UMN & LMN Disorders

Weakness (Loss of Voluntary Movement)

Treatment

Muscle Tone Abnormalities

Treatment of Spasticity

Fatigability

Pyramidal UMN lesion

Pyramidal UMN lesion

Sensory neuron lesion

Spontaneous Movements

- Fasciculations, Fibrillations
- Myokymia
- Cramp (p. spasms)
- Tetany

Muscle stiffness

Contracture

Lesion Localization Guide

Patterns of weakness

Paraparesis

Hemiparesis

Monoparesis

Bibibachal Paresis

Neck Weakness (“floppy head” syndrome)

Hyporeflexia

Bilateral Hyperreflexia

Voice

Acute Generalized Weakness

Epidemic Weakness

Drop Attack

Decerebrate/Decerebrate Rigidity

Motor system generates three general types of movements:

- Reflex response - simplest form of coordinated movement - rapid, stereotyped involuntary movements elicited by sensory stimuli that require quick reaction at involuntary level.
- Rhythmic movements - ex. 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.

Lower Motoneuron (LMN) - o motoneuron directly innervating striated skeletal muscles.

Upper Motoneuron (UMN) - term used in two senses:

- senso stricto - cortical neurons forming tractus pyramidalis.
- senso lato - all neurons forming descending tracts that ultimately play on LMN (tr. pyramidalis, tr. reticulospinalis, tr. rubrospinalis, tr. vestibulospinalis, etc.)

Weakness (Loss of Voluntary Movement)

- Muscle cannot exert normal force - most important clinical feature of motoneurone (UMN, LMN) disorders.

- Paralysis - 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 prefrontes:
  - 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 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.

Muscle Tone Abnormalities

Muscle tone changes usually accompany weakness:

- Tone is elevated by passive movements of limbs. see p. D1 >>

- Main components of muscle tone:
  1. Low level background γ-activity.
  2. Alteration in stretch reflex (most important determinant of pathological alterations in tone!) - via changes in rate of discharge in γ-neurons ± changes in general excitability of motor neuron pool.
  3. Vicissitudes properties of muscle & tendons (contribute to increased tone in chronic spasticity and rigidity).

Tonus - Spasticity (s. Flexibility) - LMN disease, cerebellar disease, sensory nerve damage.

Tonus:

- 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 hypertonic muscle
5. Treatment of spasticity; from bed to chair, facilitate hygiene, a

Drugs with systemic effects

- 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.
  - fill as increase in tone that fluctuates with repetitive passive movements.
- often accompanied by tonic grasp reflex.
- is involuntary to extent that patient has great difficulty in voluntarily suppressing urge to resist.

TREATMENT OF SPASTICITY

Spasticity may be helpful

- for CP kids, ortho procedures are delayed after spasticity is addressed.
- if fixed contracture has developed → insert temporary IT catheter!
- if cerebral palsy patient is in wheelchair or bed (drugs allow easier transfers from bed to chair, facilitate hygiene, alleviate painful spastic flasms).
- intrathecal baclofen delivery system
- electrical stimulation of antagonist muscles
- keep feet flexed at 90° (use pillow between soles and bed foot).
- keep hyperextension.
- electrical stimulation of antagonist muscles and splitting may help.

2. Drugs with systemic effects (sedation is usually limiting barrier, esp. for school-age children!)

- particularly used in patients who are confined to wheelchair or bed (drugs allow easier transfers from bed to chair, facilitate hygiene, alleviate painful spastic flasms).
- (20-240 mg/d in divided doses q6hr*) - most effective drug available.
  - single oral max dose is 70-80 mg
  - in severe cases - intrathecal via implanted pump: patients are very sensitive to small changes in microdoses of baclofen.

3. Intrathecal baclofen

- titlych, twibitty, bitchy) (itching, seizures, mood fluctuation, spasticity!); potentially lethal (vs. opioid withdrawal) - resembles septic shock; if due to IT system failure and unable to replace dose orally → insert temporary IT catheter!

2) GABA agonist - DIAZEPAM (2-4 mg at bedtime) - for leg spasms that interrupt sleep, CNS depressant!

