# Sickle cell retinopathy

Sickle cell retinopathy is an eye complication of sickle cell disease. It is caused by loss of blood supply to certain parts of the retina. This is because the red blood cells in sickle cell disease are sticky and less flexible. They can block the tiny blood vessels that supply nutrients and oxygen to keep the retina healthy.

## What is sickle cell disease?

Sickle cell disease is the name for a group of inherited conditions that affect the red blood cells. They are a serious and lifelong health condition, although treatment can help manage many of the symptoms.

These conditions cause the red blood cells to form a ‘C’- shape rather than ‘O’ shape. The ‘C’ shape (which is where the name ‘sickle’ comes from) makes the red blood cells less flexible and sticky. This makes it difficult for them to pass through small blood vessels. This causes episodes of severe pain, increased risk of severe infection and anaemia, amongst other problems.

Red blood cells contain the protein that carries oxygen around your body known as haemoglobin. In sickle cell disease, the haemoglobin is irregular and so it cannot carry as much oxygen.

Sickle cell disease most commonly affects people of African or Caribbean descent. It is also seen commonly in people of Asian, Middle Eastern and Eastern Mediterranean descent. Many of the symptoms can be managed with treatment which includes keeping well hydrated, reducing infection risks and medical treatments.

## What causes sickle cell disease?

Sickle cell disease is a genetic condition caused by a fault (mutation) in a gene. You inherit genes from your parents. Your genes give the cells in your body the instructions they need to work well and stay healthy. If a gene has a mutation, there is a fault in their instructions and the cells using those instructions don’t work as they should. In sickle cell disease, there is a fault in the gene that controls how haemoglobin is formed. Haemoglobin is the protein responsible for carrying oxygen in red blood cells.

Sickle cell disease is inherited in an autosomal recessive pattern. This means that to have sickle cell disease, you need to inherit a copy of the faulty haemoglobin gene from both parents. If both parents have a faulty gene, then there is a 1 in 4 chance of inheriting the condition. If you inherit one faulty gene then you will be a ‘carrier’ of sickle cell disease, but not have the symptoms. A carrier is referred to as having the sickle cell trait.

## Types of sickle cell disease

There are several types of sickle cell disease. The different types depend on the genes you inherit from your parents.

Red blood cells with normal haemoglobin-A are smooth and round and glide through blood vessels. The two most common gene mutations causing sickle cell disease are:

* haemoglobin-S (HbS)
* haemoglobin-C (HbC)

The following are types of sickle cell disease and depend on which of the gene mutations have been inherited:

### HbSS

HbSS is the most common and most severe type of sickle cell disease. It’s caused when you inherit haemoglobin-S (HbS) from both parents. Red blood cells with haemoglobin-S stick to one another and form long, rod-like structures. These structures cause red blood cells to become stiff, assuming a sickle shape. These sickle cells are destroyed rapidly in the bodies of people with the disease, causing anaemia. Anaemia is a term to describe lack of healthy red blood cells. This anaemia is what gives the disease its commonly known name – sickle cell anaemia.

### HbSC

HbSC is the type of sickle cell disease caused by inheriting the haemoglobin-S (HbS) gene from one parent and the haemoglobin-C (HbC) gene from the other parent. People with HbSC often have milder symptoms of the disease generally, but this type is more likely to affect the eyes.

### HbS beta thalassemia

HbS beta thalassemia is caused by inheriting the haemoglobin-S (HbS) gene from one parent and the ‘beta thalassemia’ gene from the other parent. The haemoglobin-S gene means that the red blood cells take on a sickle shape, making them unable to flow through the blood vessels smoothly. The beta thalassemia gene reduces the amount of normal haemoglobin a person has in their blood.

### Other types

There are other types of faulty haemoglobin genes that can cause a type of sickle cell disease, but they are far less common and often less severe.

More information about sickle cell disease can be found on the NHS website and on the Sickle Cell Society website.

## How the eye works

When you look at something, light passes through the front of your eye and is focused by the cornea and lens onto your retina at the back of your eye.

