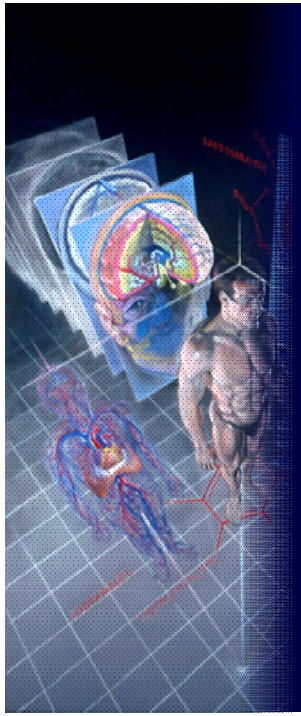

MOVEMENT DISORDERS - BASICS

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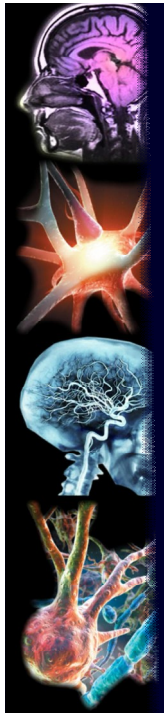
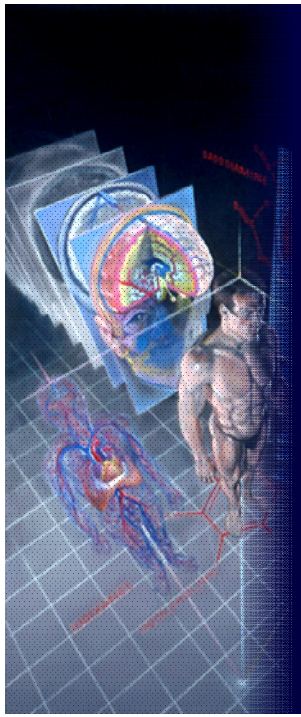


