

Neurofibromatosis Type 2

- Bilateral vestibular schwannomas
- Schwannomas of cranial nerves (CN) and spinal nerve roots
- Meningiomas on dural surfaces (up to 50%)
- Ependymomas in spinal cord and brainstem (6%)
- Use high-resolution T1 C+ FS MR through basal cisterns to evaluate cranial nerves

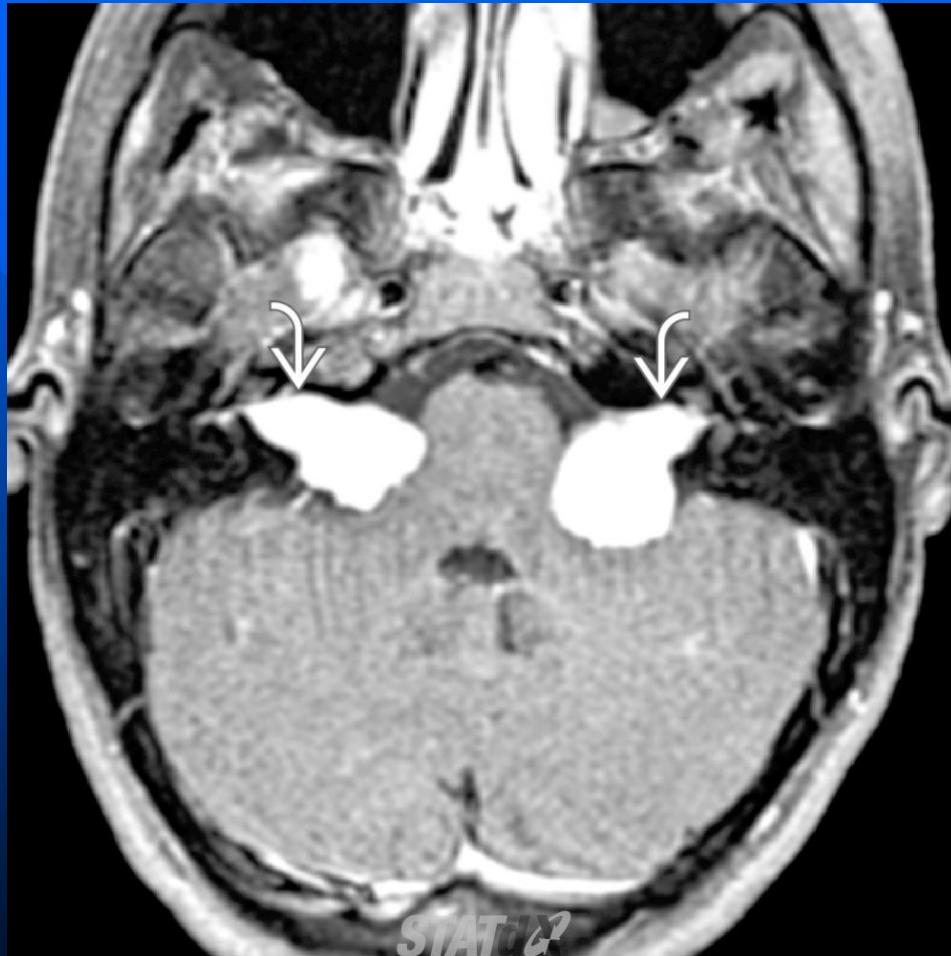
Clinical issues

- Usually presents between 2nd and 4th decade with hearing loss, \pm vertigo
- Incidence: 1:25,000-30,000
- Life span substantially shortened by presence of meningiomas and by complications related to lower cranial neuropathies (i.e., aspiration)

NF 2

- All NF2 families have chromosome 22q12 abnormalities
- NF2 gene encodes for merlin protein
- Diagnostic Checklist
 - Carefully evaluate other cranial nerves in any new diagnosis of schwannoma or meningioma in child/adolescent/young adult

