Your Young Child AND Sickle Cell Disease

St. Jude Children's Research Hospital®
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Your Young Child and Sickle Cell Disease

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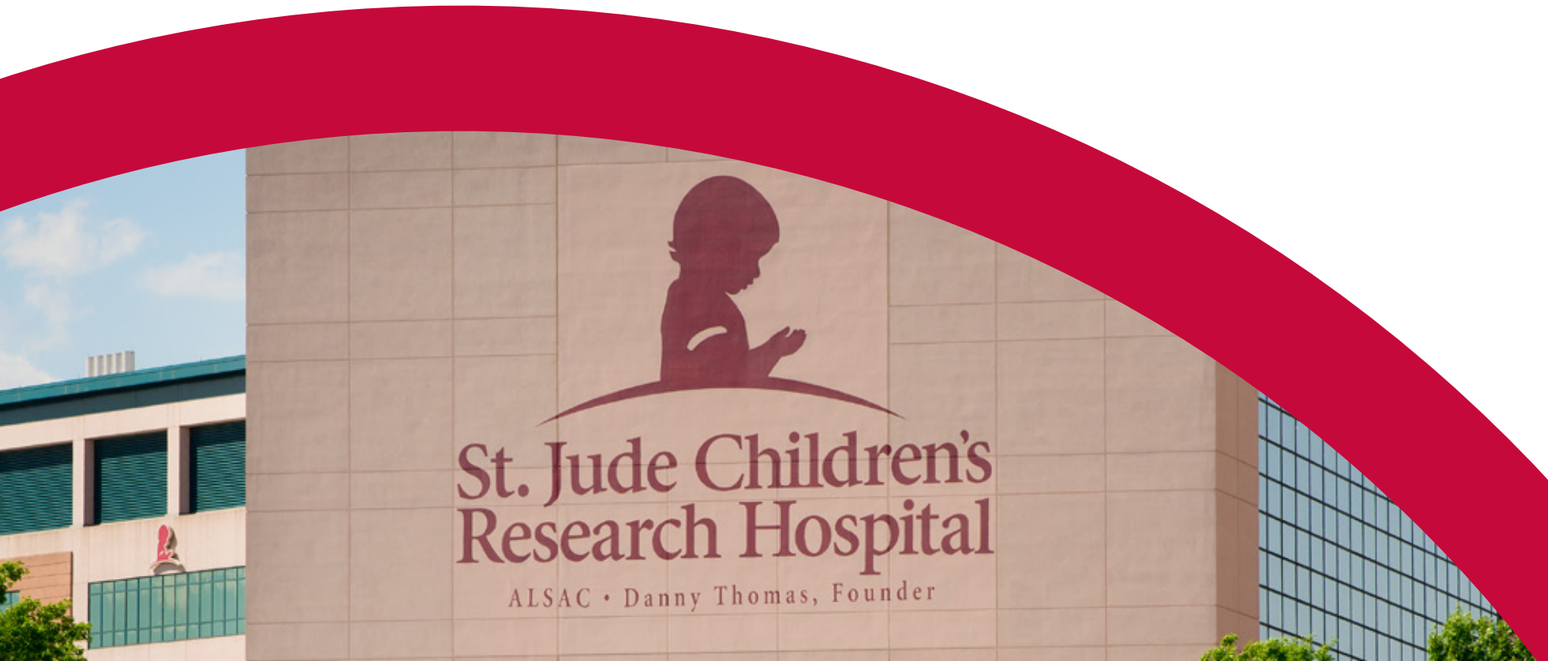
This document is not intended to take the place of the care and attention of your personal physician or other professional medical services. Our aim is to promote active participation in your care and treatment by providing information and education. Questions about individual health concerns or specific treatment options should be discussed with your physician.

Welcome to St. Jude Children's Research Hospital!

As the parent of a child with sickle cell disease, one of your greatest challenges will be learning to provide the special care your child needs. The St. Jude Hematology staff created this booklet to help you succeed in giving your child the best care possible.

Our goal is to offer complete information and easy to follow guidelines. If you have questions about anything in this booklet, please feel free to ask your child's doctor or nurse.

