Optic pathway and hypothalamic gliomas:

- 1% of brain tumours in general and 4% of brain tumours in children (75% occur in children).
- 10% confined to one optic nerve, 30% both optic nerves and 50-85% OCHG. When the tumours are restricted to the optic nerves, virtually all occur in the contest of NF1. 14-40 % of patients with NF1 develop OCHG
- Histologically the majority are benign **60% Pilocystic**, 40% fibrillary astrocytoma and ganglioglioma
- OCHG can have **variable unpredictable behaviour** from benign indolent hamartoma like growth to progressive growth leading to visual deterioration, endocrine dysfunction, neurological deficit and death in 20-30%
- Patients with NF1 have much better prognosis with the majority of tumours being indolent and asymptomatic
- Presentations: optic nerve tumours can present with progressive **visual loss and proptosis**. Patients with OCHG can present with progressive visual loss, **panhypopituitarism**, **diencephalic syndrome** "hypersomnia, obesity, anorexia, precocious puberty", **hydrocephalus**
- MRI -fusiform enlargement of the involved optic nerve with variable enhancement. OCHG are large solid and cystic tumours with variable enhancement localised on the suprasellar region. 5-12% may give CSF dissemination
- **Treatment options**:
  1. Asymptomatic patients with NF1 are followed by neuroimaging. No need for biopsy
  2. Optic nerve gliomas causing **visual loss and proptosis** are treated by surgical resection through fronto- orbital approach. The goal is to prevent intracranial extension and involvement of the chiasm or the other nerve.

3. OCHG is treated by surgical resection through **anterior interhemispheric translamina terminalis approach, Transcallosal approach, pterional and subfrontal approaches** for lateral and posterior extension. One should be careful to avoid injury to the optic nerves, chiasm, ACA, ICA, A COM, P COM arteries and the hypothalamic, diencephalic, and chiasmatic perforators

- Radiotherapy conventional and stereotactic for **residual and recurrent tumours in children older than 5 years of age** (behavioural and intellectual deficit, hypopituitarism, radiation necrosis, malignancy 4/30 developed high grade glioma, and Vasculopathy "Moyamoya variant".
- Chemotherapy results are not encouraging. It may stabilise the growth of the tumour temporarily in some Patients until they are old enough to tolerate radiotherapy. Currently it is given in the contest of clinical trials
- Differential diagnosis for OCHG: **craniopharyngioma, germinoma, teratoma, choroid carcinoma.**