7.11.7. Facial Dystonia

Hemifacial Spasm (HFS)

- intermittent synchronous gross contractures of the entire side of the face
  - begins in periocular region and progresses to involve entire face
- etiology
  - vascular compression of the facial nerve at the brain stem
  - other cerebellopontine angle lesions
    - <1%
- rarely bilateral
  - unlike BEB
- spasms are present during sleep
  - unlike BEB
- associated with ipsilateral facial nerve weakness
- MRI
  - documents the ectatic vessel
  - rules out other cerebellopontine angle lesions
- treatment
  - neurosurgical decompression of the facial nerve
    - exposes patient to relative risk of neurosurgical intervention
  - periodic injection of botulinum toxin
  - oral medications
    - carbamazepine
    - clonazepam
    - low efficacy
- differential diagnosis
  - aberrant regeneration after facial nerve palsy
    - history
      - previous Bell palsy
      - trauma
    - hemifacial contracture
    - aberrant synkinetic facial movements
    - respond well to botulinum toxin injection at very low doses

Benign Essential Blepharospasm (BEB)

- General
  - epidemiology
    - >40 years
    - F>M
    - 30/100,000 people
  - pathogenesis
    - basal ganglia disease
  - clinical presentation
    - bilateral
      - unlike HFS
    - increased blinking
    - involuntary spasms of the periocular protractor muscles
      - start as mild twitches
        - progress to forceful contractures
    - other muscles of the face may also be involved
    - spasms typically abate during sleep
      - unlike HFS
    - may severely limit patient's ability to perform activities of daily living
    - can progress until the patient is functionally blind
  - diagnostic workup
    - clinical diagnosis
    - neuroimaging is generally unrevealing and rarely indicated
  - differential diagnosis
    - reflex blepharospasm
      - dry eye syndrome
    - hemifacial spasm
  - medical treatment
    - eyelid crutches
      - attached to eyeglass frames
    - oral medications
      - limited usefulness
    - neurotoxin injections
      - primary treatment for blepharospasm
  - references
- Botulinum toxin injection
  - treatment of choice for BEB
  - botulinum toxin type A
  - improvement is temporary
    - onset of action
      - 2–3 days
    - average peak effect
      - 7–10 days
    - duration of effect
      - 3–4 months
  - complications
    - bruising
    - blepharoptosis
    - ectropion
    - epiphora
    - diplopia
    - lagophthalmos
    - corneal exposure
  - references
  -
    Figure 11-20 (© 2020 American Academy of Ophthalmology)
- Surgical myectomy
  - indications
    - poorly responsive to botulinum therapy
    - incapacitated by spasms
  - technique
    - meticulous removal of orbital and palpebral orbicularis muscle in upper (and sometimes lower) eyelids
  - complications
    - lagophthalmos
    - chronic lymphedema
    - recurrence of spasm
    - periorbital contour deformities
  - more limited myectomy
    - helpful in patients with less severe disease
    - may result in improved responsiveness to botulinum toxin therapy
  - many patients with BEB have an associated dry eye
    - may be aggravated by any treatment modality that decreases eyelid closure
    - artificial tears
    - ointments
    - punctal plugs or occlusion
    - moisture chamber shields
    - tinted spectacle lenses
    - minimize discomfort
- surgical ablation of the facial nerve
  - has been largely abandoned
    - recurrence rates ≤ 30%
    - frequent hemifacial paralysis from facial nerve dissection
  - microsurgical ablation of selected branches of the facial nerve
    - some surgeons have had greater success
- muscle relaxants and sedatives
  - orphenadrine, lorazepam, or clonazepam
  - rarely effective in primary treatment of BEB
    - sometimes prolong interval between botulinum toxin injections in mild BEB
  - dampen lower facial dystonia (Meige syndrome) associated with BEB
  - psychotherapy
    - little or no value

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