TONUS disorders

- Rhythmical movements
  - Motor system generates three general types of movements:
    - Reflex responses: simplest form of coordinated movement - rapid, stereotyped involuntary movements elicited by sensory stimulus that requires quick reaction at involuntary level.
    - 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.

**TONUS: Acute UMN Lesion**

- Chronic stage of UMN lesion
- Pseudobulbar Paralysis
- Locked in Syndrome (s. Pseudocoma)

**TONUS: Decerebrate Rigidity**

- Increase in tone (spasticity and rigidity).
- Viscoelastic properties of muscle & tendons (contribute to increased tone in chronic spasticity).

**TONUS: Decorticate/Diabrenergic Rigidity**

- Decorticate rigidity results from damage to the cerebral cortex.
- Diabrenergic rigidity results from damage to the brainstem.

**Weakness (Loss of Voluntary Movement)**

- Muscle cannot exert normal force - most important clinical feature of motoneuron (UMN, LMN) disorders.
- Paralysis - reduced voluntary movement,
- Paralyzing (s. -plegia) - complete loss of voluntary movement.
- Palsy - 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),
  - para- (both legs),
  - hemi- (limbs on one side of body),
  - quadri- or tetra- (all four limbs),
  - alternating (s. crossed) hemiplegia - hemiplegia on one side with contralateral cranial nerve palsies.
- Bilateral paralysis - 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.

**Muscle Tone Abnormalities**

- Muscle tone changes usually accompany weakness!
  - Muscle tone = resistance to passive muscle stretch.
  - Tone is evaluated by passive movements of limbs; see p. D1 >>
  - Main components of muscle tone:
    1. Low-level background on-off-activity.
    2. Alteration in stretch reflexes (most important determinant of pathological alterations in tone!) - 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!)

**Tone:**

- **Kypnotonia (s. plactility)** - LMN disease, cerebellar disease, sensory nerve damage.
- **TONUS:**
  - Tone is resistance to passive motion - if limb is rapidly moved: free interval → gradual resistance → spasticity.
  - Direction of passive motion - tone is greater in antagonistic muscles (arm flexors, leg extensors & adductors).
  - Spasticity is characterized by "scissoring" over good leg (most easily seen in spastic paraplegia).
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) \textbf{SPTICITY} - extrapyramidal UMN disease

Symmetry - signature resistance at all angles of motion; doesn’t depend on passive motion or velocity; constant through range of motion. - \textit{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).

- \textbf{RIGIDITY} - often accompanied by pseudovoluntary* - tonic spasms.

- \textbf{GEGENHALTEN} (German "hold against") - diffuse forebrain dysfunction (dementia, frontal lobe or thalamic disorders).

- \textbf{PARATONIA} - tremor, but does not feel hypertonicity! - tonic spasms.

- \textbf{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.

- \textbf{LIMB TONE} - felt as increase in tone that fluctuates with repetitive passive movements.

- octone accompanied by tonic grasp reflex.

- \textit{it} is involuntary to extent that patient has great difficulty in voluntarily suppressing urge to resist.

\textbf{TREATMENT OF SPASTICITY}

\textbf{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. \textbf{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

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

- \textbullet{} avoid knee hyperextension.

- \textbullet{} keep feet flexed at 90\(^\circ\) (use pillow between soles and bed foot).

- \textbullet{} electrical stimulation of antagonist muscles and splitting 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) \textbf{GABA\textsubscript{a} agonist} - \textbf{BACLOFEN} (20-240 mg/d in divided doses q8hrs* - most effective drug available)

- \textbullet{} single oral max dose is 70-80 mg

- \textbullet{} \textit{originally synthesized as an anticonvulsant but it was found to have no significant anticonvulsant activity.}

- \textbullet{} in severe cases - intrathecal use (impaired renal function).

- \textbullet{} 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.

- \textbullet{} activation of the GABA\textsubscript{a} receptors reduces the influx of calcium into the presynaptic terminals, the result being a reduction in the release of excitatory transmitters.

