A Practical Approach to the Neurologic Exam in Children

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Practical Day in Pediatrics

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Learning Objectives

- Demonstrate exam techniques for different developmental stages
- Describe normal and abnormal findings
- Review differential and red flags for abnormal findings

The Neuro Exam in Children

- The nervous system is a dynamic, developing system in children
- Normal findings can change throughout childhood
- The neurologic exam is important to detect developmental, structural and acquired disorders
- Pre/perinatal history, developmental history, family and social history affect interpretation of the neuro exam
- Multiple challenges:
  - Cooperation with exam varies by age
  - Children cannot always express themselves
  - Developmental milestones affect exam expectations and can be variable

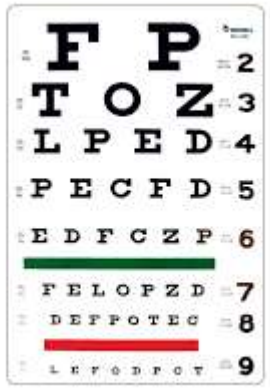
Tips for the Neuro Exam in Children

- Create a calm environment, involve parents for positioning/comfort
- Special techniques and a playful attitude are helpful
  - Infants: more observation, infant reflexes and passive movements
  - Toddlers: play-based approach
  - Older children: structured exam commands
- Perform less invasive components first (observation)
- Be prepared to be patient and flexible
- When in doubt, describe what you see!

Red Flags

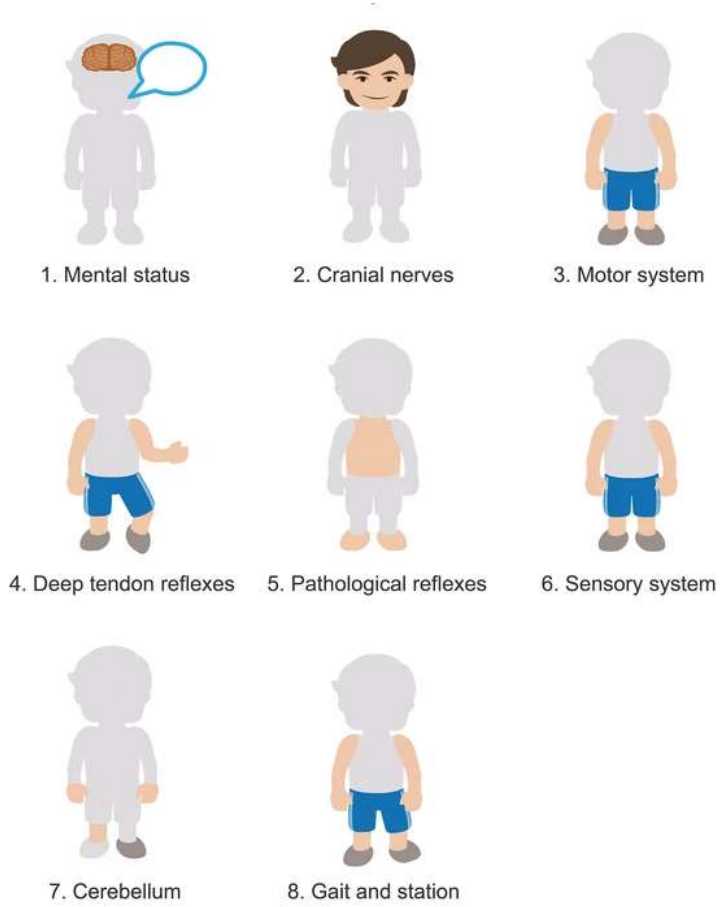
- Loss/regression of developmental milestones
- Abnormal tone (increased/decreased)
- Altered mental status
- Asymmetric findings
- Gait abnormalities

Neuro Exam Tools



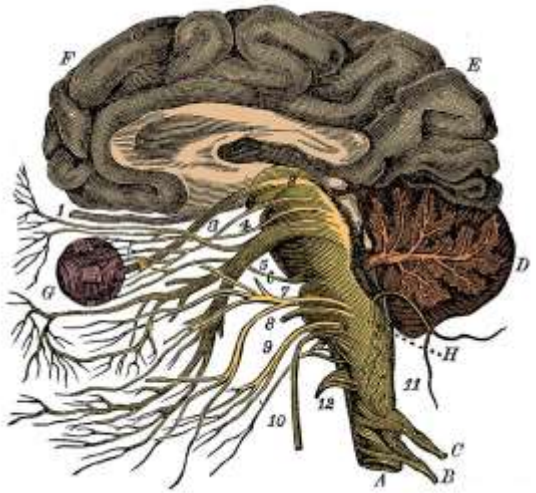
Components of the Neuro Exam

1. Mental Status
2. Cranial Nerves
3. Motor
4. Sensory
5. Reflexes
6. Coordination
7. Gait

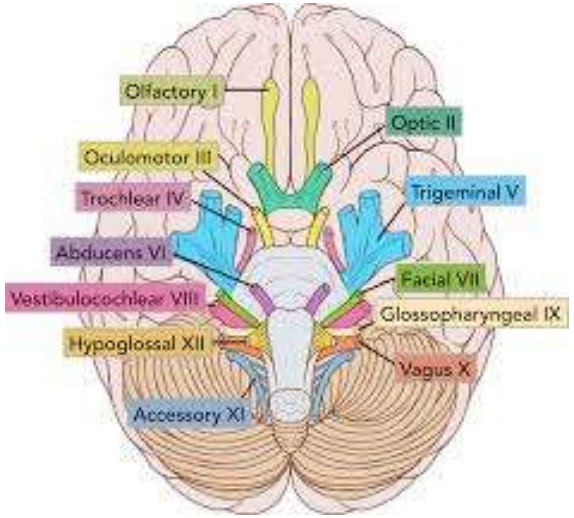


Mental Status

- Most aspects of mental status can be observed during clinic visit:
  - attention, brisk/delayed responses, ability to answer questions, overall appearance, cooperation with exam prompts
- Mental status varies drastically depending on developmental stage!
  - Infants: awake, alert, responsive to examiner
  - Toddler: alert, follows simple exam prompts, answers questions in 2-3 word phrases, names body parts
  - Older children: awake, alert, oriented to time, place, person and situation. Intact remote and recent memory. Follows two step commands. Normal attention, language and affect for age.



