acute Chest Syndrome and Pneumonia**
- **Leg Ulcers**

Infections

As you have gotten older, and have taken steps to manage your health, your body has gotten stronger. You hopefully have been able to fight infections better. You may not need to go to the hospital for infections as often. You can be treated at home and get well. Even though things may be under control, you still have to remember that, with Sickle Cell Disease, some infections can cause problems and you still need to watch out for fevers. If you ever have a fever of 101.4°F or more, call your doctor's office and get ready to go to the emergency room or clinic for antibiotics.

If you are told that you can stay home with fever of 101.4°F, be sure to remind them that you have sickle cell disease and have to come in for lab tests and antibiotics.

When you go to the emergency room, make sure that you tell the check-in desk that you have a fever and sickle cell disease, and that your hematology doctors have instructed you to always come in urgently for fever of 101.4°F to get lab tests and antibiotics.

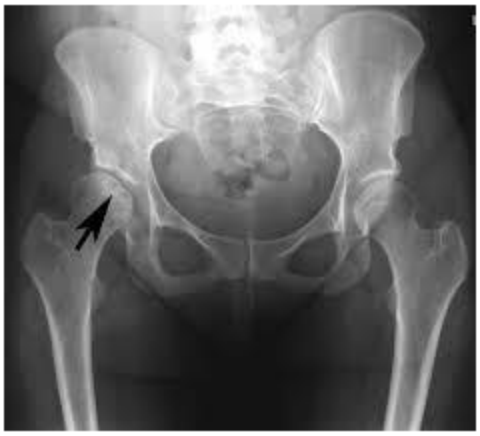
Aseptic Necrosis (Bone Changes)

Having Sickle Cell Disease can cause a lot of issues with your body including problems with tissues, organs, and bones. Aseptic Necrosis is the term for damage to the bones caused by sickle cell disease. When the hipbone is involved, it can cause chronic pain in the hip joint when a person walks. When the blood flow to the hip joint is slowed by sickle cells blocking blood vessels, the bone in the joint becomes flat and crooked. The hip can't move freely, and walking puts pressure on the joint, which can lead to more damage.

One sign to look for to know if you have hip joint damage is pain when you walk or run. Make sure to tell the doctor you have hip pain because early treatment can help

There are different types of treatment and the treatment you are prescribed depends on the extent of the problem. Sometimes a person needs to use crutches for a few months to take the weight off the joint. Other times, your doctor may suggest surgery or transfusion to stop the hip from changing shape.

Under extreme circumstances, if you can't walk without severe pain, the hip may need to be replaced. This can only be done when the bones have stopped growing, meaning this treatment is not available to children with Sickle Cell Disease. This can be something scary to hear and you may ask advice of people that have had hip replacements done. Do not rely only on advice from people who had hip surgery years ago. Artificial hip joints have improved a lot in the past few years, so make sure you talk with an orthopedist to learn the most up-to-date information.



Gallstones

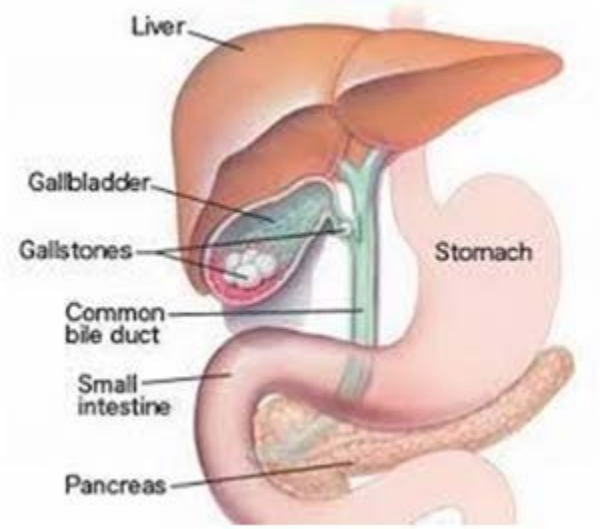
Another complication that Sickle Cell patients can suffer from is gallstones, which are small, hard masses formed in the gallbladder. About a third of children with sickle cell disease have gallstones by the age of seven and many others develop them later. Gallstones are formed from the waste products of broken-down red blood cells.

Gallstones collect in the gallbladder and form thick sludge or stones. Gallstones are not harmful but they can be extremely painful. If they get stuck in the gallbladder duct, they can cause a serious infection. Emergency surgery is then needed to take out the gallbladder.

There are symptoms to look for before gallstones get stuck in the duct. When the stones pass through the duct, they may cause pain in the right side of the abdomen. Also, when gallstones are stuck in the duct, a person's skin may become very yellow.

Because the gallbladder squeezes in response to a fatty meal, you might have gallstone pain after a meal with a lot of fried, oily, or creamy food. For example, a meal that could trigger gallstone pain might be fried chicken, French fries, creamy Cole slaw, a milkshake, and cream pie. One or two of these might be OK for the gallbladder, but the combination might cause gallstone pain.

