

Sickle Cell Disease: Retinopathy

Patient and Family Education

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.

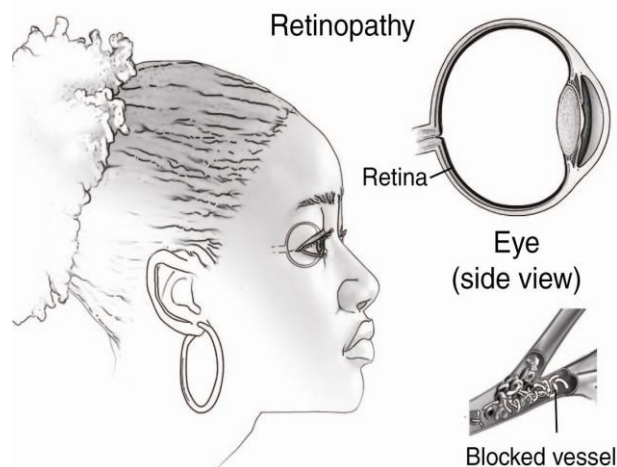
What is sickle cell retinopathy?

Retinopathy is damage to the retina in the eyes. The retina is the “seeing” part of the eye. It is in the back of the eye and it:

- Contains nerves and small blood vessels
- Captures light and images so that we can see

Eye problems caused by the sickled blood cells can be found in all types of sickle cell disease. Eye problems are more common in older children and adults with HbSC and HbSbeta+Thalassemia.

- This is thought to be because patients with these types of sickle cell disease often have a higher hemoglobin, which makes the blood thicker.
- As this “thicker” blood travels through the tiny blood vessels in the eyes, it causes damage to these vessels and can lead to vision loss if not treated early.



What causes sickle cell retinopathy?

Sickle red blood cells can get trapped inside the small blood vessels in the retina. This can cause:

- A decrease in bloodflow, leading to damage of the retina. The damage may be permanent.
- Bleeding in the eye.
 - When blood vessels are blocked, the eye makes new vessels to replace the blocked vessels.
 - The new vessels are thinner and weaker. They may break open and bleed.
 - The bleeding causes damage to the retina. Bleeding can also cause the retina to become loose from the rest of the eye. This is called a detached retina.

Damage to the retina can cause changes in eyesight. If the damage is not treated, blindness may occur.

What are the symptoms of sickle cell retinopathy?

At first, there may be no symptoms. **This is why an eye doctor needs to check your child's eyes each year.**

When retinopathy becomes worse, your child may have these problems:

- Flashes and dark shadows in some parts of his vision (floaters)
- Blurred vision
- Sudden loss of vision
- Pain in the eyes

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

Sickle Cell Anemia: Retinopathy, continued

What should I do if my child has a change in his vision?

If your child has changes in his vision, call your child's doctor **right away**.

- If you already have an eye doctor (ophthalmologist), call him **right away**.
- If you cannot see an eye doctor right away, **go to the Emergency Department**.
- Changes in vision can also occur with a stroke. Either way, your child needs to be checked right away.

What tests could my child need?

Your child needs a complete eye exam by an eye doctor who knows about sickle cell disease.

- The eye doctor will put drops in your child's eyes. This allows the doctor to see the entire eye better.
- Always tell your eye doctor that your child has sickle cell disease. Your child will need a more careful exam.

How is sickle cell retinopathy treated?

Your child will need treatment if his retina shows signs of damage.

- If caught early, most problems can be treated by the eye doctor.
- Your child may need eye exams more often.
- Some problems can be treated with a laser.
- Others may need to be treated with surgery.

What type of follow-up care does my child need?

- Make sure you understand and follow the instructions the eye doctor gives you. Always ask questions if you are not clear on any instructions given by a healthcare provider.
- Your child needs regular follow-up care with your eye doctor and your sickle cell provider.

How can I help prevent retinopathy?

Early stages of sickle cell retinopathy do not normally cause changes in eyesight. You can help prevent eye problems by doing these things:

- When your child is 7 or 8 years old, schedule him for an eye exam with an eye doctor **each year**. Treatments work better if the eye changes are found early.
- Your health insurance may require a referral from your child's pediatrician before being seen by the eye doctor. Be sure to discuss this with your health insurance company, including Medicaid.
- Follow any treatments the eye doctor gives.
- Give your child plenty of fluids to drink each day. Do this from the time he is 6 months old. Fluids help keep the sickled cells from sticking together and causing problems.
- Ask your child often if he has any changes in his vision. If he does, schedule an eye exam **right away** even if he is younger than 7 or 8 years of age.
- A simple vision screen using the eye chart is not the same as a full eye exam by an eye doctor. Ask your sickle cell provider if you are unsure of the type of eye exam that is needed.

What should I do if my child is sick?

- Follow your child's doctor's advice for what you should do.
- Call your child's health care provider anytime that your child has a fever of 101°F (38.3°C) or higher or is sick. Your child will need to be seen **right away**.

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

Sickle Cell Anemia: Retinopathy, continued

- During the time when the clinic is closed (on weekends and holidays and between 5PM and 8AM during the week), call the hematologist on call. They will notify the Emergency Room and help with your child's care.
- **ALWAYS** tell the Emergency Room doctors and staff that your child has sickle cell disease. Also tell them which sickle cell clinic he visits.

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

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