### UMN & LMN Disorders

**Last updated: April 12, 2020**

**WEAKNESS (Loss of Voluntary Movement):**
- Treatment:
- Muscle Tone Abnormalities: 1
- Treatment of Spasticity: 1
- Fatigability: 3

**PYRAMIDAL UMN LESION:**
- Acute UMN lesion
- Chronic stage of UMN lesion
- Pseudobulbar Paralysis
- "Locked in" Syndrome (s. Pseudocoma)

**LMN LESION:**
- Bulbar Paralysis
- Primary sensory neuron lesion
- Spontaneous Movements

**LESION LOCALIZATION GUIDE**
- Acute Generalized Weakness
- Epidemic Weakness
- Drop Attack

**DECORTICATE/DECEREBRATE RIGIDITY** → see p. A61 >>

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**WEAKNESS (LOSS OF VOLUNTARY MOVEMENT):**

- **Muscle cannot exert normal force** – most important clinical feature of motor neuron (UMN, LMN) disorders:
  - **PARESIS** – reduced voluntary movement;
  - **PARALYSIS** (s. **PLEGIA**) – complete loss of voluntary movement.

- **PALSY** is older term (has been used interchangeably with either paralysis or paresis); currently, its use is confined to historical diagnoses (e.g. Bell’s palsy, cerebral palsy).

- **distribution of paralysis / paresis** is defined by prefixes:
  - **mono**- (one limb); 2
  - **para**- (both legs); 2
  - **hem**- (limbs on one side of body); 2
  - **quadri**- or **tetra**- (all four limbs); 2
  - alternating (s. crossed) hemiplegia – hemiplegia on one side with contralateral cranial nerve palsy.
  - bilateral paresis – both arms.

- if clinical evaluation of weakness is limited by pain or lack of patient effort, needle EMG can provide objective information.

**TREATMENT:**
- occupational therapist and physical therapist:
  1. Strengthening & stretching exercises - maintain weak muscles in maximum tone, keep joints from developing contractures.
  2. Patient is trained to use adaptive movements - to facilitate function, to use canes and walkers.

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**MUSCLE TONE ABNORMALITIES**

Muscle tone changes usually accompany weakness!

- **muscle tone** = resistance to passive muscle stretch

  - **tone is evaluated by passive movements of limb.** see p. D1 >>
  - **main components of muscle tone:**
    1. **low-level background m. co-activation.**
    2. **alteration in stretch reflexes** (most important determinant of pathological alterations in tone!): via changes in rate of discharge in **α**-neurons → changes in general excitability of motor neuron pool
    3. **viscoelastic properties** of muscle & tendons (contribute to increased tone in chronic spasticity and rigidity!).

**TONUS** - *KREPSIONIA (S. FLACCIDITY) - LMN disease, cerebellar disease, sensory nerve damage.*

**TONUS:**
- a) **SPASTICITY** - UMN disease: resistance depends on:
  - **velocity of passive motion** - if limb is rapidly moved: free interval → gradual increase in tone (lengthening reaction, s. spastic catch due to hyperactive muscle stretch reflex) → sudden decrease (inversus muscle stretch reflex) as limb goes through its range of motion ("cheap-knife").
    - "It is important to move rapidly because rate of stretch is important in eliciting maximum stretch reflex response."
  - **direction** of passive motion - tone is greater in **antigravity muscles** (arm flexors, leg extenders & adductors).

**Modified Ashworth scale (MAS)** measures resistance during passive soft-tissue stretching and is used as a simple measure of spasticity (Bohannon and Smith, 1987).

- 0 No increase in muscle tone

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**Rhythmic movements** (e.g. walking, running) require stereotyped sequence of muscle activation.

**Voluntary movements** - most complex - goal-directed, initially require conscious direction.

- **spinal cord** contains circuitry for reflex responses and some rhythmic motor patterns.
- **brain stem** contains circuits for more complex patterns of motor movements including rhythm generators.
- **cortex** is command center that plans and initiates movements and uses reflex and patterned responses of brain stem and spinal cord to generate details of movement.

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**LOWER MOTOR NEURON (LMN)** → motor neuron directly innervating striated skeletal muscles.

**UPPER MOTOR NEURON (UMN)** → term used in two senses:
- **sensory stricts** – cortical neurons forming tractus pyramidalis.
- **sensory late** – all neurons forming descending tracts that ultimately play on LMN (tr. pyramidalis, tr. reticulospinalis, tr. rubrospinalis, tr. vestibulospinalis, etc.)

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1. Slight increase in muscle tone, manifested by a catch and release or by minimal resistance at the end of the range of motion when the affected part(s) is moved in flexion or extension.

