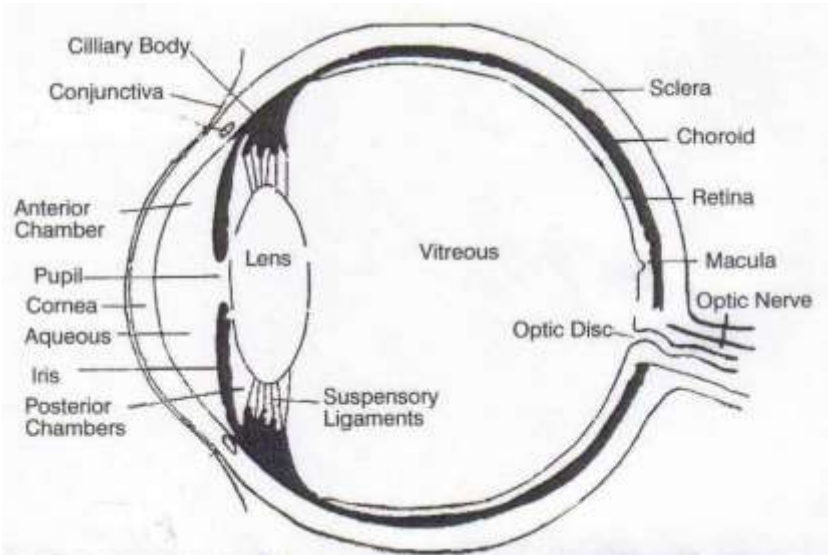


Eyes see all sorts of things — small things near to us and large things at a distance, colours and textures, even dimensions and moving objects. To do this, all the different parts of the eye need to be working correctly. A problem in one area of the eye can disrupt the messages passed on to the brain. Visual information received by the retina travels from the eye to the brain via the optic nerve. The brain then makes sense of the images it receives.

Retina The inner layer at the back of the eye, containing cells sensitive to light. These cells collect the light signals and send them as electrical messages to the optic nerve.

Optic nerve The optic nerve is made up of thousands of nerve fibres. These fibres pass electrical signals along to the brain where they are processed into an image.



This fact sheet summarises some of the basic facts about Optic Pathway Gliomas (OPGs) occurring in Neurofibromatosis Type 1 (NF1). These are sometimes called optic nerve tumours or optic gliomas but we will refer to them here as Optic Pathway Gliomas.

Up to 20% of individuals with NF1 may develop an OPG and in about a quarter of these people, it may lead to problems with their vision. It is, therefore, important that both patients and their doctors know about OPGs.

What is an Optic Pathway Glioma?

An OPG is a tumour (growth) occurring along the nerve supplying the eye, which is called the optic nerve. The optic nerve extends from the back of the eye into the brain and an OPG may occur at any point along the nerve and often can involve both optic nerves.

Although an OPG is a potentially malignant tumour it is one of the mildest forms in its class. For most people the main impact results from where it is located rather than from its cancer-like properties. In a few cases the tumour can behave more aggressively and if untreated lead to death, but this is very rare.

Who can get an Optic Pathway Glioma?

OPGs can occur in anyone, but are very much more common in NF1. In NF1 they tend to occur in early childhood, being seen most commonly at about 5 years. However, they can also occur later in childhood and occasionally even in adults.

What happens if you have an Optic Pathway Glioma?

In many cases the OPG may not cause any symptoms. If an OPG does cause symptoms, these may be related to its position along the optic nerve but can be variable. If an

OPG develops towards the front of the optic nerve, nearest to the eye itself, this may occasionally lead to a visible change. For instance: the eye may gradually become more prominent over time or the tumour may interfere with the normal movements of the eye and cause a squint. More often there is no outward sign of the OPG.

The main effect of an OPG is on vision. In about a quarter of those with the tumour there may be some degree of visual deterioration which either they or, more frequently, the parents of an affected child may detect. This deterioration can range from very mild to so severe that the individual loses all sight from that eye. In a larger number there may be either mild changes in vision or changes at the back of the eye which are only detectable by an eye specialist (ophthalmologist).

Sometimes the tumour may cause more general symptoms such as nausea, headaches or vomiting, due to its position in the brain. If the tumour involves some of the neighbouring brain structures there can be additional features such as hormone changes related to growth or puberty.

How do we detect an Optic Pathway Glioma?

If an individual with NF1 has any of the changes described above (such as change in eye movements, a prominent eye, altered vision or possible related symptoms) the doctor will investigate for the presence of an OPG if the features indicate this as a possible cause. This may include a visual examination and a brain scan.

However, as we know that there is an increased risk of developing OPGs in NF1 and that for a small number of affected individuals this may impair their vision, we recommend looking for early signs in those most at risk, especially children. Studies have shown that the best way to look for those tumours which are likely to cause problems is to look for changes in the vision or features seen at the back of the eye. If no abnormalities are detected we recommend that children should have their eyes reviewed yearly by an ophthalmologist until the age of 7 years. From 7 to 16 years this yearly eye assessment can be performed by an optician. As there is a small risk to older NF1 patients we recommend they continue to have their eyes checked from age 16 by an optician every two years.

If an OPG is suspected, more detailed investigations may be performed. A brain scan (MRI) may help to define the features of the tumour and determine the best way in which it should be managed. The ophthalmologist may perform tests to define exactly how the vision has been affected. These will all enable us to see if things change over time.

How do we treat Optic Pathway Gliomas?

It is important to note that not all OPGs will require treatment. Often, although it may have caused some impairment of vision, the OPG may not increase or may even shrink a little and treatment may not be necessary. Sometimes the position of the tumour or associated symptoms may determine whether treatment is necessary.

There are three main ways in which an OPG can be treated:

1. The tumour may be removed by surgery.
2. Radiotherapy was often used in the past but this method of treatment may be associated with further complications in NF1 patients and should only be used where it is the only option.

3. Chemotherapy is now being used more frequently, either alone or together with the other methods and there are ongoing studies evaluating the long-term effects of this treatment.

It is important that the OPG is treated by doctors who have experience of OPGs in NF1.

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