Your Young Child and Sickle Cell Disease offers many details that are crucial to your child's health. For this reason, it is best to read only one unit of the booklet at a time. Then, write down any questions you have in the "Notes" section on the last four pages. This booklet also has a glossary to help explain uncommon words. When you see a word with this symbol * next to it, the word is defined in the glossary.



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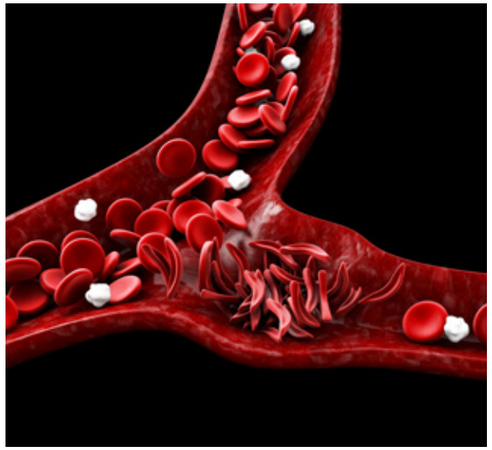
Unit 1: Sickle Cell Disease Facts

Who is affected by sickle cell disease?

About one out of every 375 African-American babies is born with sickle cell disease. Sickle cell disease affects about 70,000 people in the United States and more than a million people worldwide. Around the world, the disease affects people of all races and is heavily concentrated in Central and South America, southern Europe, central and southern India, and Africa.

A sickle cell and a normal red blood cell



What is sickle cell disease?

Sickle cell disease affects the red blood cells (RBC). The main purpose of red blood cells is to deliver oxygen to the body. Blood flows through the lungs and collects oxygen. Then the blood travels to the heart and is pumped to the rest of the body. Blood delivers oxygen to all of the tissues in the body. The part of the blood that carries oxygen is called hemoglobin.

Sickle cell disease is a blood disorder that affects the hemoglobin* (HEE-muh-glow-bin) within the red blood cells. The main role of hemoglobin is to deliver oxygen to the cells of the body.

Normal red blood cells have hemoglobin A. Normal red blood cells are shaped like discs or doughnuts. They are flexible and flow smoothly through small blood vessels.

Children with sickle cell disease have red blood cells that contain mostly hemoglobin S. This type of hemoglobin causes red blood cells to become sickle or banana shaped. Unlike normal red blood cells, sickle cells are not flexible. They are sticky, stiff, and rigid. Their shape and texture make it hard for them to flow through small blood vessels.

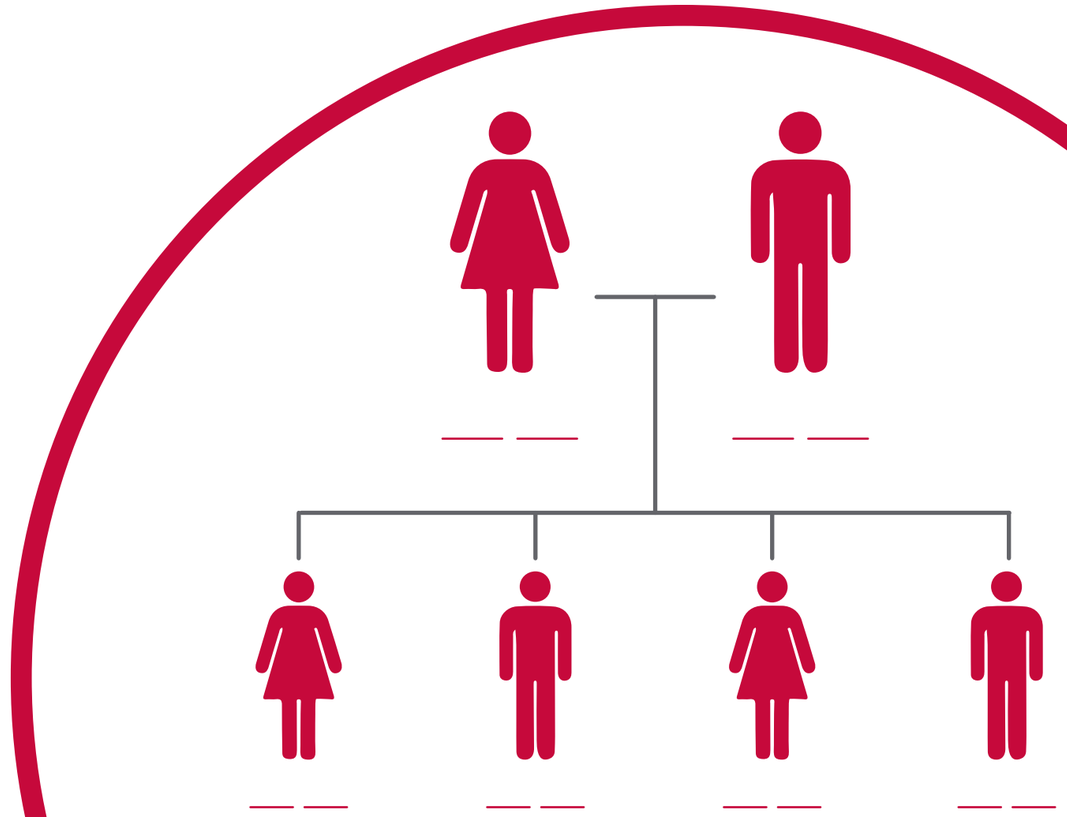
Sickle cells cause red blood cells to stick together and slow the flow of oxygen to the tissues. This causes pain and over time causes damage to the vessels.