2. More marked increase in muscle tone through most of the ROM, but affected part(s) easily moved.

3. Considerable increase in muscle tone, passive movement difficult.

4. Affected part(s) rigid in flexion or extension.

b) RIGIDITY - extrapyramidal UMN disease

- Symmetric, symmetric resistance at all angles of motion; doesn’t depend on passive motion velocity; constant through range of motion.
- "lead-pipe" rigidity (plastic type of rigidity – limbs accept positions they are left in) or "cogwheel" rigidity (superimposed on tremor).
- Cogwheeling is enhanced by voluntary movement of contralateral limb (reinforcement).
- N.B. cogwheeling can occur in patients with essential or familial tremor (may be confused with parkinsonism), but clinician feels alternate activation of flexors and extensors that produce tremor, but does not feel hypertoncity!

PARATONIA

-produce tremor, but does not feel hypertonicity!

with parkinsonism), but clinician feels alternate activation of flexors and extensors that produce tremor, but does not feel hypertoncity!

c) PARATONIA / GEGENHALTEN (German "hold against") – diffuse forebrain dysfunction (dementia, frontal lobe or thalamic disorders).

- pseudovoluntary* resistance by patient (against any passive movement of limb (i.e. not true increase in muscle tone)) - each attempt at moving limb by examiner is met with equal and opposing force.
- felt as increase in tone that fluctuates with repetitive passive movements.
- often accompanied by tonic grasp reflex

*It is involuntary to extent that patient has great difficulty in voluntarily suppressing urge to resist.

** TREATMENT OF SPASTICITY **

**Spasticity may be helpful in compensating for weakness, especially in gait - overzealous treatment of spasticity (esp. with systemic drugs) may in fact cause decrement in function!**

1. Stretching exercises (to maintain joint mobility)

- at least passive range of motion (ROM) activities (to prevent contractures) are started in immediate setting if patient cannot participate actively

- full range motion exercises; avoid over-stretching of soft tissues (deformity may result!).

- avoid knee hyperextension.

- keep feet flexed at 90° (use pillow between soles and bed foot).

- electrical stimulation of antagonist muscles and splinting may help.

2. Drugs with systemic effects (sedation is usually limiting barrier, esp. for school-age children!) - primarily used in patients who are confined to wheelchair or bed (drugs allow easier transfers from bed to chair, facilitate hygiene, alleviate painful flexor spasms).

1) GABA antagon - BACLOFEN (20-240 mg/d in divided doses q8hs*) - most effective drug available.

- originally synthesized as an anticonvulsant but it was found to have no significant anticonvulsant activity.

- in severe cases - intrathecal administration (pump).

- intrathecal baclofen given to normal patients does not interfere with movement or decrease strength, but the same dose given to a spastic patient markedly decreases spasticity and muscle tone.

- activation of the GABA receptors reduces the influx of calcium into the presynaptic terminals, the result being a reduction in the release of excitatory transmitters:

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+----------------+----------------+
| Intercostal nervous | Excitatory terminal |
| neuron           |                |
| BACLOFEN interacts with release of excitatory transmitters |
+----------------+----------------+
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Baclofen withdrawal – "itchy, twitchy, bitchy" (severe itching without a rash, excessive sweating, pruriasis, mood fluctuation, rebound spasticity) can progress to severe rigidity, fever from increased muscle activity, irritability/confusion/agitation/hallucinations, labile blood pressure, seizures; potentially lethal - can lead to rhabdomyolysis, DIC, organ failure, and can look like autonomic dysreflexia, malignant hyperthermia (vs. opioid withdrawal), septic shock.

Treatment: oral baclofen, IV benzodiazepines (DIAZEPAM, 2-5 mg 6 hours) or CYCLOSPORINE (6 mg 6 hours for 24 hours), if due to IT system failure and unable to replace full dose orally (try oral baclofen 20 mg q6-8hrs).

**Baclofen overdose may result in somnolence, respiratory depression, hypotension, seizures, rostral progression of hypotonia, coma.**

Treatment: no antidote (if no heart conduction defects), PHENYTOIN 0.5-2 mg often reverses the somnolence and respiratory effects), aspirate drug from pump reservoir, aspirate 30-40 mL of CSF. The central effects of an overdose should clear in 24 to 48 hours.
2) GABA agonists - diazepam (2-4 mg at bedtime) - for leg spasms that interrupt sleep; CNS depressant!

- unlike baclofen, which directly activates GABA receptor; diazepam works only when GABA is released, and it enhances response to the transmitter.

