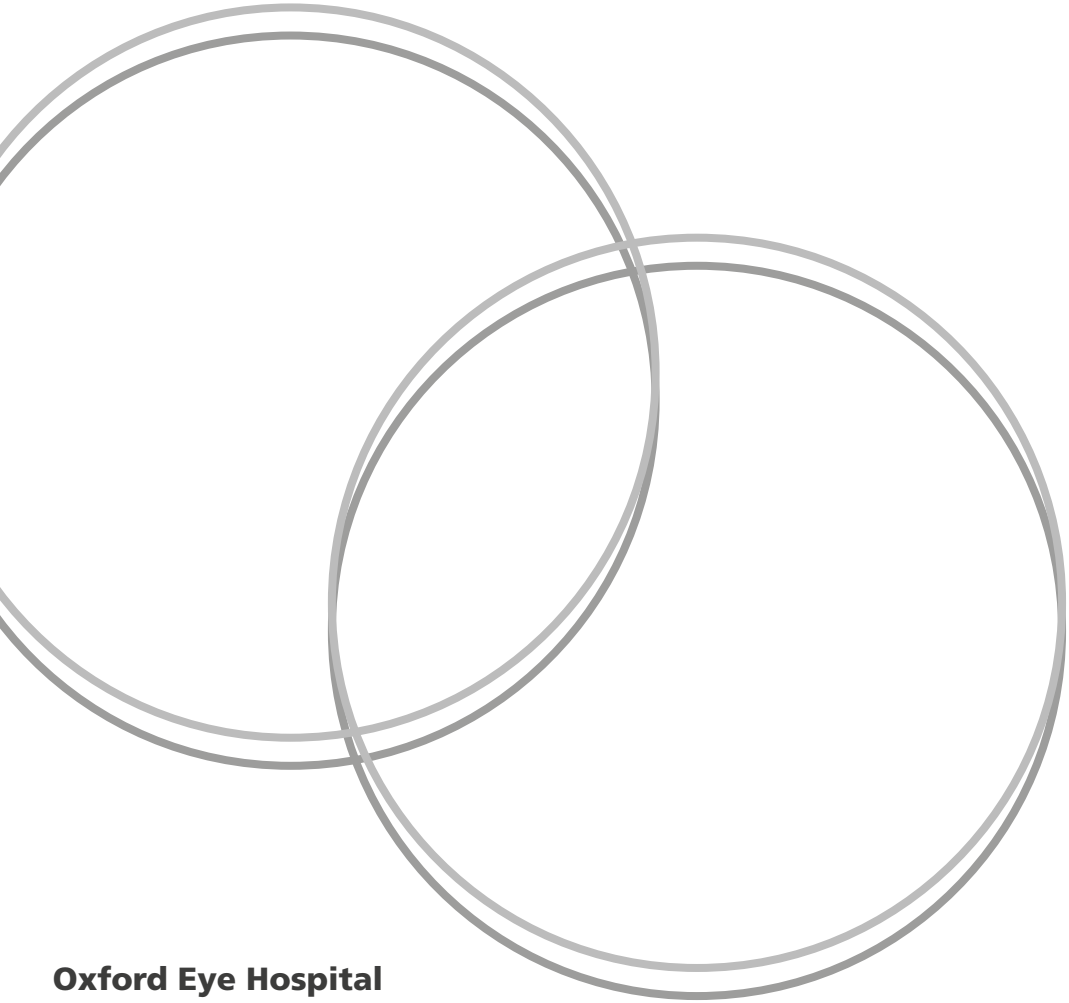
Oxford University Hospitals
NHS Foundation Trust