Sickle-shaped red blood cells ❱ slower blood flow ❱ less oxygen ❱ damaged cells

How did my child get sickle cell disease?

Children with sickle cell disease are born with it, and they will have it all of their lives. They will not outgrow sickle cell disease. Sickle cell disease cannot spread like a cold. No one can catch sickle cell disease from another person.

Sickle cell disease is inherited (in-HAIR-uh-ted), like hair color or eye color. Each child has 2 hemoglobin genes – one from the mother and one from the father. To have sickle cell disease, a child must inherit a hemoglobin S gene from one parent and another abnormal hemoglobin gene from the other parent.



A staff member will help you complete this graph so you can see how sickle cell disease might occur in your family.

Are there different types of sickle cell disease?

Yes.

There are 3 common types of sickle cell disease.

1

▶Sickle Cell Anemia* (Hemoglobin SS Disease)

This is the most common type of sickle cell disease. Children with sickle cell anemia (uh-NEE-mee-uh) have almost 100 percent hemoglobin S in their red blood cells. They inherit a hemoglobin S gene from each parent.

2

▶Sickle-Hemoglobin C Disease (Hemoglobin SC Disease)

Children with sickle-hemoglobin C disease inherit a hemoglobin S gene from one parent and a hemoglobin C gene from the other parent.

Hemoglobin C is a type of abnormal hemoglobin.

3

▶Sickle Beta-Thalassemia Disease

Children with sickle beta-thalassemia (BAYT-uh-THAL-uh-see-mee-uh) inherit a hemoglobin S gene from one parent and a beta gene from the other parent.

Beta is another type of abnormal hemoglobin. Children with sickle beta have mostly hemoglobin S in their red blood cells.

There are many other types of abnormal hemoglobin and many types of sickle cell disease, but these three are the most common.

Why do children with sickle cell disease have anemia?

Anemia* (uh-NEE-mee-uh) occurs when the blood has less than the normal number of red blood cells.

A normal red blood cell lives about 3 to 4 months. A sickle cell has a lifespan of 10–20 days. Sickle cells are destroyed early because of their odd shape. The body constantly makes new red blood cells. But, it cannot make red blood cells as fast as they are destroyed, so a person with sickle cell disease is usually anemic.

Infants are not anemic because they are born with hemoglobin F or fetal red blood cells. Hemoglobin F acts like normal hemoglobin. Infants are born with mostly hemoglobin F, but soon after birth, the body stops making hemoglobin F. By one year old, an infant usually has less than 9 percent hemoglobin F.

When a person without sickle cell disease has anemia, the doctor might prescribe iron supplements to help boost the person's red blood cell count. However, this does not work for children with sickle cell disease. You cannot correct or cure sickle cell disease by giving your child iron supplements.

Unit 2: Complications of Sickle Cell Disease

Because sickle-shaped red blood cells can slow blood flow to many parts of the body, a number of complications* (health problems) can occur. You need to be aware of these problems and how to treat them. Each child is different. Not every child with sickle cell disease will have all of these health problems. However, knowing about these complications in advance can help prevent them and may even save your child's life.

