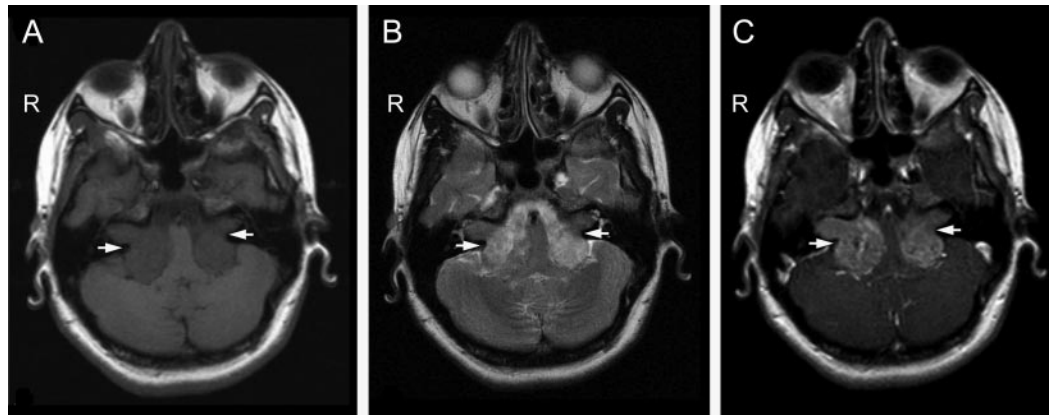


# Teaching NeuroImages: Vestibular schwannomas in neurofibromatosis type 2

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Figure Brain MRI



MRI brain (A-C) demonstrating bilateral vestibular schwannomas (acoustic neuromas, arrows) compressing the pontomedullary region. These extraaxial lesions (arrows) are hypointense on T1-weighted axial image (A), are hyperintense on T2-weighted axial image (B), and show enhancement after gadolinium administration on postcontrast T1-weighted image (C).

An 18-year-old woman with a history of juvenile cataract and hearing impairment presented with gait ataxia of 2 years' duration and demonstrated bilateral pyramidal signs in the legs. Brain MRI (figure) revealed extraaxial lesions in the cerebellopontine angles with brainstem compression. These bilateral vestibular schwannomas were pathognomonic of neurofibromatosis type 2 (NF2).<sup>1</sup> In addition, there were multiple spinal tumors that could also account for the pyramidal signs.

The estimated birth incidence of NF2 is 1 in 25,000.<sup>2</sup> Unlike neurofibromatosis type 1, these patients do not demonstrate extensive cutaneous findings. A positive family history along with the

presence of juvenile cataract and hearing impairment provide clinical clues for triaging patients for early neuroimaging to establish the diagnosis before the onset of brainstem compression.

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