- Baclofen withdrawal - "itchy, twitchy, bitchy" (severe itching without a rash, excessive sweating, priapism, mood fluctuation, rebound spasticity?) can progress to severe rigidity, fever from increased muscle activity, irritability/insomnia/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.

\textbf{Treatment: oral baclofen, IV benzodiazepines (DIAZEPAM, 2-5 mg q 6 hours) or CYPREDDP (6 mg q 6 hours for 24 hours), if due to IT system failure and unable to replace full dose orally (try oral baclofen 20 mg po q6-8hrs) \rightarrow insert temporary IT catheter and post for surgery.}

\textbf{Baclofen overdose} may result in somnolence, respiratory depression, hypothermia, seizures, rostral progression of hypotonia, coma.

\textbf{Treatment:} no antidote (no heart-conduction defects), \textit{PHYSOSTIGMINE} 0.5-2 mg often reverses the somnolence and respiratory effects, aspirate from pump reservoir, aspirate 30-40 mL of CSF. The central effects of an overdose should clear in 24 to 48 hours.
3. Local injections of neuromuscular 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/3965108.
3) gastrocnemius-soleus muscle - to convert toe walking to plantigrade foot placement.

4. SURGICAL MEASURES – ORTHOPAEDIC
- if fixed contracture has developed → surgical tendon release (most commonly - Achillels, 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!
- patients sometimes use some spasticity for support during ambulation
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 laminectomy).
- after anatomic identification of I1 root at its exit foram., 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 tetric or multisegmental motor responses or any contralateral motor responses*, are sectioned.
- *intrathecal clinical responses are correlated with intrathecal 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 limb):
- a) selective posterior rhizotomy of sacral segments (for spastic 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)."
Pyramidal UMN lesion

ACUTE UMN LESION

CEREBRAL SHOCK - tautaneous depression of reflex activity below level of injury; in addition to PARALYSIS:

1. Hypotonia of muscles
2. Absence of reflexes (muscle stretch, plantar, abdominal & cremasteric)

If lesion transects spinal cord (SPINAL SHOCK -- see p. Spinal 1), it is also accompanied by:

1. Hypotonic paralysis of bowel & bladder
2. Hypotension, anhydrosis

CHRONIC STAGE OF UMN LESION

1. Paralysis involves large areas (hemi-, para-, quadriplegia) – at and distal to capsula interna

- cortical lesions: 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; extracorporeal, upper facial, pharyngeal, and jaw muscles are almost always spared (but with bilateral corticobulbar lesions → PSEUDOBULBAR PALSY; see below) (hemiparesis)

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, tr. pyramids usually elicit muscle tone!

- resistance depends on velocity and direction of passive motion -- "clasp knife" phenomenon.

If patient can walk, spasticity causes SCISSORS GAIT (in bilateral lesions), leg circumduction (in unilateral lesions). see p. MS! >>

- pure pyramidal tract lesions cause mild pyramidal without spasticity – because control of tone is mediated by other tracts (particularly corticobulbar and corticoreticulospinal) - 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)

- abnormal reflexes -- see p. A18 >>

- only minimally increased clonus suggests UMN damage!

3) BABINSKI sign and other pathologic withdrawal reflexes (normally, they are inhibited by intact pyramidal system)

- normally only painful stimulus elicits withdrawal reflex.

- in UMN is damaged (but also in normal infants - immature CNS), lighter nonpainful stimulus may elicit withdrawal reflex - strength of response parallels extent to which UMN lesion has allowed upregulation of reflex: small hemispheric lesion - small fragment of reflex may be elicited (i.e. extension of great toe - Babinski sign; complete spinal cord transection - entire withdrawal reflex with flexion at hip, knee, and ankle) may be seen.

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

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

6. Synkinesias

7. Movements are slow, coarse but with normal rhythmicity and coordination (e.g. finger-noise-finger and heel-knee-shin are performed slowly but adequately); "incoordination" is obvious

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).
"Locked-In" Syndrome (Locked-In Syndrome, LIS) - 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 deglutition).