Movement in all conditions

- 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

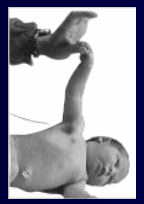


1

MOTOR DISORDER' BASICS

Inborn reflexes


Blink reflex


Grasp reflex


Stepping reflex

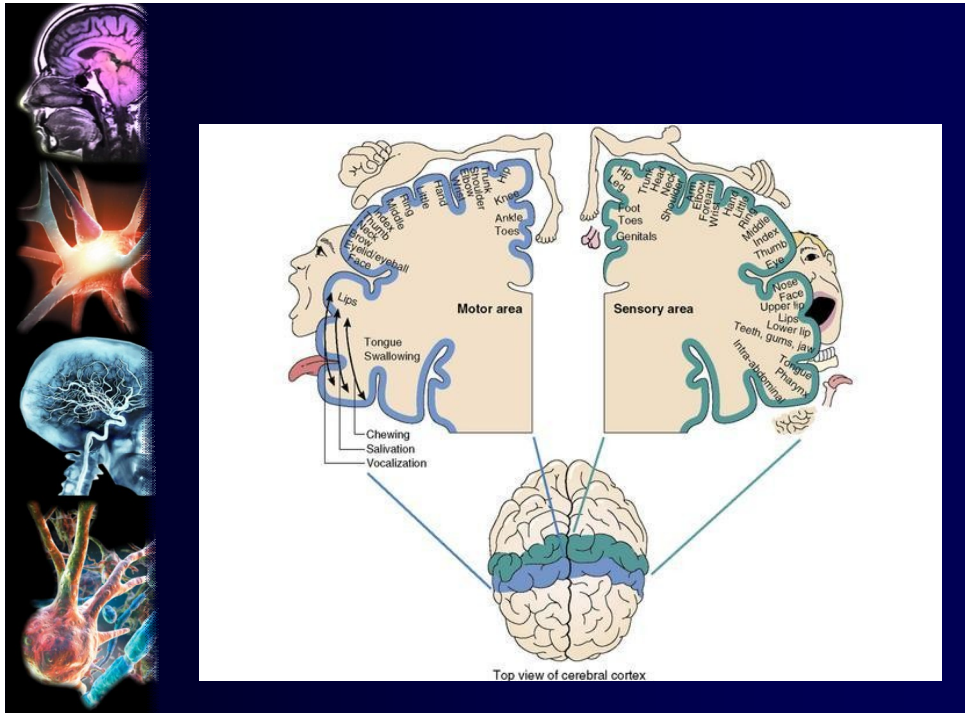

Diving reflex

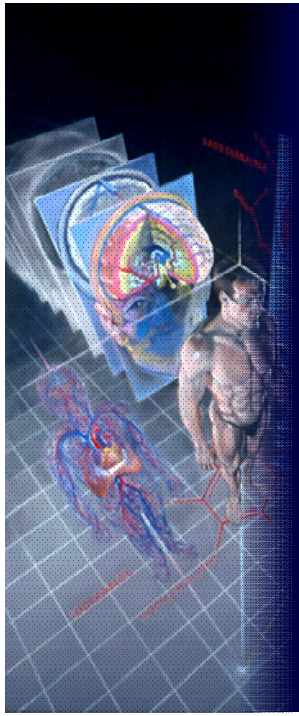

Rooting reflex


Sucking reflex


Tracking reflex


Startle reflex





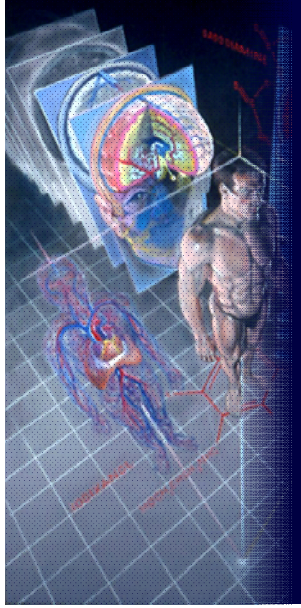
Clinical assessment

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



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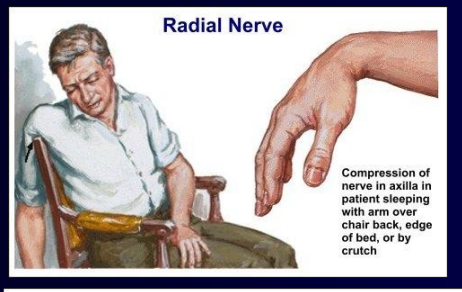
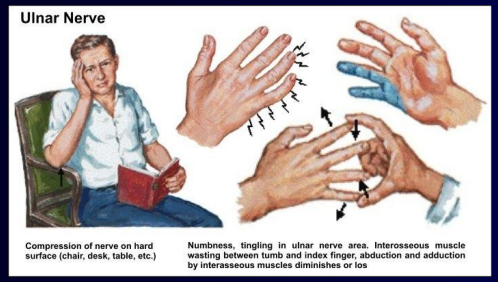
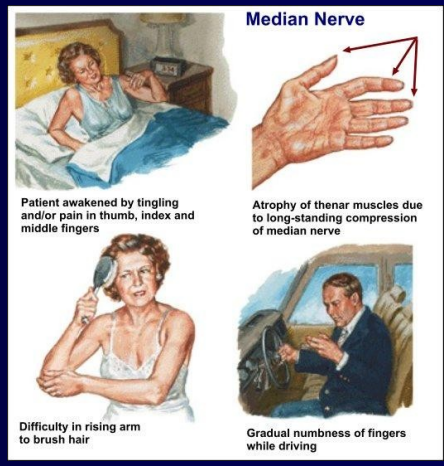
PARALYSIS (PALSY)

Upper extremity







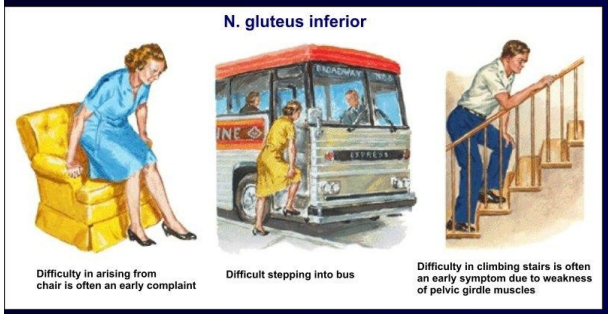
Lower motoneuron syndrome – peripheral palsy (weakness)

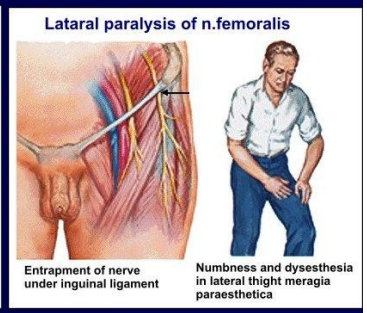
- Weakness (palsy) in one or more muscles, groups
  - watershed of nerve, plexus, root, anterior horn
- Hypotonia, atonia (flaccidity) - floppy
- Hyporeflexia, areflexia
- Muscle atrophy
- Fasciculations, fibrillations
  - mostly under motoneuronal damage (cord, brainstem)
- Spasms, cramps
  - in unaffected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

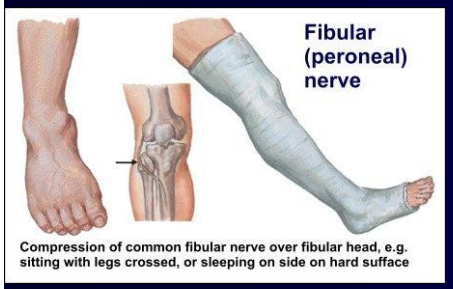
Causes:
- damage to the nerve, plexus,
- damage to the anterior horn of spinal cord (trauma, ischaemia) or ventral roots

