Chiari-1 Malformation

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Chiari-I Malformation

Downbeat nystagmus (occasionally with a torsional component), worse on lateral gaze and with convergence
Divergence nystagmus
Convergence nystagmus
Chiari-I Malformation

Horizontal nystagmus (unidirectional, present with eyes in central position)
Periodic alternating nystagmus
Gaze-evoked nystagmus
Rebound nystagmus including torsional rebound
Seesaw nystagmus
Impaired pursuit (and VOR cancellation)
Impaired OKN
Strabismus, esotropia
Divergence paralysis
Skew deviation accentuated or alternating on lateral gaze

Clinical Features of Downbeat Nystagmus

Best evoked on looking down and laterally; often in association with horizontal gaze-evoked nystagmus, and so may appear oblique on lateral gaze.

Slow phases may have linear-, increasing- or decreasing-velocity waveforms

Poorly suppressed by fixation of a visual target
Clinical Features of Downbeat Nystagmus

May be precipitated or exacerbated or changed in direction, by altering head position, vigorous head-shaking (horizontal or vertical), or hyperventilation

Convergence may increase, suppress or convert to upbeat nystagmus

Associated with other signs of vestibulocerebellar involvement

Etiology of Downbeat Nystagmus

Cerebellar degeneration
Craniocervical anomalies, including Arnold-Chiari malformation
Infarction of brainstem or cerebellum
Rotational vertebral artery syndrome
Etiology of Downbeat Nystagmus

Dolichoectasia of the vertebrobasilar artery or compression of the vertebral artery
Multiple sclerosis
Cerebellar tumor, including hemangioblastoma
Etiology

Encephalitis
Head trauma
Increased intracranial pressure and hydrocephalus
Toxic-metabolic
  Anticonvulsant medication
  Lithium intoxication
Alcohol intoxication and induced cerebellar degeneration
Neuroimaging

**Figure 1:** Sagittal T1WI shows a classic Chiari I malformation with “peglike” tonsils extending inferiorly through the foramen magnum.

**Figure 2:** Sagittal T2WI shows exquisite detail of the low-lying tonsils. Note vertically-oriented cerebellar folia. There is no associated syrinx in this case.
Neuroimaging

Figure 3: Sagittal FLAIR shows no signal abnormality in either the tonsils or medulla

Courtesy of Anne Osborn, M.D.
References