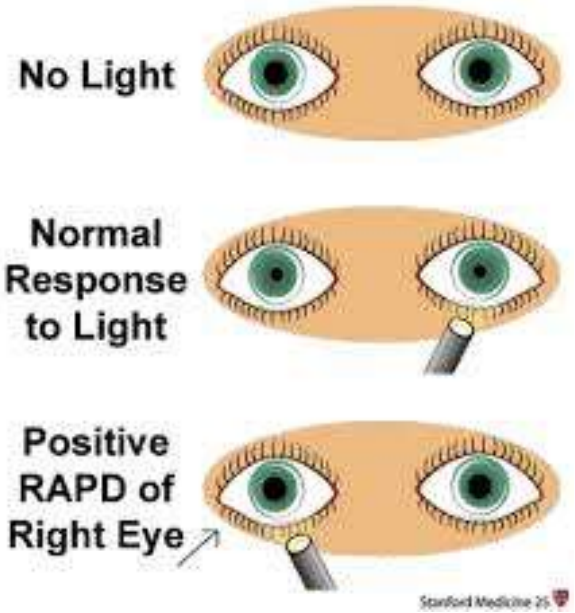
Cranial Nerves

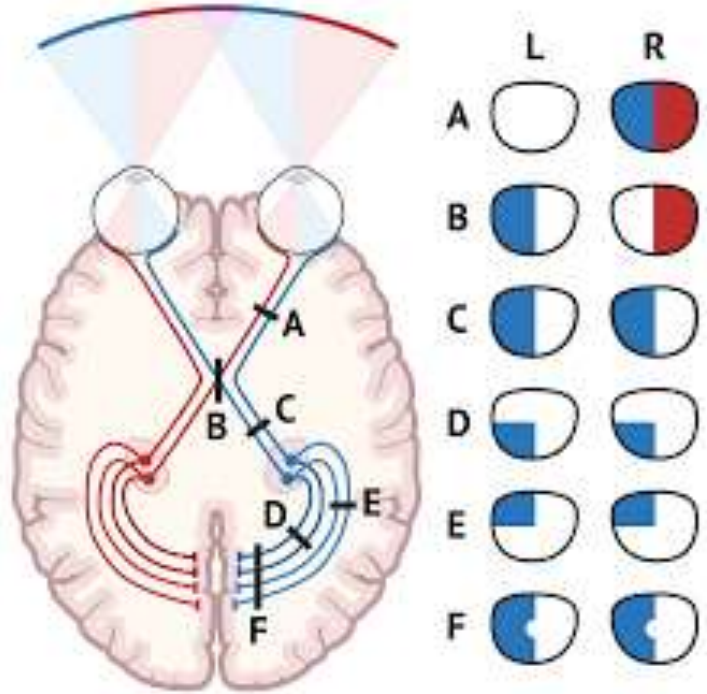


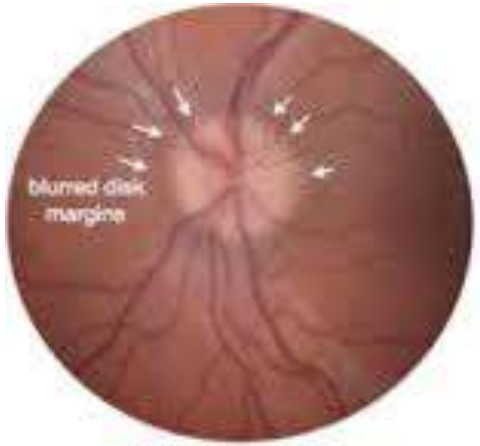
Number	Name	Function
I	Olfactory	Smell
II	Optic	Vision – acuity, fields, fundoscopy, afferent pupillary response
III	Oculomotor	Pupillary constriction, lid elevation, most EOM
IV	Trochlear	Downward, internal rotation of eye
V	Trigeminal	Facial sensation (V1 ophthalmic, V2 maxillary, V3 mandibular). Motor - temporal and masseter muscles
VI	Abducens	Abduction of eye
VII	Facial	Facial movements (facial expression, closing eye, closing mouth). Taste (anterior 2/3 tongue)
VIII	Acoustic	Hearing (cochlear) and balance (vestibular)
IX	Glossopharyngeal	Motor – pharynx. Sensory – eardrum/canal, pharynx, posterior tongue
X	Vagus	Motor – palate, pharynx, larynx. Sensory – pharynx, larynx
XI	Spinal accessory	SCM and upper trapezius movement
XII	Hypoglossal	Tongue movement

Cranial Nerves

- CN II – blurry vision
  - Refractive error
  - Uveitis, glaucoma, corneal clouding
  - Unilateral – optic neuritis
- CN II – (relative) afferent pupillary defect (CN II and III)
  - Congenital anisicoria
  - Ipratropium nebulizer
  - optic neuritis, mass
- CN II – visual field defect
  - Optic tract lesion
  - Pituitary mass (bitemporal)
- CN II – papilledema
  - Pseudopapilledema/Drusen
  - Increased ICP (mass, IIH)





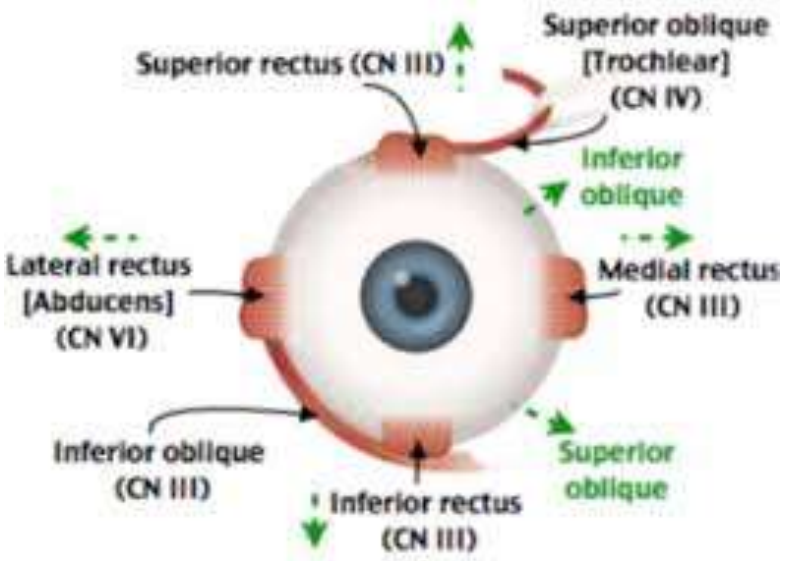


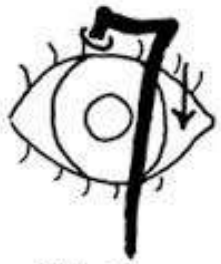
Papilledema

Normal Optic Disk

Cranial Nerves

- CN III – “down and out”
  - Mass, vascular disease, brainstem lesion
- CN III – ptosis only
  - peripheral injury
  - CNS lesion, myasthenia
- CN IV – torsional diplopia, “bad guy on top”
  - Mass/brainstem lesion
- CN VI – abducens palsy
  - strabismus
  - Mass/brainstem lesion

