The ROYAL MARSDEN
NHS Foundation Trust

Optic pathway glioma

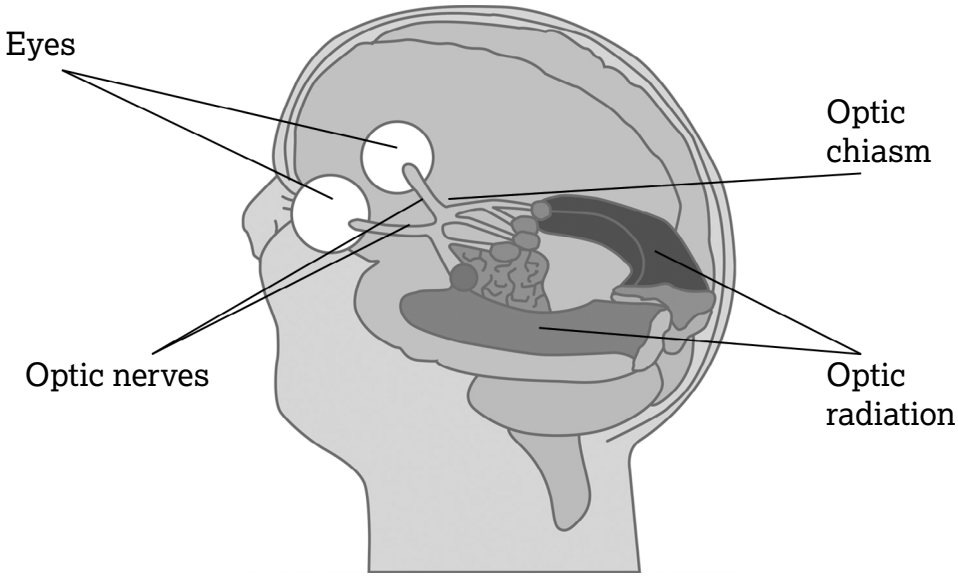
Oak Centre for Children and Young People

Information for patients and their families

NHS

What is an optic pathway glioma?

Childhood optic pathway glioma is a type of brain tumour which is usually benign (non-cancerous) and slow-growing. It is also called a low grade glioma. Gliomas are tumours that originate from brain cells called astrocytes. An optic pathway glioma occurs along the nerves that send messages from the eye to the brain (the optic pathway) and can occur anywhere along its path.



Who gets an optic pathway glioma?

Although the cause of most brain tumours is not known, we do know that a genetic condition called neurofibromatosis (type 1) can lead to these particular tumours. For children under five years old, the condition may not be obvious to anyone until the time of diagnosis of the optic pathway glioma. Neurofibromatosis type 1 (NF-1) can be inherited from a parent, but half the time the child will be the first affected in the family.

The condition varies widely but the following problems can occur throughout life:

- Tendency to develop both benign (non-cancerous) and occasionally cancerous tumours
- Skin lesions (several types from lesions similar to freckles to large lumps in the skin [neurofibromata])
- Skeletal problems (for example, curvature of the spine or scoliosis)
- Behavioural and learning difficulties (ranging from none to severe).

A child/family with NF-1 will be offered specialist counselling and will be followed up by a paediatrician with experience in the condition.

What are the signs and symptoms?

The first problems a child may suffer from are usually related to vision, such as:

- Reduced vision
- Squints
- Flickering eyes
- Eye protruding forward
- Double vision
- Head tilt
- Blind spots.

If there is increased pressure in the head (raised intracranial pressure), symptoms will also include:

- Nausea and vomiting (most common)
- Lethargy and irritability
- Headaches.

These symptoms may show up as problems in school, apparent clumsiness and sitting closer to watch television. Part of the brain that helps to control the hormones in the body (hypothalamus) is sometimes affected by optic pathway tumours. This can lead to hormonal problems such as early puberty or weight changes (either loss or gain).

What tests (investigations) will be carried out?