3) e2-agonists (central muscle relaxants):
   a) tizanidine (Zanaflex®) (4-8 mg q4h)
   b) clonidine.

4) direct muscle inhibitor - dantrolene (25 → increase up to 100 mg qid) - for nonambulatory patients; no cognitive / sedative adverse effects!

5) cyclobenzaprine (Flexeril®, Amrix®)
   - dosage: 10 mg q 6 h (5 and 10 mg tablets).
   - chemical relation to first-generation tricyclic antidepressants.
   - mechanism of action is unclear; studies from 1980s in rats indicate that drug activates locus ceruleus → release of norepinephrine in ventral horn of spinal cord → inhibitory action on alpha motor neurons.
   - decreases pain in first two weeks, peaking in first few days, but has no proven benefit after two weeks (therapy should not be continued long-term).
   - not useful for spasticity due to neurologic conditions such as cerebral palsy.
   - adverse effects: drowsiness (38% of patients), dry mouth (24%), urinary retention (in males with large prostates).

6) Methocarbamol (Robaxin®) - central muscle relaxant.
   - dosage: 1500 mg q 6 h for 2-3 days then decrease to maintenance 1000 mg q 6 h.
   - adverse effects: CNS depressant, may cause urine to turn black, blue, or green.

7) Carenopropranol (Somadine) - not recommended! (converted into benzdiazepines → addictive potential?)

8) Chlorzoxazone (Lorzone®, Paralon®)

9) Metaxalone (Skelaxin®)

10) Orphenadrine

3. Local injections of motoludum toxin (effect for 3-6 months) - no deleterious effects on helpful spasticity; target muscles:
   1) leg adductors - to facilitate nursing care.
   2) arm muscles - to relieve painful spasms; article about arm spasticity and botulinum toxin injections: http://www.medscape.org/viewarticle/865168
   3) gastrocnemius–soleus muscle - to convert toe walking to plantigrade foot placement.

4. Surgical measures – orthopedic
   - if fixed contracture has developed → surgical tendon release (most commonly - Achilles, thigh adductor, hamstring tendons).
   - for CP kids, ortho procedures are delayed after spasticity is addressed.

5. Surgical measures – neurosurgical
   - N.B. ablative procedures (convert spastic into flaccid paralysis) - reserved for extensive or complete loss of cord function!

   1) Intrathecal baclofen delivery system: see Op220 →
   2) Selective posterior rhizotomy (procedure of choice for spasticity due to cerebral palsy).
      - exposure of cauda equina (through L5 laminctomy).
      - after anatomic identification of L1 root at its exit foramen, S1, anterior root is identified by low-frequency stimulation.
      - fascicles of each of L1-S1, posterior roots are isolated and stimulated - those fascicles, stimulation of which causes ipsilateral tetanic or multisegmental motor responses or any contralateral motor responses*, are sectioned.
      - *intraoperative clinical responses are correlated with intraoperative EMG.
      - usually, 60-80% fascicles are sectioned (diminished sensation lasting no longer than several weeks) - if patients are young, have adequate cognitive function and aggressive physical therapy is carried out postoperatively, results are excellent.
      - spare fascicles innervating sphincters!
      - can be done percutaneously - may be performed at any segment (e.g. RF, local or general anesthesia, fluoroscopy and low-frequency stimulation to verify electrode positioning, therapeutic response may last several years)
   3) Limited ablative procedures (for spasticity confined to bladder or to single limbs)
      a) Interhemispheric spinal cordotomy of sacral segments (for spinal bladder).
      b) Selective peripheral neurotomy (sectioning nerve fascicles - identified by intraoperative stimulation - which maintain spastic tone; e.g. tibial nerve at popliteal region for spastic foot; obturator neurotomy).
   4) Spinal cord stimulation (SCS) - better when stimulating epidural electrode is implanted caudal to level of injury.
      - also benefits reflex and voluntary bladder control in MS.
   5) Bisphosphonates (longitudinal myelotomy) - dividing cord into anterior and posterior halves over segments involved in flexor spasms (typically L5-S1) - interrupts local reflex arcs; may not prevent spasms triggered by stimuli from segments rostral to L1 or caudal to S1.
   6) Stereotactic denervation (limited usefulness in management of spasticity)
      - N.B. selective posterior rhizotomy provides much higher success rate.
      - indication: severe spasticity with congenital choreoathetosis; thalamotomy controls choreoathetosis, if subsequently worse spasticity develops → ipsilateral denervation.

FATIGABILITY
- Patriot: feeling of being tired and not being able to put out full effort.

