

## **COMPLICATIONS OF SICKLE CELL DISEASE**

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### **WARNING SIGNS AND SYMPTOMS**

- Fever – 38.5C or above
- Irritability – readily excited to impatience or anger
- Pallor – looks pale in the face or tongue
- Breathing – trouble breathing, short of breath, fast breathing
- Headache – sudden onset or constant
- Heartbeat – very fast or pounding
- Pain – head, chest, joints, abdomen, penis (with prolonged unexpected erection)
- Swelling – hands, feet, joints
- Hemiparesis – paralyzed on either side of the body

### **ACUTE CHEST SYNDROME**

Acute chest syndrome occurs when lungs are deprived of oxygen during a crisis. It can be very painful, dangerous and even life threatening. It can be caused by infection and/or infarction (blockage in blood vessels that cut off oxygen). To identify acute chest syndrome, look for the following symptoms:

- Fever of 38.5C or above
- Rapid or hard working breathing
- Wheezing or coughing

Acute chest pain often lasts for several days and, in about half of patients, severe pain develops about two and a half days before there are any signs of lung abnormalities.

Acute chest syndrome must be treated immediately. Treatment can include supplemental oxygen, administration of fluids, pain medications, steroids, antibiotics, transfusions, and special diagnostic tests.

### **INFECTIONS**

Infections are a common and important cause of severe complications in sickle cell disease patients. Children with sickle cell disease have less resistance to infection and develop infections more easily and more frequently than other children. Serious infections can include respiratory infections such as pneumonia, kidney infections, osteomyelitis (serious bone infections) and meningitis. To identify an infection, look for the following symptoms:

- Fever is the number one indication for the presence of an infection, and temperatures over 38.5C warrants a call to the physician
- Unexplained tiredness
- Pain, swelling, warmth, reddened skin over a joint or bone
- Painful urination

A fever in a patient with sickle cell disease of 38.5C or higher is a medical emergency, and these patients must be seen by health care professionals as soon as possible. Treatment includes antibiotics and special diagnostic tests.

## **SPLenic SEQUESTRATION**

Splenic sequestration occurs most commonly in children under 5 years of age, but can occur at any age. It occurs when there is sickling in the splenic vein resulting in the sudden entrapment of a large amount of blood in the spleen. The spleen suddenly becomes very large and complications can occur. To identify a splenic sequestration, look for the following symptoms:

- Sudden weakness
- Paleness (especially lips, gums and nails)
- Abdominal pain
- Increase in abdominal and spleen size
- Fever

Splenic sequestration must be treated immediately, and these children should go directly to the hospital. Treatment can include administration of fluids, pain medication, antibiotics, special diagnostic test, blood transfusions and surgery.

## **ANEMIA AND APLASTIC CRISIS**

Anemia is a significant characteristic in sickle cell disease because of a shortened life span of the sickled red blood cells. Episodes of a sudden severe drop in hemoglobin (ie. the child becomes more anemic than usual) is called an aplastic crisis. In about 80% of cases aplastic crises are triggered by a virus called human parvovirus B19 which is common and usually harmless in healthy individuals. To identify an aplastic crisis, look for the following symptoms:

- Sudden tiredness
- Paleness
- Lack of interest in play
- Shortness of breath
- Fast heartbeat

If you notice any of these symptoms, you should call your health care provider immediately. Shortness of breath and a fast heartbeat should be evaluated in an emergency setting as soon as possible. Treatment can include blood transfusion and special diagnostic tests.

## **STROKE**

Stroke is a serious complication of sickle cell disease. It occurs when a part of the brain does not get as much blood as it needs. Between 8 to 10% of patients suffer stroke, typically at age 7. There are no tests that can definitely determine which children are at highest risk for a first stroke. It is important to recognize the following symptoms:

- Sudden loss of or blurred vision
- Persistent headache or sudden strong headache
- Dizziness
- Fainting
- Trouble with speech
- Sudden weakness or tingling of an arm, leg or whole body
- Paralysis of one side of the body

If any of these symptoms are present in your child, seek out emergency medical care immediately. The goal of treatment is to prevent a second stroke. The child may get a blood transfusion, be on anti-blood clotting medications, and undergo special diagnostic tests.

## **KIDNEY**

Damage to the kidneys of children with sickle cell disease occurs because of repeated sickling process. Problems with urination are very common, and children urinate frequently due to an inability to concentrate urine. Uncontrolled urination during sleep is also a particular issue. As well, these children are more prone to kidney infections. To identify difficulties with kidneys, look for the following symptoms:

- Frequent urination (more than usual)
- Painful urination
- Bloody urine
- Fever greater than 38.5C

All children with sickle cell disease need to get plenty of fluids (preferably water and juice) throughout the day, everyday to help prevent complications with the kidneys and dehydration. The child should be allowed to use the bathroom frequently and be encouraged to tell you if their urine becomes bloody. Treatment for kidney problems can include bed rest, plenty of fluids, antibiotics and special kidney medications.

## **GALLBLADDER DISEASE**

About 30% of children with sickle cell disease have gallstones (cholelithiasis) and by age 30, 70% of patients have them. The gallstones are a result of the excess bilirubin (product of red blood cell breakdown) caused by sickling and cell destruction. Gallstones do not cause symptoms in most cases but when symptoms occur they can include:

- Feeling overly full after meals
- Pain in the upper right portion of the abdomen
- Nausea and vomiting
- Fatty food intolerance
- Increased yellow appearance

Treatment is not necessary unless symptoms and pain are severe. Let your health care professional know and an ultrasound is usually used to confirm the diagnosis. If necessary, the gallbladder may be surgically removed.

## **PRIAPRISM**

Priapism is a prolonged and painful erection in males without sexual desire and occurs when sickle cells block blood circulation in the penis. Erections can last from several hours to days. It is important to counsel children and adolescents the importance of reporting priapism. If left untreated, partial or complete impotence can occur in 80% of cases. Symptoms to monitor for include:

- Painful, prolonged swelling of the penis
- Sustained unwanted erection

Seeking out immediate treatment is important to prevent permanent damage. Blood transfusions may be used to reduce the sickling that caused this condition and pain medications to help control the pain.