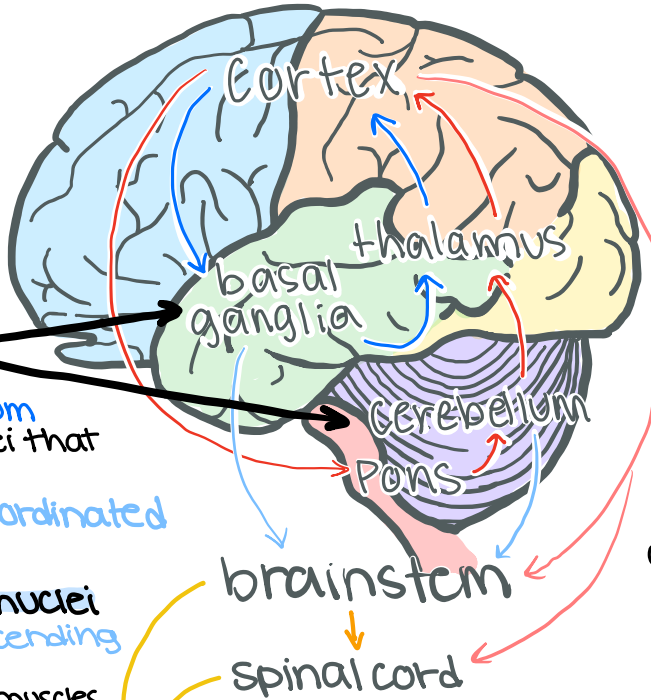
MOTOR SYSTEM



MOVEMENT DISORDERS:
disease of the **extrapyramidal motor system**

basil ganglia and cerebellum are large collections of nuclei that modify movement
↳ allows for smooth, coordinated movements

Also includes **brainstem nuclei** with their associated descending tracts (spinal tracts)
- proximal axial and girdle muscles
- postural tone and balance
- orienting movements of head/neck
- automatic gait-related movement

Disfunction → abnormal tone and posture

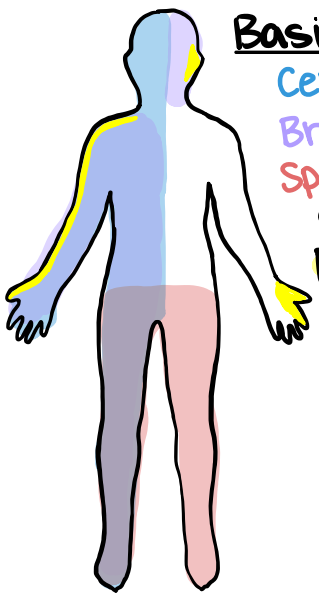
Pathways of Basal Ganglia
- **Direct**: stimulated by substantia nigra → **stimulation** of motor cortex
- **Indirect**: inhibited by substantia nigra → **inhibition** of motor cortex

Upper motor neurons: first order neurons in cortex
Lesions → weakness, ↑tone, ↑reflexes
- involvement of ONLY lower face

NEUROMUSCULAR DISORDERS
disease of the **pyramidal motor system** → peripheral nervous system and muscle
Hallmark: **weakness**

lower motor neurons: second order neurons in brainstem or spinal cord
Lesions → weakness, atrophy, fasciculations, ↓reflexes/tone
- involvement of upper AND lower face

skeletal muscle



Basic Patterns of Focal Weakness:
Cerebrum: contralateral hemi-body
Brainstem: opposite sides of head and body
Spinal Cord: bilateral, entire body below specific horizonal line
- weakness consistent w/ "myelopathy"

Peripheral NS
nerve root, cranial/peripheral nerve → unilateral in stripe or blob

Diffuse weakness:
Anterior Horn: no symmetry, affects some distal, proximal, bolbar
Peripheral nerves: symmetric, distal and progresses proximal
NMJ/muscle: symmetric, proximal/bulbar involvement

Bulbar weakness: emanating from brainstem → dysarthria, dysphagia, dysphonia, weak respiration