Bilateral vestibular schwannomas



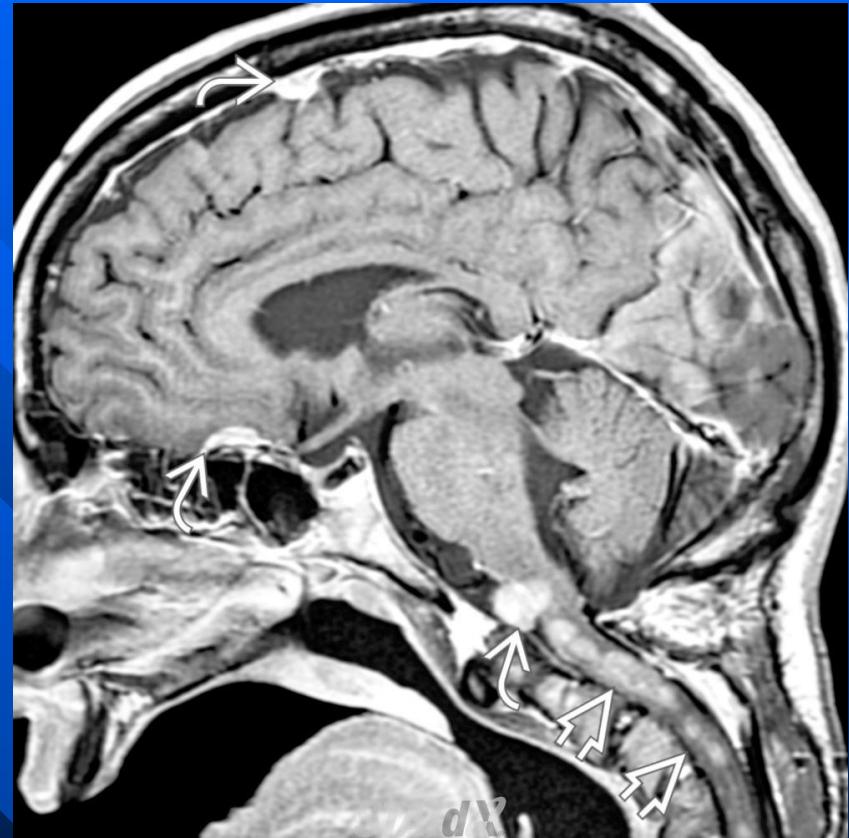
NF 2

- Coronal T1WI C+ MR shows the
- Dural-based meningiomas in the posterior fossa (white solid arrow) and along the falx and convexities (white open arrow). In this patient, the meningiomatosis predominates over the schwannomas.



NF 2

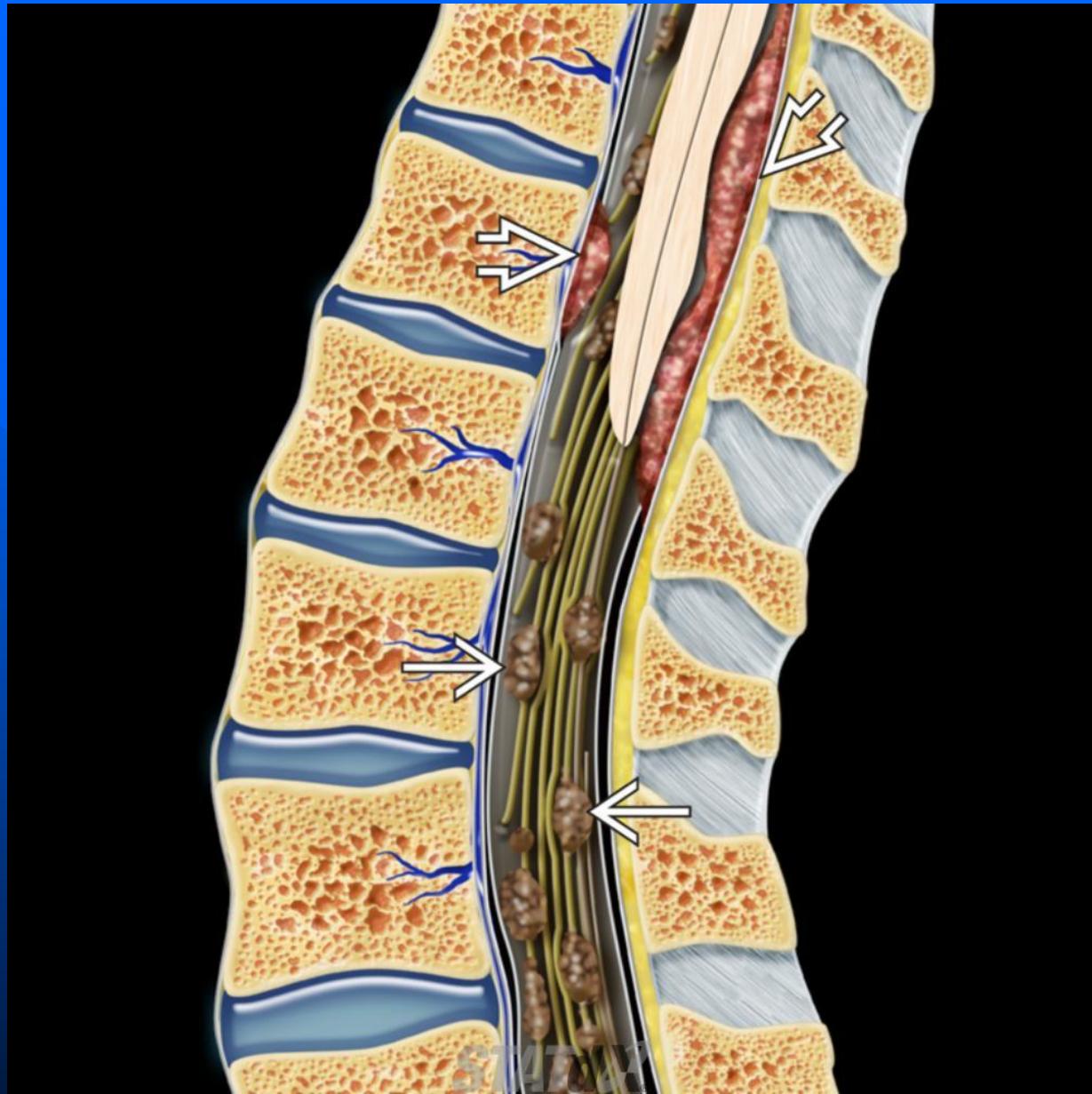
- Sagittal T1WI C+ MR in the same patient shows multiple enhancing meningiomas (white curved arrow) and enhancing ependymoma of the upper cervical cord (white open arrow). A "swan neck" deformity is noted in the upper neck, from prior multilevel laminectomies.



NF 2



Enhancement of right CN V (schwannoma) NFII



Sagittal graphic illustrates multiple rounded schwannomas (brown) (white solid arrow) along the cauda equina, as well as flat dural-based meningiomas (red) (white open arrow) impinging the conus.



Sagittal T1WI C+ MR shows multiple enhancing intramedullary masses throughout the cervical and thoracic cord due to ependymomas (white open arrow) in this patient with NF2. Note the enhancing meningioma at the foramen magnum (white solid arrow). Patient shows severe "swan neck" deformity as complication of prior multilevel laminectomies for tumor resection.