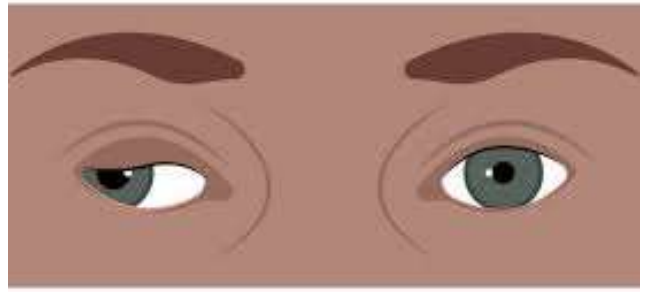




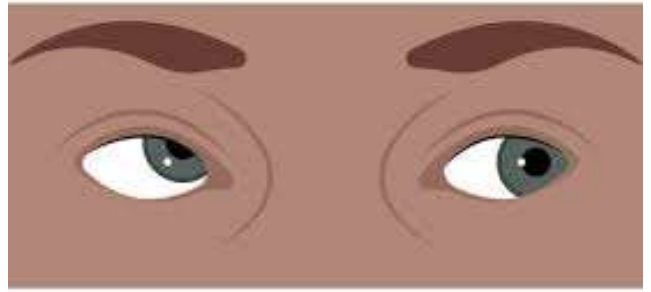
CN 7
A hook;
closes eyes

CN III
3 pillars;
opens eyes

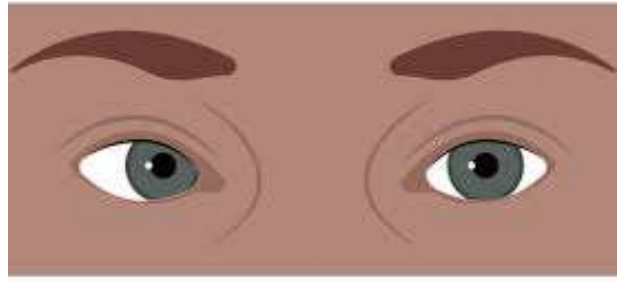
RIGHT 3rd NERVE PALSY



RIGHT 4th NERVE PALSY

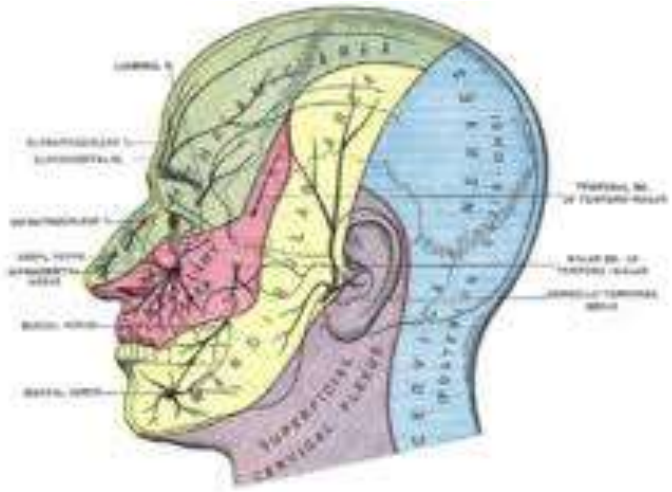


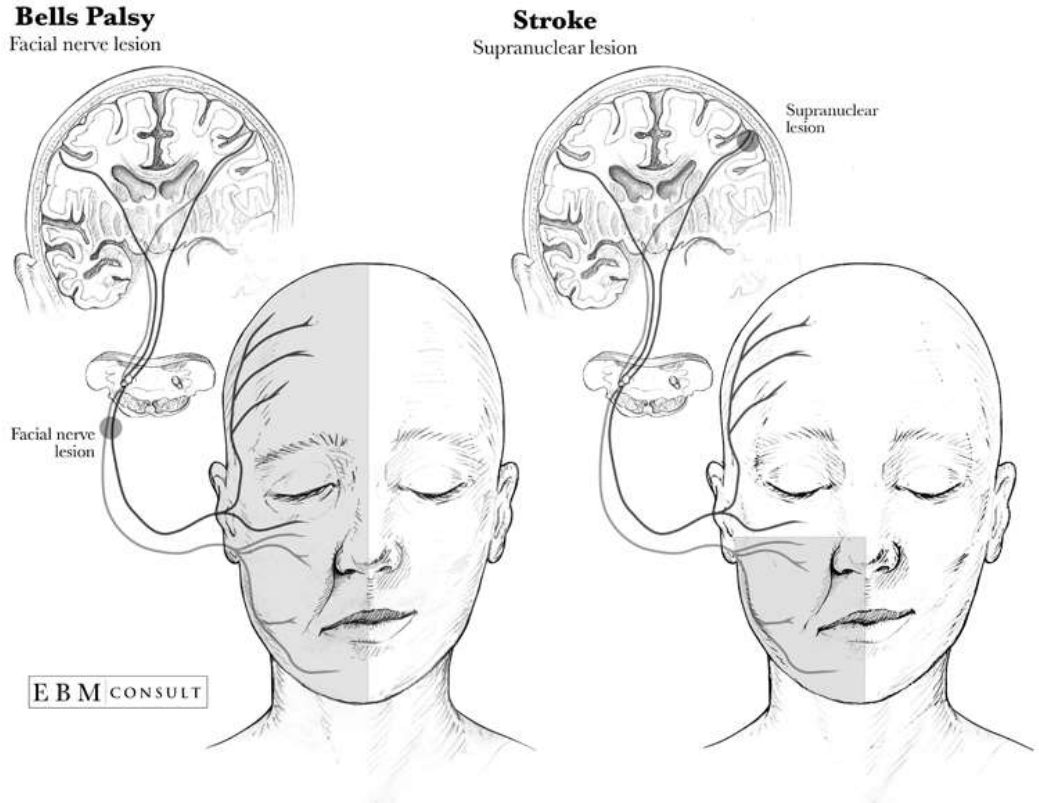
RIGHT 6th NERVE PALSY



Cranial Nerves

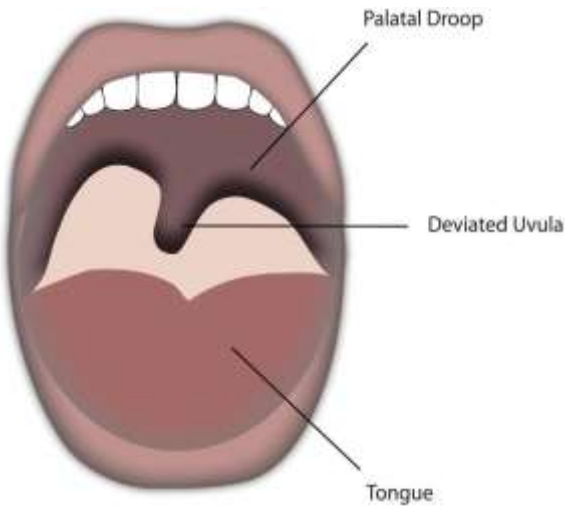
- CN V – facial numbness/pain
  - Trigeminal neuralgia
  - Peripheral nerve injury
  - Brainstem lesion
- CN VII – facial weakness (inability to close eye)
  - Bell's palsy (peripheral)
  - Stroke, mass, brainstem lesion

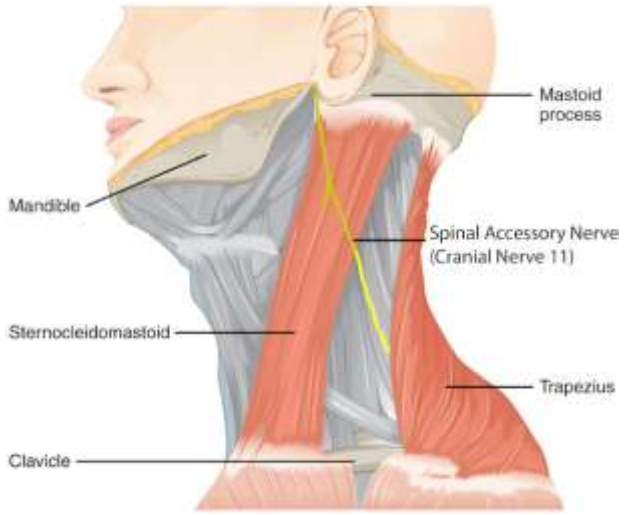


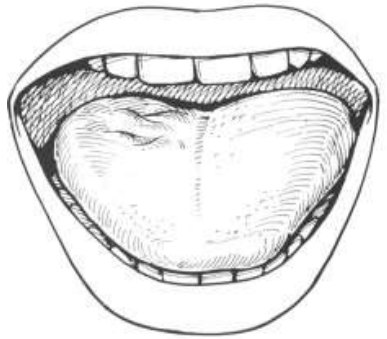


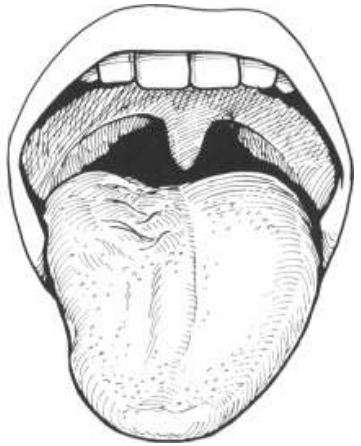
Cranial Nerves

- CN VIII – hearing loss, tinnitus, vertigo
  - Acute/chronic otitis media
  - Brainstem lesion
- CN IX/X – dysphagia/dysarthria
  - GI dysfunction
  - Brainstem lesion
  - Myasthenia (NMSK junction)
- CN XI – trapezius/SCM strength
  - Peripheral nerve/NMSK disorder
  - Brainstem lesion
- CN XII – tongue deviation (to weak side)
  - Tongue injury
  - Brainstem lesion



