Ocular Myasthenia Gravis

Clinical Features

- Triad: Ptosis, Oculomotor paresis and Obicularis oculi weakness
    - **Ptosis** may be unilateral or bilateral
        - Alternating ptosis is specific for this condition
        - **Eyelid fatigue**
            - Ptosis worsens with prolonged upgaze or upon return to primary gaze
        - **Cogan's lid twitch**
            - After brief but sustained downgaze, a saccade to primary gaze results in the eyelid quickly rising and falling (1 mm or more typically)
        - **Eyelid Curtaining**
            - When lifting the more ptotic eyelid, the contralateral eyelid will droop more because of Hering's law
        - Resting for 5 minutes with the eyes closed will cause improvement in ptosis
    - Strabismus that fluctuates or appears fatiguable
        - Any type of strabismus possible
        - prolonged or sustained gaze in the field of action of the affected muscle may show increasing paresis
    - Lagophthalmos is rare
        - attempt to open the eyelid against forced closue- if the eyelid can be opened it suggest obicularis weakness

Diagnostic Tests

Antibody Tests

- **Acetylcholine Receptor Antibody**
    - sensitivity 50% in Ocular myasthenia vs 90% in Generalized myasthenia
    - The most specific test, no false positives have been reported
- Muscle specific tyrosine kinase antibodies (MuSK)
    - Rare cares of this antibody present in the setting of ocular myathenia without acetylcholine receptor antibodies
- LRP4 antibodies
    - 3 cases reported these present in setting of ocular myasthenia

Other tests

- **Ice Pack Test**
    - Place a bag of ice on the ptotic lid for 1 minute

    - Immediately asses ptosis when ice removed
    - improvement is transient (<1 min)
    - sensitivity 80% if prominent ptosis present
- **Tensilon test**
    - Edrophonium- inhibits acetylcholinesterase
    - transiently reverses muscle weakness
    - Sensitivity 85-95%
- Electrophysiology
    - Repetitive nerve stimlation- reduction in amplitudes with repetitive stimulation
    - Single-fiber EMG- temoral variability of adjacent motor nerve fibers known as “jitter”.