Surgery for these tumours is rarely an option for OPG, and in general is only used for cosmetic purposes to remove large tumours in the orbit, which have caused a blind eye.

Radiation therapy should be avoided in children with NF1 except in extreme circumstances due to the significant risk of causing damage to blood vessels in the brain, or secondary cancers caused by radiotherapy.

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

Outcomes for Children with OPG?

The main goal of treatment is preserving vision. Research into the vision of children with NF1 associated OPG has shown that 32% had improved vision after therapy, stabilised in 40% and declined in the remainder.

There is more research to be done, and improvements to be made in treatment of OPG, but these results are encouraging.

How can the Children's Tumour Foundation support you?

The Children's Tumour Foundation (CTF) can provide information and support. We employ support coordinators and host support groups around the country in conjunction with local peer support contacts and organise a range of events including camps and information seminars.

For all NF news, subscribe to the monthly e-Bulletin at
https://bit.ly/ctfsubscribe

Contact Us

National Support: 02 9713 6111
VIC/TAS Support: 03 9936 6268
Email: info@ctf.org.au
Web: www.ctf.org.au

www.facebook.com/CTFAus

@ctfaustralia

www.youtube.com/NFAUSTRALIA

Reviewed by:

Dr Jonathan Ruddle
Prof Kristi Jones
Dr Yemima Berman
Dr Maree Flaherty

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Optic Pathway Gliomas (OPG) in NF 1

Children's Tumour Foundation
CONQUERING NF

Optic Pathway Gliomas (OPG)

Optic Pathway Gliomas (OPG) are growths (tumours) that can grow anywhere along the optic nerves that extend from the back of the eyes to the brain, including the point at which they meet (chiasm). These nerves allow our brain to form images of the things we are seeing.

Issues arise due to the location of the tumour in some children as opposed to the way it grows. It is very rare that one of these tumours acts aggressively as seen in non-NF1 associated OPG.

How common are they?

Anyone can develop an OPG, but it is more common in NF1. As many as 15 - 20% of children affected by NF1 will develop an OPG. However, only ¼ to ½ will develop symptoms such as any impairment of vision.

When are they diagnosed?

An OPG is usually detected in children in the pre-school years (ages 3-5). Children over the age of 10 are unlikely to develop an OPG, but there are reports of this occurring and so it is important to continue to get eye screening until at least 16 years of age .

How are they detected?

Changes that are looked for when determining whether a child has an OPG include:

- A more prominent eye
- Changes in eye movements (eg one eye turning in or out)
- Altered vision

If a child/parent notices any of these changes the doctor will conduct an eye examination to see if any changes in vision or the optic nerve at the back of the eye are present.

Because we know that OPG is more common in children with NF1 and that a small number of those affected will have a change in their vision it is recommended that screening for early signs of these tumours occurs 6 monthly – yearly in children up to 6 years, and yearly from age 6 – 16. After this time it is recommended that adults have their eyes checked at least every 3-5 years by an ophthalmologist if no signs of OPG have arisen during childhood.

What happens next?

For many children diagnosed with an OPG no treatment is required, but your child's OPG will continue to be monitored via eye examinations and in some cases MRI (magnetic resonance imaging) as it is hard to predict what will happen with these tumours. The intervals at which these occur may vary between clinics and based upon the individual case.

Often, even though it may have caused some impairment of vision, the OPG may not increase in size, or may even shrink. This may mean that treatment is not necessary even if vision impairment is seen. The position of the tumour or its associated symptoms will often determine whether treatment is needed.

What about MRIs?

Parents often ask whether their child should be having regular MRI. Since approximately 50% of children with OPG never have symptoms at all, and the majority with symptoms never progress, the findings on MRIs may cause unnecessary alarm. Practices vary from place to place but most NF1 specific clinics agree that "screening" MRI scans in children who have normal eye exams and their height charted yearly are not necessary.

How are they treated?

For the people affected by OPG which requires treatment (approximately 1/3) the treatment most often used now is chemotherapy involving a combination of 2 drugs: carboplatin and vincristine. This combination is usually well tolerated and hospital admission is rare. However, sometimes children can develop a hypersensitivity to carboplatin requiring a switch to a different agent, and sometimes vincristine can cause peripheral (not brain or spine) nerve damage, which is often reversible.