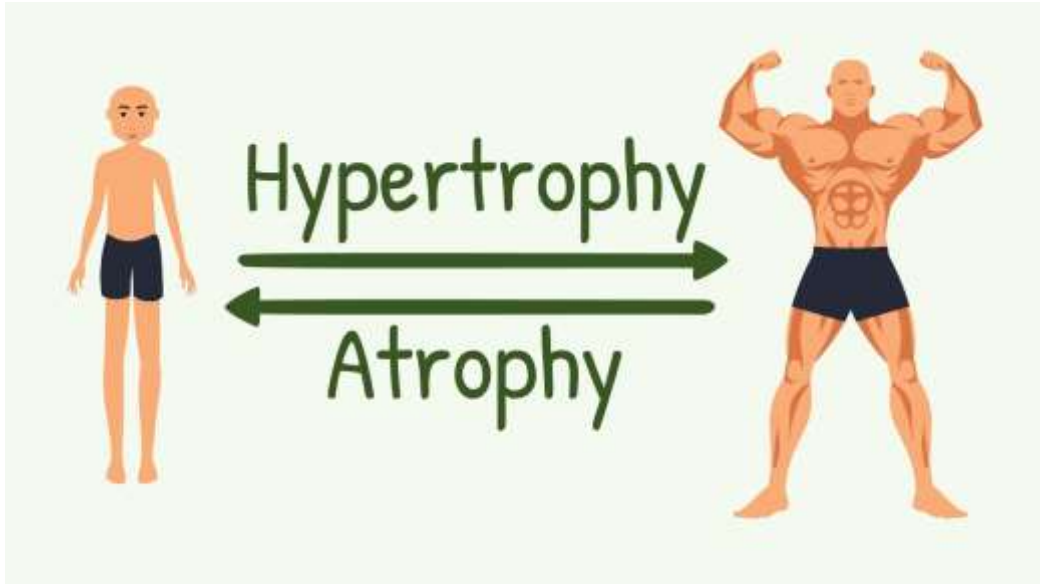






Motor Exam

- Bulk
  - Hypertrophic vs atrophic
- Tone
  - Hypotonia
  - Hypertonia
- Strength
  - Head control, truncal strength
  - Upper extremities:
    - Deltoids, biceps, triceps, hand intrinsics, grip
  - Lower extremities:
    - Iliopsoas, hamstrings, quadriceps, gastrocs, anterior tibialis
- Abnormal movements
  - Inspection
  - Arm extension, reaching



Motor Exam

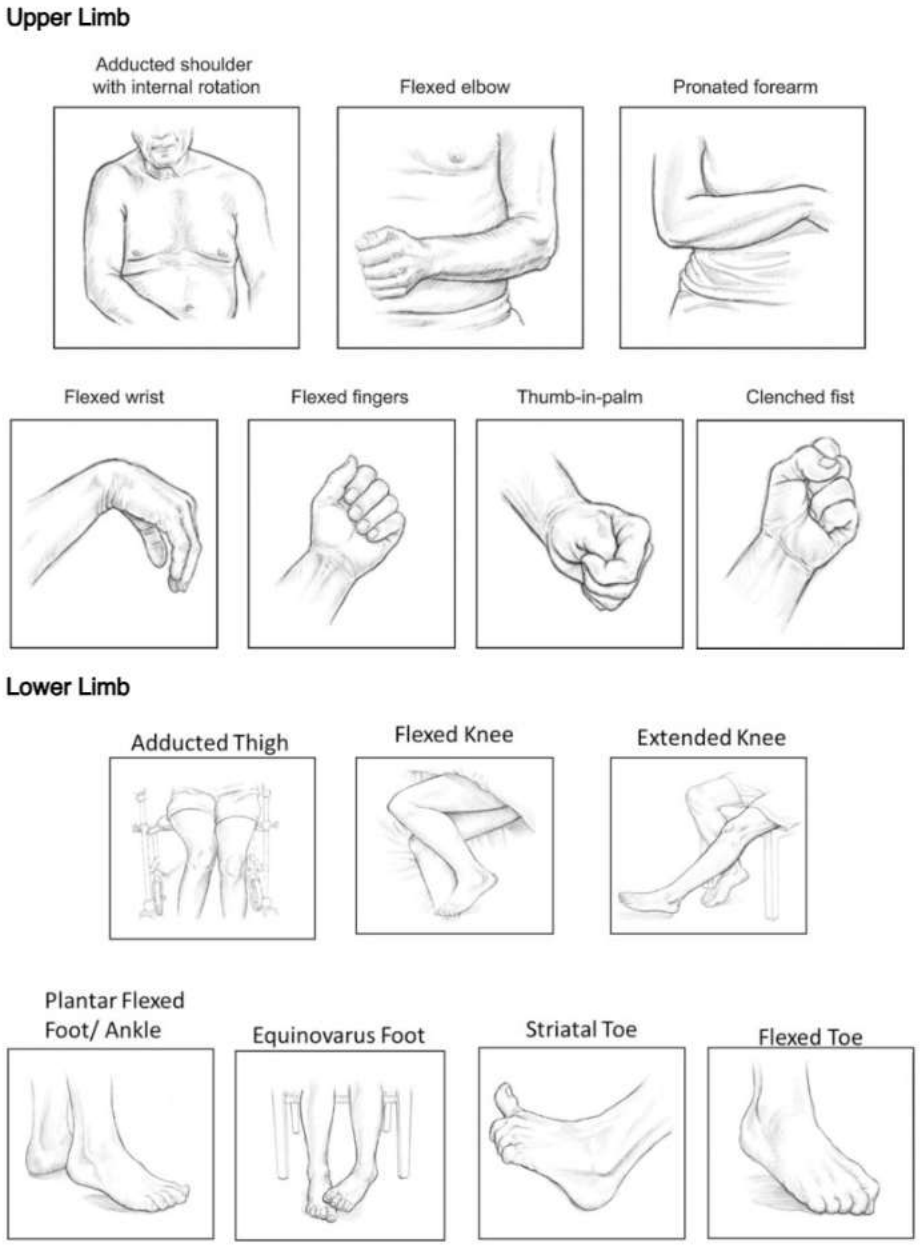
Tone

- Passive movements, posture
- Correlate with reflexes!
- Hypotonia - exam
  - Infant:
    - head lag
    - frog leg positioning
    - “floppy”
  - Toddler/older children:
    - Gross motor delay (sitting, standing, walking)
    - joint laxity/instability (pronation)
    - exercise intolerance

- Central hypotonia:
  - dev delay, dysmorphia/microcephaly, normal/brisk reflexes, axial>appendicular weakness
  - Genetic/metabolic disorders, congenital infections
- Peripheral hypotonia:
  - nl mental status, hypo/areflexic, axial=appendicular weakness, atrophy, fasciculations, resp/feeding difficulties
  - Muscular dystrophies, SMA, congenital myopathies, peripheral neuropathies

