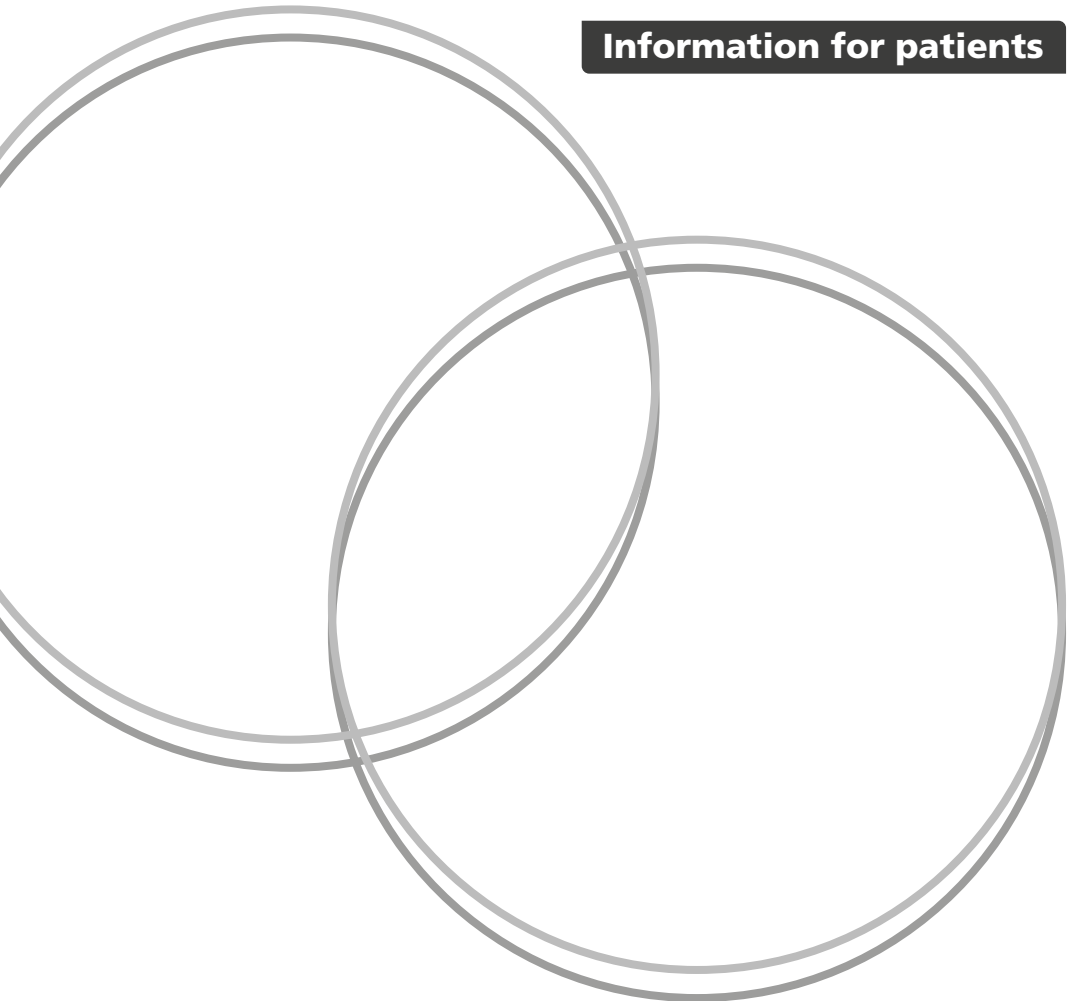
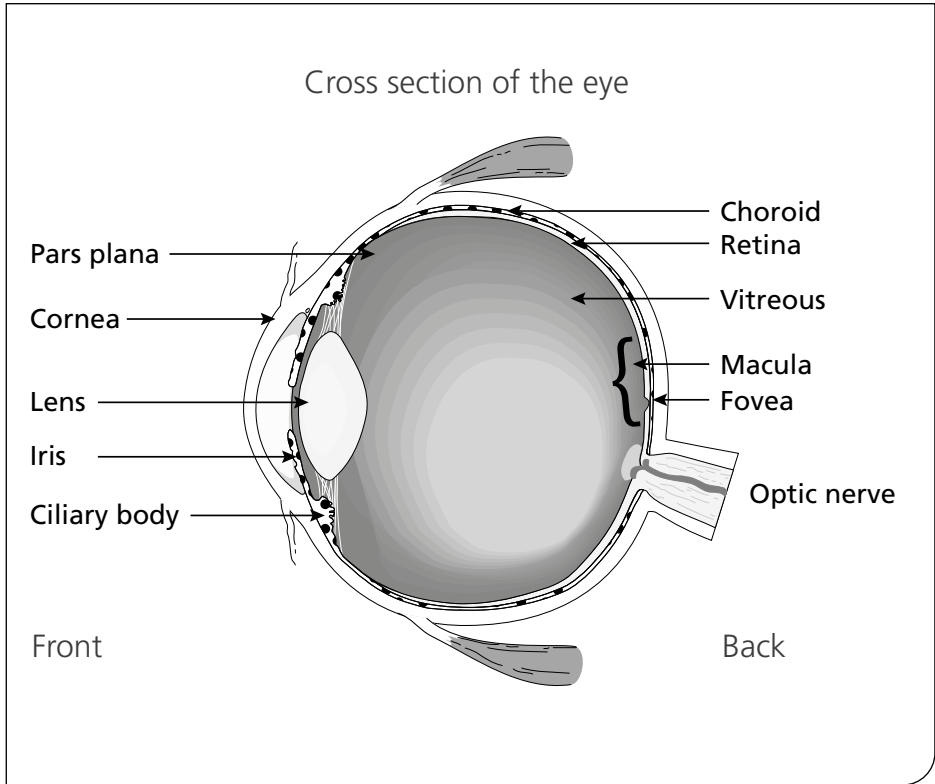


