6.13.3. Childhood Nystagmus (III)

Treatment

Prisms

- to improve head position
  - if this technique improves head position, strabismus surgery is also likely to be effective
  - each prism is mounted with the apex pointing toward the null point
  - shifts the retinal image toward the null point
  - decreases the amount of head turn required
- to improve vision
  - base-out prisms are placed in front of both eyes
  - induces convergence
  - damps the nystagmus
  - improves vision
  - amounts determined by trial and error
- Fresnel press-on prisms
  - inexpensive plastic pieces that can be cut and then applied to glasses
  - powers ranging from 1Δ–40Δ
  - powers of 10Δ–20Δ are often required
- ground-in prisms
  - less distortion
  - preferred for patients who require smaller amounts of prism

Surgery for Nystagmus

- indications
  - shifting the null point closer to the primary position
  - to correct abnormal head position
  - increases foveation time
  - decreasing nystagmus intensity
  - to improve vision
- technique
  - conjugate recessions in each eye
  - Anderson procedure
  - Table 13-3 (Courtesy of American Academy of Ophthalmology)
  - recession-resection on both eyes
  - Kestenbaum procedure
  - recession of all 4 horizontal rectus muscles
- eyes are surgically rotated toward the direction of the head turn and away from the null point
  - eyes are surgically rotated to the left
    - recession of the right lateral and left medial rectus muscles
    - resection of the right medial and left lateral rectus muscles
  - left head turn and null point in right gaze
- amount of surgery
  - total amount of surgery for each eye (as measured in millimeters) is equal in order to rotate each globe an equal amount
  - 40% augmentation
    - head turns of 30°
  - 60% augmentation
    - turns of 45°
  - augmented procedures may cause restriction of motility
- vertical torticollis
  - chin-up or chin-down posturing
  - apex toward the null point
    - vertical prism
  - surgery on vertical rectus muscles or oblique muscles
    - eyes are rotated away from the null point
    - usually 8–10 mm of recession and resection of the vertical rectus muscles of each eye
    - combined weakening of a vertical rectus muscle and an oblique muscle in each eye
- head tilt
  - torsional surgery
    - oblique muscles
    - transposition of vertical rectus muscles
- nystagmus with no eccentric null point
  - Kestenbaum procedure is not indicated
  - 8–10 mm of recession of both medial rectus muscles and 10–12 mm of recession of both lateral rectus muscles
    - improved foveation time
    - effect on visual acuity is modest
    - average improvement of about 1 line
  - merely disinserting and reattaching horizontal rectus muscles produces similar results with presumably less risk of inducing new strabismus
  - nystagmus surgery in absence of abnormal head position is controversial
- nystagmus blockage syndrome
  - recession of medial rectus muscles
    - with amounts that are larger than normal for the amount of esotropia
    - posterior fixation sutures
- strabismus
  - surgery to shift the null point must be performed on the dominant fixating eye
  - surgery on the nondominant eye is then adjusted to account for the remaining strabismus

Acquired Nystagmus

Spasmus nutans

- idiopathic
- triad
  - small-amplitude, high-frequency (shimmering) nystagmus
    - binocular
    - asymmetric
    - may appear monocular
    - can vary with gaze position
    - can be horizontal, vertical, or rotary (torsional)
    - occasionally intermittent
  - head nodding
  - torticollis
- appear to be compensatory movements that maximize vision
- course
  - manifests during the first 2 years of life
  - benign disorder in most cases
  - abnormal head and eye movements to diminish by 3–4 years of age
- associated conditions
  - strabismus
  - amblyopia
  - developmental delay
- spasmus nutans–like nystagmus
  - chiasmal or suprachiasmal tumors
  - retinal dystrophies
  - congenital stationary night blindness
- diagnostic workup
  - neuroradiologic investigation is warranted with any evidence of optic nerve dysfunction
    - optic disc pallor
    - relative afferent pupillary defect
    - any sign of neurologic abnormality
  - some investigators believe that neuroimaging is indicated for all children with presumed spasmus nutans

See-saw nystagmus

- both vertical and torsional components
  - eyes make alternating movements of elevation and intorsion followed by depression and extorsion
    - high eye intorting
    - low eye extorting
- often associated with a lesion in the rostral midbrain or the suprasellar area
  - children
    - craniopharyngioma
- congenital form of see-saw nystagmus
  - seen with complete or partial achiasma
- diagnostic workup
  - confrontation visual fields
    - bitemporal defect
  - neuroradiologic evaluation is necessary

Vertical nystagmus

- congenital
  - uncommon
  - inherited retinal dystrophies
  - downbeat nystagmus
    - associated with good vision and normal neurologic findings
    - jerk nystagmus
    - downward fast component
    - null point in upgaze
- acquired
  - most common
  - secondary to
    - Arnold-Chiari malformation
    - medications
      - codeine
      - lithium
      - tranquilizers
      - anticonvulsants
  - neurologic evaluation is usually indicated

Monocular nystagmus

- oscillations are pendular, chiefly vertical, slow, variable in amplitude, and irregular in frequency
- severely amblyopic and blind eyes
- Heimann-Bielschowsky phenomenon

Nystagmus-Like Disorders

Convergence-Retraction Nystagmus

- etiology
  - congenital aqueductal stenosis
  - pinealoma
- part of the dorsal midbrain syndrome
  - paralysis of upward gaze
  - defective convergence
  - eyelid retraction
  - pupillary light–near dissociation
- nystagmus is best elicited on attempted upgaze saccades
  - tracking a downward-rotating OKN drum
  - co-contraction of all horizontal extraocular muscles occurs
    - retraction of the globe
    - medial rectus muscles overpower the lateral rectus muscles
      - eyes converge on attempted upgaze
- abnormal eye movements are actually saccades
  - not a true nystagmus!

Opsoclonus

- etiology
  - acute postinfectious cerebellar ataxia
  - viral encephalitis
  - paraneoplastic manifestation of neuroblastoma
- involuntary saccades
  - rapid and multidirectional
  - eye movements with very high frequency and large amplitude
  - can occur intermittently
  - not a true nystagmus!
- accompanied by somatic dyskinesias

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