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- disorders

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Muscle tone is evaluated by passive movements of limb and provides objective information.

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  - 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.
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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 of 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 agonist - BACLOFEN (20-240 mg/d in divided doses q8hr*) - most effective drug available!
- i.e. single oral max dose is 70-80 mg
  - originally synthesized as an anticonvulsant but it was found to have no significant anticonvulsant activity.
  - in severe cases - intrathecal via implanted pump; (see p: Op240 >>
  - 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:

   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/confusion/ agitation/hallucinations, labile blood pressure, seizures; potentially lethal - can lead to rhodanomylolysis, DIC, organ failure, and can look like autonomic dysreflexia, malignant hyperthermia (vs. opioide withdrawal), septic shock.

   Treatment - oral baclofen, IV benzodiazepines (DIAZEPAM, 2-5 mg q 6 hours) or CYCLOPHENAZINE (6 mg q 6 hours for 24 hours); if due to IT system failure and unable to remove full dose orally (try oral baclofen 20 mg po q8-hrs) – insert temporary IT catheter and post for surgery.

2) GABA agonist - DIAMETHAZEPAM (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:

   Baclofen overdos e may result in somnolence, respiratory depression, hypothermia, seizures, restral progression of hypotonia, coma.

   Treatment - no antidote (if no heart conduction defects, PHYSOSTIGMINE 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.

3) α2-agonists (central muscle relaxants):
   a) TIZANIDINE (Zanaflex®) (4-8 mg q8h)
   b) CYCLOBENZAPRINE (Flexeril® Amrix®)
- dosage: 10 mg q 6 h (5 and 10 mg tablets).

4) direct muscle inhi bitors - PANTOLOLENE (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 structure related 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 fatigue due to neuromuscular conditions such as cerebral palsy.
• adverse effects: drowsiness (38% of patients), dry mouth (24%), urinary retention (in males with large prostates).

6) METHOCARBAMOL (Rexona®) - 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) CARISOPRODOL (Soma®) – not recommended! (converted to benzodiazepines – addictive potential!)

8) CHLORZOXAZONE (Loozone®, Parafon®)

9) METAXALONE (Skelaxin®)

10) ORPHENADRINE

3. Local injections of BUTYLIMIDAZINE (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.neurosurgeryadvisor.com/.
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 - 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! - 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 L2 root at its exit foramini, S1 anterior root is identified by low-frequency stimulation.
• fascicles of each L2/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.

*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.
• rare fascicles innervating splinters!
• 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) posterior root ganglioneuromy of sacral segments (for spastic bladder).
   b) selective peripheral neurotomy (sectioning nerve fascicles - identified by intraoperative stimulation - which maintain spastic tone; e.g. tubal 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) Bischof myelotomy (longitudinal myelotomy) - dividing cord into anterior and posterior halves over segments involved in flexor spasms (typically L2-S2); interrupts local reflex arcs; may not prevent spasms triggered by stimuli from segments caudal to L2 or caudal to S1.

6) stereotactic denervation (limited usefulness in management of spasticity)

N.B. Selective posterior rhizotomy provides much higher success rate.
• indication - congenital spasticity with congenital chorea athetosis: thalamotomy controls chorea-thetosis; if subsequently worse spasticity develops → ipsilateral demeanors...

FATIGABILITY

PATIENT - feeling of being tired and not being able to put out full effort:
NORMAL PATIENT - results from intense muscular contraction:
• accompanied by firing frequency of motor unit - result of reduced excitatory drive to motoneurons (central mechanism!).

FATIGABILITY (disfunction at neuromuscular junction) - muscles become weaker and weaker with repetitive but normal use (inability to sustain performance of activity).
• accompanied by decrease in amplitude of muscle action potentials.
• N.B. with exception of neuromuscular junction disorders, fatigue is rarely complaint of diseases of muscle itself!!!