Sometimes it is best to have the gallbladder removed to avoid serious problems in the future. Your doctor may suggest surgery to remove the gallbladder before an emergency happens. Taking out the gallbladder is the most common surgery in people with sickle cell disease. Most people can get along well without a gallbladder, but they may have trouble eating a lot of fatty foods at one time.



Problems with Kidneys

Another complication of Sickle Cell Disease is that it can damage the kidneys which means it takes more fluid to get rid of the body's wastes. People with sickle cell disease drink more fluids and pass urine more often than others. Remember that naturally you are more likely to become dehydrated, and it is important to know that when you become sick and may drink less than usual or lose more fluid by vomiting, diarrhea or fever, you can get dehydrated much faster than other people without SCD.

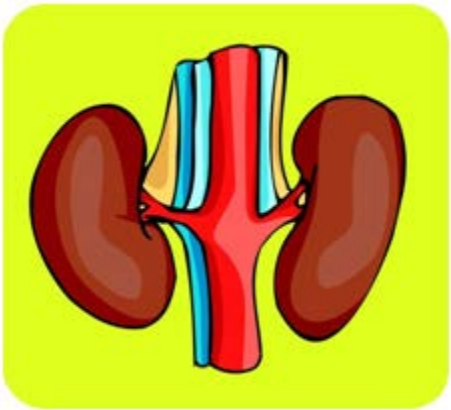
The first sign of dehydration will be darker urine color.

Late signs of dehydration:

- Very dark orange urine color
- Urinating much less than usual
- Dry, sticky mouth and lips
- Sunken eyes

Sometimes, the skin may feel different: if you pinch it, it doesn't return to normal right away.

If you notice any of these signs, you probably need to get IV fluids. Call your doctor and drink plenty of extra fluids!



Problems with Bladder

Another organ affected by Sickle Cell Disease is the bladder. With sickle cell disease, bladder infections are fairly common. If they are not treated promptly, they can spread from the bladder up to the kidneys and cause kidney damage. Call your doctor or nurse if you notice any of these signs:

- Foul smelling or cloudy urine
- Fever
- Burning and pain when urinating
- Abdominal or back pain
- An increase in the number of times you go to the bathroom
- Bedwetting after you've stopped wetting the bed
- Being unable to hold the urine

If you have a bladder infection, you will need to have your urine tested again from time to time. If the infection returns often, you may need to take pills every day so that the infections won't keep coming back.

Strokes

A stroke can be one of the worst problems caused by sickle cell disease. Early treatment can help prevent serious damage. A stroke occurs when part of the brain doesn't get as much blood as it needs. This is caused by sickled cells blocking a blood vessel in the brain, or bleeding in the head.

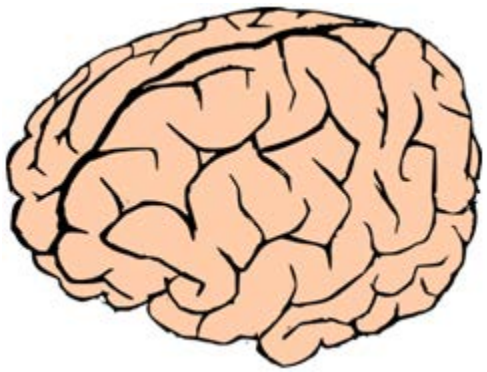
Watch for these signs of a stroke:

- Sudden weakness or tingling of an arm, leg or the whole body
- A difference in the way one side of the face or one eye moves compared to the other side
- Seizures (shaking that can't be stopped)
- Speech trouble
- Sudden, strong headache (“the worst headache of my life”)

If you see any of these signs, call your doctor and have someone take you to the hospital right away. The sooner you get help, the better.

Sickle cell stroke should not be managed in a hospital that is not used to caring for sickle cell patients, so it is important to know which hospitals around you are able to provide you the best care. Take time to do research on each hospital to find out your best options in case of an emergency.

If you are suspected of stroke or get diagnosed with stroke in a hospital that is new to you and that does not have your medical record on file, you and your family should insist on having your hematologist contacted. You should also consider being transferred to your hospital that knows your blood transfusion history (if you have had one) and that can provide you with sickle cell specialist care.



Other Brain Problems

Minor strokes and other brain problems can happen without having any signs or symptoms. The only way to know that a small stroke has happened is through some special tests. There are some tests, which check whether you are having any other types of brain problems due to sickled cells. The results of these tests will tell you if you need to take special steps to prevent strokes.

MRI (Magnetic Resonance Imaging): An MRI is done to look at the brain. Pictures of the inside of the brain are taken by a computer. It shows where an injury is in the brain. It usually takes about an hour and doesn't hurt. No X-rays or needles are used.

CAT Scan: When an MRI is not available, a CAT Scan can be used instead. It uses X-rays to make a computerized picture of the brain.

Arteriogram (Angiogram): This test shows the blood vessels in the brain. A needle is placed in a large blood vessel, and dye is injected. The dye shows which blood vessels are blocked. Medications are often given to help people relax during this test. Usually this requires preparation with blood transfusion and IV fluids, planned out with your hematologist.