We will need to carry out some tests to find out as much as possible about the type, position and size of the tumour. This will help us to decide on the best treatment for your child. These tests include:

- **CT scan** – it is likely a CT scan of your child's brain was the first specific test carried out at your local hospital. Although MRI scans are usually the best way of seeing the tumour and the effects of treatment, sometimes CT scans are also useful. For more information, please see The Royal Marsden leaflet *CT scan*.
- **MRI scan** – this scan allows us to see the brain and spine in great detail and is used regularly to diagnose and follow the effect treatment is having on your child's tumour. For more information, please see The Royal Marsden leaflet *MRI scan*.
- **Ophthalmic assessment** – a full assessment of your child's vision will need to be recorded at the time of diagnosis, during and after treatment. This is painless but sometimes difficult for young children.
- **Endocrine assessment** – if there is evidence of the tumour pressing on the hypothalamus or pituitary gland, a full test of your child's hormones will be needed. This usually involves simple blood tests.

Staging

Staging is a measure of how far the tumour has spread beyond its original site.

How do you treat an optic pathway glioma?

Observation

If a child's symptoms are very mild (children with known NF-1 will have visual screening to pick up early problems) and the optic glioma is small and growing very slowly, observing the tumour only, is an option. Optic gliomas associated with NF-1 can be very benign (non-cancerous) and can occasionally stop growing, or even shrink without treatment. However, regular eye checks and brain scans are necessary for observation.

Surgery

Surgery may be considered after the diagnosis of an optic pathway glioma. The purpose of this may be a biopsy to confirm the type of tumour or to try and remove part of the tumour to relieve pressure. As the main aim is to preserve vision, it is rare for a neuro-surgeon to try and remove all of the tumour, as this operation could damage the nerves supplying the eye.

Chemotherapy and radiotherapy

Both chemotherapy and radiotherapy can be used to treat the tumour with the aim of stopping the growth so that any further loss of vision is stopped (sometimes vision is improved). The age of the child and how fast the tumour is growing are important in deciding whether the child needs further treatment and which type of therapy will be used.

In older children, radiotherapy will be considered as a first treatment. This treatment takes up to six weeks as an outpatient and is very effective at controlling the tumour but does have some potential significant long-term side effects.

Proton beam therapy

Children with optic pathway glioma may benefit from receiving proton beam therapy. Proton beam therapy delivers radiotherapy in a way that may reduce the radiation to healthy tissue outside the area needing treatment. This can mean that children have fewer long-term side effects.

In the UK, proton therapy is delivered at University College Hospital, London and The Christie Hospital, Manchester. If your child is eligible for NHS funding, you will be informed by the medical team and have the opportunity to discuss this in detail.

Chemotherapy is usually given as an outpatient and continues for up to 18 months. It is usually given by injections and drips into a vein (intravenous infusion). If your child takes part in a clinical trial, the treatment is explained in more detail in the specific trial information sheet. You will be given an outline of the treatment in the form of a 'road map', known as a protocol.

What are clinical trials?

Clinical trials are medical research trials involving patients that are carried out to try and find new and better treatments. In cancer, clinical trials are most commonly used to try and improve different forms of treatment such as surgery, radiotherapy or chemotherapy.

The treatment being tested may be aimed at:

- improving the number of people cured (for example, trying new types of surgery or chemotherapy)
- improving survival
- relieving the symptoms of the cancer
- relieving the side effects of treatment
- improving the quality of life or sense of wellbeing for people with cancer.

Clinical trials may also involve research aimed at understanding more about the tumour's biology. You may be asked to allow us to do research on the tumour sample removed at surgery, or on blood samples.

Well-run clinical trials have led to a significant improvement in the treatment of children with cancer. You can find more general information on clinical trials in The Royal Marsden

booklet *Clinical trials*. If you are asked to consider entering your child into a clinical trial, you will be given specific information about the trial before deciding whether to take part. Please note that there is no obligation to take part and the care your child receives will not be affected by your decision.