# Sickle Retinopathy

## Information for patients



Oxford Eye Hospital

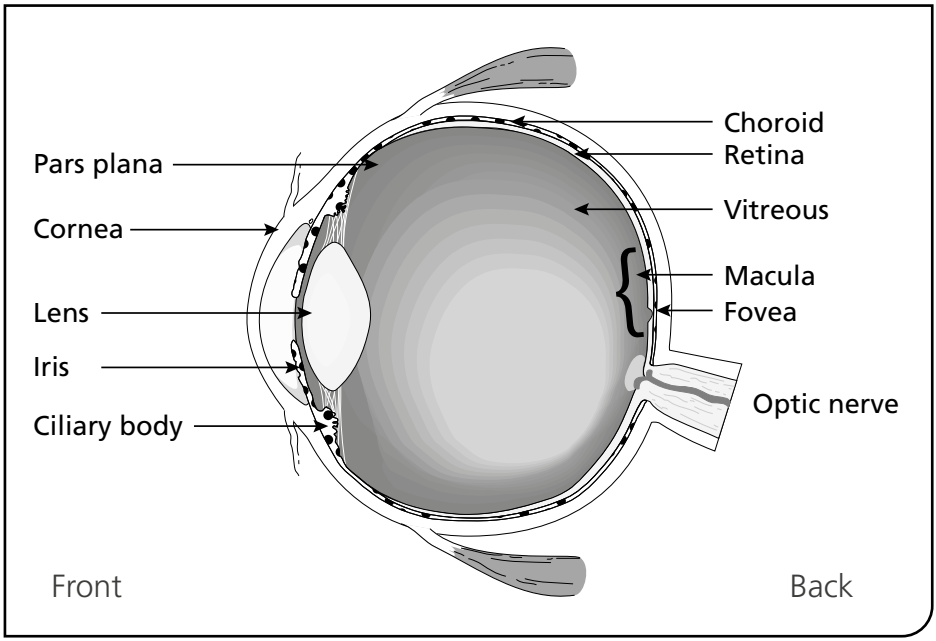
**This information leaflet will provide you with information regarding how sickle can affect the eye.**

# What is the retina and why is it important?

The retina lines the back of the eye. It is made up of delicate cell layers which work together to capture images. These images are then communicated to the brain by the optic nerve; this allows us to see. If you think of the eye as a camera, then the retina is like the film in the camera which captures the images.

The macula is the part of the retina responsible for your central vision (sight straight in front of you). This is particularly important for reading and detailed vision. The rest of the retina is responsible for your peripheral vision (sight on the edge of your vision).

### Cross section of the eye



# What is Sickle Retinopathy?

Sickle retinopathy is an eye complication of sickle cell disease. It causes damage to the back of the eye (retina) and can cause loss of vision in a few individuals.

In the early stages of the condition there are usually no symptoms. So, without regular examination of the eye by an eye specialist it may go undetected. However, sickle retinopathy may progress and affect the vision, and in some cases cause loss of vision.

The following is a brief description of the most common features that are seen in sickle retinopathy:

- Small obstructions of blood vessels can cause retinal disturbances, which are referred to as haemorrhages ('salmon patches') or old pigment changes ('sunburst' patches).
- Changes to peripheral (edge) vision may occur when retinal vessels are affected.
- Some individuals develop proliferative sickle retinopathy; this is the development of abnormal blood vessels in the retina. These are known as 'sea fans' because of their appearance. These can leak blood into the vitreous cavity in the eye (vitreous haemorrhage). See diagram of eye on page 2.
- In some cases, a retinal tear or a retinal detachment may occur. A retinal detachment is when the retina comes away from its normal position in the eye. This can lead to permanent loss of vision if appropriate treatment is not received.
- Other features include obstructions of small vessels in the macula leading to a deterioration in vision and occasionally similar blockage of small vessels may occur on the optic nerve head.
- Other causes of visual impairment include obstruction of vessels which can affect the central part of the vision.

It is important for anyone who has a haemoglobinopathy (sickle cell disease, thalassemia) to be aware that if they receive a knock or blow to their eye, that there is a risk that blood can collect in the front part of their eye, like a bruise. This is called a 'hyphaema'. The presence of a hyphaema can lead to high pressure in the eye, which can lead to loss of vision. It is often painful. It is important that after significant trauma to the eye that an urgent review is carried out by the eye service.

# Who can develop Sickle Retinopathy?

Anyone with a haemoglobinopathy can develop sickle retinopathy.

**The symptoms listed below are an eye emergency. If you experience any of these symptoms, please go to your nearest Eye Emergency Department as soon as possible for urgent assessment (see contact details for Oxford Eye Emergency Department at the end of this leaflet). If these symptoms occur out of hours, please call NHS 111 who will advise you where to go. Please do not wait for your next clinic appointment.**

Symptoms:

- Flashes of light
- New 'floaters' (black dots, lines)
- Sudden onset of blurred vision
- Loss of peripheral or central vision, a 'curtain' or shadow obscuring part of the vision in one or both eyes.