Normal fatigue - results from intense muscular contraction.
   - accompanied by firing frequency in motor-unit - result of reduced excitatory drive to motoneurons (central mechanism).

Fatigability (dysfunction at neuromuscular junction) - muscles become weaker and weaker with repetitive but normal use (inability to sustain performance of activity).
Pyramidal UMN lesion

Acute UMN lesion

Cerebral shock - transient depression of reflex activity below level of injury; in addition to paralysis:
1. Hypotonia of muscles
2. Absence of reflexes
3. Movements are slow, coarse
4. Paralysis involves large areas

If lesion transects spinal cord
- CEREBRAL SHOCK
- CORTICOSPINAL lesion: distal muscle groups are affected more severely than proximal ones, and axial movements are spared unless lesion is severe and bilateral.
- CORTICOBULBAR lesions: weakness only in lower face and tongue; extracular, upper facial, pharyngeal, and jaw muscles are almost always spared (but with bilateral corticobulbar lesions → PSEUDOBULBAR PALSY).

Bilateral pseudobulbar palsy may be erroneously regarded as reactive depression because of diagnosis.

Chronic stage of UMN lesion

1. Paralysis involves large areas (hemi-, para-, quadriplegia) – at and distal to capsula interna
   - small lesion affects large body regions; rostral to capsula interna, pyramidal neurons are dispersed (e.g. small stroke in arm area of motor cortex can produce brachial monoparesis).
   - CORTICOSPINAL lesion: distal muscle groups are affected more severely than proximal ones.
   - CORTICOBULBAR lesions: weakness only in lower face and tongue.
   - Extracular, upper facial, pharyngeal, and jaw muscles are almost always spared (but with bilateral corticobulbar lesions → PSEUDOBULBAR PALSY).

2. Muscle atrophy of disease only (late and slight).

3. Spasticity (muscle tone)
   - spasticity is more pronounced in ANTIGRAVITY MUSCLES – areflexia, leg extensors (WERNICKE-MAN posture).
   - N.B. when intact, tri. pyramidalis constantly inhibits muscle tone!
   - resistance depends on velocity and direction of passive motion – “clasp knife” phenomenon.
   - if patient can walk, spasticity causes SCIALLY DRAGGED (in bilateral lesions), leg circumduction (in unilateral lesions).
   - pure pyramidal tract lesions cause mild paralysis without spasticity – because control of tone is mediated by other tracts (particularly corticorubrospinal and corticobulbar). This may explain why degrees of weakness and spasticity often do not correspond.

4. Hyperreflexia (lost UMN inhibition on various reflexes):
   - 1) muscle stretch reflexes
   - 2) CELOWS (rhythmic, rapid alternation of muscle contraction and relaxation caused by sudden, passive tendon stretching)

5. Absent normal skin reflexes (abdominal, cremasteric).

6. Sympathetic

7. Movements are slow, coarse but with normal rhythmicity and coordination (e.g. finger-nose-finger and heel-knee-shin performed are slow but adequately; “incoordination” is obvious with rapidly repeated movements (e.g. tapping index finger on thumb).

EMG - normal number of motor units are activated at given frequency but in which maximum discharge frequency is decreased (vs. LAMN lesions - number ↓, frequency normal).

Pseudobulbar Paralysis

- bilateral corticobulbar tract lesion (i.e. central-supranuclear palsy of CN 7, 9, 10, 12):
  1) spastic 3D (dysarthria, dysphonia, dysphagia)
  2) Incomplete palsies
  3) Hyperactive gag reflex, hyperactive facial and jaw jerks (CN 5 → CN 7);
     uvula movements are more vigorous on reflex than on volition (i.e. uvula does not move well (or at all) on phonation, but vigorous response is seen in pharyngeal or gag reflex).
  4) Oral automatisms (snout, suck, etc)
  5) Emotional incontinence (reflexive crying) and spasmodic, mindless laughing with minimal provocation – release of limbic functions; patient is aware of lack of control!

*can be erroneously regarded as reactive depression because of diagnosis.

H. SSRIs (FLUOXETINE, SERTRALINE, NORFLOXACIN, LEVOFLOXACIN, NUEDEXTA (DEXTROMETHORPHAN HYDROMORPHONE + QUINIDINE SULFATE)) capsules - FDA approved first treatment for pseudobulbar affect.

- most common causes: bilateral hemisphere lesions, bilateral lacunar infarctions in internal capsule.
- patients may have dementia (due to pathology involving bilateral frontal areas).

accompanied by amplitude of muscle action potentials. N.B. with exception of neuromuscular junction disorders, fatigue is rarely complaint of diseases of motor unit!!