The retina is a delicate nerve tissue that is sensitive to light. It converts the light into electrical signals that travel along the optic nerve to your brain. The brain interprets these signals to “see” the world around you. The retina is supplied with blood by a delicate network of blood vessels.

Light entering your eye is focused onto a tiny area of your central retina called the macula. The macula is vital in allowing you recognise colours and see the fine detail needed to carry out activities such as reading and writing. The rest of your retina, called the peripheral retina, gives you peripheral vision (also known as side vision).

The following diagram shows the cross‑section of an eye. From the front to the back of the eye, it is labelled cornea, pupil, iris, lens, vitreous gel, optic nerve, macula, fovea and retina.



**Fovea**

**Macula**

**Optic**

**nerve**

**Vitreous gel**

**Ciliary body**

**Sclera**

**Choroid**

**Pupil**

**Lens**

**Retina**

**Cornea**

**Iris**

## What is sickle cell retinopathy?

Sickle cell retinopathy is an eye complication of sickle cell disease that causes damage to the retina. It is caused by loss of blood supply to certain parts of the retina (known as ischaemic retina). This is because the red blood cells in sickle cell disease are sticky and less flexible. They can block the tiny blood vessels that supply nutrients and oxygen to keep the retina healthy.

Sickle cell retinopathy can affect the retina in different ways. The type that is most likely to affect sight is known as proliferative sickle cell retinopathy (PSR).

If there is a reduced blood supply to the retina, the ischaemic retina produces an increase in a protein called vascular endothelial growth factor (VEGF). This protein stimulates growth of new blood vessels in the outer edges of your retina (peripheral retina). The new blood vessels are weak and leaky and can cause further damage to your retina and vision. When new blood vessels grow, it is known as proliferative sickle cell retinopathy (PSR). The new blood vessels form fan-shaped networks (known as sea-fan) along the surface of the retina and back of the vitreous gel inside the eye. Movement between the two surfaces can cause the new blood vessels to leak, resulting in a bleed into the vitreous gel, known as vitreous haemorrhage.

The development of these networks can vary from very small sea-fans that resolve on their own, to large fast developing sea-fans that join up with other sea-fans and expand.

In about four out of 10 people with PSR, the new blood vessels block up and the sea-fan resolves on its own. In other cases, the sea-fans do not grow much bigger and do not bleed either.

Many people with PSR have no symptoms in the early stages. However, some people with PSR can have temporary or irreversible sight loss. This affects roughly 10-12 out of every 100 people with sickle cell disease over their lifetime.

PSR is more common in people with HbSC type of sickle cell disease than those with HbSS. Some large studies have estimated it affects almost five in 10 people with HbSC and about two in 10 people with HbSS.

PSR usually starts between the ages of 15 and 24 in males and 20 and 39 in females. However, it can start as early as 8 years old in those with HbSC and 13 years old in HbSS.

The following factors can increase the risk of PSR causing sight loss:

* Age – increasing age can increase the risk of sight loss due to PSR.
* Male gender – males are more likely to experience sight loss due to PSR than females.
* HbSC – people with HbSC type of sickle cell disease are more likely to experience sight loss from PSR than people who have other types.

People with PSR can have sight loss from the following complications:

### Vitreous haemorrhage

A vitreous haemorrhage is where blood from new blood vessels leak into the vitreous gel. The vitreous gel is normally clear to let light pass through to the retina. If there is blood in the vitreous gel, this can block the light and cause vision to become cloudy.

### Retinal detachment

A retinal detachment is where the retina separates from the inside of the eye. This can occur due to the new blood vessels pulling on the retina (tractional retinal detachment) and/or due to a small tear occurring in the retina. A retinal detachment can cause sight loss in the affected area and needs to be treated very quickly. More information about retinal detachment can be found on our website or by calling our Helpline.

Some of the other ways that sickle cell disease can affect the retina and cause reduced vision include:

### Sickle cell maculopathy

Sickle cell maculopathy happens when there is a loss of blood supply to the macular area (central area of the retina). It can cause the macula area to become thin which affects how well it works to give you detailed vision. Rarely, the very centre of the macula, known as the fovea may be affected and cause reduction in vision.