"Fatigue" “tiredness”; “lack of energy” are common complaints in following disorders:
1) [MS 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
Pyramidal UMN lesion

**Adult 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 (muscle stretch, plantar, abdominal & cremasteric)

If lesion transects spinal cord (SPINAL SHOCK) → see p. Spinal 1 below, it is also accompanied by:
3. Hypotonic paralysis of bladder & bowel
4. Hypotension, anhidrosis

**CHRONIC STAGE OF UMN LESION**

1. Paralysis involves large areas (hemi-, para-, quadriplegia) – at & distal to capsula interna small lesion affects large body parts; rostral to capsula interna, pyramidal neurons are dispensed (e.g. small stroke in arm area of motor cortex can produce brachial monoplegia).

**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 & tongue; extracranial, upper facial, pharyngeal, and jaw movements are almost always spared (but with bilateral corticobulbar lesions → SEVERE PSEUDOBULBAR PALSY), see below

2. Muscle atrophy of disease only (late and slight).
3. Spasticity (muscle tone)
   - spasticity in presence of **antigravity muscles** – arm flexors, leg extensors, **WERNICKE-MAN posture**.
   - resistance depends on velocity and direction of passive motion → "clasp knife" phenomenon, see above
   - if patient can walk, spasticity causes **ICSHORS GAIT** (in bilateral lesions), leg circumduction (in unilateral lesions). see p. Mov 7 below
   - pure pyramidal tract lesions cause mild paraparesis without spasticity – because control of tone is mediated by other tracts (particularly corticobulbospinal and corticocerebellospinal) - 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. CLEANS (rhythmic, rapid alternation of muscle contraction and relaxation caused by sudden, passive tendon stretching) about mechanisms see p. A18

N.B. only exaggerated clonus suggests UMN damage?

3. BARBINI sign and other pathologic withdrawal reflexes (normally, they are inhibited by intact pyramidal system):
   - normally only painful stimulus elicits withdrawal reflex.
   - when UMN is damaged (but also in normal infants – immature CNS), lighter nonpainful stimulus may elicit withdrawal reflex - strength of **o naso-parallels extend to which UMN lesion has allowed upregulation of reflex:**
     - small hemispheric lesion - only small fragment of reflex may be elicited (i.e. extension of great toe - Barbini sign).
     - complete spinal cord transaction - entire withdrawal reflex (with flexion at hip, knee, and ankle) may occur

Symmetrical hypertensive reflexes in presence of down-going box are usually normal!

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

6. Synkinesis

7. Movements are slow, coarse but with normal rhythmicity and coordination (e.g. finger–nose–finger and heel–knee–shin are performed slowly 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 PALSY** - bilateral corticobulbar tract lesion (i.e. central-supranuclear palsy of CN 7, 9, 10, 12):
1. apraxic 3D (dyssartha, dysphonia, dysphagia)
2. infrequent paresis
3. hyperactive gag reflex, hyperactive facial and jaw jerk (CN 5 – CN 7).
4. 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).
5. extraocular, upper facial, pharyngeal, and jaw muscles are almost always spared (but with bilateral corticobulbar lesions → PSEUDOBULBAR PALSY).

Nuedexta (DEXTROMETHORPHAN HYDROBRIDE + 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)

**LOCALIZED IN**-**NEUROHORME (OR PHYSIOLOGICAL) PALSY** - bilateral basis pontis lesion: i.e. damage to corticopontine-corticospinal-corticobulbar tracts below reticular formation (therefore sparing consciousness) but above ventilatory nuclei of medulla (therefore, precluding death by respiratory arrest).
• most commonly due to basilar artery infarction; other causes: central pontine myelolysis.
• almost complete de-extermination:
  1. quadriplegia
genuinely corticospinal tracts damage.
  2. paralysis of horizontal eye movements (horizontal ophthalmo-plegia) – due to PPRF and CN6 nuclei, corticobulbar tracts damage.
  3. paralysis of jaw, face, bulbar musculature (facial & bulbar diplegia, no-volitional vocalization?).
• 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, ophthalmoplegia, hyperthermia, progression to death.
- patients must be identified rapidly for intravenous rtPA treatment.
- mortality rate is high (50%-70%); 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. Areflexia (all reflexes ↓↓↓ or absent – grade 1 or 0) – last effertent 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 summation does not cause weakness but decreases tension on muscle spindles → tendon reflexes]
2. Areflexia
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 months, 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).

