Sickle cell disease: packed red blood cell transfusion

What is a blood transfusion?

A blood transfusion is when your child gets blood from another person. When a person donates blood, the blood is separated into different parts, including the red blood cell (RBC) part. When your child gets RBC transfusion, they get just the RBC part of the blood.

Transfusions are given:

- In a vein through an I.V.
- 1 or more times.

RBC transfusions are given to children with sickle cell disease for different reasons. Some of those reasons are:

- To treat urgent sickle cell problems in the hospital.
- To help prevent sickle cell problems by getting scheduled transfusions in the clinic.
- Before surgery or anesthesia to help prevent sickle cell-related problems.

When does my child need a blood transfusion?

Blood transfusions are often used to treat children with sickle cell disease. Normal RBCs carry oxygen to the entire body. Sickle RBCs do not carry oxygen very well. Sickle cells can also get stuck in the blood vessels and block the flow of blood.

A blood transfusion gives your child healthy, normal RBCs. This can help to get more oxygen to the body and to unblock the blood vessels. Your child may need a blood transfusion:

- To improve anemia if their hemoglobin is very low.
- To treat many problems in sickle cell disease, such as:
  - Stroke
  - Enlarged spleen (splenic sequestration) or enlarged liver (hepatic sequestration)
  - Long-lasting or frequent pain crises
  - Acute chest syndrome
- To help prevent future problems from sickle cell disease, such as:
  - Sickle cell-related problems with surgery or anesthesia, like acute chest syndrome
  - Stroke or other serious sickle cell problems

How often will my child need a blood transfusion?

The number of times your child will get a transfusion depends on the type of sickle cell disease they have. Most children with sickle cell disease get a blood transfusion at least 1 time in their life. A child

with hemoglobin SS or Sβ° may need a transfusion more often than a child with hemoglobin SC or $S\beta^+$ thalassemia.

Acute blood transfusions are:
- Given 1 to a few times. This depends on your child's needs.
- Needed to treat acute sickle cell problems like acute chest syndrome or aplastic crisis.
- Used before surgery to prevent sickle cell problems.

Chronic blood transfusions are:
- Given every month for several months to many years, or for their entire life.
- Given to increase the number of healthy RBCs in your child's blood and to lower the amount of sickle RBCs. Your sickle cell disease providers measure the amount of sickle hemoglobin (hemoglobin S) in your child's blood to know how well the chronic blood transfusions are working.
- Given to children who have had a severe problem, like a stroke, or those at high risk for having a severe problem.
  - If your child has had a stroke, they may need a transfusion each month for their entire life. This helps lower the risk of another stroke.
  - If your child has had an abnormal TCD ultrasound test, they will need chronic transfusions for at least 1 year. This helps to prevent a stroke . Your child may be able to stop transfusions in the future if they have another normal TCD ultrasound test.

Using a port-a-cath for chronic blood transfusions

If your child needs a transfusion on a regularly , sometimes their veins may get weak from so many blood draws and I.V. lines. This can make it hard to start an I.V.
- Your child may need a special I.V. line called a port-a-cath (port). This will provide easy access for your child to get blood and to have blood drawn for tests.
- The port may stay in for years.
- Please ask your child's sickle cell provider for more details.

What kind of blood transfusion will my child get?

There are 3 types of RBC blood transfusions a child with sickle cell disease may get:

1. **Simple blood transfusions**
   Your child gets a blood transfusion into their vein. They do not have any of their blood removed by the transfusion. The blood comes in a unit.
   - A unit is a pint-sized bag of blood that takes from 2 to 4 hours to infuse (go into the I.V.).
   - Your child may get 1, 2, 3 or more units of blood. Thisdepends on how big your child is and how low their hemoglobin level is.

- The purpose is to raise your child's hemoglobin level and to lower the amount of sickle hemoglobin.

2. **Exchange blood transfusions (erythrocytapheresis)**
   This removes sickle RBCs and replaces them with normal, healthy RBCs.
   - It is done with a special machine. Your child will need a special type of port or I.V. to remove their blood and to give them new blood at the same time.
   - It is a faster way to lower the sickle hemoglobin level.
   - It can help stop the buildup of iron in the body.

3. **Partial manual exchange transfusions**
   Your child will have some of their sickle RBCs removed through an I.V. before getting a regular blood transfusion.
   - Less sickle RBCs are removed than in an exchange blood transfusion.
   - It helps to lower the sickle hemoglobin level and slow the buildup of iron in the body (although not as much as an exchange blood transfusion).
   - It can be done without a special port.

Please ask your sickle cell provider for more details about the kind of transfusion your child needs.

How safe is a blood transfusion for my child?

- Blood transfusions are proven to help children with sickle cell disease.
- Special testing is done to make sure the blood type matches your child and that your child will not react against the blood.
- The U.S. Food and Drug Administration (FDA) regulates all U.S. blood banks. All blood centers must pass regular inspections.
- Each unit is carefully screened to prevent infections, such as HIV and hepatitis. This is done to make sure the blood is as safe as possible for your child.

What are the risks with blood transfusions?

Although blood transfusions can help to treat sickle cell disease, problems may happen.

Blood transfusion reactions

- Your child's immune system may react against the blood. This can cause swelling or itching, trouble breathing, fever or pain. These reactions could happen during the transfusion or at the end of the transfusion.
- Your child's body may make antibodies against the donor blood. These antibodies may show up a few days or a few weeks after apheresis. If a child forms antibodies, it may be harder to find matched blood to give them.

Iron overload

- This happens when there is too much iron in the body.
- Each unit of blood contains iron. When your child gets a blood transfusion, that iron stays in the body.
- Your child's body cannot get rid of extra iron, so the iron levels increase with each transfusion.
- Too much iron can be harmful to the body, especially to the liver.
- If your child gets many blood transfusions, their iron levels will be tested.
- If your child's iron levels are too high, they will need medicine to lower their iron level. This may be a medicine that they will need to swallow every day.

Infection

- The chance of getting an infection from a blood transfusion is very low all donor blood is tested for infection.
- The chance of getting HIV from a blood transfusion is less than 1 in 2 million.

Please talk with your child's sickle cell provider for more details about problems that may happen.

What should I do if my child is sick?

- Follow the sickle cell provider's advice for what you should do.
- Call your child's provider anytime your child has a fever of 101°F (38.3°C) or higher and is sick. Your child will need to be seen **right away**.
- During the time when the clinic is closed (between 5 p.m. and 8 a.m. during the week and on weekends and holidays), call the hematologist on call. They will contact the emergency department (ED) and help with your child's care.
- **ALWAYS** tell the ED doctors and staff that your child has sickle cell disease. Also tell them which sickle cell clinic your child visits.

For more details on sickle cell disease and services, visit the Children's Healthcare of Atlanta website at choa.org/sicklecell.

This teaching sheet contains general information only. Talk with your child's doctor or a member of your child's healthcare team about specific care of your child.

In case of an urgent concern or emergency, call 911 or go to the nearest emergency department right away.

