

Disorders of motility

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Definitions

- **Paralysis (plegia)**: total loss of voluntary movement due to damage of the motor pathways at any point: cortical motoneuron, lower motor neuron, axons – neuromuscular transmission – muscle diseases...
- **Paresis**: a lesser degree of paralysis
- **Plegia** comes from a Greek word meaning „to strike” (e.g. hemorrhagic stroke – ictal plegia)
- **Palsy** comes from an old French word „same meaning as ” *paralysis*”

GRADES: 0 TO 5

0/5: No motion ==Paralysis (plegia)

1/5: Slight muscle motion, but minimal motion at joint

2/5: Full motion parallel to ground (can't move against gravity)

3/5: Full motion against gravity, but not against resistance

4/5: Full motion against some resistance

5/5: Full motion against full resistance; normal

Lower motor neuron

- Motor neuron, its axon and the muscle fibers it innervates constitute the motor unit
- Muscle fibers innervated by a single motor neuron can be only a few or up to 1000
- Delicate movements involve few motor units
- Powerful movements involve many motor units

Interruption of a motor nerve

- Denervated muscle fibers (by **axonal – Wallerian degeneration**) contract spontaneously (within a few days of after the damage of the axon)
- This is called **fibrillation**, it is so fine that it is not visible_ (probably isolated muscle fibers are not able to maintain a stable membrane potential)

Fibrillation: 1-2 msec, 100-300 mV, (-)

Fasciculation

- Spontaneous, involuntary contraction of the motor unit, or part of the motor unit
- **Visible** dimpling or twitching under the skin
- Evidence of motor nerve fiber irritability
- Typically points **to reinnervation following a nerve or motor neuron damage**

Fasciculation: polyphasic, once or two in every sec., duration 5-15 msec., amplitude several millivolts

Fasciculation

- Fasciculation is typical for **slowly advancing, destructive diseases** of spinal motor neurons (amyotrophic lateral sclerosis, spinal muscular atrophy)
- **Fasciculation** occasionally develops due to anterior root lesion (disc herniation), compression- and other neuropathies
- If large number of axons are affected **fasciculation** may be more prominent (even cramps can appear)

benign fasciculation

- *benign fasciculation occur in **many healthy person** occasionally, even for many years without weakness or wasting*
- In calves, hands, periocular paranasal muscles

Combination of fibrillation and fasciculation

- active denervation (**fibrillation**) combined with reinnervation (**fasciculation**) of muscle *points to a long standing but still ongoing process* (e.g. amyotrophic lateral sclerosis (ALS), chronic inflammatory demyelinating polyneuropathy (CIDP))

Signs of the lesions of the
lower motor neurons
(brain stem, spinal cord)

- Paresis
- Flaccidity (hypotonia, atonia)
- Tendon reflexes are lost (or diminished)
- Muscle atrophy (can be extreme: 20-30% loss of original bulk within 3-4 months)
- Fibrillation, fasciculation
- *Flaccid paresis, loss of tendon reflexes, muscle atrophy sometimes fasciculation can be the consequence of anterior root lesions, or (pure motor) neuropathies too*

Upper motor neuron and (pyramidal) corticospinal tract

- **Pyramidal tract:** in strict sense those fibers that course longitudinally in the pyramid of medulla oblongata.
- It descends from the cortex, crosses corona radiata, posterior limb of internal capsule, cerebral peduncle, ventral pons, pyramid of upper medulla, **crosses in lower medulla** and continues in the lateral funiculus of the spinal cord = **corticospinal tract**
- **corticospinal tract is the only direct long fiber connection between cerebral cortex and spinal cord.** Only 25-35 000 large motor Betz cells are in the fifth layer of the primary cortex – pyramid in the medulla contains about **1 million axons**, thus the pyramidal tract contains fibers from Br 4, 6, even 3,1,2 (primary somatosensory cortex), superior parietal cortex (Br 5,7). Only 10-20% of corticospinal fibers (of Betz cells?) establish direct synapses with spinal motor neurons
- **Indirect tracts:** rubrospinal, tectospinal, vestibulospinal; these tracts do not run in the pyramid of medulla (hence '**extrapyramidal**') (functions: stability of the head /labyrinth/, **postures of body, axial stability** (Parkinson diseases, Parkinson 'plus' diseases))