### Retinal artery occlusion

A retinal artery occlusion is where the blood vessels delivering blood and oxygen to the retina becomes blocked, causing a sudden loss of sight. The amount of sight lost depends on where the blockage has occurred. More information about retinal artery occlusion can be found on our website or by calling our Helpline.

### Epiretinal membrane (ERM)

ERM is a condition where a sheet of naturally occurring cells develops on or above the surface of the macula. ERM can affect vision if this sheet of cells starts to shrink, causing the retina to wrinkle up under it. This wrinkling of the retina can then cause distortion and blurring of your vision, as well as a possible reduction in your level of sight. More information about ERM can be found on our website or by calling our Helpline.

### Macular hole

A macular hole describes a small gap which develops in the centre of the macula. A macular hole affects your central vision. It can make your vision distorted and blurred and affect how well you can see detail. You may also have a blank patch in the centre of your vision. More information about macular hole can be found on our website or by calling our Helpline.

## How is PSR diagnosed?

### Early detection during regular checks by your ophthalmologist

Early detection of PSR at the back of the eye can prevent sight loss. It is important for anyone with sickle cell disease to have regular eye examinations.

The current NICE guidelines recommend that everyone with sickle cell disease should be seen every two to three years by an ophthalmologist (eye doctor) if they have no sickle cell retinopathy and every year if there is sickle cell retinopathy present.

In an eye examination the ophthalmologist will be able to examine the health of the retina. They can detect the early stages of sickle cell retinopathy before and if PSR (new blood vessel growth) develops as well as any other associated complications.

They will use a technique called ophthalmoscopy to view the retina directly and photograph the retina. Ultra-widefield retina cameras have been shown to pick up most changes in the far edges of the retina (peripheral retina) that occur with sickle cell disease.

It is also possible to have a 3D scan of the inside of the eye called Optical Coherence Tomography (OCT). This test can check the health of the macula, as well as detecting any thinning of the macula (known as sickle cell maculopathy). Sickle cell maculopathy does not often affect the vision, but research is ongoing to determine whether it might affect other aspects of how the eye functions. Research is also looking at whether it can help haematologists (doctors specialising in conditions relating to the blood) know whether treatments being prescribed for sickle cell disease are working.

To get the best view of the retina, eye drops are used to make the pupils larger. They make the vision blurry for a few hours and it is important not to drive home from the appointment.

The ophthalmologist may recommend imaging of the retina using a special dye. This is called fluorescein angiography. This test allows the ophthalmologist to see sea-fans clearly and determine how leaky they are.

Optical coherence tomography angiography (OCTA) is a specialised form of OCT. This is used to monitor for blocked and abnormal blood vessels in the early stages of the condition at the macula. Some hospitals now have ultra-widefield OCTA which may one day allow the blood supply of the peripheral retina to be assessed without the need for fluorescein angiography.

### Being aware of the symptoms

Sometimes, PSR is diagnosed because you suddenly notice signs that your eyesight has changed. These signs may include:

* Floaters: this is where you can see small dark dots, squiggly lines or cobwebs, or a ‘dark shadow’ or a curtain moving across your vision.
* Blurred vision
* Flashing lights: sudden flashes of light

It is important that you ask for an urgent appointment with your optometrist (optician) or your eye clinic (if you are already looked after by an ophthalmologist), if you notice any of these.

## How is PSR treated?

Sea-fans resolve on their own in around four out of 10 people. Therefore, your ophthalmologist may want to monitor them for some time before considering any treatment. There is ongoing research to understand better which sea-fans are likely to bleed and which ones can be monitored.

The aim of treatment is to reduce the risk of any sea-fan causing vitreous haemorrhage and reduce the traction between the vitreous and the retina.

### Laser treatment

Laser treatment may be used when sea-fan is present and thought to be at risk of causing a vitreous haemorrhage or retinal detachment. Applying laser to the areas around the sea-fan helps to reduce the production of proteins such as VEGF. The result is that the sea-fan may be more likely to shrink and less likely to bleed or pull on the retina.