Motor Exam

- Hypertonia – exam
  - Spasticity – velocity dependent
    - Early hand preference (<18 mos), fisting (cortical thumb), elbow flexed at rest, toe walking
  - Rigidity – velocity independent
    - Rare in children
    - cogwheeling
- Hypertonia – spasticity vs rigidity
  - Spasticity:
    - **Cerebral palsy** (perinatal injury, infection, malformation, leukodystrophies)
    - hereditary spastic paraplegia
    - spinal cord injury
  - Rigidity:
    - Serotonin syndrome, genetic/metabolic disorders, brain malformations



Motor Exam

- Muscle strength
  - extremity vs truncal strength
  - 0-5/5 rating vs descriptive (antigravity, resists examiner, sits unassisted)
    - 0/5 – no muscle twitch
    - 1/5 – muscle twitch
    - 2/5 – movement of joint, gravity eliminated
    - 3/5 – antigravity
    - 4/5 – some resistance
    - 5/5 – full resistance

- Weakness – exam
  - Distal (hyporeflexic) – grip weakness, foot drop
    - GBS
    - peripheral neuropathy
  - Proximal (hyporeflexic) – difficult w/stairs, standing up
    - muscular dystrophy
    - myopathy
  - Unilateral (hyperreflexic/nl)
    - Stroke
    - spinal/brain lesion
    - Todd's paralysis, hemiplegic migraine
  - Fluctuating – worse w/fatigue, later in day
    - Myasthenia (w/ptosis, dysphagia)
    - Systemic/metabolic disease
    - Functional neurologic disorder
  - Weakness with pain
    - Rhabdo, myositis
    - Peripheral neuropathy

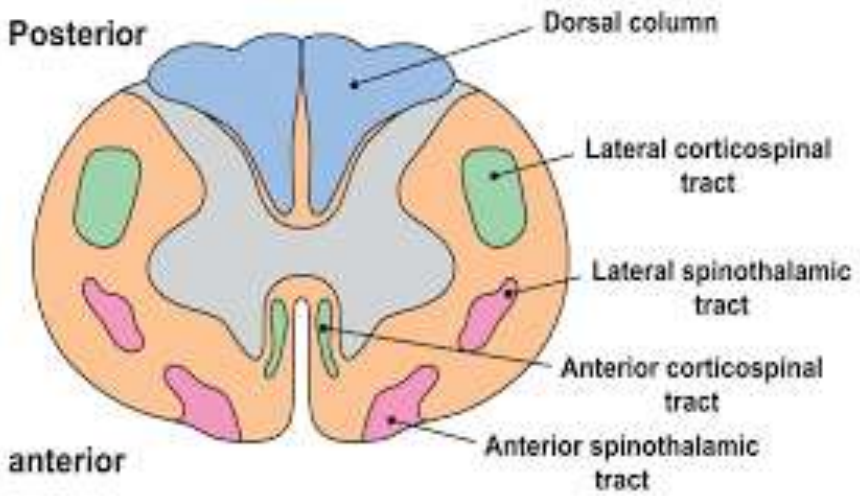
Motor Exam

- Abnormal movements
  - Tremor
    - frequency, amplitude?
    - Extremity, head, trunk, generalized?
    - Resting, postural, action?
  - Tics
    - Brief, stereotyped, suppressible
    - Provoked by stress, boredom
    - Vocal vs motor
  - Dyskinesias, chorea, dystonia, stereotypies….

- Tremor
  - Action/postural tremor
    - Physiologic tremor
    - high adrenergic state
    - Medications
    - Nutritional deficiencies
    - Essential tremor
    - Cerebellar, brainstem lesion
  - Rest tremor
    - Parkinsonism
    - Medications
    - Functional tremor
- Tics
  - Transient tic disorder
  - Tourette syndrome
  - Functional tics

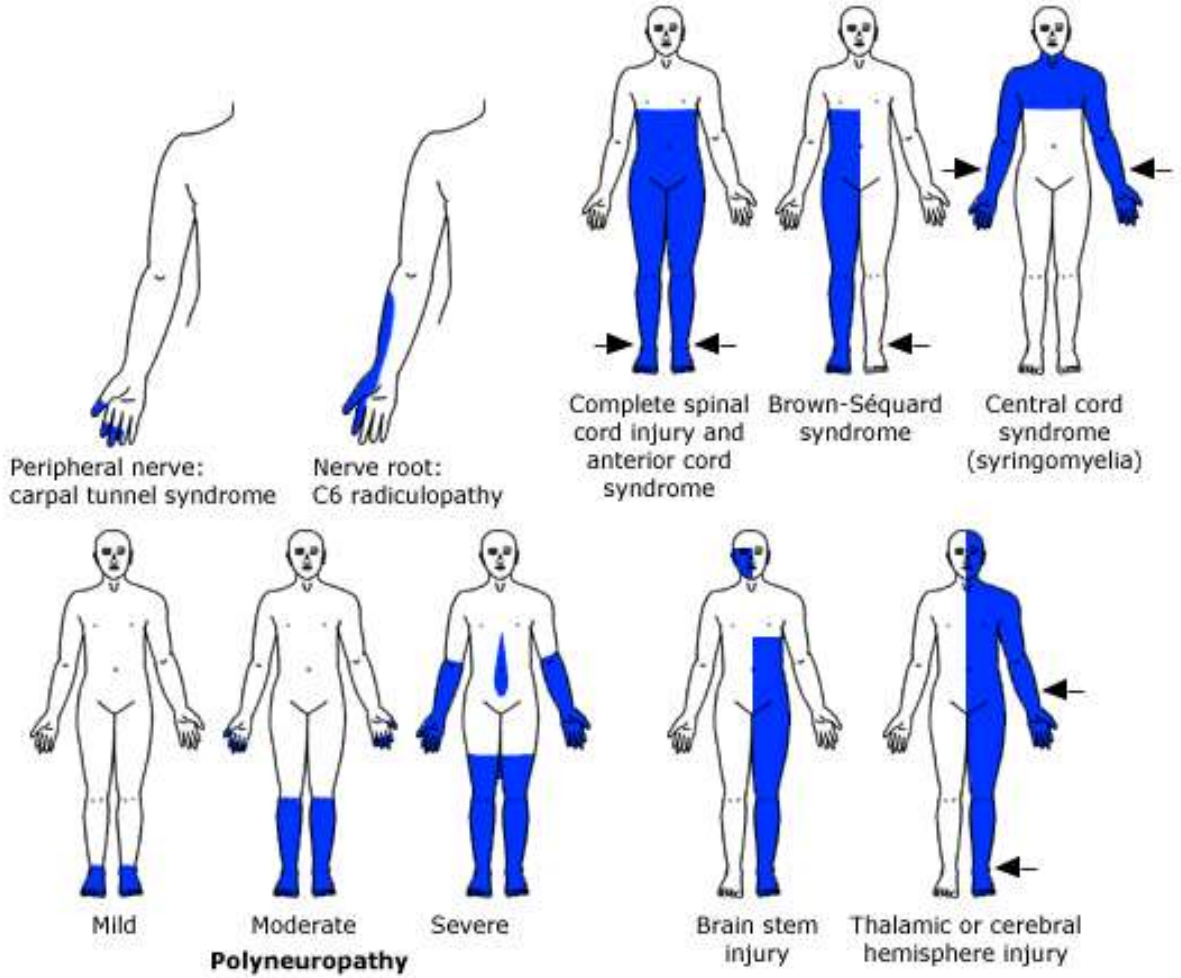
Sensory Exam

- Compare left to right, upper to lower extremities, proximal to distal
- Light touch – does not localize, but acceptable if low suspicion
- Pinprick, temperature
  - spinothalamic tract (anterior spinal cord)
- Vibration, proprioception
  - dorsal columns (posterior spinal cord)