- most commonly due to basal artery infarction; other causes: central pontine myelolysis.
- almost complete de-energization:
  1. quadriparesis - due to corticospinal tracts damage.
  2. paralysis of horizontal eye movements (horizontal ophthalmpoplegia) - due to PPRF and CN6 nuclei, corticopontine tracts damage.
  3. 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! when patient is not actively moving eyes, spontaneous ocular bobbing may occur.
  4) eyes are open and partially blink (via inhibition of levator palpebrae) – another way to communicate!

- if lesion also affects dorsal pontine tegmentum → sudden coma, pinpoint pupils, ophthalmpoplegia, hyperthermia, progression to death.

- patients must be identified rapidly for intravenous rt-PA treatment.
- mortality rate is high (40-50%); survival in locked-in state has lasted as long as 18 years.
- 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:**

1. A(hyporeflexia) (all reflexes 2 or absent – grade 1 or 0) – lost afferent portion of reflex arc!

- N.B. reflexes present only with reinforcement (grade 1) imply intact reflex pathway and may or may not be abnormal!
  - loss of γ- motoneurons does not cause weakness but decreases tension on muscle spindles → tendon reflexes;

2. A(hypotonia)

3. Atrophy of denervation (early & severe – in 2-3 months muscle loses 50% of its mass!), abnormal electrical activity.

- maximum degree of denervation atrophy after acute injury to axons occurs in 90-120 days and reduces muscle volume by 75-80% (vs. disease atrophy does not reduce muscle volume by more than 25-30%); in 3-4 years, most of denervated fibers will have degenerated.

4. Paralysis of individual muscles (or groups of muscles)

5. Fasciculations, fibrillations see below.>>

EMG - recruitment of motor units is delayed / reduced (fewer than normal are activated at given discharge frequency).

**BULBAR PARALYSIS**

Peripheral (LMN)palsy 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 "aah"; 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.

**SPOONTAENOUS MOVEMENTS**

Cause of spontaneous movements can reside at any level of nervous system, movements that occur in entire limb or in more than one muscle group concurrently are caused by LMN disease

- a) extrapyramidal see p. Mov1 >>
- 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!
- etiology - LMN disease

1. Disease motor neuron 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 (MINISP.PMYELOCLONE).


EMG: 03.120 >>

LMN & LMN DISORDERS Mov3 (5)
**FEATURES**

<table>
<thead>
<tr>
<th>Benign fasciculations (Denny-Brown, Foley’s syndrome)</th>
<th>Malignant (neuropathologic) fasciculations</th>
</tr>
</thead>
</table>

- **Gender predilection**
  - males*

- **Predominance for certain muscle groups**
  - calves and thighs

- **Nature**
  - repetitive twitch in same muscle fascicle; may be accompanied by frequent cramps

- **Associated weakness or atrophy**
  - –

- **Electrophysiology**
  - appears like normal motor unit; no features of muscle denervation

---

**MYOKYMIA**

- 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**

  a) lesions of pons (e.g. neoplasms or multiple sclerosis) - FACIAL MYOKYMIA - nearly continuous twitching of facial muscles (palpebral fissure narrowing, continuous undulation of facial skin surface: “bag of worms” appearance)

  b) defects of nerve K⁺ channels (e.g. neurogenic atrophy).

  c) amyotrophic lateral sclerosis.

---

**CRAMP VS. SPASM**

- **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).

  **etiology**

  1) **ORDINARY MUSCLE CRAMP**
  
  2) myoneurologic disease of LMN (e.g. ALS), nerve roots, peripheral nerve.

  3) **myogenic disease** - muscle ischemia, myopathy (e.g. phosphorylase deficiency, phosphofructokinase deficiency).

  4) **dehydration**, hypokalemia, pregnancy, hypothyroidism, uremia

  **EMG** - brief, periodic bursts of motor unit potentials at 200-300 Hz (much higher than with voluntary contraction), intermingling with similar discharges from adjacent motor units.