3) u2-agonists (central muscle relaxants): a) TRANZAX® (Zanaflex®) (4-8 mg q8h)

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

5) CYCLOBEPRIDE - Flexeril® (Anxure®)

- dosage: 10 mg q 6 h (5 mg 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 coeruleus; → 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 neurological conditions such as cerebral palsy.
- adverse effects, drowsiness (38% of patients), dry mouth (24%)
- central muscle relaxant
- may cause urine to turn black, blue, or green.

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) CARISOPRODOL (Soma®)

8) CHLOROZAZONE (Lorzone®, Parasorb®)

9) METAXALONE (Skelaxin®)

10) ORPHENADRINE

3. Local injections of BOTULINUM TOXIN (effect for 3-6 months) - no deleterious effects on helpful spasticity: target muscle: spasticity

1) leg adductors - to facilitate nursing care.

2) arm flaps - to relieve painful spasms.


- 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, thigh adductor, hamstrings).
- 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 → Spasticity is usually limiting barrier, esp. for school-age children!

2) selective posterior rhizotomy (procedure of choice for spasticity due to cerebral palsy).

- exposure of cauda equina (through L3-L5 laminectomy).
- after anatomic identification of L2 root at its exit foramen, S: anterior root is identified by low-frequency stimulation.
Pyramidal UMN lesion

A. PYRAMIDAL LESIONS

1. **CEREBRAL STROKE** - Widespread destruction of the ipsilateral motor cortex 
   - **Diagnosis**: by neuroimaging 
   - **Symptoms**: usually appears slowly over days to weeks 
   - **Treatment**: few effective treatment options, rehabilitation

2. **SPINAL SHOCK** - Initial inactivity and flaccidity 
   - May last several weeks to months 
   - Recovery follows a predictably progressive course 

3. **SPINAL STANZ** - Persisting signs of pyramidal tract lesion
   - Motor examination reveals hyperreflexia
   - Increased muscle activity in the lower limbs

4. **COMA** - Severe brain injury leading to unconsciousness
   - Altered level of consciousness 
   - Important to distinguish from other causes of altered consciousness 
   - Treatment: supportive care, management of complications

5. **WHITE)?$? INCLUDING** - Remote effects of stroke 
   - May include weakness, numbness, or sensory changes

6. **RADIATION LESIONS** - Effects of radiation therapy
   - Common for brain tumors 
   - Radiation-induced damage to the motor cortex 

7. **METABOLIC AND ENDOCRINE DISORDERS** - Various metabolic and endocrine conditions
   - Can cause weakness, atrophy, and loss of motor function
   - Treatment: management of underlying disorder

8. **INTRAVENTRICULAR HEMORRHAGE** - Bleeding into the ventricular system 
   - Immediate neurological deterioration 
   - May cause motor deficits and cognitive impairment

9. **INFECTIOUS DISEASES** - Bacterial, viral, or parasitic infections affecting the CNS
   - Neurological symptoms such as weakness, ataxia, or altered consciousness
   - Treatment: antibiotics, antiviral agents, or antiparasitic drugs

10. **NEURODEGENERATIVE DISEASES** - Progressive deterioration of neural function
    - Commonly associated with motor symptoms
    - Treatment: symptomatic management, disease-modifying therapies

11. **MULTIPLE SCLEROSIS (MS)** - Autoimmune disease affecting the CNS
    - Motor symptoms including weakness, ataxia, and spasticity
    - Treatment: disease-modifying agents, immunomodulatory therapy

12. **MUSCULAR DYSTROPHY** - Genetic disorder affecting muscle function
    - Progressive weakness, fatigue, and muscle wasting
    - Treatment: supportive care, enzyme replacement therapy

13. **HYPOTHALAMIC SYNDROME** - Dysfunction of the hypothalamus
    - May cause hyperactive or hypoactive motor behavior
    - Treatment: pharmacological intervention, behavioral therapy

14. **HYPOFUNCTION OF THE MOTOR SYSTEM** - Overall reduction in motor activity
    - Common in conditions affecting the motor cortex or basal ganglia
    - Treatment: rehabilitation, pharmacological agents

15. **HYPERFUNCTION OF THE MOTOR SYSTEM** - Increased muscle tone and spasticity
    - Common in conditions affecting the pyramidal tract
    - Treatment: antispasmodic medications, physical therapy