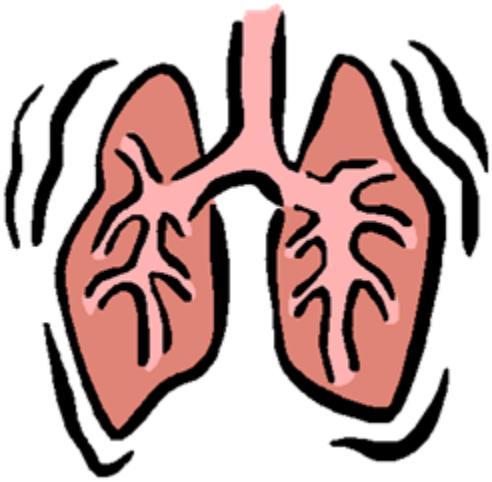
Acute Chest Syndrome and Pneumonia

Acute Chest Syndrome is chest pain that can be caused by an infection or sickle cells in the lungs. This can be a life-threatening complication of sickle cell disease. It often happens suddenly, when the body is under stress from infection, fever, or dehydration, so it is important to take preventive measures to make sure your body stays hydrated and well-taken care of. The sickled cells stick together and block the flow of oxygen in the tiny vessels in the lungs. Early treatment will keep it from getting worse.

Watch for these warning signs:

- Fever
- Coughing
- Rapid breathing
- Shortness of breath
- Difficulty breathing or "grunting"
- Severe chest pain

If you see any of these signs, go to the doctor right away. Call first and describe the signs you are showing. Multiple episodes of acute chest syndrome can cause permanent lung damage.



Sickle cell Acute Chest Syndrome should not be managed in a hospital that is not accustomed to caring for sickle cell patients, so it is important to know which hospitals around you are able to provide you the best care. Take time to do research on each hospital to find out your best options in case of an emergency.

If you get diagnosed with Acute Chest syndrome in a hospital that is new to you and does not have your medical history on file, you and your family should insist on having your hematologist contacted. You should also consider being transferred to

your hospital with sickle cell specialist care and that knows your blood transfusion history (if you have had one).

Leg Ulcers

Sometimes we are not able to see the changes that Sickle Cell Disease has on our bodies and other times the signs are right in front of us. One sign to keep a lookout for is ulcers on the legs.

Ulcers usually start as a small sore on the ankle, but they can grow large and get infected. Some sickle cell leg ulcers heal quickly, while others can sometimes take a very long time, even months, to heal.

Sickling of red blood cells happens more in the lower legs and ankles because of the pressure of standing. With sickling, small blood vessels get blocked, and blood can't get through to all parts of the body. An ulcer forms when lack of blood flow to the ankle skin kills the skin cells.

Go to see the doctor if you see either of these signs:

- A cut or wound that doesn't heal
- A patch of dry, itchy skin
- Swollen, shiny ankles

It is much better to treat leg ulcers when they are small than when they are larger. These are the basics for proper treatment for ulcers:

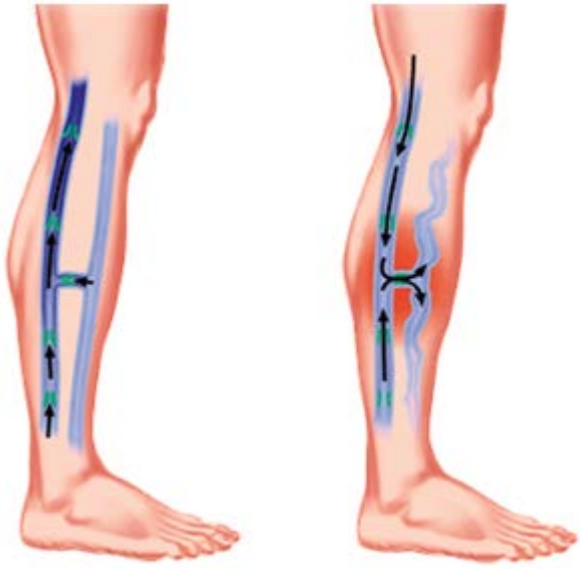
- Keep the area very clean and put on a fresh bandage twice a day or as often as advised.
- Stay off the feet and elevate them as much as possible.
- Wear clean white cotton socks and flat shoes until the ulcer is healed.
- Use lotion or ointment to keep the skin moist.
- Get it seen early by the doctors before it gets severe
- If the ulcer looks infected, you must see your doctor for antibiotics soon.

If the ulcer is large or has not started to heal in a few days, you may need to go into the hospital.

Keys to Prevention:

- Avoid getting IV needles placed in near the ankles. The damage to the blood vessels could increase your risk of developing leg ulcers.
- Avoid work that involves standing all day, because this will be hard on the ankle skin.

- Avoid walking with bare ankles and legs through weeds or brush that could injure and infect the skin above your ankles.



Great job! You're another step closer to finishing the CHECK Sickle Cell Online Program. Hopefully you now know more about problems you might have with sickle cell.

Now you can move on to the next topic, which talks about how you can stay healthy with sickle cell.