# Important Contact Information

<table>
<thead>
<tr>
<th>Office/Unit</th>
<th>Phone Number:</th>
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<tbody>
<tr>
<td>Outpatient Clinic:</td>
<td>708.684.3898</td>
</tr>
<tr>
<td>Make an appointment:</td>
<td>708.684.3113</td>
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<tr>
<td></td>
<td><em>Please leave a message and a receptionist will call back to schedule</em></td>
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<tr>
<td>4th floor nursing station:</td>
<td>708.684.5880</td>
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<tr>
<td>2nd floor nursing station:</td>
<td>708.684.1300</td>
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<tr>
<td>Pediatric Intensive Care Unit:</td>
<td>708.684.1308</td>
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**Clinician:**

<table>
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<tr>
<th>Program Coordinator: Fran Majca, RN, Sickie Cell Nurse Coordinator – Call Mon, Tues, Thurs and Fri between 8 and 5pm for general questions of coordination related concerns.</th>
<th>Phone Number: 708.684.4247</th>
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<tbody>
<tr>
<td>Social Worker: Maria Hartleben, LCSW Call with any social work concerns</td>
<td>708.684.4213</td>
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<tr>
<td>Psychologist: Dr. Gabrielle Roberts</td>
<td>708.684.3590</td>
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<tr>
<td>School Coordinator: Deide Fischer</td>
<td>708.684.5335</td>
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<tr>
<td>Child Life Specialist: Mary Butler</td>
<td>708.684.9151</td>
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<tr>
<td>Manager of Keyser Center: Tammy Klapp, RN</td>
<td>708.684.4208</td>
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<tr>
<td>Advanced Practice Nurse: Kim Duback, APN</td>
<td>708.684.4207</td>
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Our Team

As a parent, you are part of the team that will take care of your child. Depending on your child’s needs, any of the following specialists may be part of your child’s care team:

**Attending Physician:** a doctor who specializes in blood and manages the medical care of your child.

**Clinical Nurse Practitioner:** a nurse who has completed a master’s program and specialty training in caring for children with sickle cell disease. He or she coordinates the medical and nursing care of your child.

**Nurse Coordinator:** a person who collaborates with the physician and health care team to develop a treatment plan of care, assesses the needs of the child and family, provides patient/family education, monitors all clinical activities and evaluates the effectiveness of interventions.

**Resident:** a doctor who has completed medical school and is now receiving specialty training in pediatrics.

**School teacher/liaison:** a teacher who works in the hospital to help your child keep up with school work during the hospital stay. This person will also help communicate with school any accommodations or medical needs your child may need when they return.

**Social Worker:** a person who is trained to help you and your child cope with illness and hospitalization through counseling, support groups, financial assistance and resource referral.

**Child Life Specialist:** a person with special training in child development, giving them an understanding of how children react to illness hospital stays. They help children cope with their medical condition and the treatment they receive.

**Chaplain:** A person who is trained to offer spiritual care, support and prayer according to each family’s individual needs.

**Nutritionist:** a person who is trained to evaluate your child’s nutritional needs and weight. They can help provide teaching and support about eating and drinking when your child goes home.

**Pharmacist:** a person who is trained to prepare the medicines and nutritional support that your child needs.

**Physical therapist:** a person who works with your child to maintain and restore a level of fitness through strength and endurance activities.
Sickle Cell Frequently Asked Questions:

What causes sickle cell anemia?
Sickle cell anemia is an inherited condition, like hair color or eye color. People with sickle cell disease inherit two defective hemoglobin genes – one from each parent – that affect the way their red blood cells are formed. The abnormal hemoglobin gene causes a distorted shape of red blood cells, making them form a sickle or C shape.

What is sickle cell trait?
Sickle cell trait is when somebody inherits a copy of the sickle cell gene from only one parent, and gets a normal hemoglobin copy from the other parent. People with sickle cell trait do not have sickle cell anemia, but they can pass the sickle cell gene to their children. If you have sickle cell trait, it may be helpful to meet with a genetics counselor before having children of your own.

Who is at risk for sickle cell anemia?
In the United States, sickle cell anemia affects about 70,000 people and the major population affected is African Americans. The disease occurs in about one out of every 500 African-American births and one in every 1,000 to 1,400 Hispanic births.