Von-Hippel Lindau (VHL) Disease – Screening of the Retina for Retinal Angiomas

Information for patients



You have been asked to attend for retinal screening to check the back of your eyes as VHL can affect the retina (see diagram below to show the location of the retina). This leaflet will give you more information about what to expect at this appointment.



The retina is a delicate layer at the back of the eye which is important for vision. Individuals with VHL have a high chance of developing retinal angiomas. Retinal angiomas are a benign (non-cancerous) type of tumour formed from an overgrowth of small blood vessels within the retina. If retinal angiomas are not treated, they usually continue to grow and can cause visual loss. This is why screening for retinal angiomas has been recommended for you.

If retinal angiomas are found while they are still small, they can be treated early to stop them growing, thus preventing them from affecting vision. It is recommended that individuals with VHL have a screening check of the retina every year by an Ophthalmologist (eye doctor), or to have retinal photographs reviewed by an Ophthalmologist each year. If one or more angiomas are identified, then treatment will usually be recommended. The type of treatment recommended will depend on which part of the retina the angioma is found in and its size. This will be explained to you at an appointment with the Ophthalmologist.

What happens at your screening appointment?

First, your vision will be checked. After this, drops will be put into your eyes to dilate your pupils so that the ophthalmologist can see the whole of your retina using a special piece of equipment called a 'slit lamp'. We recommend that you do not drive to your appointment, as after the drops have been put in your vision will be blurred, and your eyes may be sensitive to light. If you do need to drive home, you will need to wait for about 4 hours after drops were put in for the effect to wear off, in some people this can take longer.

At the appointment, the Ophthalmologist will examine your retina. If no new angioma(s) are seen, another appointment for eye screening will be offered for the following year. A letter will be sent to you with the results of your screening tests. If there are changes suggestive of an angioma, then a follow-up appointment will be organised in the Eye hospital for more tests or for treatment. Alternatively, instead of seeing an Ophthalmologist at your appointment, retinal photographs will be taken in the Eye Hospital, which the Ophthalmologist will review at a later time, screening the images for angioma(s). Likewise if changes are seen, a follow up appointment will be organised in the Oxford Eye Hospital for more tests or for treatment.

What treatment might be recommended?

If angioma(s) is/are found, a clinic appointment with a specialist Ophthalmologist will be arranged, usually within 2 to 4 weeks to discuss options for treatment. The most common treatment is laser to the angioma. The type of treatment and amount of treatment required will depend on which part of the retina the angioma is located in and the size of the angioma(s). The aim of laser treatment is to destroy the angioma.