Blood Infections

One of the most serious problems facing young children with sickle cell disease is a blood stream infection* (in-FEK-shun). The infection risk is greatest during your child's first 3 years of life. Blood infections can be deadly, and they require treatment right away.

What is the main symptom of a blood stream infection?

Fever is one of the first symptoms and sometimes the **only** symptom of a blood stream infection*.

If your child has a fever of 100.4 degrees F (38 degrees C) or higher, seek medical treatment right away. If you delay treatment for even a few hours, you will put your child in danger.

Do not forget to tell the health care team that your child has sickle cell disease. Fever **must not** be ignored in a child with sickle cell disease.

Oral and rectal temperature readings are the most correct because they measure internal body temperature. The rectal method is the best for infants, but it is hard to perform on a toddler. In this case, the underarm method can be used.

One of the easiest ways to take a young child's temperature is by placing a digital thermometer under the arm. This method works well, but you must add a degree to obtain an accurate reading.

Follow these steps:

- **1.** Remove your child's shirt from one arm, so skin surfaces will touch the thermometer.
- **2.** Place the tip of thermometer high into the center of the armpit.
- **3.** Hold your child's arm tightly against his side to keep the thermometer in place.
- **4.** Remove the thermometer when it beeps.
- **5.** Read the temperature and add a degree (101 degrees + 1 degree = 102 degrees).

Why do children with sickle cell disease have an increased risk of serious infection?

In children with sickle cell disease, the spleen* does not work properly. The spleen is an organ in the upper left part of the abdomen. It helps protect against infection by filtering bacteria from the bloodstream and by producing antibodies* (AN-tih-bah-deez). In a person with sickle cell disease, the spleen is damaged early in life. In turn, this causes the child to have fewer antibodies and less protection against infection.

What can be done to help prevent a serious infection?

A child with sickle cell disease is 400 times more likely to get a bacterial infection, because the spleen* does not work properly. Penicillin (peh-nuh-SIH-lun) kills bacteria. Before penicillin was given to infants with sickle cell disease, bacterial infection was the greatest cause of death in this age group. Penicillin reduces the risk of bacterial infection by more than 85 percent. Infants should begin taking penicillin by 3 months of age. Penicillin taken 2 times a day every day can greatly reduce the risk of infection in children with sickle cell disease.

Facts about Penicillin:

Penicillin is available in liquid or tablet form.

Liquid penicillin is often easier to give to young children. However, liquid penicillin must be stored in a refrigerator and must not be kept for longer than 2 weeks. Also, you must obtain a fresh bottle every 2 weeks.

Always measure liquid penicillin with the dosing syringe your nurse or pharmacist provides.

Do not use a kitchen teaspoon because it is not accurate.

Store penicillin tablets in a cool, dry place. If stored correctly, penicillin tablets last for 4 months.

Your child's doctor will decide when and if your child should stop taking penicillin. At present, children with sickle cell disease stop taking penicillin at age 5. In some cases, children with sickle cell disease will keep taking penicillin after that age.

Penicillin only protects against bacterial infection. Your child can still catch a viral infection like the flu or a cold.

Follow these guidelines to help keep your child as healthy as you can:

- Wash your hands before feeding your baby and after diaper changes. When children are older, teach them to wash their hands before eating and after using the bathroom.
- Do not take your child around people with illnesses that spread easily (cold, flu, etc).
- Dress your child correctly for the weather. Children lose most of their heat through their heads. So, make sure your child has a warm hat and gloves in the winter.
- Get all childhood immunizations (vaccines) on time.
- Every member of the family should have the flu vaccine every year.
- Seek medical treatment right away if your child has a temperature of 100.4 degrees F (38 degrees C) or higher.

