Sickle Cell Disease: Complications

Seek medical care immediately for any of the following:

- Fever ≥38.5°C in the mouth or ≥38°C under the arm
- Chest pain and/or shortness of breath
- Sudden severe abdominal pain and/or swelling
- Unexpected erection lasting >30 minutes
- Extreme fatigue, rapid heartbeat, and looking pale
- Inconsolable crying/irritability in infant
- Severe persistent pain that does not respond to medication
- Sudden slurred speech, seizures or paralysis/weakness

SICKLE CELL EMERGENCIES

Below are 6 sickle cell complications that require action quickly.
If your child experiences any of these symptoms, seek health care immediately.

1. Infections

Infections can cause severe complications in sickle cell disease patients. For example, viruses that may only cause a cough in healthy individuals may cause pneumonia in patient with sickle cell. Also, people with sickle cell disease do not have functioning spleens so they are at higher risk for more serious bacterial infections like meningitis. These infections can be fatal and must be treated promptly. Signs of infection include:

- Fever ≥38.5°C in the mouth or ≥38°C under the arm
- Pain, swelling, warmth or reddened skin over a joint or bone
- Painful urination
- General irritability or feeling unwell

2. Acute chest syndrome

Acute chest syndrome occurs when lungs are deprived of oxygen during a crisis. It is a life-threatening complication, usually caused by an infection or sickling in the lungs. Acute chest syndrome requires intense treatment with oxygen, IV fluids, pain medications, antibiotics, and sometimes transfusions. To identify acute chest syndrome, look for the following symptoms:

- Fever ≥38.5°C in the mouth or ≥38°C under the arm AND
- Increased rate and/or work of breathing
- Wheezing or coughing
- Chest pain

Not every episode of acute chest will show all of the symptoms listed, so it is important to seek help immediately with any of these.

3. Splenic Sequestration

Splenic sequestration occurs most commonly in children under 5 years of age, but can also occur in older children and teens. When sickle cells block blood flow away from the spleen, blood becomes trapped in the spleen and causes it to become very large very quickly. Less blood is available for the rest of the body and the patient can experience shock. An enlarged spleen can also rupture and cause internal bleeding. Splenic sequestration must be treated immediately in a hospital. Treatment includes IV fluids, pain medication and antibiotics. Blood transfusions may be required to restore blood volume, and surgery may be required to remove the spleen. The following are symptoms of splenic sequestration:

- Sudden weakness
- Paleness, especially of the lips, gums and nails
- Increasing abdominal pain and/or abdominal swelling
- Increasing spleen size
- Fever ≥38.5°C in the mouth or ≥38°C under the arm

It is important that patients and parents know how to feel for the spleen so that enlargement may be detected early.

4. Priapism

Priapism is a prolonged and painful erection of the penis *without sexual desire*. It occurs when sickle cells block the blood circulation in the penis, causing an erection that lasts several hours to days. Priapism is extremely painful and if untreated can cause long term damage and impotence. Treatment includes pain medications, IV fluids, and blood transfusions. Sometimes more invasive measures are required. From a young age, boys should understand the importance of reporting priapism as seeking immediate treatment can prevent permanent damage. Signs of priapism are:

- Painful, prolonged swelling of the penis
- Unwanted erection that lasts longer than 30 minutes

5. Aplastic Crisis

Certain viruses such as parvovirus can cause the bone marrow to "shut down" and stop producing red blood cells. This causes a

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sudden severe drop in hemoglobin, and is called an *aplastic crisis.* Treatment usually involves blood transfusions until the body recovers. Symptoms of an aplastic crisis may include:

- Sudden fatigue or lack of interest in playing
- Paleness
- Shortness of breath
- Rapid or pounding heartbeat

6. Stroke

In sickle cell patients, stroke can occur when sickle cells gather and block the blood flowing to the brain, causing part of the brain tissue to die from lack of oxygen. About 10% of children with sickle cell disease suffer stroke. Transcranial Doppler (TCD) testing should be done annually to detect risk for stroke early. Signs of a stroke include:

- Sudden blurred vision or loss of vision
- Persistent or sudden strong headache
- Dizziness and/or fainting
- Garbled speech or inability to speak
- Sudden weakness or tingling of an arm, leg or body
- Paralysis of one side of the body

Anyone experiencing these symptoms should call 911 immediately. Children that have had a stroke or are at high risk for stroke may get monthly blood transfusions to reduce the risk of future episodes.

OTHER SICKLE CELL COMPLICATIONS

Sickle cell patients may experience other complications as well. These are less urgent, but still require treatment.
Be sure to notify your hematology team if you have signs of any of the following.

Gallstones

The gallbladder is an organ that stores bile, a fluid that aids in digestion. Bile is made from bilirubin, which is released from red blood cells when they break down. Because sickle cells break down so frequently, people with sickle cell disease often have high levels of bilirubin. Bilirubin can build up in the gallbladder and form crystals called *gallstones*. Symptoms of gallstones include:

- Feeling overly full after meals
- Pain in the upper right portion of the abdomen, especially after a fatty meal
- Nausea and vomiting
- Increased yellowing of the skin and eyes (jaundice)

Treatment is not necessary unless pain is severe. Any patient experiencing these symptoms should let their hematology team know. An ultrasound is used to look for gallstones, and if necessary the gallbladder may be surgically removed.

Impaired Kidney Function

People with sickle cell disease are unable to concentrate their urine fully. They may urinate more frequently than other people, and many children have difficulties with bedwetting. Patients with sickle cell are also more prone to kidney infections and long term kidney damage. The treatment for kidney problems varies depending on cause and severity of the problem. Infections can be treated with rest, fluids, and antibiotics. Kidney damage may require stronger medications, and kidney failure requires dialysis and a kidney transplant. The following symptoms indicate there may be issues with kidney function:

- More frequent urination than is usual
- Painful urination and or blood in urine (red or brown)
- Fever ≥38.5°C in the mouth or ≥38°C under the arm

Other Long Term Complications

Sickle cells continuously form small blockages in vessels throughout the body, even when there are no overt symptoms such as pain. This is often referred to as “silent sickling.” Over time, silent sickling can cause damage to various organs such as the heart, liver and lungs. It can also cause visual and hearing impairments, pulmonary hypertension, joint deterioration and skin ulcers on the lower legs. These complications take years to develop and usually occur in adulthood, though more severe cases may develop earlier. Your hematology team will perform various tests throughout your life to monitor for the development of these complications.

For more information, contact:

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