MOVEMENT DISORDERS

HYPOKINETIC

Bradykinesia: slowness of movement
- Akinesia, freezing

Rigidity: stiffness/resistance to movement

PARKINSONS

patho: degeneration of nigrostriatal pathway sets off cascade resulting in decreased stimulation of motor cortex
- core pathology affects dopamine-producing neurons of substantia nigra

Clinical: 4 cardinal features
1. Tremor (pill-rolling, pronation)
2. Rigidity
3. Akinesia (bradykinesia)
4. Postural instability

Asymmetric onset, "frozen shoulder", hypophonia (softer speech), micrographia (smaller handwriting), decrease arm swing

Non-motor sx: orthostatic hypotension, constipation, depression (50% pts), dementia (6x more common in PD)

Treatment: achieve symptomatic benefit, ↓disability, maintain QoL

Tremor predominant
- <60 amantadine, artane
- >60 dopamine agonist

Akinetic-rigid
- <60 dopamine agonist
- >60 levodopa, dopamine agonist

Other Causes of Parkinsonism

Vascular AKA "multi-infarct"
Parkinsonian sx produced by one or more small strokes → involves more than lower body
Treatment: ↓stroke risk, levodopa, PT/OT

Dementia pugalistica occurs in people who have suffered multiple concussions
- may not experience signs until later

Neuropathologic: tau-protein abnormalities, substantia nigral degeneration

HYPERKINETIC

ESSENTIAL TREMOR Most common movement disorder
- familial in 50% of cases → autosomal dominant w/ variable penetrance

Clinical: postural and intention tremors of the arms and hands (↑frequency, ↓amplitude)
- causes head nodding, voice changes, and difficulty w/ eating, drinking, fine motor tasks

Aggravating → stress
Alleviating → alcohol

Treatment: propranolol (30 mg BID)
primidone (50 mg daily)

HUNTINGTONS

epi: 4th-5th decades of life

patho: fatal autosomal dominant CAG trinucleotide expansion condition → unstable

Clinical: behavioral changes (depression, psychosis), Chorea, dementia

diagnostics: MRI → atrophy of the caudate nucleus and dilatation of the anterior horns of the lateral ventricles (advanced cases)

TOURETTES tic disorder syndrome

defined by:
- multiple motor AND at least one vocal
- tics occur many times a day nearly every day OR intermittently for a year w/out 3 consecutive tic-free months
- onset before age 18

Cause: unknown

Treatment: CBT and meds.
- only when child is bothered by them

CEREBELLAR Dysfunction → ataxia

involved in coordination of movement → coordinates motor cortex with proprioceptive feedback
- partly responsible for motor learning

ATAXIA three main types

1. Spinal/sensory: lesions interrupt sensory input to cerebellum
2. Cerebellar: cerebral cortex pathology → incorrect execution of cortical signs
3. Spinocerebellar: combination of both

NEUROMUSCULAR DISORDERS

Disease of the peripheral nervous system and muscle

Bell's Palsy

epi: pregnancy, diabetes

cause: unknown. Viral or inflammatory.

clinical: sudden, unilateral facial paralysis (recovery within 6months)
- eyebrow sagging, inability to close eyes, inability to activate smile
- occurs within hours-days