Rapid Enlargement of the Spleen*

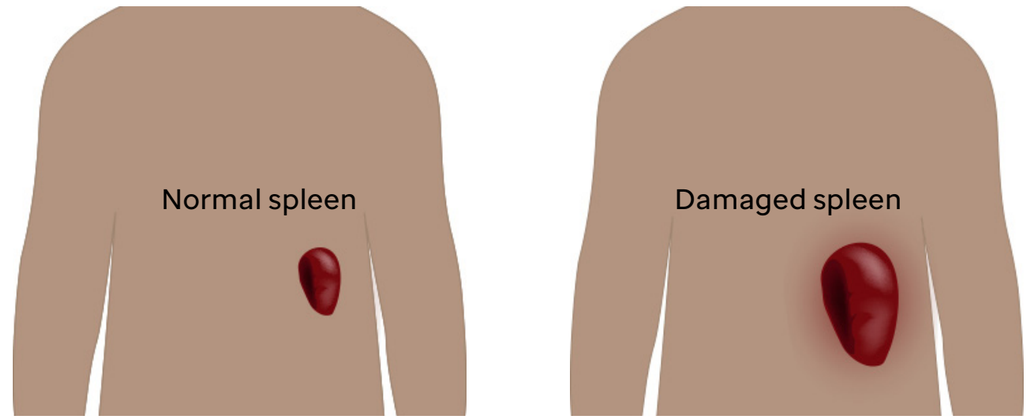
What is a splenic sequestration crisis?

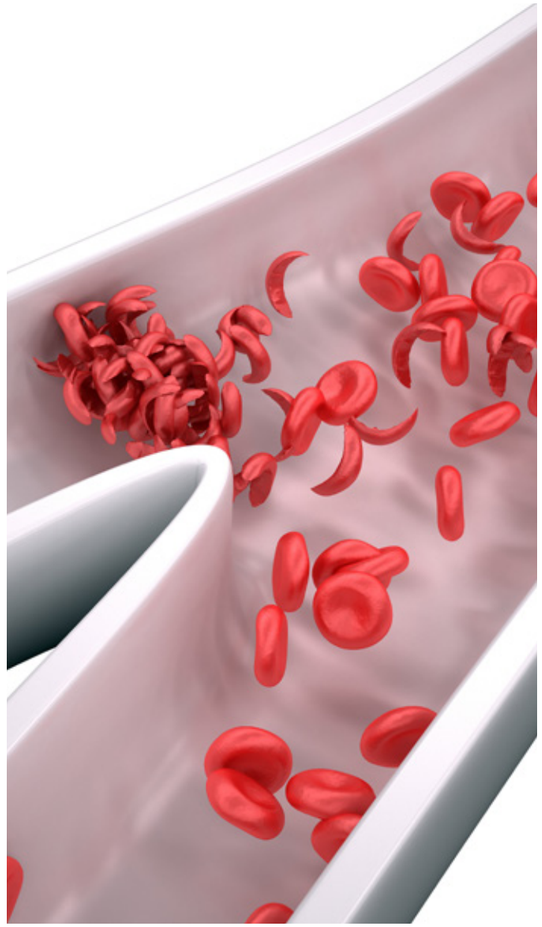
This complication (health problem) occurs when blood becomes trapped in the spleen. These are the symptoms of a splenic sequestration (SEE-kwes-tray-shun) crisis:

- Feeling weak
- Feeling tired
- Skin appears pale or washed out
- Enlarged or tender spleen

A splenic crisis is not common, but it **is** serious and requires medical treatment right away. Take your child to the emergency room if you notice these symptoms. Children who have had one (1) splenic sequestration crisis are at greater risk for having another one.

Damaged spleen ❱ fewer antibodies ❱ higher risk of infection





Pain

What causes sickle cell pain?

When sickle cells slow the flow of blood to an area of the body, that area does not receive enough oxygen. This lack of oxygen causes pain. Pain episodes occur most often in the arms, legs, chest, back, and abdomen.

Young children can have pain and swelling of their hands and feet. This is called dactylitis* (DAK-tih-ly-tus) or “Hand-Foot Syndrome.” Dactylitis can last for a few hours or several days. Tell the doctor if your child has an episode of dactylitis at home.

How often will a pain episode occur?

This varies greatly with each child. Some children have pain 1 or 2 times a year, others even less.

A few children have pain episodes quite often (3 or more times a year).

Certain stresses to the body can trigger a pain episode. Infection, extreme heat or cold, a poor diet, not enough liquids, and lack of sleep all cause stress to the body.

Sickle-shaped cells ❱ slow blood flow ❱ less oxygen ❱ pain

Can we do anything to prevent a pain episode?

Yes.