#### What happens in laser treatment?

As laser treatment is performed at an outpatient clinic, you won’t need to stay in hospital. You’ll be given eye drops to widen your pupils, so your ophthalmologist can look into your eyes more easily.

Your eye is then numbed with anaesthetic drops so that you don’t feel any pain, and a small contact lens is put on the surface of your eye to keep it open. During the treatment, you’ll be asked to move your eyes in certain directions so the correct part of your retina can be treated. You’ll be able to do this easily with the contact lens in place. Ask your ophthalmologist how long each session of laser surgery is likely to last. Some people need more than one treatment session.

#### Is laser treatment painful?

Laser treatment for PSR does not usually cause discomfort because it doesn’t take long and only treats a small area of your retina.

#### Does laser treatment have any side effects?

No treatment is possible without some side effects. However, your vision may be at greater risk by not having the laser treatment. The short-term effects of laser treatment happen because of the brightness of the laser beam. It can reduce your vision for an hour or two after the treatment. You may also temporarily lose a little of your central vision or see small black spots, all of which should get better with time. As laser treatment is applied only to specific areas of affected peripheral retina, it is unlikely to affect your vision at all.

It’s important to remember that laser treatment aims to prevent your vision from getting worse. It cannot make your vision better. However, if you do not have laser treatment when recommended, you may end up losing a lot more of your sight. Ask your ophthalmologist to talk you through your treatment plan, the advantages and disadvantages of the treatment and its possible side effects, temporary or permanent, for your vision.

### Anti-VEGF treatment

Anti-VEGF medications are used commonly to reduce new blood vessel growth in several different conditions. They work by blocking the action of VEGF. VEGF is a protein produced by the retina when there is not enough oxygen or blood flow to an area. VEGF stimulates the growth of new blood vessels. Anti-VEGF medications work by helping to reduce new blood vessel growth.

In many countries, anti-VEGF treatments are used alone or in addition to localised laser to reduce the risk of bleeding or to help promote clearing of vitreous haemorrhage. Sometimes these injections will be used for people who have previously had laser treatment or before they have a treatment called vitrectomy. These injections are not currently licensed for PSR and are rarely used in the UK. Large studies are still required to understand when these treatments are most beneficial in the treatment of PSR.

### Vitrectomy

The middle part of your eyeball is filled with a clear gel (called the vitreous gel). If you have a bleed from your retina into the vitreous gel, your vision will become cloudy. Often, this blood is reabsorbed by your body and your vision gets better on its own over a few months.

However, if your vision doesn’t improve, you may need to have vitrectomy surgery. In this surgery, the cloudy vitreous gel is removed and replaced with a clear liquid, which usually helps to improve your vision.

Your ophthalmologist may advise you to wait for up to six months before carrying out a vitrectomy, and this might be frustrating. However, over this time, your ophthalmologist will monitor how the bleeding changes, to see if there are any new bleeds, and whether your bleeding has begun to be reabsorbed.

If your retina detaches, then vitrectomy is needed to reattach your retina and restore vision.

A vitrectomy is a specialised and complicated operation, and your ophthalmologist would only carry out this surgery when it is needed, and the benefits outweigh the risks involved. They should discuss the advantages and disadvantages of the procedure with you before scheduling the surgery.

## Driving and PSR

If you drive and have had a diagnosis of PSR in both eyes you must inform the Driver and Vehicle Licensing Agency (DVLA). Your eye doctor may also advise you to inform the DVLA.

The DVLA may ask that you have a detailed eye examination to make sure your peripheral and central vision is good enough for safe driving.

## Coping

It’s completely natural to be upset when you’ve been diagnosed with sickle cell retinopathy and it’s normal to find yourself worrying about the future of your sight.

It can sometimes be helpful to talk about these feelings with someone outside your circle of friends or family. At RNIB, we can help with our telephone Helpline and our Counselling and Wellbeing Team. Your GP or social worker may also find a counsellor for you if your feel this might help.