16. **HYPERREFLEXIA** - Exaggerated muscle stretch reflexes
    - Common in conditions affecting the pyramidal tract
    - Treatment: antispasmodic medications, physical therapy

17. **HYPERKINESIA** - Excessive voluntary or involuntary movements
    - Common in conditions affecting the basal ganglia
    - Treatment: antiparkinsonian medications, deep brain stimulation

18. **HYPOKINESIA** - Reduced voluntary or involuntary movements
    - Common in conditions affecting the basal ganglia
    - Treatment: dopamine agonists, deep brain stimulation

19. **HYPOFUNCTION OF THE SOMATOSENSORY SYSTEM** - Reduced sensation
    - Common in conditions affecting the sensory cortex or spinal cord
    - Treatment: rehabilitation, pain management

20. **HYPOFUNCTION OF THE AUTONOMIC NERVOUS SYSTEM** - Reduced autonomic function
    - Common in conditions affecting the autonomic nervous system
    - Treatment: pharmacological agents, rehabilitation
5. Absent normal skin reflexes (abdominal, cremasteric).
6. Synkinesias
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. LMN lesions - number 3, frequency normal).

**PSEUDOBULBAR PARALYSIS**
- bilateral corticobulbar tract lesion (i.e. central-supranuclear palsy of CN 7, 9, 10, 12):
  1. Absent swallowing & gag reflexes
  2. Absent normal skin reflexes
  3. Absent tendon reflexes
  4. Absent swallowing & gag reflexes

**Locked-in** Syndrome (or Pseudobulbar Paralysis)
- bilateral basis pontis lesions: i.e. damage to corticospinal-corticopontine-corticobulbar tracts below reticular formation (therefore sparing consciousness) but above ventilatory nuclei of medulla (therefore, precluding deep inspiration).
- most commonly due to basilar artery infarction: other causes - central pontine myelinolysis
- almost complete de-afferentation
  1. Spastispasms – due to corticospinal tracts damage.
  2. Bilateral horizontal eye movements (horizontal ophthalmoplegia) – due to PRP 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 spinal pontine tegmentum – sudden coma, pinpoint pupils, ophthalmoplegia, hyperthermia, progression to death
- patients must be identified rapidly for intravenous n-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’s”**

1. **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
2. **Atrophy of denervation**
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4. **Paralysis of individual muscles** (or groups of muscles)
5. **Fasciculations, Fibrotications** see below >>

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

- 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, TETANUS**

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

- fibrillation - 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) diseased anterior horn cell may spontaneously discharge → fasciculations.
  2) Ach 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.
  3) fasciculations are seldom seen with peripheral nerve lesions (atrophy without fasciculations is more compatible with peripheral nerve lesion).
- in long-standing muscle denervation and reinervation, motor unit size enlarges and fasciculations may be so large as to produce movement of limbs, particularly of fingers (MINIPOP/MIYOCLEONS).

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

**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 MYOKYMS - nearly continuous twitching of facial muscles (palpable fissure narrow, continuous undulation of facial skin surface = "bag of worms" appearance).
  b) defects of nerve K+ 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) ORINARY MUSCLE CRAMPS.
  2) neurogenic disease of LMN (esp. ALS), nerve roots, peripheral nerve.
  3) myogenic disease - muscle ischemia, myopathy (e.g. phosphofructokinase deficiency, phosphofructokinase deficiency).
  4) dehydration, hypotremia, pregnancy, hypothyroidism, uremia
  5) 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.
  6) 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.

**FREQUENT VIOLENT CRAMPS** - 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 knot-like as involuntary contraction wages on 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.
  - prophylaxis:
    a) avoid caffeine and other stimulants.
    b) bedtime QUININE SULFATE 300 mg!!! - FDA warns against use of this drug for this unapproved indication "Qualquin should not be used for night time leg cramps - may result in thrombocytopenia, HUS/TTP".
    c) calcium supplements (CALCIUM GLUTONATE 1-2 g bid) - effectiveness is doubtful.
**Lesion of**

- Intense toxic painful muscle cramps (e.g. carpopedal spasms, laryngospasm, opisthotonus).

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

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

**Lesion**

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)

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

**Lesion of**

- State of continuous muscle contraction at rest.

