Movement in all conditions

- Standing up, sitting, lying – just to fix up the skeleton
- Movement of the limbs – hands, legs
- Breathing – diaphragm & others
- Old implicit acts „reflexes“ – sniffing, swallowing

Motor processes always comprise 2 qualities, although not equally and not always visible
- Phasic activity - episodic, contractions, spikes in EMG, prevail in kinetic muscles, mostly flexors, red muscle fibres, energy demanding, rapid fatigue.
- Tonic activity (tone) - sustained, tightening of segments, muscle resistance & turgor, no evident EMG, prevail in postural muscles, mostly extensors, white muscle fibers, slower fatigue
- Isometric – muscle length is kept stable, change the tone
- Isotonic – tone maintained stable, change the length

Inborn reflexes
- Blink reflex
- Grasp reflex
- Stepping reflex
- Diving reflex
- Rooting reflex
- Sucking reflex
- Tracking reflex
- Startle reflex
Clinical considerations - terminology

- Paralysis, paresis, palsy
  - Hemiparesis, quadruparesis, monoparesis, paraparesis,
- Hypokinesia, bradykinesia
- Hyperkinesia, dyskinesia
  - Chorea, athetosis, tics, ballism, tremor, akathisia, myotonia, myokymia, myorhythmia
- Hypotonia - flaccidity
- Hypertonia
  - Spasticity, rigidity
- Dystonia, spasms
- Ataxia (dystaxia)

Clinical evaluation and terminology

- Ability to move, muscle force:
  - Muscle weakness (Paresis)
  - Paralysis (Plegia, Palsy)
  - Distribution of these:
    - Hemiparesis, quadruparesis, monoparesis, biplegia, paraparesis, etc.
- Appropriate amount or pattern of movement:
  - Hypokinesia, bradykinesia
  - Hyperkinesia, dyskinesia
    - Chorea, athetosis, tics, ballism, tremor, akathisia, myotonia, myokymia, myorhythmia
- Muscle tone:
  - Hypotonia - flaccidity
  - Hypertonia
    - Spasticity and rigidity
  - Dystonia and spasms
- Gait – standing, walking (narrow base, wide-base):
  - Ataxia (dystaxia)
    - Spinal (posterior collums)
    - Cerebellar
    - Frontal
    - Vestibular
PARALYSIS (PALSY)

Lower motoneuron syndrome – peripheral palsy (weakness)

- Weakness (palsy) in one or more muscles, groups
- watershed of nerve, plexus, root, anterior horn
- Hypotonia, atonia (flaccidity) - floppy
- Hyporeflexia, areflexia
- Muscle atrophy
- Fasciculations, fibrillations
  - mostly under motoneuronal damage (cord, brainstem)
- Spasms, cramps
  - in unaffected antagonistic muscles

Causes:
- damage to the nerve, plexus,
- damage to the anterior horn of spinal cord (trauma, ischaemia) or ventral roots

Symptoms are homolateral to the site of damage

Upper extremity

- Radial Nerve
- Median Nerve
- Ulnar Nerve

Lower extremity

- N. gluteus inferior
- Lateral paralysis of n. femoralis
- Sciatic Nerve

Fibular (peroneal) nerve
- Compression of common fibular nerve over fibular head, e.g. sitting with legs crossed, or sleeping on side on hard surface
- Posterior dislocation of hip. Fibular head may impinge on sciatic nerve, leading to palsy.
**Upper motoneuron syndrome – central paralysis (weakness)**

**A. Acute stage, B. Chronic stage – after 7-14 d.**

- Hemiparesis (-plegia), paraplegia, monoparesis
  - Loss of fast, delicate, (flexor) movements
  - Face (mouth, tongue), hand (arm swing, grasp, fingers), internal rotation, foot – external, dorsal flex.
- Hypertonia - spasticity (clasp-knife)
  - Acutely little evident, mostly late sign
  - Always occurs in groups of muscles, not individual muscles.
- Hypereflexia – brisk UE, LE reflexes
- Spasms in affected muscles
- Pathological reflexes – Babinski - extensor plantar response
  - Main finding in acute stage compared to LMNS

**Causes:** damage to motor cortex and along the pyramidal pathway (capsula interna, brainstem, spinal cord)

Symptoms are contralateral to damage.