# How is Sickle Retinopathy monitored and why is this important?

Regular examination of the retina allows the eye specialist to check for the presence of sickle retinopathy and whether there have been any changes since your last appointment. If sickle retinopathy has developed which requires treatment this will be discussed.

It is very important to attend screening appointments as progression in sickle retinopathy can occur without any noticeable changes to your vision, so without review it is not possible to detect these. If you are unable to attend, please call us to cancel your appointment and to reschedule your appointment for the next available date.

The current recommendations are that screening should occur once a year after the age of 18. However, if changes are detected in your retina, your doctor may suggest more frequent reviews.

Children aged 12 to 18 should have screening every 2 years.

After the age of 50, if there are no signs of sickle retinopathy, it is likely that yearly screening will not be required as the chances of developing sickle retinopathy are very low. You would then be discharged from our screening service to your community optometrist for regular check-ups.

# What happens when I come to the clinic?

At your appointment you will have your vision checked. Please bring your glasses if you wear any.

Eye drops will be used to dilate your pupils, this will allow us to see the back of your eye clearly when the eye is examined and during imaging of the eye. These drops will make your vision blurred for 4 to 6 hours and you will not be able to drive during this time. Please arrange someone to drive you home, or accompany you home if you are using public transport or a taxi.

During the appointment, a picture and a scan will be taken of the back of your eye with a special camera. This is a quick and painless procedure.

In some cases, you may need to have further imaging of the back of your eye.

Very occasionally, it may be necessary to carry out a type of imaging which involves injecting dye into your arm to highlight any abnormal vessels when the retinal imaging is performed (fluorescein angiography). If this type of test is required at your appointment, you will be given an explanation of what this will involve so that you are fully informed to give your consent for the procedure.

The imaging will be reviewed by the Ophthalmology team, and the results fed back to you, usually within 10 days. Occasionally it will be necessary to be seen by an eye specialist (Ophthalmologist). If this is the case you will be sent an appointment.

For many individuals who have developed sickle retinopathy, laser or surgery is not required, but this is reviewed at each visit depending on the stage of the retinopathy.

Treatment of seafans (proliferative sickle retinopathy) may be indicated in the presence of recurrent vitreous haemorrhages or severe proliferative retinopathy. Laser will be required for retinal tears, and surgery for retinal detachments.

If laser or surgery is required, this will be explained at your appointment. For most individuals who undergo screening, although some features of sickle retinopathy may be identified, most do not affect vision and do not require intervention.

Of note there is further research ongoing into sickle retinopathy which may inform how we screen and treat this condition in the future.

# How to contact us

## Eye Emergency Department

Do not attend the eye emergency department without calling by telephone first.

If you have an eye emergency, please call **01865 234 567 option 1** followed by **option 1**

You will be able to speak to an eye specialist who will advise you on what action to take.

Opening Hours Monday to Friday – 8:30am to 4:30pm
Saturday, Sunday and Bank Holidays – 8:30am to 3:30pm
Closed Christmas Day

The Eye Emergency Department is in the Oxford Eye Hospital in the West Wing on level LG1.

If you have an eye emergency outside of these hours, please call NHS 111 who will advise you.

# Adult Red Cell Service Oxford Triage Assessment Team

If you are worried that you need urgent attention for your blood disorder, or have a medical concern (e.g. if you have a sickle cell disease and think you might be developing a crisis), we have a dedicated triage assessment team.

Telephone: 01865 572 192
Open 8am to 8pm, 7 days a week
Outside of these hours please call 01865 235 048/049

# Useful information

### Sickle Cell Society UK

Telephone: 020 8961 7795

Email: info@sicklecellsociety.org

Website: www.sicklecellsociety.org

## Further information

If you would like an interpreter, please speak to the department where you are being seen.

Please also tell them if you would like this information in another format, such as:

- Easy Read
- large print
- braille
- audio
- electronic
- another language.

We have tried to make the information in this leaflet meet your needs. If it does not meet your individual needs or situation, please speak to your healthcare team. They are happy to help.

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