What are the different forms of sickle cell anemia?
Sickle cell anemia (hemoglobin SS disease): This is the most common and severe type of sickle cell anemia. These patients have a large amount of hemoglobin S in their red blood cells and no normal hemoglobin.

Sickle-hemoglobin C (hemoglobin SC disease): Typically a milder form of sickle cell disease. These patients have inherited two abnormal hemoglobin genes in their red blood cells - one hemoglobin S and one hemoglobin C.

Sickle thalassemia disease: Patients have a large amount of hemoglobin S in their red blood cells, combined with a second type of abnormal hemoglobin. Severity of this illness can vary. Beta-thalassemia is another type of anemia that causes the red blood cells to be smaller and contain less hemoglobin.
Newborn Screening:

In the United States, newborn screening for sickle cell disease is performed in all states. The newborn screen is performed on all babies before they leave the hospital for the first time and results can show whether the baby has sickle cell anemia, sickle cell trait, or other hemoglobin problems.

Hemoglobin Electrophoresis:

A hematologist can perform a blood test called hemoglobin electrophoresis, which can measure how much sickle hemoglobin is in the baby’s red blood cells.

It also is possible to diagnose sickle cell anemia before a baby is born by testing the amniotic fluid or tissue from the placenta.
Common Lab Tests

**Complete Blood Count (CBC):**
This blood test can tell you the number, shape and size of red blood cells and the hemoglobin. The normal hemoglobin level for patients with Sickle Cell Anemia is between 6-10 g/dl. Children without sickle cell anemia usually have a hemoglobin between 11-14 g/dl.

**Reticulocyte Count (blood test):**
Reticulocytes are young red blood cells. The number of these cells shows whether the bone marrow is doing its job well, making and releasing young red blood cells into the bloodstream.

**Kidney and liver function tests (blood test):**
These tests show if the kidneys or liver have been damaged by sickle cell anemia.

**Urine test (Urinalysis)**
This test can determine if there is a bacterial infection in the urine. It can also tell if there are red blood cells in the urine, which can be caused by slight bleeding in the kidneys.

**Ferritin**
A simple blood test to measure iron levels in the body and determine whether there is enough iron in the body to make new red blood cells. As patients undergo repeated blood transfusions, iron can build up in the body and iron deposits in many main organs can cause damage. A medication can be prescribed to treat patients with chronic iron overload.
Common Medical Tests

**Pulse Oximetry**
A test used to measure the oxygen level in the blood. A plastic band is placed around the patient’s index finger and a machine then measures the oxygen going through the blood.

**X-Ray:**
A picture taken of parts inside of the body. X-rays can show if there is avascular necrosis of a bone or if an infection or fluid is in the lungs. X-rays do not hurt.

**MRI (Magnetic Resonance Imaging):**
A test that produces detailed images of organs and bones. There is no radiation exposure. Test used to look at the brain or at bones. The pictures are taken with a computer.

**Cardiac Echocardiography (Echo):**
This test is used to determine heart function. It can tell when the heart is not working well or when the heart is working too hard to pump blood through the lungs due to lung damage.

**Computed Tomography (CT) Scan:**
A CT scan uses a special X-ray to make a three-dimensional picture of the inside of the body. The scan is done in the radiology department. Contrast dye may be given in a vein and/or by mouth.

**Transcranial Doppler Ultrasound**
An ultrasound works by bouncing sound waves off parts of the body. An ultrasound does not hurt. A “T.C.D.” is used to evaluate a child’s risk of developing stroke.
Signs and Symptoms of Sickle Cell Anemia

Signs and symptoms of sickle cell disease vary from patient to patient and can range in severity. Common symptoms and complications may include:

- **Anemia** – The most common feature of sickle cell disease. Anemia can cause tiredness, headaches, dizziness, shortness of breath, pale skin or chest pain.

- **Pain** – Pain episodes occur most often in the arms, legs, chest, back and abdomen. Certain stressors like an infection, rapid change in temperature, stress or lack of sleep can trigger a pain crisis. Frequency of pain crises will vary with each child.

- **Infections** – One of the most serious problems in children with sickle cell anemia are infections in the bloodstream, lungs or bone. The risk is greatest during the first three years of life.

- **Fever** – one of the first symptoms of an infection. A fever of 101 degrees Fahrenheit (38.3 degrees Celsius) or higher requires immediate attention from a physician. Delayed treatment by even a few hours is dangerous!