Clinical signs of (cortical) **upper motor neuron** lesions

- Spastic hemiparesis (one sided hemispheric lesion)
- *Spastic paraparesis (spinal cord injury)*
- *little or NOT effected* : motor cranial nerve functions (**except lower part of the face (central facial palsy), tongue (central hypoglossal palsy)**), movements of the neck, thorax, abdomen, diaphragm

Spasticity and rigidity

- **spasticity**: velocity dependent increase in the resistance of muscles to a passive stretch stimulus: „clasp knife phenomenon”
- **spasticity** affects predominantly the antigravity muscles: flexors of the arms, extensors of the legs
- **rigidity**: tone is increased both in agonists and antagonists muscles (‘led pipe character’) – one of the major sign of Parkinson disease (extrapyramidal origin)

Note for your practice

- *Acute, profound spinal cord lesions, profound corticospinal lesions in the brainstem or cerebrum may temporarily abolish tendon reflexes and muscle tone due to the interruption of descending tonic excitatory impulses (from reticular formation)*

Clinical signs of upper motor neuron lesions

- Spasticity and paresis depend on separate mechanisms: selective blocking of gamma motor neurons abolishes spasticity, as well as hyperactive tendon reflexes, but do not have effect on motor performance

if there is **no** contraction
of the intrafusal fibers by gamma motor
neurons there is no stretch the nuclear
bag region. → No afferent fiber
activation → no α -motor neuron
depolarisation → **no muscle tone** :

Clinical signs of upper motor neuron lesions

- The **most sensitive indicator** of an upper motor neuron (corticospinal tract) lesion is **Babinski sign** (described in 1896)
- It can appear in normal infant, but later in life it is an **invariable indicator** of a lesion in the corticospinal tract
- **Important: extension of the large toe is a component of triple flexion (nocifensive) response**
- With incomplete corticospinal lesion, response may be fractionated: the hip and knee may flex, but the foot may not dorsiflex, or vice versa
- **Clonus:** rhythmic involuntary muscular contractions to an abruptly applied and sustained stretch

Clinical signs of upper motor neuron lesions

- **Spread of reflex activity:** knee reflex can be elicited by tapping the tibia
- Hoffmann-Trömner sign are useful only in cases of clear-cut asymmetry

Differences between upper (central) and lower motor neuron (peripheral) paresis

Upper (central, corticospinal) lesion

Lower motor neuron, peripheral lesion

- **Never individual** muscle is affected ↔ **Individual** muscle **can** be effected (e. g. C5 lesion-deltoid)
- **Slight** atrophy (inactivation can result in) ↔ **Atrophy profound** up to 70% of the original bulk
- **Hyper-reflexia, Babinski sign (triflexion!), spasticity** ↔ **Hypo-reflexia, loss of tendon reflexes, flaccidity**
- Fasciculation **absent** ↔ Fasciculation **characteristic**
- EMG: normal ↔ EMG: **fibrillation, fasciculation** (if nerve conduction impaired =polyneuropathies)

- APPENDIX

- **Dystonias** are agonists-and antagonists muscle contracts simultaneously. Examples include blepharospasm, spasmodic torticollis, oromandibular, spasmodic dysphonia, and writer's cramp.
- The term **chorea** means dance. Quasi-purposeful movements are observed that affect multiple joints with a distal preponderance.
- **Hemiballismus** is a violent flinging movement of half of the body. It is associated with lesions of the subthalamic nucleus (ie, body of Louis).

- **Tics** are involuntary contractions of single muscles or groups of muscles that result in stereotyped movements. Gilles de la Tourette syndrome can manifest with multiple tics and elaborate, complex movements and vocalizations.
- **Myoclonus**, as the word implies, is a muscle jerk; it is a brief (<0.25 s), generalized body-jerk, which at times is asymmetric.
- **Asterixis** can be elicited by having the patient extend both arms with the wrists dorsiflexed and palms facing forward and eyes closed; brief jerky downward movements of the wrist are considered a positive sign.