"Fatigue,” “tiredness,” “lack of energy” are common complaints in following disorders:
1) UMN disease (bilateral corticospinal tract or extrapyramidal disease)
2) multifocal CNS disease (e.g. established MS)
3) sleep disorders, psychiatric and behavioral disorders
4) chronic fatigue syndrome
5) fibromyalgia
6) renal, hepatic, cardiac, pulmonary diseases, anemia
7) hyperventilation, hypoglycemia

Symmetrical hyperactive reflexes in presence of down-going toes are usually normal!

N.B. when intact, tri. pyramidalis constantly inhibits muscle tone!

\[\text{Symmetrical hyperactive reflexes in presence of down-going toes are usually normal!}\]

\[\text{N.B. when intact, tri. pyramidalis constantly inhibits muscle tone!}\]
**“Locked-In” Syndrome (S, Pseudobulbar)**

- Bilateral basis pontis lesion; i.e. damage to corticospinal-corticopontine-corticobulbar tracts below reticular formation (therefore sparing consciousness) but above ventilatory nuclei of medulla (therefore, precluding defecation).

  - Most commonly due to basal artery infarction; other causes: central pontine myelinolysis.
  - Almost complete de-afferentation.
  - Causes of spontaneous movements can reside at any level of nervous system.

1. Paralysis of individual muscles
2. Areflexia
3. Fasciculations

4. Cause of spontaneous movements can reside at any level of nervous system.

5. Paralysis of jaw-face-bulbar muscles (facial & bulbar diplegia; no volitional vocalization?) – due to CN7 nuclei, corticobulbar tracts damage.

   - Very resembles coma, but
   - 1) Fully conscious and mentally intact
   - 2) can feel, see, hear
   - 3) preserved vertical eye movements – the only way to communicate!

   - If lesion also affects dorsal pontine tegmentum – sudden coma, pinpoint pupils, opisthotonus, hyperthermia, progression to death.

   - Patients must be identified rapidly, because recovery to independence can occur over weeks to 3-4 months (magnetic stimulation of motor cortex producing motor evoked potentials is positive prognostic feature).

Similar state may occur in severe Guillain-Barré syndrome, but vertical eye movements are not selectively spared.

**LMN lesion**

*“Three A’s”*

1. **Areflexia** (all reflexes ↓↓↓ or absent – grade 1 or 0) – lost afferent portion of reflex arc!

2. Areflexia

The absence of tendon reflexes does not cause weakness but decreases tension on muscle spindles → tendon reflexes.

3. Areflexia

**Bulbar Paralysis**

Peripheral (LMN) palsies of CN 9, 10, 12:

1. **3D**. dysphagia, dysphonia, dysarthria

2. Absent swallowing & gag reflexes (vs. in pseudobulbar paralysis!)

3. Tongue atrophy and fasciculations

4. Velum palatii does not elevate during “ahaa!”; uvula deviates to intact side.

**Primary sensory neuron lesion**

1. Hypotonia

2. Areflexia (absent all reflexes) – lost afferent portion of reflex arc!

3. Volitional movements and their strength remain normal!

4. Patient’s appearance looks normal.

5. Coordination normal only with eyes open.

**Spontaneous Movements**

- Movements that occur in entire limb or in more than one muscle group concurrently are caused by LMN disease

  a) extrapyramidal

  p. Movie>>

  b) seizure disorders

- Movements confined to single muscle are likely to be reflection of disease of motor unit (LMN of brain stem and spinal cord = muscle).

**Fasciculations, Fibrillations**

Fasciculations - visible fine, rapid, flickering / twitching movements in small group of muscle fibers (fascicles or bundles).

Fibrillations - invisible contractions of individual muscle fibers - can be detected by EMG

- vary irregularly in frequency and extent.
- do not move joint!

- EMG:

  1) Disease anterior horn cell may spontaneously discharge → fasciculations.

  2) Aech receptors in denervated muscle fibers fail to cluster at motor end plate and become spread across muscle membrane → muscle fibers may then discharge spontaneously → fibrillations.

  - Fasciculations are seldom seen with peripheral nerve lesions (atrophy without fasciculations is more compatible with peripheral nerve lesion).

  - In long-standing muscle denervation and reinnervation, motor unit size enlarges and fasciculations may be so large as to produce movement of limbs, particularly of fingers (minispiomyoclonus).
### MYOKYMBIA
- continuous involuntary quivering or rippling (numerous, repetitive fasciculations) of muscles at rest.
- caused by spontaneous, repetitive firing of groups of motor units – specific EMG pattern.