Primary Sensory Neuron lesion
1. Hypotonia
2. Areflexia (absent all reflexes) – last afferent portion of reflex arc! 
- Volitional movements and their strength remain normal!
- Patient’s appearance looks normal!
- Coordination normal only with eyes open.

SPONTANEOUS 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
  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 (fascicules or bundles).

FIBRILLATIONS - invisible contractions of individual muscle fibers - can be detected by EMG see p. D20 >>
- vary irregularly in frequency and extent.
- do not move joint!
- source: LMN disease:
  a) extrapyramidal.
  b) seizure disorders

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 (MINIOPHTALMOCYCLOYES).

N.B. fasciculations are commonly experienced as benign phenomenon in absence of any disorder! (e.g. in incompletely relaxed muscles)

<table>
<thead>
<tr>
<th>Features</th>
<th>Benign Fasciculations (Drummond, Fales syndrome)</th>
<th>Malignant (neuropathologic) fasciculations</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gender predilection</td>
<td>males*</td>
<td></td>
</tr>
<tr>
<td>Predilection for certain muscle groups</td>
<td>calves and thighs</td>
<td></td>
</tr>
<tr>
<td>Nature</td>
<td>repetitive twitch in same muscle fascicle; may be accompanied by frequent cramps</td>
<td>random nonstereotyped twitches of many parts of muscle</td>
</tr>
<tr>
<td>Associated weakness or atrophy</td>
<td>-</td>
<td>*</td>
</tr>
<tr>
<td>Electrophysiology</td>
<td>appears like normal motor unit; no features of muscle denervation</td>
<td>complex, longer duration, higher amplitude</td>
</tr>
</tbody>
</table>
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. see p. D20

- etiology
  a) lesions of pons (e.g. neoplasms or multiple sclerosis) - FACIAL MYOKYMIA - newly continuous twitching of facial muscles (pupillary fissure narrowing, continuous undulation of facial skin surface – “bag of worms” appearance).
  b) defects of nerve C-channels (e.g. neuromyotonia).
  c) amyotrophic lateral sclerosis.

CRAMP (S. 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) neurogenic disease of LMN (esp. ALS): nerve roots, peripheral nerve.
     3) myogenic disease - muscle ischemia, myopathy (e.g. phosphorylase deficiency, phosphofructokinase deficiency).
     4) dehydration, hypothyroidism, pregnancy, hypothyroidism, uremia
     5) EMG - brief, periodic bursts of motor unit potentials

- 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.

PREVENTION & TREATMENT - 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 waves and waves, 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.
- prophylaxis:
  a) avoid caffeine and other stimulants.
  b) bedtime QUININE SULFATE 500 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 hemodynamic and respiratory impairment. HUNTS TTD***
  c) calcium supplements (CALCIUM GLUCONATE 1-2 g bid) - effectiveness is doubtful.
  d) MAGNESIUM OXIDE 100-200 mg bid.
  e) low doses of benzodiazepines.
  f) PHENYTOIN, CARBAMAZEPINE
  g) MIXEDLINE 150 mg tid - effective when increased LMN irritability is suspected.

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

- pathophysiology - hyperexcitability* of LMN or peripheral nerves → spontaneous firing of peripheral nerves.
* demonstrated by reactions to ischemia [Trousseau sign] and percussion [Chvostek sign [Chvostek sign]

- etiology
  1) hypocalcemia, hypomagnesemia
  2) tetanus toxin (GABA receptor blocker) - causes TETANUS
  3) strychnine (glycine antagonist)
  4) black widow spider toxin.
  5) latent tetany (s. normocalcemic tetany, spasmophilia)
  6) EMG - individual motor units discharge independently at 5-25 Hz; each discharge consists of group of 2-10 identical potentials.

MUSCLE STIFFNESS - state of continuous muscle contraction at rest.

- etiology
  1) malignant hyperthermia. see p. 3190
  2) neuroleptic malignant syndrome
  3) stiff-man syndrome. see p. 3180
  4) myotonic disorders – myotonic dystrophy, channelopathies. see p. MUS5 & MUS7

MYOTONY - impaired muscle relaxation after forceful voluntary contraction (painless muscle stiffness).