General side effects of chemotherapy

- Bone marrow suppression (myelosuppression) – chemotherapy drugs decrease the production of blood cells by the bone marrow for a variable period of time. This results in low red blood cells (anaemia), low white blood cells (neutropenia) and low platelets (thrombocytopenia). Your child may need blood or platelet transfusions and will be at increased risk of infections. The doctors and nurses caring for your child will tell you more about these side effects.
- Nausea and vomiting – some of the chemotherapy drugs used may make your child feel sick or vomit. We will give anti-sickness drugs at the same time to stop nausea and vomiting which are usually very effective.
- Sore mouth (mucositis) – some of the chemotherapy drugs make the lining of the mouth and throat very sore and ulcerated. We will give your child painkillers for this and explain how to care for your child's mouth during treatment.
- Weight loss – nausea, vomiting, mucositis and taste changes may result in your child losing weight. A dietitian will help you to support your child's nutrition during this time.
- Hair loss – temporary hair loss is common.

Chemotherapy drugs

For details of the side effects of individual drugs, please see the Macmillan individual drug information factsheets. Your Clinical Nurse Specialist (CNS) may print these for you or can access them via the Macmillan website *www.macmillan.org.uk*

What is the outlook (prognosis)?

Optic pathway gliomas are not usually life threatening (although aggressive tumours which grow quickly can be) and the aim is to preserve sight or function. In up to half of children, although controlled at first, optic pathway gliomas can grow back and may need further treatment.

What are the possible long-term effects?

A child may become visually impaired or blind, needing help for their disability including suitable educational support. They may need hormone replacement therapy throughout their lifetime. They may develop learning difficulties as a result of the tumour or its treatment.

Children with NF-1 may have other problems in addition to those from the tumour and these can impact on schooling.

All children are followed up after treatment is finished in the 'long-term brain tumour follow up clinic'. Your child will be seen at regular intervals in this clinic, indefinitely, so that we can help with any long-term effects of the treatment.

Contact details

Children and Young People's Unit

24 hour Helpline: 020 8915 6248

Other sources of information and support

General information on chemotherapy, hair loss and side effects is also available from the PALS Help Centre or via *www.royalmarsden.nhs.uk* and in the 'parent held record'.

Macmillan Cancer Support

Macmillan Support Line: 0808 808 0000

Website: *www.macmillan.org.uk*

Macmillan provides a range of free information and support on all aspects of cancer including:

- advice on benefits and other kinds of financial support
- information on local cancer support groups and organisations near you.

The Children's Cancer and Leukaemia Group (CCLG)

Website: *www.cclg.org.uk*

CCLG is a national professional body responsible for the organisation of the treatment and management of children with cancer in the UK. They coordinate national and international clinical trials. They also provide information for patients and families and produce a quarterly magazine called Contact.

The Brain Tumour Charity

Website: *www.thebraintumourcharity.org*

The Brain Tumour Charity is a UK dedicated brain tumour charity, committed to fighting brain tumours. They fund scientific and clinical research into brain tumours and offer support and information to those affected, whilst raising awareness and influencing policy.

Young Lives vs Cancer

Website: *www.younglivesvscancer.org.uk*

Support line: 0300 303 5220

Young Lives vs Cancer is a UK charity for children, young people, and their families, which provides clinical, practical, financial and emotional support to help them cope with cancer.

Brainstrust

Website: *www.brainstrust.org.uk*

Brainstrust is a UK brain tumour charity which is dedicated to helping people affected by a brain tumour. They aim to provide support and advice from the point of diagnosis.

Nerve Tumours UK

Website: *www.nervetumours.org.uk*

Nerve Tumours UK provides information and support to patients and their families who have Neurofibromatosis type 1 (NF1).

References

This booklet is evidence based wherever the appropriate evidence is available, and represents an accumulation of expert opinion and professional interpretation.

Details of the references used in writing this booklet are available on request from:

The Royal Marsden Help Centre

Telephone: Chelsea 020 7811 8438 / 020 7808 2083

Sutton 020 8661 3759 / 3951

Email: patientcentre@rmh.nhs.uk

No conflicts of interest were declared in the production of this booklet.

Should you require information in an alternative format, please contact The Royal Marsden Help Centre.

The patient information service is generously supported by The Royal Marsden Charity.

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Revised January 2023. Planned review January 2026