  - several foci within same muscle may discharge independently.

  - **electrical activity clearly arises within LMN** (whether it occurs in soma, in peripheral nerve, or in intramuscular nerve terminals is still debated); chemical mechanisms are not understood.

**ORDINARY MUSCLE CRAMP**

- cramps in normal persons.

  - can affect almost any voluntary muscle; most frequently in lower extremities (e.g. nocturnal calf cramps).

  - often starts with fasciculations → muscle becomes intermittently hard and knotty like as involuntary contraction waxes and wanes, passing from one part of muscle to another.

  - particularly common in older patients.

  - provoked by trivial movement or by contracting shortened muscle; may occur during vigorous exercise, but are more likely to occur after exercise ceases.

  **treatment**

  - stretching affected muscle.

  **pathophysiology**

  a) avoid caffeine and other stimulants.

  b) bedtime **QUININE SULFATE** 300 mg↑ – FDA warns against use of this drug for this unapproved indication "Quinidine should not be used for night time leg cramps – may result in thrombocytopenia, HUS/TTP"*

  c) calcium supplements ([CALCIUM GLUCONATE] 1-2 g bid) - effectiveness is doubtful.

  d) **MAGNESIUM CITRATE** 100-200 mg bid.

  e) low doses of benzodiazepines.

  f) **MEXILETINE**

  g) **MISULTINE** 150 mg tid → effective when increased LMN irritability is suspected.

**TETANY**

- intense tonic painful muscle cramps (e.g. carpopedal spasms, laryngospasm, opisophonia).

  - pathophysiologically – hyperexcitability* of LMN or peripheral nerves → spontaneous firing of peripheral nerves.

  *demonstrated by reactions to ischemia [Trousseau sign] and percussion [Chvostek sign]

  **etiology**

  1) hypocalcemia, hypomagnesemia

  2) tetanus toxin (GABA receptor blocker) – causes TETANUS.

  3) strychnine (glutamate antagonist)

  4) black widow spider toxin.

  5) latent tetany (s. normalcalcemic tetany, spasmodial)

  **EMG** - individual motor units discharge independently at 5-25 Hz; each discharge consists of group of ≥ 2 identical potentials.

---

**MUSCLE STIFFNESS**

- state of **continuous** muscle contraction at rest.

  **etiology**

  1) malignant hyperthermia. see p. 3910 >>

  2) neuromyopathic syndrome

  3) stiff-man syndrome see p. S27 >>

  4) myotonic disorders – myotonic dystrophy, channelopathies see p. Mus5 >>, p. Mus7 >>

**Contracture**

- prolonged severe, exercise-provoked tonic muscle shortening* (massassociated with muscle membrane depolarization)

  **etiology** – **glycolytic enzyme deficiencies** that interfere with substrate utilization as fuel (e.g. McArdle disease)

  - intensely painful, and result in muscle damage → myoglobinuria → renal failure.

  - contractures are electrically silent by EMG (vs. cramps - intense motor unit activity).

N.B. disorders of muscle contractile system cause electrically inactive contractions!
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 palsy.
  *In some cases only paresis (esp. in trunk muscles) – due to contralateral tr. corticospinalis ant. (if well-developed – may account for some degree of recovery).
B) below DECUSATIO PYRAMIDUM (anterior and lateral corticospinal tracts) → IPSILATERAL plegia;

<table>
<thead>
<tr>
<th>Lesion Location</th>
<th>Pattern of Signs</th>
</tr>
</thead>
<tbody>
<tr>
<td>Medial hemispheres (leg area)</td>
<td>Spastic leg paresis with no sensory level</td>
</tr>
<tr>
<td>Thoracic spinal cord</td>
<td>Spastic leg paresis, thoracic sensory level</td>
</tr>
<tr>
<td>Lumbar spinal cord</td>
<td>Flaccid paralysis, double incontinence (flaccid bladder and sphincters)</td>
</tr>
</tbody>
</table>

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 – parietal lesion in interhemispheric fissure (most commonly parasagittal meningioma; other → ACA 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 (parasagittal 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, HIV myelopathy.