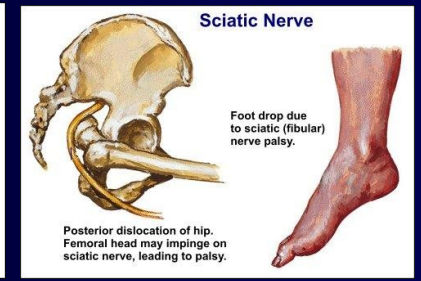
Symptoms are homolateral to the site of damage

Lower extremity









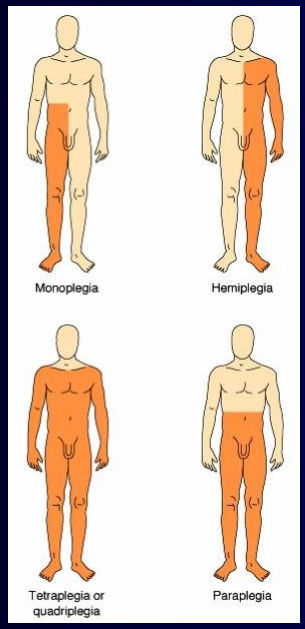
Upper motoneuron syndrome – central paralysis (weakness)

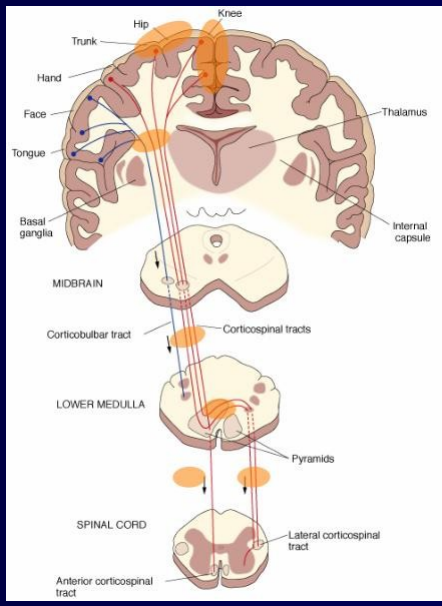
A. Acute stage, B. Chronic stage – after 7-14 d.

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

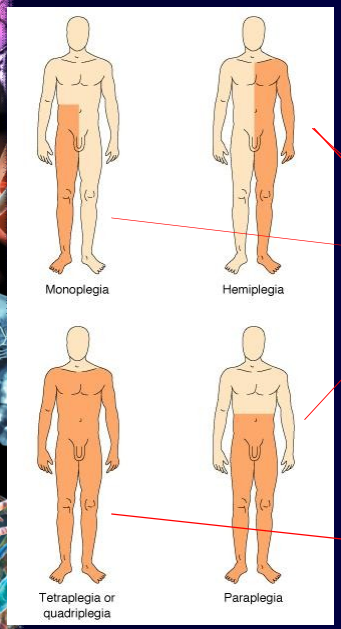
Causes: damage to motor cortex and along the pyramidal pathway (capsula interna, brainstem, spinal cord)

