

Sickle cell disease: retinopathy

What is sickle cell retinopathy?

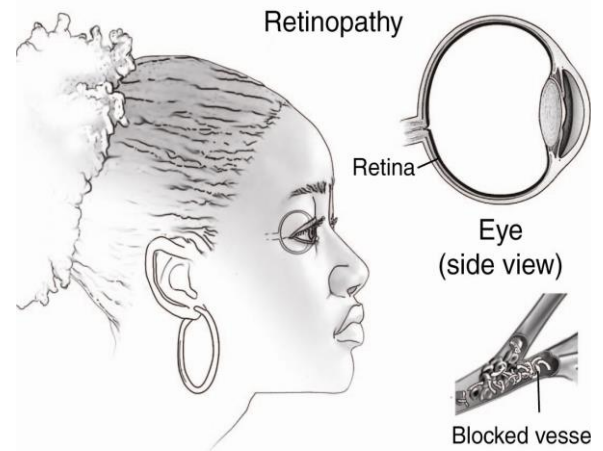
Retinopathy is damage to the retina in the eyes.

The retina is the “seeing” part of the eye. It is in the back of the eye and:

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

Eye problems are more common in older children and adults with HbSC and HbSbeta+Thalassemia.

- This may be due to a higher hemoglobin level in patients with these types of sickle cell disease. This makes the blood thicker.
- As this “thicker” blood travels through the small blood vessels in the eyes, it causes damage to these vessels and can lead to vision loss if not treated early.



What causes it?

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

- A decrease in blood flow. This can lead to damage of the retina. The damage may be permanent.
- Bleeding in the eye.
 - When blood vessels get 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 loosen 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 happen.

What are the symptoms ?

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

When retinopathy worsens, your child may have these problems:

- Flashes and dark shadows in some parts of their 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 disease: retinopathy, continued

What should I do if my child has vision changes?

Call your child's doctor **right away** if your child has vision changes.

- If your child already has an eye doctor (ophthalmologist), call them **right away**.
- **If your child cannot see an eye doctor right away, take your child to the nearest emergency department (ED).**

Vision changes can also happen 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 helps the doctor see the entire eye better.
- **ALWAYS** tell the eye doctor that your child has sickle cell disease. Your child needs a more careful exam.

What is the treatment?

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

- If caught early, your child's eye doctor can treat most problems.
- 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 follow-up care does my child need?

- Make sure you understand and follow the instructions the eye doctor gives you. Always ask questions if the instructions are unclear.
- Your child needs regular follow-up care with the eye doctor and your sickle cell provider.

How can I help prevent retinopathy?

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

- When your child is 9 or 10 years old, start scheduling them for an eye exam with an eye doctor **every year**. Treatments work better if eye changes are found early.
- Your health insurance may need a referral from your pediatrician before your child can see an eye doctor. Talk about this with your health insurance company (including Medicaid) ahead of time.
- Follow treatments as advised by the eye doctor.
- Give your child plenty of fluids to drink each day. Do this from the time they are 6 months old. Fluids help keep the sickle cells from sticking together and causing problems.

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

Sickle cell disease: retinopathy, continued

- Ask your child often if they have any vision changes. If they do, schedule an eye exam **right away** even if they are younger than 9 or 10 years old.
- 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 your child needs.

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 or 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.