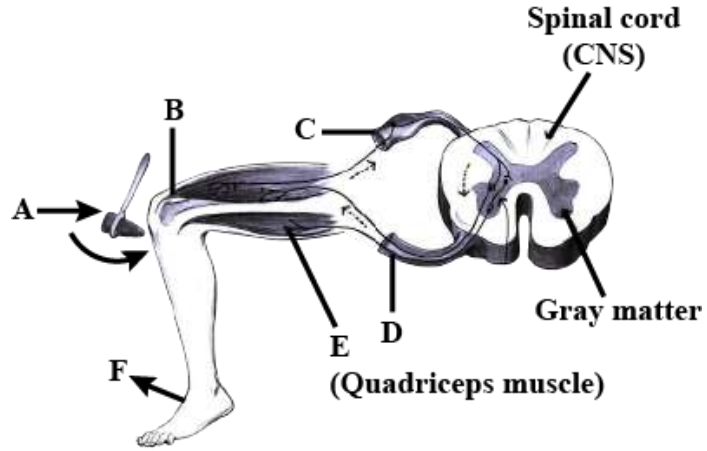
Sensory Exam

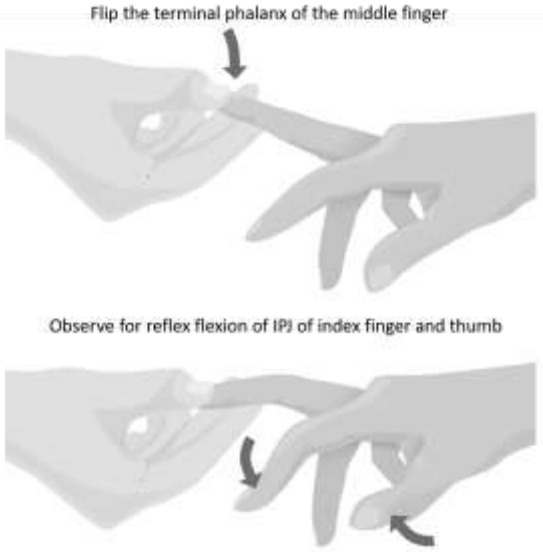
- Unilateral
  - Spinal cord, brain lesion
- Bilateral
  - Spinal cord
- Distal
  - Neuropathy
  - radiculopathy
- Proximal – uncommon!
  - syringomyelia
- Focal/segmental
  - Focal neuropathy
  - Radiculopathy

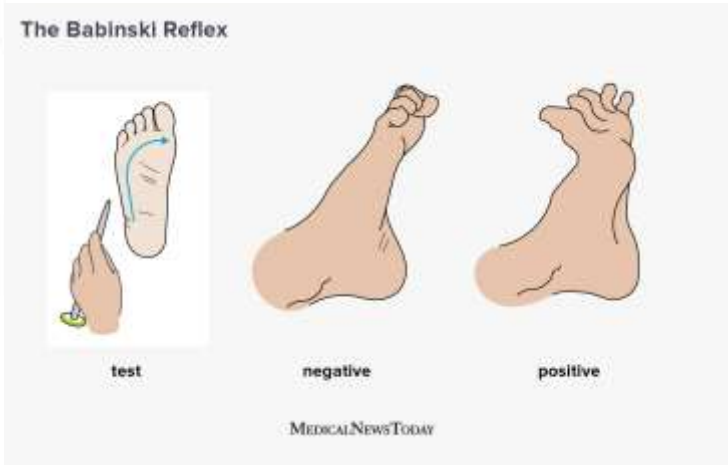


Reflexes

- Rating
  - 0 = Absent
  - 1+ = hypoactive
  - 2+ = normal
  - 3+ = hyperactive
  - 4+ = hyperactive with clonus
- Pathologic reflexes:
  - Hoffman's
  - Crossed adductors
  - Babinski/upgoing toes (over age 2-3)

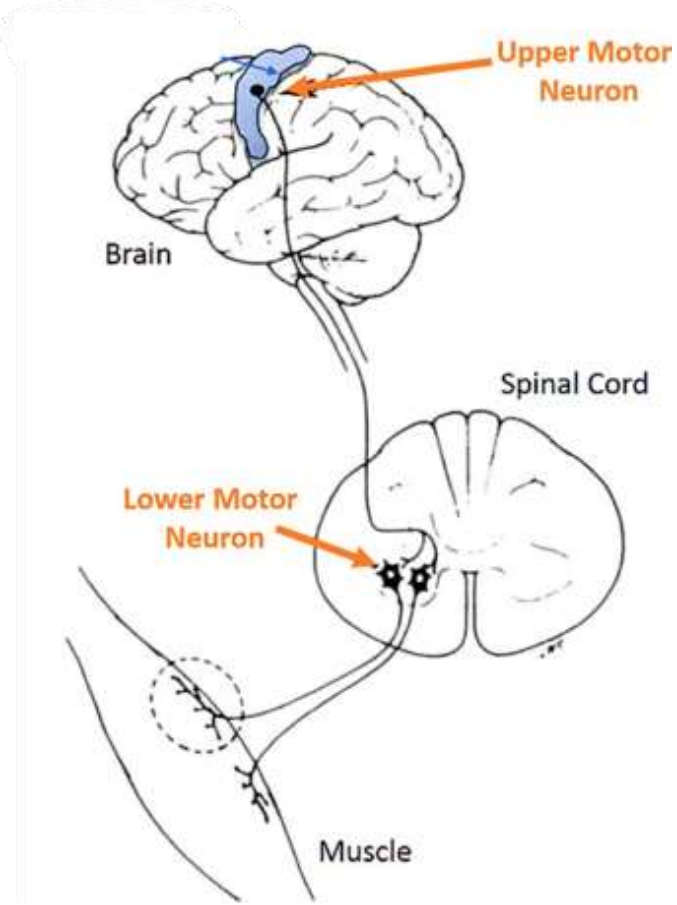






Reflexes

- Spectrum of normal reflexes
  - mildly hypoactive->mildly hyperactive
  - Often correlates with baseline joint flexibility
- Absent/hypoactive reflexes = lower motor neuron
  - Hypotonia?
  - Sensory loss?
  - Weakness?
- Hyperactive reflexes/clonus = upper motor neuron
  - Spasticity/Contractures?
  - Bladder/bowel dysfunction?
  - Cognitive delays?

