Optic Nerve Glioma

- Optic pathway glioma (OPG) Primary neuroglial tumor of optic pathway
- 3 broad subtypes
  - Childhood syndromic [neurofibromatosis type 1 (NF1)], childhood sporadic, adult
- **Childhood lesions with NF1 (syndromic)**
  - Anterior pathways, unilateral or bilateral
  - Bilateral highly associated with NF1
  - 50% extend to chiasm, hypothalamus, and retrochiasmal optic pathways
  - Optic radiation involvement is rare
- **Childhood lesions without NF1 (sporadic)**
  - Affects chiasm and retrochiasmal segment predominantly
- **Adult lesions**
  - Unilateral ON with posterior extension

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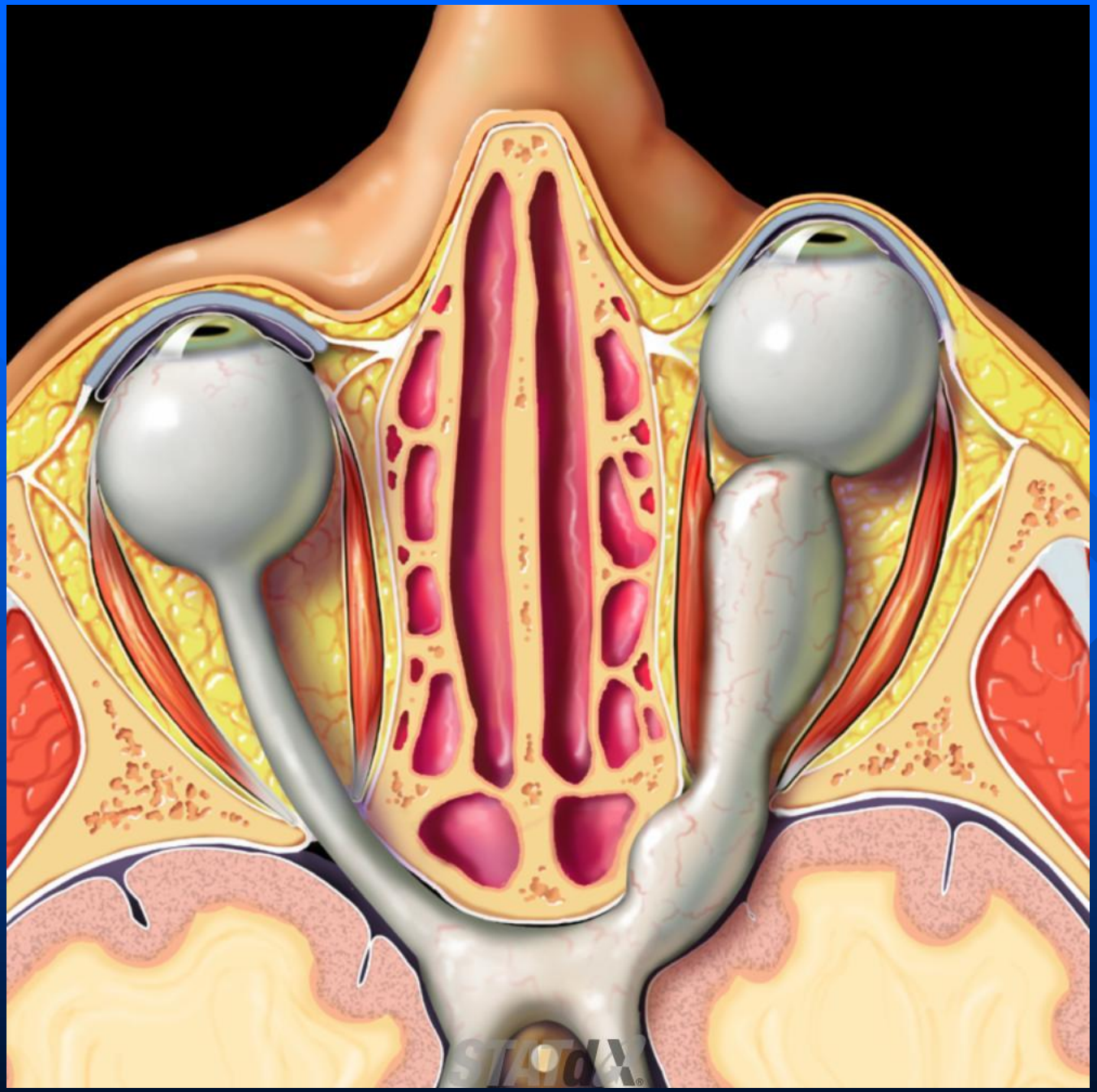
- Adult: Onset 20-80 years (mean: 50 years)
  - Aggressive course with rapid deterioration of vision
- Childhood benign lesions
  - 3% of orbital tumors; 5% of intracranial tumors
  - 30-40% of patients with OPG have NF1
  - 11-30% of patients with NF1 have OPG
  - Syndromic:
    - » Frequently asymptomatic; lesions detected on routine imaging
  - Sporadic: Larger, more aggressive