**Lesion**

1) Maligant hyperthermia. See p. 3910 >>
2) Neuroleptic malignant syndrome
3) Stiff-man syndrome. See p. 3827 >>

**Myotonia** – impaired muscle relaxation after forceful voluntary contraction (painless muscle stiffness).

**Lesion of**

Lesion of pyramidal UMN:

A) **Above Decussatio Pyramidal (pyramidal tract)** – **Contralateral hemiplegia**

- Bilateral lesions can cause PSEUDOBULBAR PARALYSIS. See above >>

B) **Below Decussatio Pyramidal (anterior and lateral corticospinal tract)** – ** Ipsilateral plegia**

- 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 **cerebral** muscles!

**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>1 (spastic)</td>
<td>+</td>
<td>−</td>
</tr>
<tr>
<td>Tone</td>
<td>±</td>
<td>⫸</td>
<td>normal / ±</td>
</tr>
<tr>
<td>Distribution of weakness</td>
<td>pyramidal/regionat</td>
<td>distal/segmental</td>
<td>proximal</td>
</tr>
<tr>
<td>Tendon reflexes</td>
<td>⫸</td>
<td>⫸</td>
<td>/ absent</td>
</tr>
<tr>
<td>Babinski sign</td>
<td>+</td>
<td>+</td>
<td>−</td>
</tr>
</tbody>
</table>

**Weakness distribution, UMN or LMN signs or Lesion location**

- **Hemiparesis** (with lower face on same side) (UMN) → Contralateral cerebral hemisphere
- **Tetraparesis** (UMN) + pseudobulbar palsy (UMN) → Bilateral cerebral hemispheres
- **Hemiparesis** (LMN) → Cranial nerve signs (LMN) → Brain stem
- **Tetraparesis** (LMN) + cranial nerve signs (LMN) → Bilateral brainstem
- **Tetraparesis** (UMN) → Mid or upper cervical cord
- **Paraparesis** (UMN) + hands (LMN) → Low cervical cord
- **Paraparesis** (UMN) → Thoracic spinal cord
- **All limbs, proximal > distal** (LMN) → Bilateral medial motor cortex
- **Upper limbs, proximal > distal (LMN)** → Muscle (myopathy or dystrophy)
- **Legs, distal > proximal (LMN)** → Nerve (polyneuropathy)
- **Ocular muscles, eyelids, jaw, face, pharynx, tongue (LMN)** → Neuromuscular junction (NMJ)
- ** Jaw, face, pharynx, tongue; sparing ocular muscles, eyelids (UMN and LMN)** → Motor neuron disease (ALS)
- **Specific muscle groups in one limb (LMN)** → Nerve root, plexus or peripheral nerve
**Paraparesis**

- Lesion location is **BILATERAL** (*):

<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 paraparesis with no sensory level</td>
</tr>
<tr>
<td>Thoracic spinal cord</td>
<td>Spastic leg paraparesis, thoracic sensory level</td>
</tr>
<tr>
<td>Lumbar spinal cord</td>
<td>Flaccid paraparesis, 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 - 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.

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;
- HTLV-I infection, HIV myelopathy.