### Etiology
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### Cramp v. Spasm
- **Cramp**: sudden *transient* (up to few minutes) intense tonic contraction of *single* *multiple* muscles.
- associated with severe pain.
- prolonged severe cramps can produce *muscle injury* (e.g. creatine kinase↑ in blood, myoglobinuria).

### Electrophysiology
- appears like normal motor unit; no features of muscle denervation
- complex, longer duration, higher amplitude

### Prevention for certain muscle groups
- calves and thighs

### Nature
- repetitive twitch in same muscle fascicle; may be accompanied by frequent cramps
- random nonsterotyped twitches of many parts of muscles

### Associated weakness or atrophy
- 
- 
- 
- 

### Electrophysiology
- muscle stiffness (e.g. in incompletely relaxed muscles)
LESION LOCALIZATION GUIDE

Lesion of pyramidal UMN:

A) above DECUSATIO PYRAMIDUM (pyramidal tract) → CONTRALATERAL hemiplegia (including lower face; lesions below pons spare face).
   • Bilateral lesions can cause PSEUDOBULBAR PARALYSIS. See above >>

B) below DECUSATIO PYRAMIDUM (anterior and lateral corticospinal tracts) → IPSILATERAL plegia:
   • in some cases only paresis (esp. in trunk muscles) – due to contralateral tr. corticospinalis and. (if well-developed – may account for some degree of recovery).

In general, only BILATERAL lesions cause UMN-type weakness in trunk and cranial muscles!

PATTERNS OF WEAKNESS

Lesion Location Pattern of Signs

Medial hemispheres (leg area) Spastic leg paraparesis with no sensory level
Thoracic spinal cord Spastic leg paraparesis, thoracic sensory level
Lumbar spinal cord Flaccid paraparesis, double incontinence (flaccid bladder and sphincters)

Paraparesis implies lesion below cervical cord; exceptions:

1) leg areas (on medial side of each hemisphere, at apex of motor strip) face each other in interhemispheric fissure - interhemispheric fissure lesion - parietal operculum lesion - motor cortex injury - ACP ischemia - superior sagittal sinus thrombosis could affect both legs - PARAPARESIS SIMULATING SPINAL CORD LESION.

N.B. this possibility seems more theoretical than real, however, because no well-documented cases have been reported!

2) HYDROCEPHALUS may be another supraspinal cause (parietal operculum leg fibers are stretched most by dilated lateral ventricles).

ETIOLOGY

In adults, most common cause of paraparesis is MULTIPLE SCLEROSIS (“spastic paraparesis of middle life”).

Other causes:

- cervical spondylotic myelopathy;
- hereditary spastic paraparesis;
- primary lateral sclerosis;
- HIV-1 infection, HV myelopathy.

DIAGNOSTIC APPROACH begins with spinal MRI or myelography.

HEMIPARESIS:

In adults, most common cause of hemiparesis is MULTIPLE SCLEROSIS (“spastic paraparesis of middle life”).
--- lesions are UNILATERAL (1): ---

**Cerebral cortex**
- **Contingual weakness** (arm, leg; face; sometimes tongue)*.
- **LEFT HEMISPHERE**: aphasia, apraxia.
- **RIGHT HEMISPHERE**: left hemi-inattention, extinction of sensory stimuli, constructional apraxia, spatial disorientation. 
- **Hemorrhagic hemispheric** on weak side. 
- **Cortical sensory loss** (decreased graphesthesia, stereognosis, point localization).
- **Horizontal eye deviation** (toward lesion side).

**Internal capsule (posterior limb)***
- **Contingual weakness** (face = arm = leg); face may be spared! 
- No sensory loss or aphasia.

**Brain stem** see p. ASH >
- **Contingual weakness** (arm = leg) + ipsilateral peripheral cranial nerve palsy.

**Midbrain (crus cerebelarum)**
- Lesion of CN3 (Weber syndrome); red nucleus, superior cerebellar pedicle (limb ataxia contingent to hemispheric side).

**Pons (basis pontis)**
- Lesion of CN6 (Foville syndrome, CN7 (Millard-Gubler syndrome); internuclear ophthalmoplegia.

**Medulla (pyramidis)**
- Lesion of CN12; face spared.

**Cervical spinal hemisensory** (Brown-Séquard syndrome)
- Ipsilateral weakness sparing face.
- Ipsilateral loss of proprioception and vibration.
- Contingual loss of pain and temperature.

* (face) & arm > leg (MCA territory); face & arm < leg (ACA territory).