Image Interpretation Pearls

- Distinct subtypes have distinct features
  - **Childhood OPG with NF1**
    - » Anterior pathways; fusiform
    - » T2 peripheral hyperintense; mild enhancement
  - **Childhood OPG without NF1**
    - » Posterior pathways (chiasm); nodular
    - » Cystic changes; prominent enhancement
  - **Adult OPG**
    - » ON and sheath, posterior spread
    - » Similar to high-grade gliomas elsewhere

DDX:

- Optic Neuritis
  - Acute-onset pain and vision loss
  - Enhancing ON with minimal nerve enlargement
- ON Sheath Meningioma
  - Slow-onset proptosis and ↓ vision in adult
  - Perineural mass, may be calcified
- Idiopathic Orbital Inflammatory Pseudotumor
  - Painful proptosis, mass-like inflammation
  - Can involve any structure in orbit
  - Variable imaging appearance, including perineural enhancement
- Sarcoidosis
  - Systemic illness, orbital inflammation
  - Predilection for lacrimal gland
  - ON, orbital, and intracranial enhancement



Axial graphic depicts a left optic pathway glioma (OPG) extending along the length of the intraorbital nerve, through the enlarged optic canal, and into the prechiasmatic segment. The fusiform pattern of enlargement is typical.

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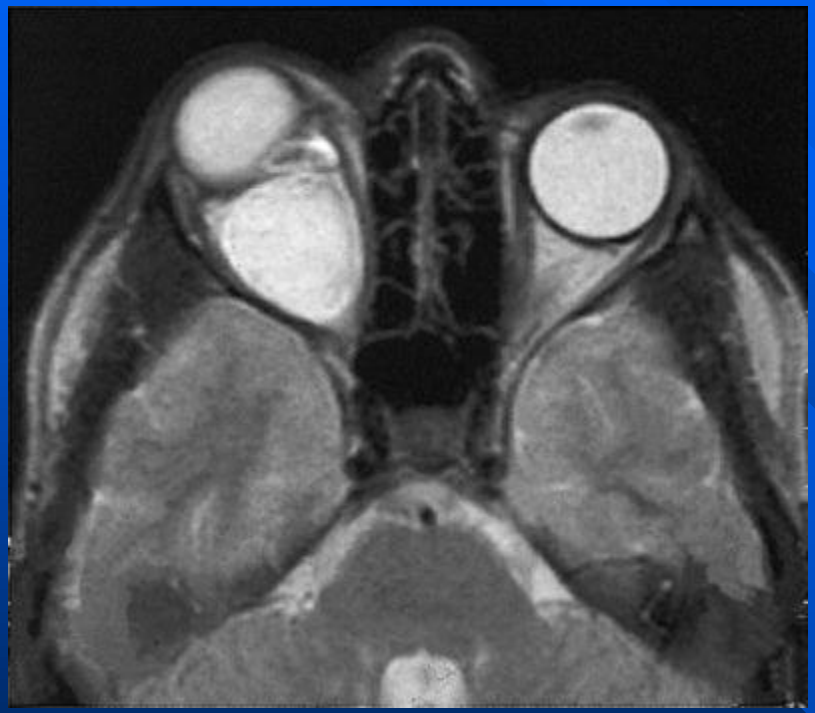
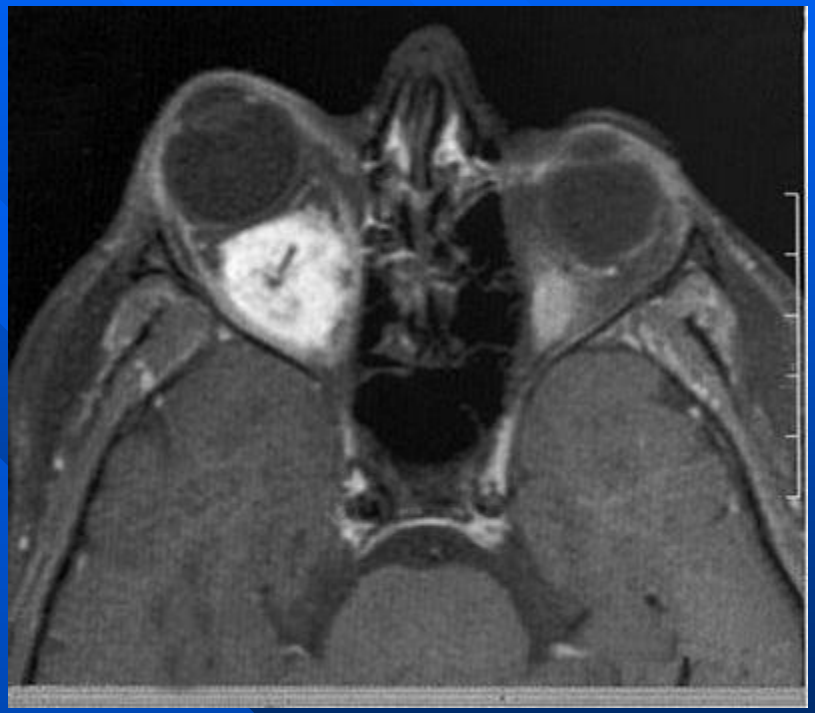
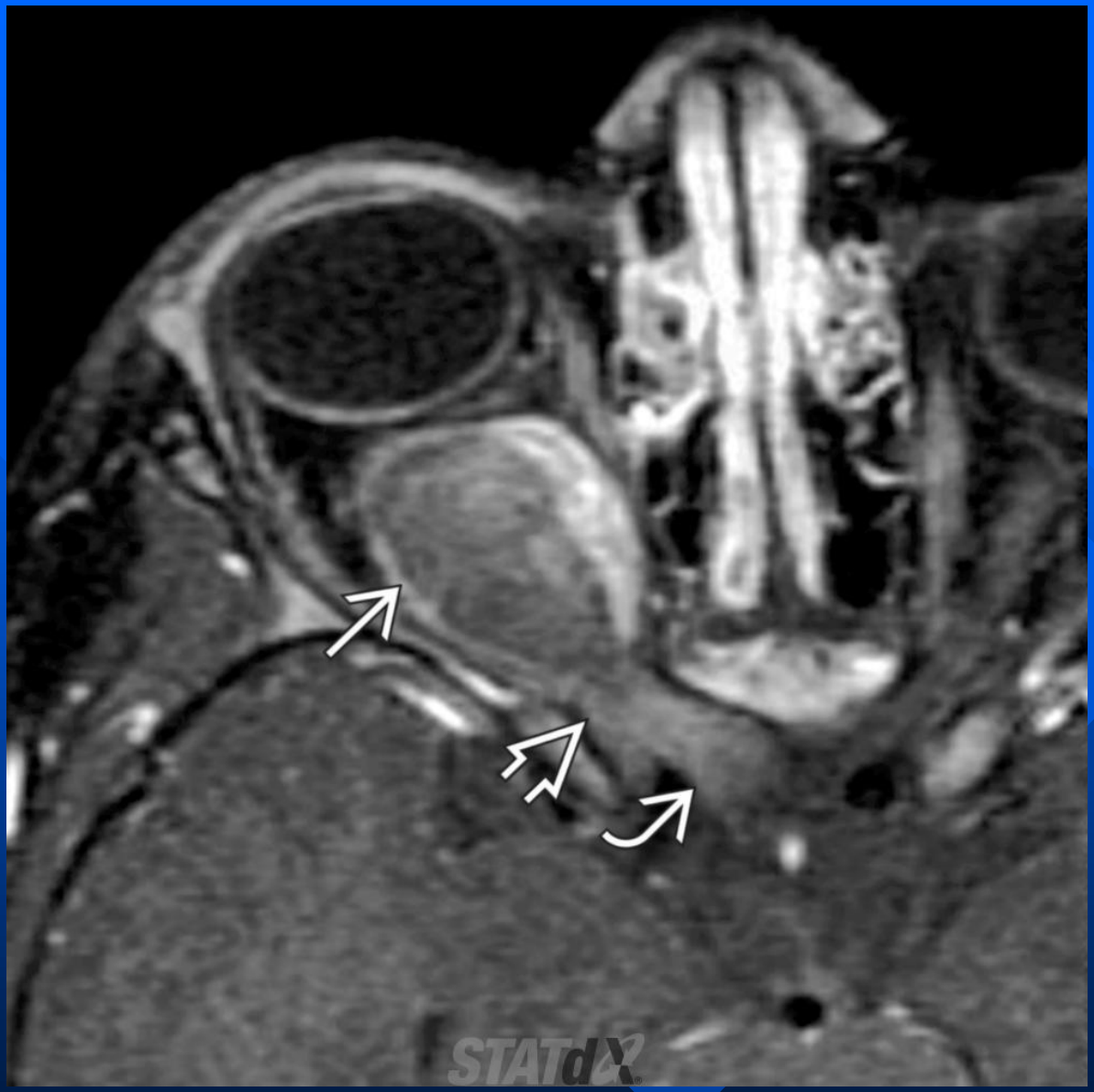
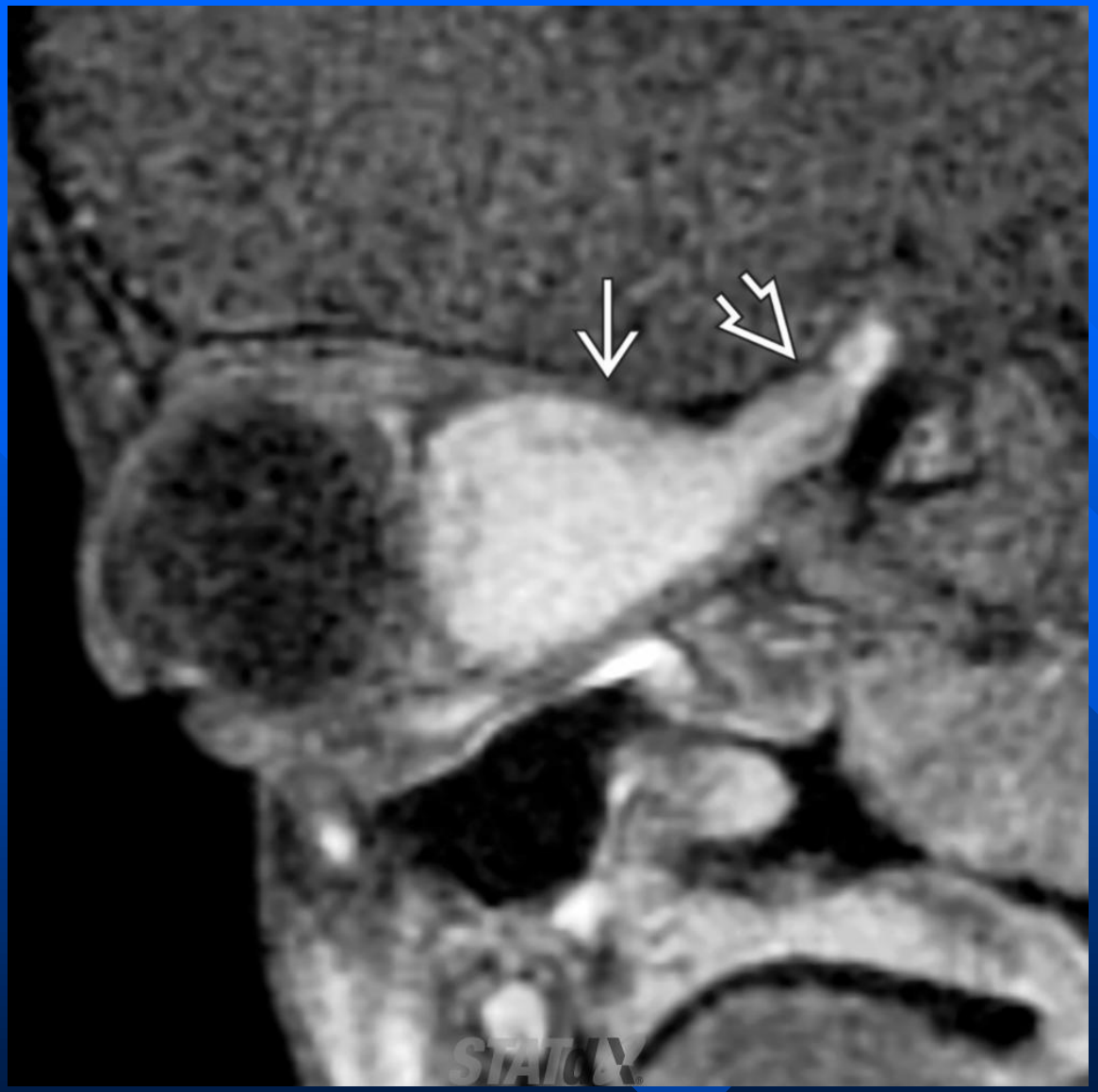