These guidelines can help

- Encourage your child to drink a lot of liquid (water and juices are best).
  - Infants should only get formula or breast milk until 4 months old. After 4 months, you can start giving your baby a small amount of juice or water.
  - After your child is 12 months old, you can start increasing the amount of fluid until they can tolerate 8 cups of liquid a day
- Do not let your child’s body become too hot or too cold.
- Provide a well-balanced diet.
- Make sure your child gets plenty of sleep.
- Avoid people who have infections.

Even if you do all these things, your child may still have a pain episode. However, pain is much less likely if you follow these guidelines.

What is the best treatment for a pain episode?

Often, you can manage mild to moderate pain episodes at home. For older children, rating pain on a scale from 1 (mild) to 10 (severe) will help you choose the best treatment. The FACES pain scale (at right) might help your younger child describe the pain.

Home Treatment Guidelines:

- Increase liquids.
- Give pain medicine as directed.
  - Mild pain is best treated with ibuprofen (Motrin® or Advil®).
  - Often, Tylenol® with codeine can be used for moderate pain.
  - Severe pain requires treatment by a doctor.
- Apply heat to the area that is hurting. Sometimes, a heating pad can help if it is not too hot. Do not put a heating pad on a child that is too young to talk.
- Provide rest! Have your child rest in bed or on a couch until the pain improves.

Prompt treatment of pain is crucial, because it may prevent the pain from becoming more severe.

When does a pain episode require treatment by a doctor?

See the doctor right away if your child has any of these symptoms:

- Trouble breathing
- Severe pain in abdomen
- Severe pain that does not respond to pain medicine

If your child needs to go into the hospital, treatment often includes pain medicine and fluids given through an IV*.

Is there a medication that can help prevent pain?

Yes, Hydroxyurea is a medication which helps reduce the production of sickle hemoglobin and has been shown to reduce pain crises in children and adults.

The FDA approved use of hydroxyurea in children in 2017. Your doctor may talk to you about starting your child on hydroxyurea to help prevent pain and some other complications of sickle cell disease.

Key Points to Remember

Your child will not have sickle cell disease complications all the time. In fact, most of the time your child will be free of symptoms. Kids with sickle cell disease can play and attend school like other children. They can grow up, go to college, and have a happy life and a successful career.

St. Jude is committed to improving the care of our sickle cell patients. We perform many types of research that will someday benefit all patients with sickle cell disease.

Because of research, the future for children with sickle cell disease looks promising. Medical advances during your child's lifetime will lead to longer, healthier lives for people with sickle cell disease.

Just keep in mind: It is important for you to be involved in your child's care. As a parent, you can do many things to make sure your child receives the best medical care:

- Learn about sickle cell disease
- Ask your child's doctors and nurses questions
- Keep all medical appointments
- Give medicines as directed
- Never delay treatment when problems occur
- Discuss any concerns with your child's doctor or nurse (even if you think they are not related to the disease, such as school issues).

Glossary

- **Anemia** (uh-NEE-mee-uh)**:** Occurs when the blood does not have enough red blood cells. The hemoglobin and hematocrit are laboratory tests used to find out if a person is anemic.
- **Antibodies** (AN-tih-bah-deez)**:** Proteins that fight bacteria and other foreign toxins in the body.
- **Bacteria** (bak-TEER-ee-uh)**:** Germs that are made up of one cell. Certain types of bacteria can cause illness when they get inside the body.
- **Blood stream infection** (in-FEK-shun)**:** When bacteria* get in the blood stream and start spreading throughout the body, making a person ill.
- **Complications** (KOM-plih-kaa-shunz)**:** In people with sickle cell disease, these are health problems caused by the disease.
- **Dactylitis** (DAK-tih-ly-tus)**:** Pain and swelling of hands and feet that is also called Hand-Foot Syndrome.
- **Gene:** A "blueprint" that is passed from parent to child. It carries the instructions for a certain trait, such as hair color, eye color, or skin color.
- **Hemoglobin** (HEE-muh-glow-bin)**:** The main substance of the red blood cell. It carries oxygen from the lungs to all parts of the body. Normal red blood cells contain hemoglobin A. Hemoglobin S and hemoglobin C are abnormal types of hemoglobin.
- **IV:** A needle placed in a vein to deliver fluids and medicines directly into the bloodstream.
- **Spleen:** An organ on the left side of the abdomen (belly). It helps protect against infection by filtering bacteria from the bloodstream. It also produces antibodies*.

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