- **Hand-foot syndrome** – Swollen hands or feet or dactylitis is a common symptom in infants up to two years of age with sickle cell disease.

- **Acute chest syndrome** – A severe complication of sickle cell disease when sickle cells in the chest cause a pneumonia with fevers, cough and difficulty breathing.

- **Splenic sequestration** - When the spleen becomes enlarged and possibly painful to touch. Call your child’s doctor right away if you think their spleen is swollen. The spleen is located on the left side of the abdomen.

- **Aplastic anemia** – A severe type of anemia when an infection temporarily stops the body from making new blood cells leading to very low levels of blood and extreme fatigue.
• **Delayed Growth and Puberty** – Children with sickle cell anemia tend to grow slower than others their age. They also are more likely to reach puberty later. Adults with sickle cell anemia are often smaller in size and more slender than other adults their age.

• **Priapism** – A condition in boys when sickle cells cause a painful and prolonged erection. Call your child’s hematologist for a painful erection lasting more than one hour.

• **Gallstones** – Sickle cell patients are at an increased risk for gallstones. Common symptoms include pain in the upper right side of the stomach, nausea and vomiting, clay-colored stools or yellowing of the skin or eyes. Occasionally, patients must have the gallbladder removed due to recurrent pain or infections of the gallbladder.

• **Eye Problems** – Sickle cells can cause damage to the eyes. Without enough blood and oxygen to the retina, damage can occur and lead to vision impairment and possible blindness. If older than 10-years-old, patients with sickle cell anemia should be seen annually by an ophthalmologist.

• **Tonsils** – Sickle cell patients commonly have enlarged tonsils, which can lead to obstructed sleep apnea. Tonsillectomy is sometimes needed.
Stroke and Sickle Cell Anemia

Stroke is the most severe complication of sickle cell disease leading to weakness, confusion, change in speech and unresponsiveness.

Signs that your child might be suffering a stroke:

- Unable to move their hand, arm or leg
- Limps without pain
- Experience a severe headache
- Dizziness
- Drooling, slurred words or a twisted, hanging mouth
- Unable to wake up

If you think your child is having or had a stroke, immediately call 911 so they can be taken to the emergency room.
Pain Management

The following are several things that your child can do at home to ease pain associated with sickle cell disease:

- Drink plenty of fluids
- Rest or play quietly
- Take warm bath
- Place heating pad or warm, moist towel on sore places
- Massage the places that hurt
- Take pain medications

There are different levels of pain medications:

For mild pain, children should take Ibuprofen. If the pain worsens or is not relieved by Ibuprofen, it may be necessary to have the physician prescribe a pain medication that contains a narcotic.

Sometimes pain is unresponsive to home therapy. During those times, the doctor should be called first, and the child should go to the clinic for treatment during the weekdays, or the emergency room on Wednesdays, evenings or weekends, for intravenous (IV) hydration and pain medication. IV morphine is usually used. Most of the time this is adequate treatment so that home therapy will then be effective.

In very severe painful crises, the child may need to be admitted to the hospital for IV therapy. Sometimes the child can be taught to give his or her own pain medication by pressing a “button” on the I.V. This is called patient controlled analgesia, or PCA.
Treatment Options

**Blood Transfusions:**
If your child needs a blood transfusion, the blood given will match your child’s blood type, and is given over several hours into a vein, through an IV in the arm. Your child will be watched closely for any signs of a reaction.

**Exchange Transfusion:**
Occasionally, patients with sickle cell anemia require an “exchange transfusion” or “apheresis procedure.” In an exchange transfusion, red blood cells are removed from the patient with a special machine and then replaced with a blood transfusion. Certain emergency situations will require an exchange transfusion (apheresis) such as a stroke.

*Complications of Blood Transfusions*

**Allo-immunization:**
Your child may develop antibodies that destroy the blood he has been transfused with. Most sickle cell patients receive “phenotypically matched” blood that will reduce the chance this will happen.

**Allergic reactions:**
These can cause rashes, hives, itching, or rarely, breathing problems. Rashes are common: fever or breathing problems are much less common.