**UMNS manifestation**

- Water-shed of ant. crebral a.
- Capsula interna
- Pons

**UMNS vs. LMNS**

**UMNS**

- Corticospinal (bulbar) + subcorticospinal tr.
- Muscles are normal
- Hypertonia (spasticity)
- Reflexes are brisk
- Spasms, cramps in affected muscles
- No irritation signs
- Pathological reflexes

**LMNS**

- Mononeurons + axons
- Muscles atrophy
- Atonia (flaccidity)
- Weak reflexes
- Spasms, contractures in unaffected muscles
- No irritation signs
- Pathological reflexes
BASAL GANGLIA DISORDERS

- **Structures:**
  - Caudate nucl., putamen, globus pallidus, substantia nigra, subthalamic nucl., red nucleus, nucl. campi Forell

- **Function of BG:**
  - Initiation of movement, Muscle tone
  - Complex motor programs, implicit memory

- **Clinical manifestations:**
  - No palsy or weakness, problem is rather in amount, fluency, smoothness, timing
  - Goal-directed movements are interrupted, uncoordinated, slow or infiltrated by escaped movement intrusions
  - Muscle tone is always affected – hypotonia, dystonia, hypertonia
  - Difficulties are ameliorated at rest, omitted in sleep, perpetuated by motion tasks, worsened by good or bad emotional excitement

HYPERKINETIC - DYSTONIC MOVEMENT DISORDERS

- **TREMOR** - a rhythmical predictable oscillation of a body part
- **DYSTONIA** - an involuntary muscle contraction causing a sustained twisted or abnormal posture
- **MYOCLONUS** - a lightening like jerk of a body part
- **STEREOTYPY** - any patterned, stereotypic movement.
- **TIC** - a stereotypic or patterned movement that is frequently preceded by an urge to need to move, transient suppressibility, and post movement relief.
- **CHOREA** - random, purposeless, fleeting movements, spreading from one body part to another.
- **HEMIBALISMUS** - A high amplitude flailing of the limbs on one side of the body.

HYPOKINETIC - HYPERTONIC MOVEMENT DISORDERS

- **PARKONSONISM**
- **Stiff Man Syndrome, Akinetic Mutism**
- **Psychomotor Retardation**

Biochemistry

- Ach> dopamine -> hypokinesia
- Dopamine> Ach -> hyperkinesia
**Parkinson's disease**

- Neurodegenerative disorder - described in 1817 by James Parkinson

**Pathology:**
- Degeneration of SNpc, symptoms evident after 80% loss,
- Degeneration of raphe nuclei (serotonin) and LC (norepinephrine)

**Course:** Insidious onset, often hand tremor and distal stiffness 10-20 years to incapacitation, symptoms can disappear for periods

**Causes:**
- Spontaneous & inherited, poisoning by Hg, Mn, Fe, Cu.
- MPTP (N-methyl-4-phenyl-1,2,3,6-tetrahydropyridine) contaminant in synthetic heroin; converted to MPP+ which is toxic to DA cells

**MPTP treated macaque monkeys (model of Parkinson's disease)**
- Hypokinesia/bradykinesia, rigidity, tremor reversed with L-DOPA
  - DA cells in ventral tegmentum & NE cells in locus coeruleus destroyed resembling human
  - Elevated levels of activity in GPi, disinhibition of STN and excessive inhibition in Gpi, excessive excitatory drive to Gpi/SNpr
  - Excessive thalamic inhibition, reduced cortical production of movements.