If a retinal angioma is not treated, it is likely to grow in size and may pull on the retina. This can lead to a retinal tear, which can develop into a retinal detachment and cause loss of vision. In addition, fluid can leak from an angioma which can also lead to visual impairment. The laser treatment is aimed at destroying the angioma so that it no longer grows or leaks.

If an angioma is large it is likely to require more extensive laser, or alternative treatment and this will be discussed with you. If the angioma has developed on the optic nerve, it might need to be managed in a different way as laser treatment might not be helpful. Again, your Ophthalmologist will discuss management options with you. The angioma may be monitored over time on a regular basis.

Retinal Laser Treatment

Retinal laser treatment is usually carried out in the Outpatients Department of the Oxford Eye Hospital by an Ophthalmologist. You will usually be awake for the procedure, which is performed while you are seated. Dilating drops and numbing (anaesthetic) drops are put into your eye, then a lens is placed on the front surface of your eye to enable the ophthalmologist to have a good view of your retina and the area to be treated.

The slit lamp machine (which is the same machine used for the original screening examination) is used to deliver the laser treatment. After the treatment you may notice blur or dazzle, but this will wear off after a few hours. If the angioma is in a more difficult position to reach with the laser, or if it is already large, a different type of laser or freeze treatment (called cryotherapy) may be used. Sometimes multiple episodes of treatment may be necessary. This may be carried out in the operating theatre rather than in the Outpatient Department, and you may be given sedation. A general anaesthetic is usually used for the treatment of angiomas in children.

The appropriate options will be discussed with you at your appointment and summarised in the letter we will send you after your appointment.

A follow up appointment will be arranged depending on your requirement for more treatment. This will usually be between one and three months after your first treatment.

What are the risks of treatment?

If the angioma is very small and has developed away from the centre of your retina, then laser treatment usually does not cause any symptoms or side effects after treatment. A pigmented (coloured) scar develops in the retina after treatment which can be seen in photographs of the eye or by your Optometrist or Ophthalmologist. This will not affect your vision if it is in the peripheral retina (the far part of your retina), away from the central retina. However, the following may occur:

- Bleeding from the angioma is uncommon, however it may make the vision blurred post treatment, but would be expected to settle within a week depending on the amount of bleeding.
- Retinal tear formation, which can lead to a retinal detachment. Retinal tears can be treated with laser in most cases, but retinal detachments will require surgical intervention. It is important to be aware that if you experience any of the following symptoms after treatment, then you should be seen urgently:
 - flashing lights
 - sudden onset of new floaters in the vision
 - loss of part of the vision (visual field loss)
 - decrease in vision.
- Exudation – the angioma may leak/ooze fluid after treatment which can affect your vision. If you have any changes in your vision you should be seen urgently.
- An epiretinal membrane may form which is a wrinkling of the surface of the retina at the centre. This usually takes weeks or months to develop and is usually associated with a gradual deterioration in vision and/or distortion of vision.

If you develop any of these, or any new symptoms affecting your vision after the treatment, it is important that you seek an urgent ophthalmology review from either the Oxford or your local Emergency Eye Care team so that your eyes can be reviewed. Contact details for the Oxford Eye Emergency department are on the next page.

This leaflet has been designed to provide information about the screening, monitoring and management of retinal angiomas. It may not address all your questions. Please do not hesitate to contact us if you require any more information. If you have VHL and have not had your annual screening appointment, please notify the VHL service coordinator, who can arrange an appropriate screening appointment.

If you have an eye emergency, please do not come to Oxford Eye Hospital straightaway. Call our specialised telephone triage number.

Telephone: **01865 234 567** option 1 followed by option 1
Monday to Friday 8.30am to 4.30pm
Saturday and Sunday 8.30am to 3.30pm (including Bank Holidays)

You will be able to speak to an ophthalmic health professional who will advise you.

If you need advice **out of hours**, please phone **NHS 111** or your out of hours GP practice

Useful contacts:

Oxford Eye Hospital Eye Clinic Liaison Officer

Telephone: 01865 231 137
Email: eyeclinicliaisonofficer@ouh.nhs.uk

Oxford Cancer Genetics Service / VHL Service Coordinator

Telephone: 01865 226 034
Email: orh-tr.churchill-clinicalgenetics@nhs.net

VHL UK /Ireland

Telephone: 0808-189-0891
Email: support@vhl-uk-ireland.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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Oxford University Hospitals NHS Foundation Trust
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