**Iron Overload:**
It is possible with multiple blood transfusions to end up with too much iron in the body. This occurs because there is extra iron in the blood that is transfused. This is treated by removing the extra iron with a medication called ExJade. It is taken by mouth once daily.
Medications

These medications may be prescribed to your child by their hematologist:

**Penicillin:**
Because infections are among the greatest dangers to those with sickle cell disease, all children from birth until age six should be given daily penicillin. Some children will need penicillin for life. The usual dose for age two months until age two years is 125 mg twice a day as a liquid. At age three, the dose goes up to 250 mg twice a day as a liquid. This simple low-dose penicillin treatment has saved many patients’ lives. Liquid penicillin must be refrigerated and replaced every two weeks.

**Folic Acid:**
Due to the fact that children with Sickle Cell Anemia have to make more red blood cells than usual, occasionally they need to supplement their diet with folic acid. Folic acid is important in the production of new red blood cells.

**Hydroxyurea:**
Hydroxyurea decreases the amount of sickle hemoglobin in the red blood cells by causing a different type of hemoglobin to be made. This hemoglobin is called Fetal hemoglobin, which is the type that is found in babies. When there is less sickle hemoglobin, there is a decreased chance of sickling to occur and therefore less pain crises and complications.

**Jadenu:**
Jadenu is indicated for the treatment of chronic iron overload due to blood transfusions. This is generally started when the blood ferritin level is greater than 1000.
When to call the doctor or visit the emergency room

Call your child's doctor or nurse to be seen immediately if your child has one of these danger signs:

- Fever of 101 degrees or higher (never ignore this!)
- Severe headache or dizziness
- No movement in face, hands, arms or legs
- Slurred speech or drooling
- Rapid breathing or coughing with chest pain
- Severe pain or swelling in the belly
- Prolonged erection (greater than 1 hour)
- Very pale skin or palms
- Swollen hands or feet

If you cannot reach your child’s doctor, go to the emergency room immediately.

Call your child's doctor or nurse for advice if any of the following occur:

- Vomiting
- Diarrhea
- Jaundice (skin or eyes look yellow)
- Extreme fatigue
- Pain
- Limps without pain when walking
- Persistent cough
- Refuses to take penicillin
- No appetite
Prepare an emergency plan

• Write down directions from your house to your child’s doctor’s office and hospital emergency room. Keep a copy of these directions with you at all times.
  o Make sure anyone who regularly cares for your child has a copy of the directions.

• Create a list of phone numbers of cab and ambulance companies.
  o Keep a copy of this list by all house and cell phones

• Arrange for back-up emergency transportation
  o Find out if your community based sickle cell program offers transportation services
  o Ask your family, friends and neighbors if they can provide transportation to the doctor or hospital in case of an emergency

• Create an emergency contact list.
  o Maintain a list of phone numbers of people who are willing to help out in an emergency

• Request a “travel letter” from your child’s doctor or hematologist.
  o If your child gets sick while you are away, the letter will help the out-of-town doctors understand your child’s condition and current treatment.
Parent Care Tips

Hydration! Hydration! Hydration!
To prevent complications of sickle cell disease, children with sickle cell anemia should be encouraged to drink the recommended daily amounts of water throughout the day, whether at school, playing sports or spending time with friends or family.

Help your child eat healthy:
- Children with sickle cell disease have more particular dietary needs than other children.
- A healthy diet for a child with sickle cell is low in saturated fats, trans fats, cholesterol, salt and sugars. Good food choices include:
  - Fruits and vegetables
  - Whole grains (wheat)
  - Lean meats (chicken, turkey) plus other sources of protein (fish, beans, eggs and nuts)
  - Fat-free or low-fat milk and milk products

Vitamins are important:
- Children with sickle cell disease should take regular daily vitamins, especially if they do not regularly eat a well-rounded meal.
- It is common for children with sickle cell to be prescribed daily folic acid to help them produce new red blood cells.
- Many children with sickle cell disease have low levels of zinc, Vitamin D and Vitamin A. Check with the physician about whether your child will need to take supplements.

Avoid letting your child get chilled:
Children with sickle cell disease are more likely to have a pain episode when it is very hot (over 80 degrees) or very cold (less than 45 degrees) outside. Sudden changes in weather condition can cause the child to develop a pain episode. We recommend:
- Dress your child warmly when the weather is cool or when the child is going to be in an air-conditioned room for a long time.
- Your child should only swim in heated pools.
- Dry your child off quickly after a swim and be sure to wrap him or her in a dry towel to keep them warm.
- Make sure your child is always well hydrated, especially when the weather is warm.