**Lesion in internal capsule** may be very small and still cause complete hemiparesis;
- **pure motor hemiplegia** - weakness that affects entire side of body equally without associated sensory signs;
- small strokes (lacunar infarcts in posterior limb near genu) can produce more focal weakness (e.g. weakness in face and arm - dysarthria-clumsy hand syndrome).

Another possible cause of dysarthria-clumsy hand syndrome - lacunar infarction in **basis pontis** (esp. at junction of upper third and lower two-thirds) – lesion of corticobulbar & corticopontocerebellar fibers.

In general, hemiparesis usually signifies cerebral lesion and etiology* is likely to be denoted by clinical course + brain-imaging.

* in adults - most commonly cerebral infarction / hemorrhage

**DIAGNOSTIC APPROACH** - brain CT; if CT normal and ischemic stroke is unlikely → MRI of brain → Imaging.

**Acute hemiparesis**
- **usually vascular pathogenesis**
- **bacterial infection** of normal vessels
- **hemorrhage into primary / metastatic brain tumors**
- focal inflammatory lesion (multiple sclerosis, sarcoidosis)
- **acute bacterial abscess**

**Subacute hemiparesis**
- **subacute subdural hematoma**
- **infection** - cerebral bacterial abscess, fungal granuloma or meningitis, parasitic infection.
- **malignant primary / metastatic neoplasms**
- N.B. AIDS may present with subacute hemiparesis due to toxoplasmosis or primary CNS lymphoma!

**Chronic hemiparesis** (slowly develops over months)
- **histologically benign neoplasm**
- **unruptured AVM**
- **chronic subdural hematoma**
- **degenerative disease.**

**TETRAPARESIS**
- lesion locations are BILATERAL (1):

**Lesion Location** | **Pattern of Signs**
--- | ---
**Cerebral hemispheres** | Pseudobulbar palsy, decorticating posturing (large acute lesions)
**Midbrain** | Coma, mid-size poorly reactive pupils, decerebrate posturing
**Basis pontis** | “Locked-in” syndrome
**Cerebromedullary junction** | Les ≥ arm, weak & facies & tongue, facial hypalgesia (descending tract of CN 5)
**High cervical** | No cerebral signs (cranial nerve palsies, etc)
**Mid cervical** | Preservation of shoulder movements
**Peripheral nerves (e.g. acute demyelinating polyneuropathy)** | Dorsal weakness
**Muscles (myopathy)** | Proximal weakness

**MONOPARESIS**
A. With pain:
- 1. Congressive lesion of spinal cord
- 3. Peripheral nerve entrapment syndromes
B. Painless:
- 1. Thoracic spinal lesions (e.g. ALS, tumor, demyelinating plaque).
- 2. Cerebral lesions (theoretically; because abnormal signs are almost always present in leg, i.e. syndrome is really hemiparesis) - weakness predominantly in distal and nonantigravity muscles.

**BIBRACHIAL PARESIS**
- arms hang limply at side while patient walks with normal movements of legs.

1. Cervical LMN lesion in some cases of ALS (with or without UMN signs in legs).
2. Myopathy of unusual distribution.

**NECK WEAKNESS (“FLAPPY HEAD”) SYNDROME**
- Never in UMN disorders!
1. ALS
2. Myasthenia gravis
3. Polymyositis
4. Tick-borne encephalitis

**HYPOREFLEXIA**

1. Normally hypoactive reflexes.
2. Hypothyroidism (delayed relaxation phase of reflex) - this unique "hypoactive" reflex is classic for this metabolic abnormality (best seen in ankle jerk).
3. Spinal shock
4. Acute stroke (initially, there is hyporeflexia on hemiparesis side; later, hyperreflexia develops).
5. Holmes-Adie syndrome (asymptomatic areflexia with large pupil that reacts to accommodation but not to direct light) see p. Eyelid >>
6. Myopathy
7. Neuropathy (incl. radicalpathy)
   (N.B. patient with no reflexes usually has neuropathy!)

**BILATERAL HYPERREFLEXIA**

1. Normal anxious patients
2. Metabolic causes (e.g. hepatic and uremic encephalopathy)
3. Spinal cord compression
4. Multiple sclerosis
5. Amyotrophic lateral sclerosis
6. Multiple small strokes (stupor laccunae)
7. Familial spastic paraplegia
8. Cerebral palsy
9. Parasagittal intracranial mass (may affect cortical leg fibers)
10. Hydrocephalus (may stretch leg fibers)

**VOICE**

- HypoLMN impairment → soft, weak, low-pitched, monotonous voice.
- UMN impairment → harsh and strained voice.

**ACUTE GENERALIZED WEAKNESS**

- pace of disease is so rapid that by time patient is seen in hospital weakness has become generalized.