You may think of further questions about your eye condition on your way home from hospital or in the days and weeks following your diagnosis. There is someone to turn to with these questions. Your eye clinic may have a sight loss advisor working alongside the doctors and nursing staff. This advisor may be known as either the Eye Care Liaison Officer (ECLO), the Vision Support Officer or the Early Intervention Support Officer and they are on hand within your hospital to provide you with further practical and emotional support about your eye condition. Alternatively, you can call our Helpline to speak to our advisors within our Eye Health Team who would be happy to discuss any questions you may have.

## Further help and support

### Help to see things better

If you have sight loss from sickle cell retinopathy, there are lots of things that you can do to make the most of the vision you have. This may mean making things bigger, using brighter lighting or using colour to make things easier to see.

A low vision assessment gives people a chance to discuss any practical problems they are having with their vision with a low vision specialist. The specialist can explore things like magnifiers, lighting, colour contrast and other adaptations that may help.

Assistive technology can also be very useful to help you with your work, hobbies and activities.

You can ask for a referral to a low vision clinic from your ophthalmologist, optometrist or GP. We have a series of leaflets with helpful information on living with sight loss, including how to make the most of your sight.

You can find out more about our range of titles by calling our Helpline.

### Support for your work or schoolwork

When you have sight changes, you may be worried about finding work, or staying in your job. Our Employment team can provide specialist support and advice about employment for people with sight loss. You can contact this team via our Helpline.

For children who have sight loss from sickle cell retinopathy, having the right support at an early age can make a big difference. Your local authority (LA) should have at least one qualified teacher of children and young people with vision impairment (QTVI) to work with you and your child both at home and at school. A QTVI is a qualified teacher who can provide support with development, play, learning and education. At an early stage, ask your local authority to put you in contact with a QTVI. They will support you and your child as soon as a visual impairment is suspected or diagnosed. If you have difficulty getting help or need the details of the specialist teacher in your area, contact RNIB Helpline.

### Support from social services

Local social services should also be able to offer you information on staying safe in your home and getting out and about safely. They should also be able to offer you some practical mobility training to give you more confidence when you are out.

Depending on how much of a person’s sight is affected by sickle cell retinopathy, they may be eligible to be registered as sight impaired (partially sighted) or severely sight impaired (blind). An ophthalmologist would be able to tell you whether you are eligible. Registration can act as a passport to help and some concessions, but a lot of this support is still available to people who aren’t registered.

You can find more information about all the support available to people with sight problems on our website or by calling our Helpline.

### Further help from RNIB

#### RNIB Helpline

If you need someone who understands sight loss, call our Helpline on **0303 123 9999**, say **"Alexa, call RNIB Helpline"** to an Alexa-enabled device, or email **helpline@rnib.org.uk**. Our opening hours are weekdays from 8am-8pm and Saturdays from 9am – 1pm.

You can also get in touch by post or by visiting our website:

#### RNIB

Grimaldi Building

154a Pentonville Rd

London N1 9JE

**rnib.org.uk**

#### Sight Advice FAQ

Ask the Sight Advice FAQ website your questions about sight loss and get helpful answers: **sightadvicefaq.org.uk**

#### Connect with others

Meet or connect with others who are blind or partially sighted online, by phone or in your community to share interests, experiences and support for each other. From book clubs and social groups to sport and volunteering, our friendly, helpful and knowledgeable team can link you up with opportunities to suit you. **Visit rnib.org.uk/connect** or call our Helpline.

## Other useful contacts

Sickle Cell Society

54 Station Road

London

NW10 4UA

Tel: **020 8961 7795**

Web: **sicklecellsociety.org**

## We value your feedback

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Send your comments to us by emailing us at **eyehealth@rnib.org.uk** or by writing to the Eye Health Information Service, RNIB, Grimaldi Building 154a Pentonville Road, London, N1 9JE.

## Information sources

This factsheet has been written by the RNIB Eye Health Information service. Our factsheets have been produced with the assistance of patient and carer input and up-to-date reliable sources of evidence. The accuracy of medical information has been checked by medical specialists. If you would like a list of references for any of our factsheets, please contact us at **eyehealth@rnib.org.uk**.

All factsheets are available in a range of formats including print, audio and braille.

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