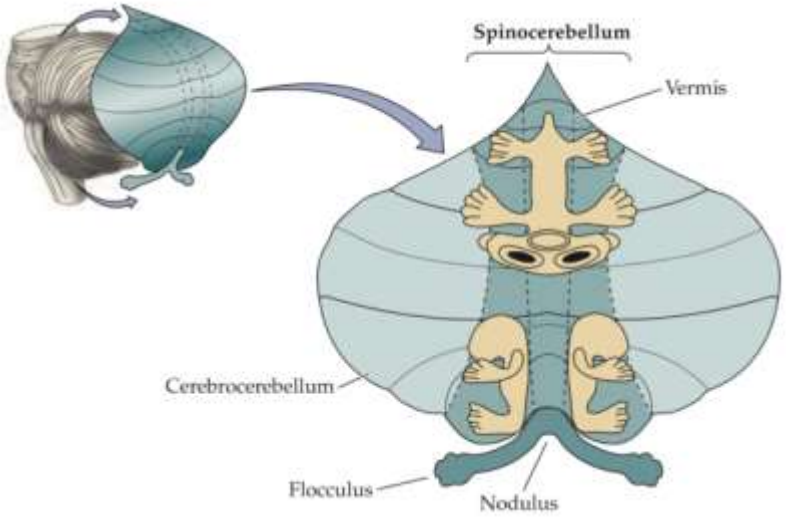


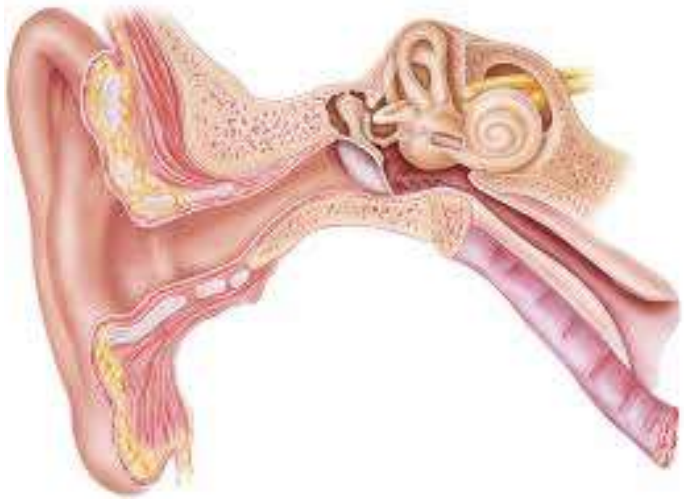
Reflexes

- Newborn reflexes - evolutionary survival function
  - should suppress with frontal lobe/pyramidal tract development
  - Persistence/recurrence in older children
    - Genetic disorders, TBI, stroke
- Moro – 6 months
  - Asymmetric -> birth trauma
- Asymmetric tonic neck reflex/fencing reflex – 4 months
- Stepping/walking reflex - 6 months
- Rooting/sucking – 4 months
- Palmar – 6 months
- Plantar – 12 months

Coordination

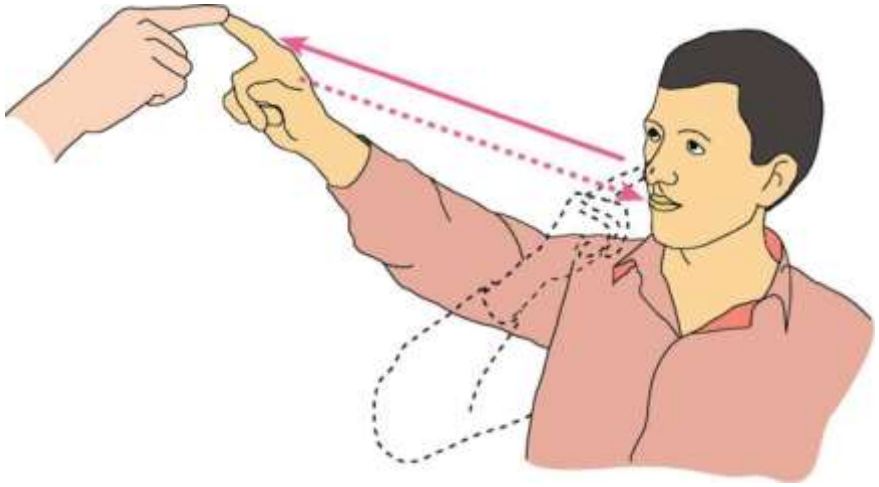
- Requires coordination of motor system, cerebellum, vestibular system, proprioception
- Truncal coordination
- Finger to nose/reaching for objects
- Heel to shin
- Finger/heel tapping
- Rapid alternating movements
- Gait

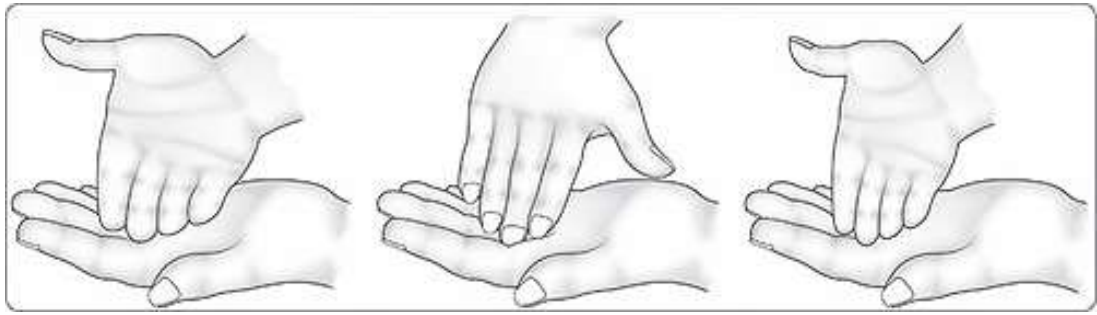




Coordination

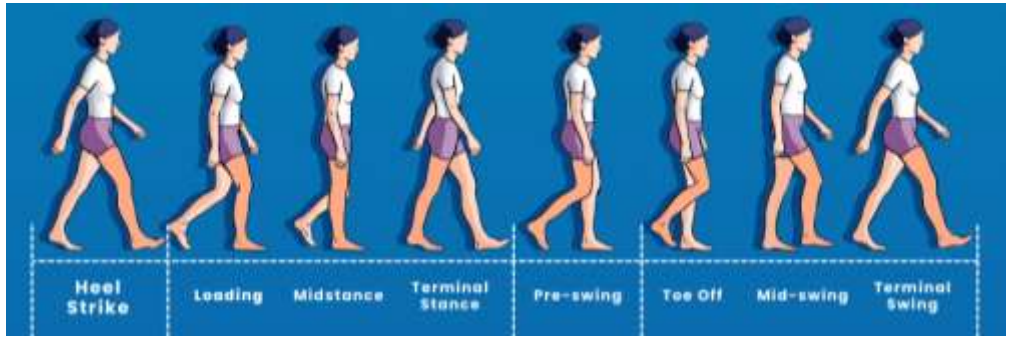
- Dysmetria (w/finger to nose or heel to shin)
  - Tremor
  - Weakness/abnormal tone
  - Cerebellar dysfunction
- Dysdiadochokinesia (clumsiness with rapid alternating movements)
  - Fine motor delays
  - Cerebellar dysfunction
- Truncal ataxia
  - Cerebellar dysfunction (vermis/midline)





Gait Exam

- Combination of motor, coordination, and sensory exams
- Unstressed gait
  - Strength, coordination, proprioception
- Heel/toe gait
  - Strength
- Tandem gait
  - Coordination
- Romberg (with pronator drift)
  - Coordination, proprioception
  - Pronator drift - weakness



Gait Exam

- Unstressed gait
  - Ataxic
    - Cerebellar dysfunction
    - Sensory – metabolic, nutritional deficiencies
    - FND
  - Spastic
    - CP, hereditary spastic paraplegia
  - Steppage gait
    - Neuropathy, foot drop
  - Waddling gait – w/Gowers sign
    - NMSK, disease, muscular dystrophy
  - Cautious, antalgic, shuffling

Exam Findings in Functional Neurologic Disorder

- FND (conversion disorder) – dysfunction with “software” rather than “hardware”
- Often presents with abnormal movements and weakness
- Functional facial numbness:
  - Check angle of jaw, earlobe
  - Vibration on forehead should transmit to contralateral side. “splitting the forehead to vibration”
- Functional weakness:
  - Variability, distractability, enhancement with attention, suggestibility
  - Hoover sign – no extension effort in contralateral leg when testing weak leg flexion
  - Give-way weakness – limb collapses with light touch/normal strength suddenly collapses
  - Drift without pronation when testing for pronator drift
  - Co-contraction – agonist and antagonist contract simultaneously->no movement
- Functional tremor:
  - Variability, entrainment, cocontraction, rest=posture=action
- Functional gait:
  - Astasia-abasia – more strength/coordination required than for unstressed gait (flailing, staggering, robotic, trembling)

Question 1

A 4 year old presents to clinic after his parents notice he has trouble standing from a sitting position on the floor. He tends to walk on his toes, falls easily, and doesn't run well. He crawled late and walked independently at 18 months. He has normal verbal and social skills. On exam his reflexes are 1+ throughout, has 4/5 weakness in his proximal leg muscles and a waddling gait. Where does his disorder localize?

