5.13.1. Childhood Nystagmus (I)

- evaluation
  - history
    - transmission
      - sporadic
      - familial
        - autosomal dominant
        - autosomal recessive
        - x-linked
    - perinatal events
      - labor and delivery
      - maternal infections
      - prematurity
      - for children >3 months old
        - parental observations about
          - head tilts
          - head movements
          - gaze preference
          - viewing distances
    - associated ocular conditions
      - congenital cataract
      - congenital glaucoma
      - iridocorneal dysgenesis
      - Leber congenital amaurosis
      - retinopathy of prematurity
      - FEVR
      - Norrie disease
      - retinoblastoma
      - albinism
      - optic nerve hypoplasia
      - infectious diseases
  - ocular examination
    - visual acuity
      - patients with nystagmus and nearly normal visual acuity usually have congenital motor nystagmus
        - benign
      - markedly decreased visual acuity usually implies either retinal or optic nerve abnormalities
      - monocular visual acuity should be tested with fogging
        - monocular occlusion may increase nystagmus intensity!
      - binocular visual acuity should be measured at distance and near with any desired head position
      - near visual acuity is usually better than distance
    - preverbal children
      - optokinetic nystagmus (okn) drum
        - if vertical rotation elicits a vertical nystagmus superimposed on the child's underlying nystagmus, visual acuity is usually ≥20/400
      - preferential looking tests
      - Teller acuity cards II
        - in patients with a horizontal nystagmus, the response can be more easily observed with the card held vertically
    - pupils
      - sluggish or absent response to light
      - afferent defect
      - paradoxical pupil constriction in reaction to darkness
      - pupil responses can be normal in mild abnormalities
        - foveal hypoplasia
        - achromatopsia
        - primary motor nystagmus
      - anatomical structure
        - iris colobomas
          - suggest optic nerve or retinal colobomas
        - aniridia
          - foveal hypoplasia
        - iris transillumination
          - sign of albinism
          - foveal hypoplasia
      - optic nerve or retinal dysfunction
    - ocular motility
      - may have strabismus for several reasons
        - poor vision
        - damping of the nystagmus by converging
        - latent nystagmus secondary to strabismus
          - children with manifest latent nystagmus
            - often fixate with the preferred eye in adduction
            - turn their heads to cross-fixate
            - look to the right with the left eye, to the left with the right eye
    - fundus
      - optic nerve hypoplasia
      - foveal hypoplasia
      - fundus may appear normal or there may be only subtle retinal pigmentary changes
        - differential diagnosis of decreased vision with minimal fundus changes
          - electrophysiologic testing may be necessary
          - electrophysiologic testing may be necessary
          - Leber congenital amaurosis
          - achromatopsia
          - blue-cone monochromatism
          - congenital stationary night blindness
          - foveal hypoplasia

- general features
  - prevalence in young children
    - 0.35%
  - involuntary, rhythmic oscillation of the eyes
  - classification
    - symmetry
      - pendular nystagmus
        - equal velocity in each direction
      - jerk nystagmus
        - unequal velocity in different directions
        - fast component defines the direction of the nystagmus
    - direction
      - horizontal
      - vertical
      - torsional
      - combination
    - frequency
    - amplitude
    - intensity
  - characteristics of nystagmus may change with gaze direction
    - pendular nystagmus can become jerk nystagmus on extreme gaze
    - gaze position can affect amplitude and frequency (intensity) of nystagmus
      - especially true of jerk nystagmus
        - can decrease in intensity with gaze opposite the fast-phase component
        - null point
          - gaze position in which intensity of oscillations is diminished and visual acuity improves
          - abnormal head position
  - nomenclature
    - committee for the classification of eye movement abnormalities and strabismus (CEMAS)

- PAN
  - dysfunction of the cerebellar nodulus and uvula
  - Arnold-Chiari type I malformation
- Conv-retraction syndrome
  - etiology
    - congenital aqueductal stenosis
    - pinealoma

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