DIAGNOSTIC APPROACH begins with spinal MRI or myelography.

HEMIPARESIS

- Lesion location is BILATERAL(!):

<table>
<thead>
<tr>
<th>Weakness distribution, UMN or LMN signs</th>
<th>Lesion location</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hemiparesis (with lower face on same side (UMN))</td>
<td>Contralateral cerebral hemisphere</td>
</tr>
<tr>
<td>Tetraparesis (UMN) + pseudobulbar palsy (UMN)</td>
<td>Bilateral cerebral hemispheres</td>
</tr>
<tr>
<td>Hemiparesis (UMN) + cranial nerve signs (LMN)</td>
<td>Brain stem</td>
</tr>
<tr>
<td>Tetraparesis (UMN) + cranial nerve signs (LMN)</td>
<td>Bilateral brain stem</td>
</tr>
<tr>
<td>Tetraparesis (UMN)</td>
<td>Medial or upper cervical cord</td>
</tr>
<tr>
<td>Paraparesis (UMN) + hands (LMN)</td>
<td>Low cervical cord</td>
</tr>
<tr>
<td>Paraparesis (UMN)</td>
<td>Thoracic spinal cord</td>
</tr>
<tr>
<td>All limbs, proximal &gt; distal (LMN)</td>
<td>Bilateral medullary motor cortex</td>
</tr>
<tr>
<td>Legs, distal &gt; proximal (LMN)</td>
<td>Nerve (polyneuropathy)</td>
</tr>
<tr>
<td>Ocular muscles, eyelids, jaw, face, pharynx, tongue (LMN)</td>
<td>Neuromuscular junction (NMJ)</td>
</tr>
<tr>
<td>Jaw, face, pharynx, tongue; sparing ocular muscles, eyelids (UMN and LMN)</td>
<td>Motor neuron disease (ALS)</td>
</tr>
<tr>
<td>Specific muscle groups in one limb (LMN)</td>
<td>Nerve root, plexus or peripheral nerve</td>
</tr>
</tbody>
</table>

PATTERNS OF WEAKNESS

<table>
<thead>
<tr>
<th>Sign</th>
<th>UMN weakness</th>
<th>LMN weakness</th>
<th>Myopathic weakness</th>
</tr>
</thead>
<tbody>
<tr>
<td>Atrophy</td>
<td>-</td>
<td>-</td>
<td>+</td>
</tr>
<tr>
<td>Fasciculations</td>
<td>+</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Distribution of weakness</td>
<td>pyramidal/regional</td>
<td>distal/segmental</td>
<td>proximal</td>
</tr>
<tr>
<td>Babinski sign</td>
<td>+</td>
<td>-</td>
<td>-</td>
</tr>
</tbody>
</table>

PARAPARESIS

- Lesion location is BILATERAL (?):

<table>
<thead>
<tr>
<th>Lesion Location</th>
<th>Pattern of Signs</th>
</tr>
</thead>
<tbody>
<tr>
<td>Brainstem</td>
<td>Paraparesis</td>
</tr>
<tr>
<td>Spinal cord</td>
<td>Quadriparesis</td>
</tr>
<tr>
<td>Meninges</td>
<td>Paraparesis</td>
</tr>
<tr>
<td>Distal</td>
<td>Proximal</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>UMN signs</th>
<th>LMN signs</th>
</tr>
</thead>
<tbody>
<tr>
<td>LMN signs</td>
<td>UMN signs</td>
</tr>
</tbody>
</table>

**Distribution of weakness**

- UMN signs
- LMN signs
- Myopathic pattern

**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, HIV myelopathy.

**Diagnosis** begins with spinal MRI or myelography.
**Lesion Location** | **Pattern of Signs**
--- | ---
Cerebral cortex | **Contusional 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)** | **Contusional weakness** (*face = arm = leg*); face may be spared! *No sensory loss or aphasia.*