**DIAGNOSTIC APPROACH** begins with spinal MRI or myelography.

**Hemiparesis**

- Lesions **are UNILATERAL** (*):**

<table>
<thead>
<tr>
<th>Lesion Location</th>
<th>Pattern of Signs</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cerebral cortex</td>
<td>Contralateral weakness (arm = leg; face, sometimes tongue)*.</td>
</tr>
<tr>
<td>Left hemisphere: aphasia, apraxia.</td>
<td>Right hemisphere: left hemi-inattention, extinction of sensory stimuli, constructional apraxia, spatial disorientation.</td>
</tr>
<tr>
<td>Right hemisphere:</td>
<td>Homonymous hemianopia on weak side.</td>
</tr>
<tr>
<td></td>
<td>Cortical sensory loss (decreased graphesthesia, stereognosis, point localization).</td>
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<tr>
<td></td>
<td>Horizontal eye deviation (toward lesion side).</td>
</tr>
<tr>
<td>Internal capsule (posterior limb)**</td>
<td>Contralateral weakness (face = arm = leg); face may be spared!</td>
</tr>
<tr>
<td></td>
<td>No sensory loss or aphasia</td>
</tr>
<tr>
<td>Brain stem: see p. A59.</td>
<td>Contralateral weakness (arm = leg) + ipsilateral peripheral cranial nerve palsy:</td>
</tr>
<tr>
<td>Midbrain ( crus cerebri)</td>
<td>Lesion of CN3 (Weber syndrome), red nucleus, superior cerebellar peduncle (limb ataxia contralateral to hemiparesis side)</td>
</tr>
<tr>
<td>Pons (basis pontis)</td>
<td>Lesion of CN6 (Foville syndrome), CN7 (Millard-Gubler syndrome); internuclear ophthalmoplegia</td>
</tr>
<tr>
<td>Medulla (pyramids)</td>
<td>Lesion of CN12; face spared</td>
</tr>
<tr>
<td>Cervical spinal hemicord (Brown-Séquard syndrome)</td>
<td>Ipsilateral weakness; sparing face.</td>
</tr>
<tr>
<td></td>
<td>Ipsilateral loss of proprioception and vibration.</td>
</tr>
<tr>
<td></td>
<td>Contralateral loss of pain and temperature.</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**

- a) usually vascular pathogenesis
- b) traumatic rupture of normal vessels
- c) hemorrhage into primary / metastatic brain tumors
- d) focal inflammatory lesion (multiple sclerosis, sarcoidosis)
- e) acute bacterial abscesses

**Subacute hemiparesis**

- a) subacute subdural hematoma
b) infection - cerebral bacterial abscess, fungal granuloma or meningitis, parasitic infection.
c) malignant primary / metastatic neoplasms
N.B. AIDS may present with subacute hemiparesis due to toxoplasmosis or primary CNS lymphoma!
d) focal inflammatory lesion (multiple sclerosis, sarcoidosis).

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

TETRAPARESIS
- lesion locations are BILATERAL (!):

<table>
<thead>
<tr>
<th>Lesion Location</th>
<th>Pattern of Signs</th>
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<tbody>
<tr>
<td>Cerebral hemispheres</td>
<td>Pernodolbal palsy, decorticate posture (large acute lesions)</td>
</tr>
<tr>
<td>Midbrain</td>
<td>Coma, mid-size poorly reactive pupils, decerebrate posturing</td>
</tr>
<tr>
<td>Basis pontis</td>
<td>“Locked-in” syndrome</td>
</tr>
<tr>
<td>Cervicomedullary junction</td>
<td>Legs &gt; arms, t weakness of pharynx &amp; tongue, facial hypalgpsia (descending tract of CN 5)</td>
</tr>
<tr>
<td>High cervical</td>
<td>No cerebral signs (cranial nerve palsies, etc)</td>
</tr>
<tr>
<td>Mid cervical</td>
<td>Preservation of shoulder movements</td>
</tr>
<tr>
<td>Peripheral nerves (e.g. acute demyelinating polyneuropathy)</td>
<td>Distal weakness</td>
</tr>
<tr>
<td>Muscles (myopathy)</td>
<td>Proximal weakness</td>
</tr>
</tbody>
</table>

MONOPARESIS
A. With pain
- 1. Compressive 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.

BIRACHIAL PARAPARESIS
- 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).
- 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

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

BILATERAL HYPERREFLEXIA
- 1. Normal axon 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)

VOICEX:
- 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, traumatic 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>Polyradiculopathy (acute inflammatory demyelinating)</td>
<td>Limb weakness (legs before arms), areflexia, absent or flexor toe response, minimal distal sensory loss without sensory level</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>
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<tr>
<td>LESS COMMON</td>
<td></td>
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<tr>
<td>Partial motor seizure, Todd's paresis (postictal 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>Tension 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>Sheep study</td>
</tr>
<tr>
<td>Sleep paralysis</td>
<td>Narcolepsy; terminated by touch</td>
<td>Sheep 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>Negative myoclonus</td>
<td>Sudden, brief, rapid, unpredictable (shocklike) inhibition of muscle tone (single muscle ÷ entire body).</td>
<td></td>
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<tr>
<td>RARE</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Periodic paralyses</td>
<td>Familial channelopathies</td>
<td>Serum K+</td>
</tr>
</tbody>
</table>

**DROP ATTACKS**

- 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.
     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 >>