6.12.3. Special Forms of Strabismus (III)

Thyroid Eye Disease

- introduction
  - lymphocytic infiltration of the EOMs
    - Edema
    - inflammation
    - fibrosis
    - restrict motility
  - thyroid-stimulating immunoglobulins (TSIs) mediate thyroid eye disease
  - some patients also have myasthenia gravis
  - association between severity of thyroid eye disease and smoking
    - hazard ratio for strabismus surgery is ≈ double in patients with thyroid disease who smoke
- clinical features
  - inferior rectus>medial rectus>superior rectus>lateral rectus
    - hypotropia
      - Figure 12-7 (Courtesy of American Academy of Ophthalmology)
      - common cause of acquired vertical deviation in adults
        - especially women
        - rare in children
    - esotropia
      - often asymmetric
        - most often bilateral
  - positive forced-duction test
  - upper eyelid retraction
  - proptosis
- management
  - botulinum toxin
    - may reduce the severity of fibrosis when injected into tight muscles in the acute phase
  - prisms
    - may alleviate diplopia
  - indications for strabismus surgery
    - diplopia
    - abnormal head position
  - surgery rarely restores normal motility
  - 1/2 patients who had surgery before stability was achieved required further surgery
  - recession of the affected tight muscles in 1 or both eyes is the preferred surgical treatment
    - slight initial undercorrection is desirable
      - late progressive overcorrection is common, especially with large inferior rectus muscle recessions
      - unlike XT repair
  - strengthening procedures usually worsen restriction
  - adjustable sutures can be helpful in these difficult cases
  - limited depression of the eyes after inferior rectus muscle recessions can interfere with patients' bifocal use
  - proptosis can become worse after EOM recessions
  - postpone strabismus surgery until decompression has been accomplished
  - perform surgery after strabismus measurements and thyroid function tests have been stable for ≥6-12 months
  - postpone eyelid surgery until the strabismus is treated
    - upper eyelid retraction may improve once the patient no longer strains to elevate the eye
  - large recessions of very tight inferior rectus muscles can cause lower eyelid retraction
    - severe lower eyelid retractors as part of strabismus surgery
    - place spacer of banked sclera or synthetic material to vertically lengthen lower eyelid tarsus

Sixth Nerve Palsy

- less common in children than in adults

Brown Syndrome

- restriction of elevation in adduction
- introduction
  - pathogenesis
    - abnormalities of the superior oblique tendon–trochlea complex
    - structural problems remote from superior oblique tendon, including instability of lateral rectus pulley, can present an identical clinical picture
      - pseudo–Brown syndrome
  - types
    - congenital
      - most cases
    - acquired
      - trauma in region of trochlea
      - iatrogenic causes
        - scleral buckles
        - glaucoma drainage devices
      - orbital tumors
      - systemic inflammatory conditions
        - often result in intermittent Brown syndrome
          - may resolve spontaneously
      - sinusitis
        - patients with acute-onset presentation of Brown syndrome of undetermined cause should undergo imaging of the orbits and paranasal sinuses
  - bilateral in ≤ 10%
  - spontaneous improvement in 75%
    - often over many years
- clinical features
  - deficient elevation in adduction
    - improves in abduction but often not completely
    - Figure 12-5 (Courtesy of American Academy of Ophthalmology)
  - positive forced-duction test demonstrating restricted passive elevation in adduction is essential for diagnosis
    - retropulsion of globe during this test stretches the superior oblique tendon and accentuates the restriction
      - in restrictions involving inferior rectus muscle the limitation of passive elevation is accentuated by forceps-induced proptosis of the eye rather than by retropulsion
  - attempts at elevation straight upward usually cause divergence ('V pattern)
    - inferior oblique muscle paralysis usually exhibits an A pattern
  - widening of palpebral fissure in adduction
    - unlike Duane retraction syndrome
  - hypotropia in primary position
    - in severe cases
    - chin-up head posture or head turn away from affected eye
  - downshoot of involved eye on gaze to opposite side (overdepression in adduction)
    - in severe cases
    - in true superior oblique muscle overaction downshoot occurs less abruptly as adduction is increased
- management
  - mild congenital cases
    - observation
  - cases secondary to rheumatoid arthritis or other systemic inflammatory diseases
    - systemic treatment of underlying disease
    - corticosteroids injection near trochlea
  - more severe congenital cases
    - ipsilateral superior oblique tenotomy nasal to the superior rectus muscle
      - complications
        - iatrogenic superior oblique muscle paresis
          - prevention
            - careful handling of intermuscular septum
            - avoidance of complete tenotomy as a sole procedure
              - insertion of an inert spacer or suture between the cut ends of the superior oblique tendon
              - Z-plasty of tendon on the medial side
              - partial (80%) tenectomy of the posterior portion of the tendon on the temporal side
            - some surgeons perform simultaneous ipsilateral inferior oblique muscle weakening
        - downgaze restriction due to adhesions
          - repair the intermuscular septum to prevent contact of spacer or suture with nearby structures

Third Nerve Palsy

- etiology
  - children
    - congenital
      - 40%–50%
    - trauma
    - inflammation
    - viral infection
    - ophthalmoplegic migraine
    - after vaccination
    - neoplastic lesion
  - adults
    - intracranial aneurysm
    - microvascular infarction
    - inflammation
    - trauma
    - infection
    - tumor
- clinical features
  - ischemic (microvascular) causes
    - a pupil-sparing
  - compressive lesions
    - cause mydriasis
  - complete paralysis
    - limited adduction, elevation, and depression of the eye
      - causing exotropia and often hypotropia
      - adults report incapacitating diplopia
        - incidence of diplopia in patients <8 years is low
      - amblyopia is common in children
      - Figure 12-6 (Courtesy of American Academy of Ophthalmology)
    - upper eyelid ptosis
      - often with pseudoptosis
  - misdirection (aberrant regeneration) of the damaged nerve
    - anomalous eyelid elevation, pupil constriction, or vertical excursion of the globe
    - on attempted rotation into field of action of EOMs supplied by injured nerve
  - mistic pupil sometimes seen in congenital cases
- management
  - wait all months (even up to 12 months) for spontaneous recovery
    - except in congenital cases
  - patients with at least partial recovery have much better functional and cosmetic results
  - prism adaptation testing
    - adult patients who can achieve single binocular vision with prisms of any power before surgery are most likely to do well
  - goal of surgery
    - adequate alignment for binocular function in primary position and in slight downgaze for reading
  - incomplete paralysis
    - large recession-resection of the horizontal rectus muscles to correct the esodeviation
    - isoplacement of both to correct the hypotropia
  - complete paralysis
    - large lateral rectus muscle recession + fixation of the globe to nasal orbital periosteum
    - disinsertion of lateral rectus muscle and reattachment to lateral orbital periosteum can maximize muscle inactivation
  - hypotropia
    - concurrent superior oblique tenotomy
    - transfer of superior oblique tendon to upper nasal quadrant of globe
      - anomalous eye movements can result from this procedure
  - reserve correction of ptosis for a subsequent procedure

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