CONTRACTION - prolonged severe, exercise-provoked tonic muscle shortening* (unassociated with muscle membrane depolarization.

*do not confuse with limitation of joint range of motion (also termed contracture).

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

- intensify painful, and result in muscle damage (→ myoglobinuria → renal failure).

- compares 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 DISECTIONS PYRAMIDUM (pyramidal tract) → CONTRALATERAL hemiplegia
  - including lower face; lesions below pons spare face.
  - bilateral lesions can cause PSEUDOBULBAR PARALYSIS. see above

B) below DISECTIONS PYRAMIDUM (anterior and lateral corticospinal tract) - ipsilateral paraparesis
  - 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).
In general, only bilateral lesions cause UMN-type weakness in trunk and cranial muscles!

### Patterns of Weakness

<table>
<thead>
<tr>
<th>Sign</th>
<th>UMN weakness</th>
<th>LMN weakness</th>
<th>Myopathic weakness</th>
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</thead>
<tbody>
<tr>
<td>Atrophy</td>
<td>+ + + +</td>
<td>+ + + +</td>
<td>+ + + +</td>
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<tr>
<td>Paraplecses</td>
<td>+ + + +</td>
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<td>+ + + +</td>
</tr>
<tr>
<td>Tone</td>
<td>![Um]; (spastic)</td>
<td>![N]; normal / +</td>
<td>+; normal / +</td>
</tr>
<tr>
<td>Distribution of weakness</td>
<td>pyramidal/regional; distal/segmental</td>
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</tr>
<tr>
<td>Tendon reflexes</td>
<td>+ +</td>
<td>+ +</td>
<td>+ +</td>
</tr>
<tr>
<td>Babinski sign</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
</tbody>
</table>

Weakness distribution, UMN or LMN signs

<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>Paraplecses</td>
<td>+ + + +</td>
<td>+ + + +</td>
<td>+ + + +</td>
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<tr>
<td>Babinski sign</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
</tbody>
</table>

### Paraparesis

- Lesion location is bilateral (!):

#### Lesion Location

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

Paraparesis implies lesion below cerebral cord; exceptions:

1. **leg areas (on medial side of each hemisphere; at apex of motor strip)**: face each other in interhemispheric fissure - parasagittal lesion in interhemispheric fissure (most commonly parasagittal meningioma, other - ACA ischemia, superior sagittal sinus thrombosis) could affect both legs - PARAPARESIS simulating spinal cord lesion.

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;
- HTLV-I infection, HIV myelopathy.

#### Diagnostic approach

begins with spinal MRI or myelography.

### Hemiparesis

- Lesions are unilateral (!):

#### Lesion Location

- **Cerebral cortex**: Contralateral weakness (arm, leg, face; sometimes tongue)*
  - Left hemisphere: aphasia, apraxia.
  - Right hemisphere: left hemianopia, extinction of sensory stimuli, constructional apraxia, spatial disorientation.
  - Homonymous hemianopia on weak side.
  - Cortical sensory loss (decreased graphesthesia, stereognosis, point localization).
  - Horizontal eye deviation (toward lesion side).
- **Internal capsule (posterior limb)**: Contralateral weakness (face = arm = leg); face may be spared!
  - Cortical sensory loss (decreased graphesthesia, stereognosis, point localization).
  - Horizontal eye deviation (toward lesion side).
- **Brain stem**: see p. A59 >
  - Contralateral weakness (arm = leg = symmetrical peripheral cranial nerve palsy)
- **Midbrain (crus cerebri)**: Lesion of CN3 (Waber syndrome), red nucleus, superior cerebellar peduncle (limb ataxia contralateral to hemiparesis side).
- **Pons (basis pontis)**: Lesion of CN6 (Foville syndrome), CN7 (Millard-Gubler syndrome); internuclear ophthalmoplegia.
<table>
<thead>
<tr>
<th>Lesion Location</th>
<th>Pattern of Signs</th>
</tr>
</thead>
<tbody>
<tr>
<td>Medulla (pyramidal)</td>
<td>Lesion of CN12, face spared</td>
</tr>
<tr>
<td>Cervical spinal hemisected (Brown-Sequard syndrome)</td>
<td></td>
</tr>
<tr>
<td>Ipsilateral weakness sparing face.</td>
<td></td>
</tr>
<tr>
<td>Ipsilateral loss of proprioception and vibration.</td>
<td></td>
</tr>
<tr>
<td>Contralateral loss of pain and temperature.</td>
<td></td>
</tr>
</tbody>
</table>