Differential diagnosis must be made with rhabdomyosarcoma and orbital pseudotumor and Lymphoma **(child)**



Axial T1WI C+ FS MR shows mild, patchy enhancement and marked fusiform enlargement of intraorbital (white solid arrow), intracanalicular (white open arrow), and prechiasmatic (white curved arrow) segments of right ON. Lesions in children with neurofibromatosis type 1 (NF1) may demonstrate only minimal contrast enhancement.



Oblique sagittal T1WI C+ FS MR reveals globular enlargement and intense homogeneous enhancement of intraorbital ON (white solid arrow) extending posteriorly into widened optic canal (white open arrow).

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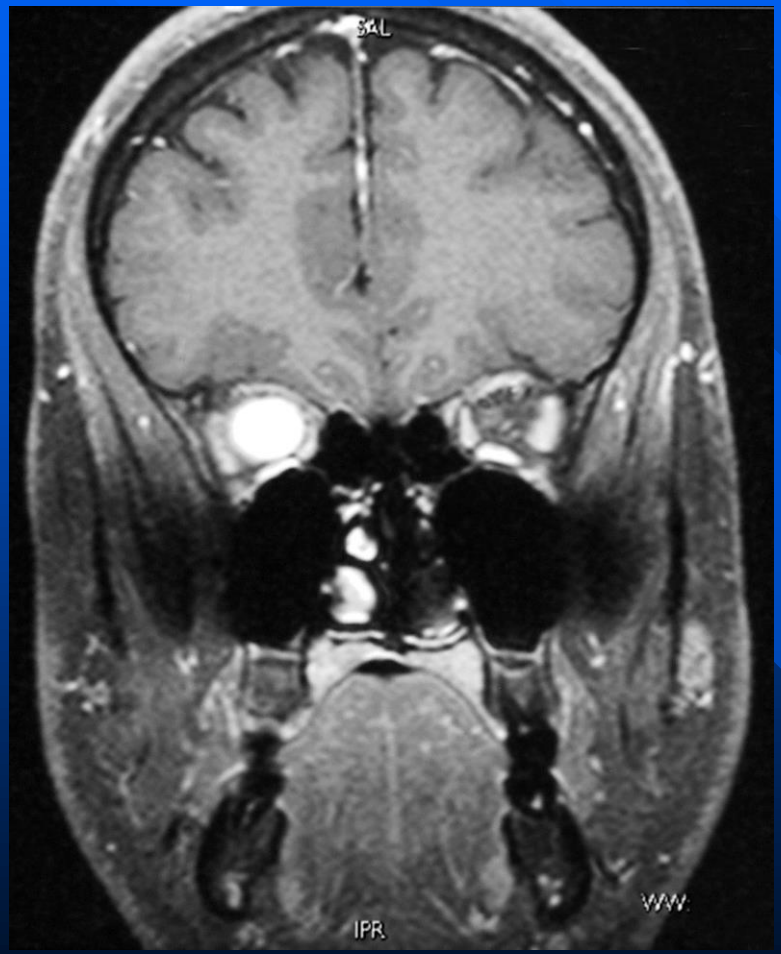

75% present < 10 yrs

Decreased vision

1/3 have NF

Look for intracranial optic pathway extension

Diff. Dx: Meningioma