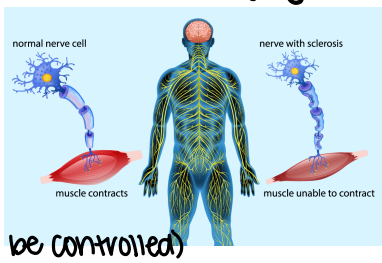
Amyotrophic lateral sclerosis (ALS | Lou Gehrigs): progressive & fatal neurologic disorder
- Path: degeneration of nerves in motor neurons of spinal cord & brain stem
  → results in muscle weakness & wasting
  → cause unknown
- assessment: progressive muscle weakness
  - wasting of extremeties & trunk develop
  - muscle fasciculations (twitching)
  - If affects brain stem: difficulty speaking & swallowing, periods of inappropriate laughter/crying, resp failure & total paralysis (3-5 years after diagnosed)
- diagnostics: no specific diagnostic (Sx) tests available - rules out other cause
  - electromyography: validates weakness in affected muscles
- medical management: no specific tx & death occurs several years after diagnosis
  - Riluzole: slows progression ALS & delays need for tracheostomy
- nursing management:
  - early: assistance ADLs - maintain patent airway!
  - late: dependence



Movement disorders - disease of extrapyramidal motor system (extrapyramidal - involuntary muscle movements that cant be controlled)

Parkinsons: deficency of neurotransmitter dopamine
→ prevelance ↑ w/ age (>50)
- cause: unknown; no cause for dopamine depletion
  - exposure to enviormental toxins such as insecticides, herbicides
  - self-administration of illegal synthetic form of heroin (MPTP)
  - sequelae of head injuries & encephalitis
  - phenothiazines (category antipsychotics)
- assessment: 4 cardinal features
  ① Tremor (pill-rolling, pronation)
  ② Rigidity (stiffness)
  ③ Akinesia (bradykinesia)
  ④ Postural instability (stooped posture)
  - others:
    - hypophonia (low volume speech)
    - depression, dementia
    - unintentional weight loss - tremors expend energy (problem for elderly & frail pts)
- diagnostics: no specific tests - based on Sx & neurologic exam
- med management:
  - selegiline, levodopa - goal to prolong indepen
  - PT, OT, education, nutritional counseling
- surgical manage:
  - stereotaxic pallidotomy, DBS, gene therapy
- nursing manage:
  - sm frequent meals (semisolid food to help w/ swallowing
  - ↑ calorie density (↑ fiber, ↓ protein)

Huntingtons: basal ganglia & portions of of cerebral cortex degenerate
→ cause: genetically inherited disorder of CNS by both genders
- assessment:
  - mental apathy, emotional disturb
    ↳ severe depression/psychosis → suicide
  - choreform movements (chorea)
    ↳ writhing & twisting of body
  - grimacing, difficulty chewing & swallowing, speech difficulty
  - early: most physical activity maintained
  - as progresses: intellectual decline, hallucinations, abn movements
    ↳ Sx develop slowly
- diagnostics: no specific test
  - based on Sx & family hx
  - genetic testing: predict which family members will develop disease
- med management:
  - tranquilizers & anti-parkinson's to relieve choreiform movements
  - genetic counseling before pregnancy
- nursing management:
  - family planning discussion (only takes on parent to pass gene)
  - Pt eventually becomes dependent

→ high protein may be indicated in case of severe weight loss (meds must be reevaluated)

extrapyramidal disorder care plan:

diagnosis: impaired physical mobility & self care deficit r/t tremors, dementia
→ assist ambulate; minimize fatigue - provide rest periods

diagnosis: impaired verbal communication r/t inability to articulate words
→ ↓ environment noise, speak slowly