**Lesions of STN result in immediate, dramatic reduction of akinesia and bradykinesia as well as tremor and rigidity in contralateral limbs.**

**Manifestation:**

**Positive symptoms** - behaviors normally inhibited
- Tremor (80%) – mostly in rest (hands, feet, chin, tongue), sparing the head; thumb slides back and forth on the index finger - "pill rolling"
- Rigidity (stiffness) - loss of arm swing with walking, lack of facial expression, fatigue, muscle pain, "cog wheel"

**Negative symptoms** – missing but normally present
- Hypokinesia
- Disorders of posture
  - Disorders of fixation - inability to maintain a part of body in normal position
  - Disorders of equilibrium - difficulty in standing or sitting
  - Disorders of righting - inability to get up
- Disorders of locomotion
  - Difficulty to start and to maintain the movement (slowing down),
  - Disorders of speech
    - Telegraphic slurred speech, soft voice.
- Akinesia, bradykinesia delayed & slowed movements
  - Facial mask, shuffling gait, messy illegible writing, drooling (wet pillow) due to difficult swallowing (50%), freezing or sudden loss of movement

**Parkinson's syndrome**

- **Manifestation:**
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    - Tremor (80%) – mostly in rest (hands, feet, chin, tongue), sparing the head; thumb slides back and forth on the index finger - "pill rolling"
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**Dystonia**

- **Manifestation:**
  - Sustained, irregular, involuntary contractures
  - Focal - cervical dystonia (torticollis), writer's cramp, blepharospasm, oromandibular dystonia, Meige's syndrome
  - Generalized – torsiospasm
- **Causes:**
  - Hereditary (focal) vs. acquired (central lesions), occupational
- **Mechanism:**
  - Cholinergic excess in striatum (anticholinergic therapy)
- **Treatment:**
  - Anticholinergics, botulinum toxin injections

**Chorea (choreos = dancing)**

- **Manifestations:** distal limbs, head
  - Irregular, fast jerky movements in distal muscles – hands, head, feet;
  - Steps are overswinging, staggering
  - Obeisance - like poses, gestures by hands, fingers), head turns and grimasing, unrest, jitterning in legs, unsettled appearance
- **Causes:**
  - Huntington's disease + other hereditary dis.
  - Sydenham's chorea – acute rheumatic fever
  - Cerebral palsy, pregnancy, etc.
- **Mechanism:**
  - Loss of cholinergic & abundance of dopaminergic effects in striatum

**Huntington's disease**

- **George Huntington – 1872 (first systematic study)**
- **Occurrence:**
  - 1.6 per million per year death rate; more common in caucasian Europeans; rare in Asians or Africans
  - Village of Bures in England in 1630 - individuals thought to be witches
  - Brought to US in 1630 among passengers of John Winthrop fleet
  - Lake Maracaibo - large incidence - one woman whose father, an English sailor, carried the gene - > 3000 descendent, 100 with Huntington's disease, 1,100 children with 50% chance of having it!
- **Etiology:**
  - Hereditary AD- transmitted disease (discovered in 1993) studying 75 families from Lake Maracaibo
  - Defective huntingtin protein (Ch4) trinucleotide repeat mutation; CAG triplet occurs 11-34 times in the normal gene, from 35 to 100 or more times in mutant
- **Pathogenesis:**
  - Degeneration of caudate nucleus - loss of cholinergic and GABA-ergic neurons in basal ganglia; relative excess of DA
  - Modelled in nonhuman primates by excitatory neurotoxins injected in the striatum
- **Manifestation:**
  1. Middle-age onset (40-50y) subtle start: absentminded, irritable, depressed, fidgeting, clumsiness
  2. Progressive chorea: violent uncontrolled overbursts - until individual confined to bed
  3. Dementia: cognitive impairment, speech is slurred, incomprehensible and finally stops; death after 15-20y
**Athetosis (athetos = fidgety)**

- **Manifestations:**
  - Irregular, twisting, revolving, turbulent, widely bursting, fidgety extra-movements of extremities
  - Walking – interrupted, staggering, swinging, rolling
  - Poses, obeisance, head turns, grimacing hands, fingers (gestures), legs (shaking)

- **Causes:**
  - Cerebral palsy, pregnancy, etc.

