Infectious Diseases of the Motor Neuron

Zakia Bell, M.D.
Associate Professor of Neurology and Physical Medicine & Rehabilitation
Virginia Commonwealth University
School of Medicine

• Poliomyelitis
  – rare vaccination associated polio
  • post polio syndrome:
    – progressive fatigue, pain and weakness occurring in survivors of the polio infection 30 or more years after the original infection

• Enteroviral infection:
  – segmental infection with:
    • varicella-zoster virus
    • rabies
      – may result in motor neuron injury

Muscles Commonly Weakened by Polio

Gait Patterns of Polio Patients

Post-Polio Syndrome
**Paralytic Polio**
- Caused by a single-stranded RNA enterovirus
- Less than 10% of those infected will have any neurologic findings
- Virus infects and destroys anterior horn cells
- Virus infects through respiratory secretions and fecal contamination

**Amyotrophic Lateral Sclerosis (ALS)**
- Amyotrophic lateral sclerosis (ALS), also known as Lou Gehrig’s disease, causes degeneration of nerve cells connected to the brain that control voluntary motor functions

**Anterior Horn**

**ALS**
- Bulbar signs
  - resulting from brain stem involvement
- Spinal signs
  - resulting from spinal cord involvement
- Mixed signs
  - resulting from brain stem and spinal cord involvement

**Adult Onset Diseases**
- Amyotrophic Lateral Sclerosis (ALS) affects:
  - lower motor neuron
    - producing weakness and atrophy
  - upper motor neuron
    - resulting in spasticity

- Progressive weakness starting in one limb to eventually involve all
- Bulbar weakness may result in difficulty speaking swallowing (progressive bulbar palsy)
  - at risk of aspiration and early respiratory failure
  - 5-10% are genetically transmitted as autosomal dominant
  - fasciculations are helpful clinical signs
Anterior Horn

Spinal Muscular Atrophy (SMA)

SMA Type-I
- Frog-leg positioning
- Bell-shaped deformity
  - narrow thorax
  - pectus excavatum
  - rib flaring
- Feeding difficulties

SMA
- Type I also known as Werdnig-Hoffman or progressive infantile SMA
- Congenital loss of the anterior horn cells
- In most cases, it is an autosomal recessive defect on 5q11
- Usually presents in first 6 months
  - can have symptoms in utero

Child Onset Diseases
- Spinal muscular atrophy(s)
  - most inherited as autosomal recessive traits
  - differ in age of onset and severity of weakness
  - weakness and atrophy of muscles results from impaired function and death of the motor neuron

Child Onset Diseases
- Type I Werdnig Hoffman:
  - presents in infancy as severe hypotonia (floppy baby)
  - marked wasting, lack of spontaneous movement evident at age 6 months
Child Onset Diseases

- Type II:
  - less severe
  - children may sit independently
  - survival variable
- Type III Kugelberg-Welander disease:
  - much less severe
  - may show no obvious symptoms until first decade; prolonged survival the rule

Tongue Fasciculations

Wasting of Hand Muscles