diagnosis: Clinical. EMG → prognosis
- If atypical → imaging

treatment: oral GC EARLY - prednisone
±self-limiting

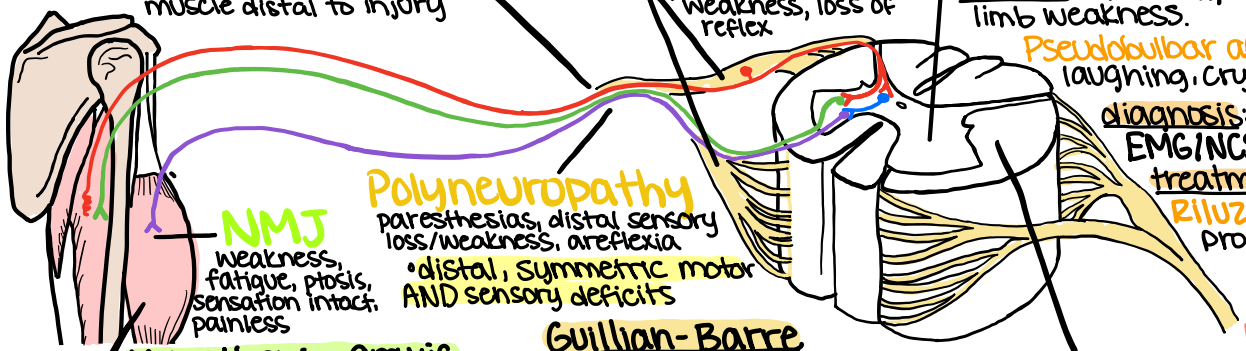
Focal Neuropathy

pain, weakness, atrophy of muscle distal to injury

- Compressive radiculopathy
- Shingles

Nerve Root (Radiculopathy)

pain, paresthesia, weakness, loss of reflex



NMJ

weakness, fatigue, ptosis, sensation intact, painless

Polyneuropathy

paresthesias, distal sensory loss/weakness, areflexia
- distal, symmetric motor AND sensory deficits

Myopathy

weakness, ± muscle pain, elevated CK

Hereditary:
- muscular dystrophies
- metabolic myopathies

Acquired:
- inflammatory

signs/sx: atrophy, calf hypertrophy, scapular winging, gower's sign
↳proximal lower limb weakness

Myasthenia gravis

patho: antibodies against ACh receptors

sx: diplopia, ptosis, bulbar and proximal limb weakness
- fatigable weakness

diagnosis: AChR and Musk antibodies
- Single-fiber EMG → evaluates NMJ activity

tx: pyridostigmine
- immunotherapy

Acute → IVIG or plasmapheresis

Guillian-Barre

patho: immune mediated NS damage commonly following infection
- campylobacter, CMV, EBV, M. pneumonia

clinical: areflexia and ascending weakness/numbness (starts distal and moves proximal)
- progresses over 2 week period

diagnosis: LP → ↑protein
EMG/NCS → prognosis

treatment: admit
IVIG or plasmapheresis can slow progression

Muscular Dystrophies

defects in muscle cellular proteins
- dystrophin stabilizes the muscle membrane glycoprotein complex and protects cell from degradation

Types: duchenne's (boys 2-3) and becker's (boys 5-15)
- X-linked deletions

Clinical: calf hypertrophy, gower's sign

Diabetic Polynephropathy

50% of diabetics

Clinical: sensory > motor, distal, burning pain

Management: treat diabetes.
Pain control → antidepressants (amitriptyline, duloxetine)
anticonvulsants (pregabalin, gabapentin)
- foot care, fall precautions

Myotonic dystrophy: MOST COMMON

- autosomal dominant

Clinical: Myotonia - delayed muscle relaxation after contraction
- Weakness/muscle wasting, frontal balding, temporal wasting, cataract, endocrinopathy, hypersomnia, low intelligence, conduction defects

↳ Spinal muscular atrophy, poliovirus

Anterior Horn cell

atrophy, weakness, fasiculations, normal sensation, painless

ALS amyotrophic lateral sclerosis

Degenerative disease of the upper and lower motor neurons
- progressive disease → fatal within 3-5 years (respiratory infection/failure)

patho: majority sporadic
- glutamate tox, oxidative stress
5-10% familial → ↓radical protection

Clinical: bilateral, asymmetric limb weakness.
Pseudobulbar affect: inapprop laughing, crying, yawning

diagnosis: no single test
EMG/NCS → denervation

treatment: no cure
Riluzole can slow progression

MS, Stroke

Corticospinal Tract

Spasticity, brisk reflexes, weakness

Cerebral Palsy: group of permanent disorders of the development of movement and posture due to non-progressive disturbances in developing brain.

patho: PVL, HIE, IVH, any causing peri/prenatal injury

Clinical: activity limitation
Spasticity is hallmark

Classified based on abnormality:
1. Spastic (pyramidal): upper motor neuron syndrome
   - Spastic hypertonia, hyperreflexia, extensor plantar responses, clonus
   Slow, effortful voluntary movement, impaired fine motor, difficulty isolating movements, fatigue
2. Dyskinetic (extrapyramidal)
3. Ataxic (cerebellum)