*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 → MRI of cervical spine:**

**Acute hemiparesis:**

- unusually vascular pathogenesis
- traumatic rupture 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
- chronic inflammatory lesion (multiple sclerosis, sarcoidosis).

**Lesion Location**

- lesion locations are bilateral, (1):**

<table>
<thead>
<tr>
<th>Lesion Location</th>
<th>Pattern of Signs</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cerebral hemispheres</td>
<td>Peridural bulbar palsy, decorticate posture (large acute lesions)</td>
</tr>
<tr>
<td>Midbrain</td>
<td>Coma, mid-size poorly reactive pupils, decerebrate posture</td>
</tr>
<tr>
<td>Brainstem</td>
<td>Locked-in syndrome</td>
</tr>
</tbody>
</table>
| Cervicomedullary junction | Lesia > arm, t weakness of phynx & tongue, facial hypagula (descending tract of CN 5) 
- No cerebral signs (cranial nerve palsies, etc) |
| High cervical | Preservation of shoulder movements |
| Mid cervical | No cerebral signs (cranial nerve palsies, etc) |
| Peripheral nerves | Distal weakness |
| Muscles (myopathy) | Proximal weakness |

**MONOPARESIS:**

- With pain
  1. Compensatory lesion of spinal cord
  2. Acute brachial plexus neuritis (neuralgic amyotrophy).
  3. Peripheral nerve entrapment syndromes

- 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.

**BRAINSTEM PARESIS**

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

1. Cerebral LMN lesion in some cases of ALS (with or without UMN signs in legs).
2. Mysopathy of unusual distribution.
3. Cerebral lesion (bilateral prerolandic) – *man-in-the-barrel syndrome* seen in comatose patients who survive bout of severe hypotension.

**NECK WEAKNESS (“FLOPPY 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 aneurysis with large pupil that reacts to accommodation but not to direct light) see p. Ey647 **>
6. Mysopathy
7. Neuropathy (incl. radiculopathy)
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 (état lacunaire)
7. Familial spastic paraplegia
8. Cerebral palsy
9. Parasagittal intracranial mass (may affect cortical leg fibers)
10. Hydrocephalus (may stretch leg fibers)

VOICE

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>COMMON</td>
<td></td>
<td></td>
</tr>
<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>LESS COMMON</td>
<td></td>
<td></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>Hemiplegic migraine</td>
<td>Gradual development over several minutes; family history</td>
<td></td>
</tr>
<tr>
<td>Myasthenia gravis</td>
<td>Fatigability, recovery with rest; predilection for ocular and cranial muscles</td>
<td>Transiton test, repetitive stimulation test</td>
</tr>
<tr>
<td>Hysteria</td>
<td>Normal reflexes, nonanatomic distribution of sensory loss</td>
<td></td>
</tr>
<tr>
<td>Cataplexy</td>
<td>Triggered by emotion; association with other features of narcolepsy; episodes very brief</td>
<td>Sleep study</td>
</tr>
<tr>
<td>Sleep paralysis</td>
<td>Narcolepsy; terminated by touch</td>
<td>Sleep study</td>
</tr>
<tr>
<td>Drop attacks see below &gt;&gt;</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>Negative myoclonus see p. Mov1 &gt;&gt;</td>
<td>Sudden, brief, rapid, unpredictable (shocklike) inhibition of muscle tone (single muscle ÷ entire body).</td>
<td></td>
</tr>
</tbody>
</table>

RARE

Periodic paralyses | Familial channelopathies | Serum K+ |

DRIP 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. verteobasilar 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.
  4) 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).