- **Mechanism:**
  - Loss of cholinergic & abundance of dopaminergic effects in striatum

**Hemiballism**

- **Manifestation:**
  - Sudden, violent, purposeless, excessive, throwing movements, gyrations (ball = throw; ballistic rocket)

- **Causes:**
  - Stroke in subthalamic nucleus

- **Mechanism:**
  - Cortical escape from basal ganglia control

**Tremor**

- **Resting 4-6 Hz (occurs with limb inactivity, chin, hand)**
  - Parkinsonism, heavy manual work, emotional distress, midbrain stroke

- **Action (intention) 3-4 Hz (exposed during movement)**
  - Cerebellar disease, midbrain stroke

- **Postural (occurs with antigravity posturing, exposed in fingers of outstretched arms, protruded tongue)**
  - Exaggerated physiologic 10-12 Hz
  - Catecholamines, sympathetic
  - Essential 4-10 Hz
    - 50% inherited familial tremor
    - Treatment: beta-blockers, primidone

**Tardive dyskinesia**

- **Etiopathogenesis:**
  - Chronic treatment (> 6 weeks) by dopamine antagonists, neuroleptics
  - Hypersensitivity of striatal DA receptors

- **Manifestation:**
  - Orofacial repetitive movements
  - Limb an trunk involuntary movements
**Tics**

- **Manifestation:**
  - Brief, stereotypic, predictable, suppressible jerks worsening with stress
  - Vocalisations, grimacing, swearing, rising eye brows, gestures, grinning
  - Worse with stress

- **Mechanism:**
  - ? Dopamine excess causing disinhibition of limbic circuit

**Cerebellar disorders**

- **Manifestation:**
  - Hypotonia - pendular knee reflex
  - Loss of elementar postural reflexes, Asynergy
  - Cerebellar ataxia – zig-zag walking, titubations, falling to back or sides
  - Adiadochokinesis
  - Hypermetria, dysmetria
  - Intentional tremor

- **Causes:**
  - Trauma, ischemia, haemorrhage, tumors, degeneration, demyelinisations affecting cerebellum, 4th- ventricle, pontocerebellar angle, pathways into and from

**Apraxia**

- **Liepmann (1900)**
  - inability to produce a movement that is not due to paresis (paralysis) - specific loss of skill
  - various forms depending on site of damage (not complete agreement on designations and criteria)
    - Ideational apraxia
      - misuse of objects due to disturbance of identification (agnosia)
    - Ideomotor apraxia
      - simple movements can be executed, but complex movements cannot
    - Limb kinetic apraxia
      - inability to make movements or use objects or the purpose intended by the will
  - clinical / experimental testing necessary to distinguish damage to a motor system from damage to areas that control it
- some tests with no bilateral impairment
  - Finger-tapping speed
  - Movement steadiness
  - Repetitive screw rotation
  - Imitation of single hand posture, imitation of single face posture
- some tests with bilateral impairment
  - Finger tapping on 2 keys
  - Finger tapping with rhythm
  - Manual sequence box
  - Imitation of multiple hand movements
  - Imitation of multiple face movements
**Apraxia 2**

- Premotor cortex (area 6 - lateral)
  - inability to produce indirect trajectories
  - ablation of premotor cortex impairs conditional motor behavior
- Supplementary motor area (area 6 - medial)
  - Brinkman, bimanual coordination deficit
  - absence of speech
- Prefrontal
  - deficits in delayed response tasks
    - dorsolateral - spatial
    - ventral - object
- Posterior parietal cortex (areas 5, 7)
  - Gerstmann's syndrome
    - Following left parietal (in normal right hand dominant)
    - left-right confusion
    - finger agnosia
    - dysgraphia, dyscalculia, apraxia
  - Balint's syndrome
    - Following bilateral damage
    - unable to make voluntary eye movements into affected hemifield
    - optic ataxia - deficit in visually guided reaching, deficit in visual attention
- Neglect
  - Following right (nondominant) hemisphere damage
  - Constructional apraxia