6.12.4. Special Forms of Strabismus (IV)

Chronic Progressive External Ophthalmoplegia (CPEO)

- mitochondrial cytopathy
- rare
- heredity
  - sporadic
  - familial
- begins in childhood
- ptosis
- slowly progresses to total paralysis of the eyelids and EOMs
- clinical features
  - constricted visual fields
  - electrodiagnostic abnormalities
  - diagnosis
    - muscle biopsy show ragged red fibers
    - specific alterations of mitochondrial DNA
  - Kearns-Sayre syndrome
    - retinal pigmentary changes
    - CPEO
    - cardiomyopathy (especially heart block)
      - life-threatening arrhythmias can occur
  - true pigmentary retinal dystrophy is usually absent
- management
  - cardiac status evaluation
  - treatment options for the ocular motility disorder are limited
    - high rate of long-term undercorrections
  - cautious surgical elevation (suspension) of the upper eyelids can lessen a severe chin-up head posture

myasthenia gravis

- introduction
  - pathogenesis
    - antibodies directed against acetylcholine receptors
  - transient neonatal form
    - caused by the placental transfer of acetylcholine receptor antibodies of mothers with myasthenia gravis
    - usually subsides rapidly
  - onset in childhood is uncommon
  - a purely ocular
  - a part of a systemic disorder
    - less common in childhood-onset than in the adult-onset form
  - myasthenia gravis foundation of america, inc
- clinical features
  - weakening of EOMs, including levator muscle
    - most cases (90%) exhibit both ptosis and limited ocular rotations
  - ocular signs can resemble any unilateral or bilateral ophthalmoplegia
  - affected muscles fatigue rapidly
    - ptosis typically worsens when the patient looks upward x 30 seconds
    - sleep test
      - patient rests in a dark room with the eyelids closed x 20–30 minutes
        - ptosis often subsequently resolves
    - external application of ice over the involved eyelids x 2–5 minutes
      - improves function of levator palpebrae superioris and other affected EOMs
  - Cogan lid-twitch sign
    - overshoot of the eyelid when the patient looks straight ahead after looking down for several minutes
    - highly suggestive
  - pharmacologic tests
    - edrophonium test
      -
      - Figure 12-8 (Courtesy of American Academy of Ophthalmology)
    - neostigmine test
      - effect of this agent begins later than that of edrophonium and is prolonged
      - allows more time to measure changes in alignment
  - electromyography
    - decreased electrical activity of involved muscles after prolonged voluntary innervations
    - increased activity (including faster saccadic velocity) after administration of edrophonium or neostigmine
    - single-fiber electromyography
  - antibodies
    - circulating anti–acetylcholine receptor antibody
    - anti–muscle-specific kinase antibody
    - negative test result does not rule out MG
- differential diagnosis of ptosis and extraocular muscle involvement
  - thyroid eye disease
  - chronic progressive external ophthalmoplegia
  - myasthenia gravis
- management
  - adults
    - ocular manifestations are frequently resistant to usual systemic myasthenia treatment
  - children
    - often successfully managed with pyridostigmine alone
  - in adults and children in whom the ocular deviation has stabilized, standard eye muscle surgery can help restore binocular function in at least some gaze positions
  - ptosis occasionally requires upper eyelid surgery

esotropia and hypotropia associated with high myopia

- highly myopic patients
  - elongated globe herniates between the superior and lateral rectus muscles
    - stretching and dehiscence of intermuscular septum between these 2 muscles
- pathogenesis
  - inferior slippage of the lateral rectus pulley
  - medial displacement of the inferior rectus
  - medial rectus is often tight
- progressively worsening hypotropia and esotropia
- treatment
  - joining of superior and lateral rectus muscles, usually with a nonabsorbable suture, to reposition the globe
  - a recession of medial rectus if muscle is tight

internuclear ophthalmoplegia (INO)

- medial longitudinal fasciculus (MLF)
  - integrates the nuclei of the cranial nerves governing ocular motility
  - has major connections with vestibular nuclei
  - intact MLF is essential for production of conjugate eye movements
- etiology
  - demyelinating disease
  - cerebrovascular accidents
  - brain tumors
- clinical features
  - eye ipsilateral to the MLF lesion adducts slowly and incompletely or not at all
  - both eyes adduct normally on convergence
  - abducting eye exhibits a characteristic horizontal jerk nystagmus
  - exotropia
  - a skew deviation
  - management
    - persistent exotropia
      - medial rectus muscle resection + unilateral or contralateral lateral rectus muscle recession
        - can help eliminate diplopia, particularly in bilateral cases

strabismus associated with other ocular surgery

- refractive surgery
  - can result in dissimilar sensory input to the 2 eyes
    - to create monovision
  - can be sufficient to cause disruption of fusion in predisposed patients
- surgery for retinal detachment
  - scarring from dissection of the EOMs and the application of devices (scleral buckle)
  - corrective surgery can be extremely difficult, especially if these elements must be removed
- macular translocation surgery can cause torsional diplopia
- glaucoma drainage devices
  - may require removal or relocation or substitution of the device
- retrobulbar injections
  - direct injury to the muscle
  - toxic effect of the injected material
  - vertical rectus muscles are most vulnerable
- injection of botulinum toxin into eyelids
  - transient paralyzing effect
- pterygium removal or endoscopic sinus surgery
  - laceration or inadvertent excision of an entire section of the medial rectus muscle

superior oblique myokymia

- rare
- etiology
  - aberrant regeneration of fourth cranial nerve fibers
  - vascular compression of the nerve
- clinical features
  - abnormal torsional movements of the eye that cause diplopia and monocular oscillopsia
  - otherwise neurologically normal
  - recurrences may persist indefinitely
- management
  - treatment is not necessary if the patient is not disturbed by the visual symptoms
  - systemic medications
    - first-line treatment
      - carbamazepine
      - phenytoin
      - propranolol
      - baclofen
      - gabapentin
      - topical timolol
    - some patients will benefit, at least in the short term
  - surgical treatment
    - disconnection of superior oblique muscle from globe by generous tenectomy
      - results in a superior oblique palsy
        - some surgeons perform simultaneous inferior oblique muscle weakening

ocular motor apraxia

- introduction
  - "saccadic initiation failure"
  - supranuclear disorder
    - rare
  - etiology
    - congenital form
      - most commonly autosomal dominant
      - a familial
    - premature birth
    - developmental delay
    - bilateral lesions of the frontoparietal cortex
    - agenesis of the corpus callosum
    - hydrocephalus
    - Joubert syndrome
      - abnormal eye movements
      - developmental delay
      - microcephaly
      - hypoplasia of the cerebellar vermis
      - retinal dysplasia
    - type 3 Gaucher disease
    - ataxia-telangiectasia
    - mass lesions of the cerebellum that compress the rostral part of the brainstem
- clinical features
  - normal voluntary horizontal saccades cannot be generated
  - changes in horizontal fixation are accomplished by a head thrust that overshoots the target
    - initial thrust serves to break fixation
      - followed by a rotation of the head back in the opposite direction once fixation is established
      - + associated blink
    - head thrust may improve in late childhood
  - vertical saccades and random eye movements are intact
  - horizontal vestibular and optokinetic nystagmus are impaired
- differential diagnosis (acquired type)
  - conditions that affect the generation of voluntary saccades
    - metabolic and degenerative diseases
      - Huntington chorea
- management
  - neurodevelopmental evaluation and imaging of the brain are advisable

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