**Brain stem** | see p. A59 >
 | **Contusional weakness** (*arm = leg*); ipsilateral peripheral cranial nerve palsy.
Midbrain ( crus cerebri) | Lesion of CN5 (Weber syndrome); red nucleus, superior cerebellar peduncle (limb ataxia contralateral to hemispheric side).
 | Pons (basis pontis) | Lesion of CN6 (Foville syndrome); CN7 (Millard-Gubler syndrome); internuclear ophthalmoplegia.
 | Medulla (pyramid) | Lesion of CN2, face spared.
Cervical spinal hemisected (Brown-Sequard syndrome) | Ipsilateral weakness sparing face.
 | *Lesion location in some cases of ALS (with or without UMN signs in legs).

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.

**Diagnostic approach** - brain CT; if CT normal and ischemic stroke is unlikely → MRI of brain → brain CT; if CT normal and ischemic stroke is unlikely → MRI of cervical spine.

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

**Subacute hemiparesis**
- *subacute subdural hematoma*
- *infection - cerebral bacterial abscesses, fungal granuloma or meningitis, parasitic infection.)*
- *malignant primary / metastatic neoplasms*
- *N.B. AIDS may present with subacute hemiparesis due to toxoplasmal meningitis or primary CNS lymphoma!*
- *focal inflammatory lesion* (multiple sclerosis, sarcoidosis).

**Chronic hemiparesis** (slowly develops over months)
- *histologically benign neoplasm*
- *unruptured AVM*
- *corticobulbar & corticopontocerebellar inflammation / hemorrhage / infarction / demyelination / nonantigravity muscular weakness.*

**Monoparesis**
- *A: With pain:
  1. Congenital lesion of spinal cord
  2. Acute brachial plexus neuritis (neuropathic amyotrophy).
  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.*

**Bilateral paresis** - arms hang limply at side while patient walks with normal movements of legs.
- *Cervical LMN lesion in some cases of ALS (with or without UMN signs in legs).
- *Myopathy of unusual distribution.*
- *Cerebral lesion (bilateral proroladic)-- “man-in-the-barrel syndrome” seen in comatose patients who survive bout of severe hypotension.*

**Neck weakness (“floppy head” syndrome)** - Never in UMN disorders!
- *ALS*
- *Myasthenia gravis*
- *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. Eyed >>
6. Myopathy
7. Neuropathy (incl. radiculopathy)
   N.B. patient with no reflexes usually has neuropathy!

**BILATERAL HYPOREFLEXIA**

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 (stac lacerane)
7. Familial spastic paraplegia
8. Cerebral palsy
9. Parasagittal intracranial mass (may affect cortical leg fibers)
10. Hydrocephalus (may stretch leg fibers)

**VOICES**

LMN 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, hyperreflexia, bilateral Babinski signs</td>
</tr>
<tr>
<td>Basis pontis (stroke)</td>
<td>“Locked-in” 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 toe 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>
</tr>
</thead>
<tbody>
<tr>
<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>
<td>Carotid ultrasound</td>
</tr>
<tr>
<td>Partial motor seizure, Todd's paresis (postural weakness)</td>
<td>Gradual “march” of symptoms in several seconds to few minutes</td>
<td>EEG</td>
</tr>
<tr>
<td>Myasthenia gravis</td>
<td>Fatigability, recovery with rest; predilection for ocular and cranial muscles</td>
<td>Tensilon test, repetitive stimulation test</td>
</tr>
<tr>
<td>Hysteria</td>
<td>Normal reflexes, nonanatomic distribution of sensory loss</td>
<td>Sleep study</td>
</tr>
<tr>
<td>Drop attacks</td>
<td>Sudden loss of postural tone without loss of consciousness</td>
<td>MRI, MRA, X-ray of cervical spine with flexion-extension, EEG</td>
</tr>
<tr>
<td>Periodic paralyses</td>
<td>Familial channelopathies</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. vertebrobasilar 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 cervicomедullary 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 >>