<table>
<thead>
<tr>
<th>Lesion Location (cause)</th>
<th>Differentiating Features</th>
</tr>
</thead>
<tbody>
<tr>
<td>Midbrain (stroke, trauma, tentorial herniation)</td>
<td>Coma, mid-size poorly reactive pupils, decorticate/decerebrate posturing, hyporeflexia, bilateral Babinski signs</td>
</tr>
<tr>
<td>Basal pontis (stroke)</td>
<td>&quot;Locked-in&quot; syndrome</td>
</tr>
<tr>
<td>Spinal cord (trauma, infarction, metastatic tumor, transverse myelitis)</td>
<td>Spinal shock</td>
</tr>
<tr>
<td>Polyradicular neuropathy (acute inflammatory demyelinating polyneuropathy, tick paralysis, poliomyelitis)</td>
<td>Limb weakness (legs before arms), areflexia, absent or flexor plantar response, minimal distal sensory loss without sensory level</td>
</tr>
<tr>
<td>Neuromuscular junction (botulism, organophosphates)</td>
<td>Ocular (incl. loss of pupillary light reflex) and pharyngeal weakness → absolutely generalized weakness with no sensory loss, no decorticate / decerebrate posturing; preserved consciousness</td>
</tr>
</tbody>
</table>

**EPISODIC WEAKNESS**

- attacks of severe weakness occurring in patient with baseline normal strength.

<table>
<thead>
<tr>
<th>Disorders</th>
<th>Key Features</th>
<th>Diagnostic Tests</th>
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</thead>
<tbody>
<tr>
<td>COMMON</td>
<td>Transient ischemic attack</td>
<td>All symptoms begin at once (abrupt and simultaneous onset of weakness in all muscles that will be affected during attack)</td>
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<td>Carotid ultrasound</td>
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<tr>
<td>LESS COMMON</td>
<td>Partial motor seizure, Todd's paresis (postural weakness)</td>
<td>Gradual &quot;march&quot; of symptoms in several seconds to few minutes</td>
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<td></td>
<td>EGG</td>
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<td></td>
<td>Myasthenia gravis</td>
<td>Fatigability, recovery with rest; predilection for ocular and cranial muscles</td>
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<td>Tension test, repetitive stimulation test</td>
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<tr>
<td></td>
<td>Hysteria</td>
<td>Normal reflexes, nonanatomic distribution of sensory loss</td>
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<td></td>
<td>Caraplexy</td>
<td>Triggered by emotion; association with other features of narcolepsy; episodes very brief</td>
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<tr>
<td></td>
<td>Sleep study</td>
<td></td>
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<tr>
<td></td>
<td>Sleep paralysis</td>
<td>Narcolepsy; terminated by touch</td>
</tr>
<tr>
<td></td>
<td>Sleep study</td>
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<tr>
<td></td>
<td>Drop attacks</td>
<td>Sudden loss of postural tone without loss of consciousness</td>
</tr>
<tr>
<td></td>
<td>MRI, MRA, X-ray of cervical spine with flexion-extension, EGG</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Negative myoclonus</td>
<td>Sudden, brief, rapid, unpredictable (shocklike) inhibition of muscle tone (single muscle or entire body)</td>
</tr>
<tr>
<td></td>
<td>RARE</td>
<td>Periodic paralyses</td>
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<tr>
<td></td>
<td></td>
<td>Serum K⁺</td>
</tr>
</tbody>
</table>

**DROP ATTACK**

- sudden falling spell (loss of postural tone);
  - no warning, no loss of consciousness!!!
  - attack is very brief; no postictal symptoms; person is immediately able to get to his feet after hitting ground.
  - pathophysiology - dysfunction of pyramidal tracts in medulla / high cervical cord.
  - etiology
    1) Brief ischemia (e.g. vertebralbasilar ischemic attack)
    2) transient MECHANICAL COMPRESSION.
a) ligament holding odontoid in place destroyed by RA or trauma: head movement (esp. extension) → excessive odontoid movement → compression of cervicomedullary junction.

b) chronic cerebellar tonsillar herniation (characteristic of Chiari malformation).

c) severe congenital cervical spinal stenosis during Valsalva maneuvers or after falls.

d) idiopathic drop attacks in elderly women; benign prognosis.

• differentiate from disorders with very brief loss of consciousness (unnoticeable by patient):
  1) akinetic seizures (H: EEG)
  2) syncope (H: history of brief warning).

BIBLIOGRAPHY for ch. “Movement disorders, Ataxias” → follow this LINK >>