1. Spinal cord
2. Peripheral nerve
3. Muscle
4. Motor cortex

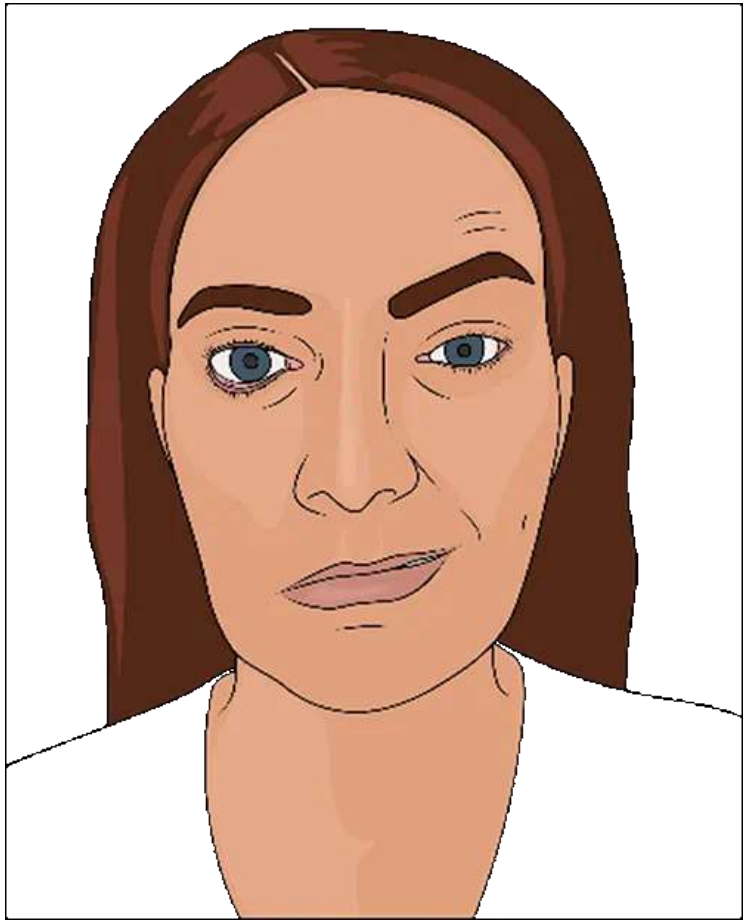
Question 1 - Explanation

1. Spinal cord
   - Cord pathology generally involves hyperreflexia, as well as bladder and bowel dysfunction
2. Peripheral nerve
   - Peripheral neuropathies cause distal rather than proximal weakness
3. **Muscle**
   - **Myopathies and muscular dystrophies present with a pattern of proximal weakness which can manifest as gross motor delay, hyporeflexia, difficulty climbing stairs, exercise intolerance, a positive Gowers sign, and a waddling gait**
4. Motor cortex
   - Weakness from a brain lesion typically affects one side of the body, would involve hyperreflexia, and would expect other domains (language, cognitive) to be affected

Question 2

A 16 year old female presents with a three day history of right facial droop after one week of URI symptoms. She has difficulty closing her right eye, and says food tastes bland. She has no facial numbness, extremity weakness or numbness. What is the best next step?

1. Send immediately to the ER
2. Prescribe a course of steroids
3. Refer to outpatient neurology
4. Order an MRI brain and spine



Question 2 - Explanation

1. Send immediately to the ER
   - Weakness involving the forehead localizes to the peripheral nerve, arguing against acute stroke
2. **Prescribe a course of steroids**
   - **Unilateral facial weakness involving the forehead, inability to close the eyelid and taste changes are consistent with CN VII palsy (Bell's Palsy). History of URI and lack of other neuro findings is reassuring against a brainstem lesion.**
3. Refer to outpatient neurology
   - A patient with history and exam consistent with uncomplicated Bell's Palsy does not need to be referred to neurology. Would consider referral for bilateral Bell's Palsy, other cranial nerve findings on exam, or complicated recurrence
4. Order an MRI brain and spine
   - Forehead involvement argues against CNS lesion, and with a recent URI and otherwise normal neurologic exam, neuroimaging is not indicated

Question 3

A 17 year old female presents with one week of progressive right leg weakness, reports she is now unable to move her right leg at all. On formal motor testing she is unable to voluntarily move her right leg at all but has full strength in her left leg and both arms. Tone, reflexes, and sensation are intact in her right leg, and she has no bladder or bowel dysfunction. You are concerned for functional neurologic disorder. What exam maneuver or finding would help you confirm this diagnosis?

1. Give-way weakness
2. Entrainment
3. Astasia abasia
4. Hoover sign

Question 3 - Explanation

1. Give-way weakness
   - Since the patient was unable to move her leg at all, you would not be able to test for give-way weakness
2. Entrainment
   - Entrainment is a technique used to elicit a functional tremor, not weakness
3. Astasia abasia
   - This is a descriptor of a functional gait, not weakness
4. **Hoover sign**
   - **The absence of leg extensor effort in the contralateral leg when attempting to raise/flex the weak leg indicates functional weakness**

Question 4

A 10 month old presenting for a well child visit is noted to not be able to sit unsupported. Which is the next best step?

1. Reassure parents that sitting independently can develop by 12 months
2. Refer to neurology for motor delay
3. Evaluate for hearing loss
4. Provide prescription for physical therapy

Question 4 - Explanation

1. Reassure parents that sitting independently can develop by 12 months
   - Although minor delays can be normal, sitting independently is a key developmental milestone that should be closely assessed if delayed
2. **Refer to neurology for motor delay**
   - **Most infants can sit unsupported by 6-8 months. Failure to achieve this milestone by 10 months is concerning for potential motor or neurologic delays, warranting further evaluation**
3. Evaluate for hearing loss
   - Hearing loss can impact language development but should not affect sitting ability
4. Provide prescription for physical therapy
   - Although PT may help, a detailed evaluation is first necessary to identify an underlying cause

Question 5

A 10 year old presents with three weeks of intermittent double vision and drooping eyelids that worsen as the day progresses. On exam, you note ptosis and difficulty maintaining upward gaze. Which additional physical exam maneuver would be most helpful to confirm the suspected diagnosis?

1. Assess for nystagmus during lateral gaze
2. Perform a Romberg test to assess balance
3. Test deep tendon reflexes in upper and lower extremities
4. Test for fatigability by having the child hold their arms outstretched

Question 5 - Explanation

1. Assess for nystagmus during lateral gaze
   - This would suggest a different diagnosis, such as vestibular or cerebellar dysfunction
2. Perform a Romberg test to assess balance
   - This tests vestibular or proprioception problems, not fatigability
3. Test deep tendon reflexes in upper and lower extremities
   - Reflexes are typically normal in myasthenia gravis
4. **Test for fatigability by having the child hold their arms outstretched**
   - **Progressive ptosis and double vision, combined with fatigability, strongly suggests myasthenia gravis. Testing for fatigability by having the patient hold their arms outstretched or maintaining upward gaze can help confirm this**

Questions?

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