Optic pathway glioma treatment

- Growth hormone excess in children with neurofibromatosis and optic pathway glioma, an underdiagnosed condition: experience with long-acting treatment
- A Rare Case of Sporadic Optic Pathway Glioma in a 7-year-old Female
- Recent Advancement of Neurofibromatosis Type 1: A Narrative Review
- Supra-sellar clear cell ependymoma in a 2-year-old female: A case report
- Neurofibromatosis Type 1 : A General Review
- Predictors in Optic Pathway Gliomas in Neurofibromatosis Type 1: A Single Center Study
- A rare presentation of pediatric germinoma mimicking optic pathway glioma: illustrative case
- A Phase 2 PBTC Study of Selumetinib for Recurrent/Progressive Pediatric Low-Grade Glioma: Strata 2, 5, and 6 with Long-term Outcomes on Strata 1, 3, and 4

The extent of resection of pediatric low-grade glioma mostly improves progression-free survival. In chiasmatic hypothalamic glioma (CHG), complete resections are limited due to the relevantly high risk of associated neurological and endocrinological deficits. Still, surgery might have its role in the framework of a multidisciplinary team (MDT) approach [1)]

If it is isolated to one optic nerve and does not extend to the chiasm, then resection is curative (albeit with the loss of vision in that eye). If the tumour extends to the chiasm or more posteriorly, then curative resection is not possible, with resection reserved for treatment of mass effects (proptosis, intracranial mass effect)

En-block removal of intraorbita tumor is recommended in cases with disfiguring exophthalmos and loss of vision.

Surgical resection of intraorbital optic nerve (ON) poses the risks of permanent ptosis and globe atrophy.

Optimal management of optic pathway/hypothalamic glioma (OPHG) remains an ongoing challenge. Little is known about the natural history, management strategies, and outcomes in adolescents.

see Anterior interhemispheric approach

Radiotherapy continues to be the mainstay of progressive OPHG management strategies. In the 2000s, chemotherapy has emerged as a therapeutic option for those <10 years of age aiming to avoid or at least postpone radiation to the immature brain [2)].

Carboplatin-based chemotherapy is a useful modality in younger children, delaying radiation to their immature brains. National trials have focused on younger children and excluded adolescents from studies evaluating the role of chemotherapy.

In fact, many institutions continue to consider irradiation as the treatment of choice for progressive OPHGs in patients >10 years of age.

Chemotherapy is a valuable treatment modality for the achievement of disease control even in adolescents; their progression-free survival compares favorably with that in younger children. Chong et al. propose that chemotherapy be considered as a first-line modality in adolescents, avoiding potential radiation-associated morbidities [3].

1)
Karbe AG, Gorodezki D, Schulz M, Tietze A, Gruen A, Driever PH, Schuhmann MU, Thomale UW. Surgical options of chiasmatic hypothalamic glioma-a relevant part of therapy in an interdisciplinary approach for tumor control. Childs Nerv Syst. 2024 Jun 25. doi: 10.1007/s00381-024-06498-2. Epub ahead of print. PMID: 38918262.
2)
Gnekow AK, Kortmann RD, Pietsch T, Emser A. Low grade chiasmatic-hypothalamic glioma-carboplatin and vincristin chemotherapy effectively defers radiotherapy within a comprehensive treatment strategy—report from the multicenter treatment study for children and adolescents with a Low-grade glioma—HIT-LGG 1996—of the Society of Pediatric Oncology and Hematology (GPOH) Klin Padiatr. 2004;216(6):331–342.
3)
Chong AL, Pole JD, Scheinemann K, Hukin J, Tabori U, Huang A, Bouffet E, Bartels U. Optic pathway gliomas in adolescence–time to challenge treatment choices? Neuro Oncol. 2013 Mar;15(3):391-400. doi: 10.1093/neuonc/nos312. Epub 2013 Jan 7. PubMed PMID: 23295772; PubMed Central PMCID: PMC3578487.