Overview:

Tumour Group:
Gliomas and Pediatric Brain Tumours

Optic pathway gliomas (OPG) are generally very low-grade tumours.

WHO Grade:
Grade I pilocytic astrocytomas and grade II fibrillary (diffuse) astrocytoma are the most common tumours affecting these structures.

It would be unusual to have a high-grade tumour in this location.

Prevalence/Incidence:
5% of all childhood intracranial tumours are optic pathway gliomas.

Typical Age Range:
Occurs most often in infants and children under the age of 10, but can occur in adults.

Description of Tumour:

Like many brain tumour types, the exact cause of an optic pathway gliomas is unknown. However, optic pathway gliomas are associated with neurofibromatosis type 1 (NF-1), also called Von Recklinhausen’s disease.

- 20% of children with neurofibromatosis (NF-1) will develop an optic glioma
- These gliomas are typically grade I pilocytic astrocytomas
- Children with optic glioma are usually screened for NF-1 for this reason
- Adults with NF-1 typically do not develop optic gliomas.

This tumour type is named for its location on or near the optic nerve pathways between the eyes and the brain. These tumours may involve any part of the optic pathway and have the potential to spread along these pathways.
Symptoms:
Common symptoms include:
• Behavioural disturbance
• Clumsy, uncoordinated walk
• Developmental delay(s), or early puberty due to hormonal disturbance
• Double vision, also called strabismus (crossed eyes)
• Headaches
• Visual disturbance/loss (children may fall or bump into things more easily because of this symptom and may not complain directly that their vision has changed)

Treatment/Standard of Care:
The treatment of an OPG depends very much on the location and extent of the tumour. Not all patients with identifiable optic pathway gliomas on imaging ever have visual symptoms (especially if the patient has NF-1).

Spontaneous regression of these tumours has been documented (in patients with and without NF-1)

In many patients with mild symptoms, observation alone may be recommended because:
• Symptoms are mild and there is no immediate threat to vision
• The natural history of disease is most likely very slow (as with patient with NF-1)
• The patient is young and the aim is to reduce the risk of treatment related morbidity.

Surgical treatment for OPG may involve either biopsy or excision of the tumour. Surgery is usually the first choice when:
• Single nerve involvement is causing progressive disfiguring proptosis (bulging out the eye) and blindness.
• An exophytic tumour involving the optic chiasm is causing mass effect or hydrocephalus (raised intracranial pressure)

Chemotherapy has the ability to shrink OPG and may even stabilize the tumour. This treatment is often given to very young children to delay giving radiation therapy and avoid damage to the developing brain.

Moderately high-dose radiation therapy can be used to control these tumours. Different techniques can be used to spare surrounding normal structures such as stereotactic radiation therapy.

Prognosis:
Optic pathway gliomas that are considered Grade I tend to have the most favourable survival rates compared to other higher grade brain tumours.

For more details, please refer to braintumour.ca.