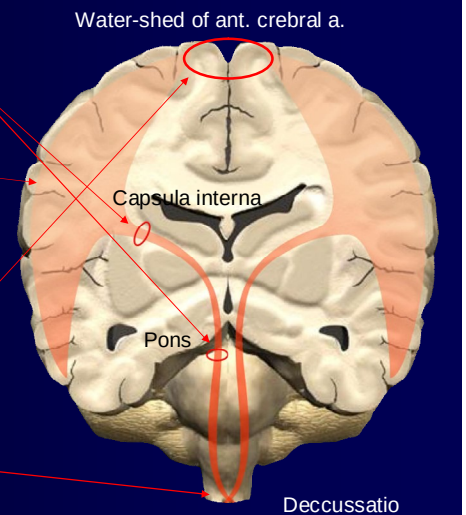
Symptoms are contralateral to damage





UMNS manifestation



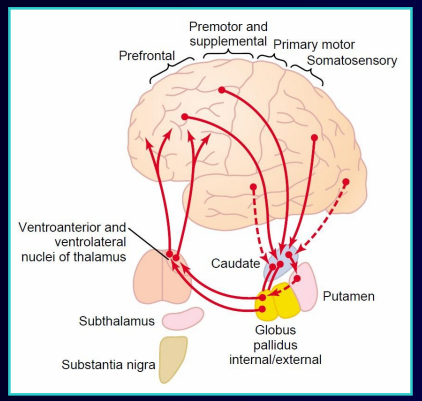


UMNS vs. LMNS

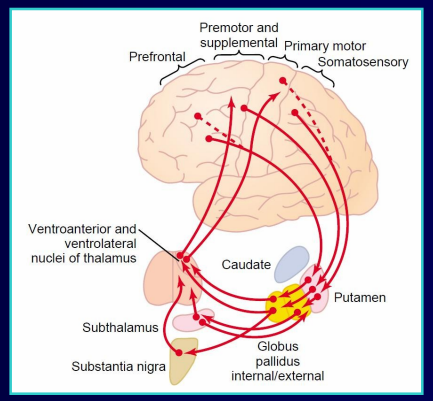
UMNS	LMNS
Corticospinal (bulbar) + subcorticospinal tr.	Mononeurons + axons
Muscles are normal	Muscles atrophy
Hypertonia (spasticity)	Atonia (flaccidity)
Reflexes are brisk	Weak reflexes
Spasms, cramps in affected muscles	Spasms, contractures in unaffected muscles
No irritation signs	Irritation signs – fasciculations
Pathological reflexes	No pathological reflexes

BASAL GANGLIA DISORDERS

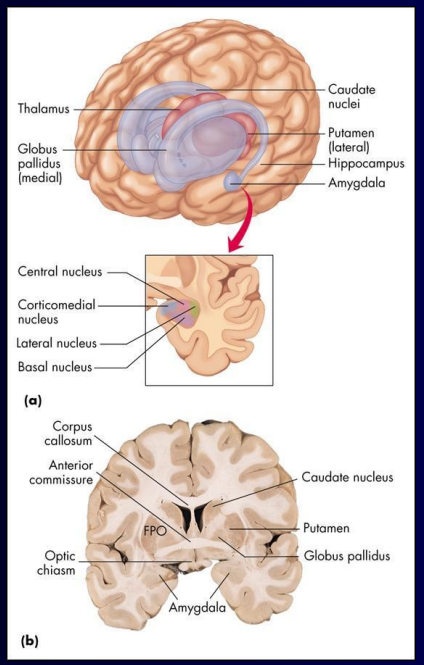
- Structures:
  - Caudate nucl., putamen, globus pallidus, substantia nigra, subthalamic nucl., red nucleus, nucl. campi Forell
- Function of BG:
  - Initiation of movement, Muscle tone
  - Complex motor programs, implicit memory
- Clinical manifestations:
  - No palsy or weakness, problem is rather 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

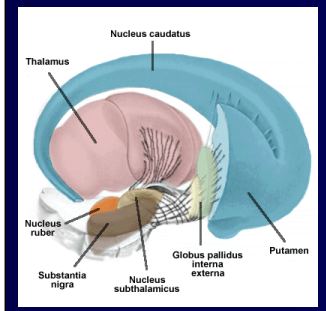


Caudate circuit for cognitive planning of sequential and parallel patterns to achieve specific conscious goals.



Putamen circuit for subconscious execution of learned patterns of movement.





BASAL GANGLIA DISORDERS

- 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' disease

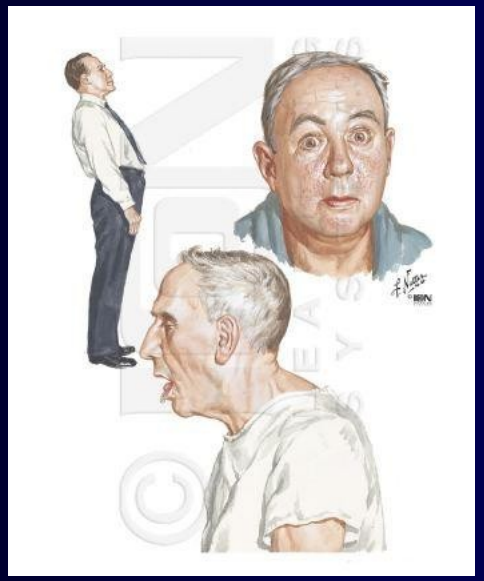
- Neurodegenerative disorder - described in 1817 by James Parkinson
- **Pathology:**
  - degeneration of SNpc, symptoms evident after 80% loss,
  - degen. of raphe nuclei (serotonin) and LC (norepinephrine)
- **Course:** Insidious onset, often hand tremor and distal stiffness 10-20 years to incapacitation, 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
    - 1982 drug addicts in San Francisco with Parkinson's sy.
    - 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 GPe, 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

Parkinson' disease

- **Mechanism:**
  - lack of dopamine in striatum (degeneration within substantia nigra)
- **Manifestation:**
  - Parkinsonism: variety in different patients
  - Cognitive defects: in some progressive cases
    - impaired ability to spontaneously generate efficient strategies when relying on self-directed, task-specific planning
    - deficits associated with frontal lobe damage
      - prefrontal caudate circuit, frontal cortex receives direct dopaminergic input from basal forebrain
    - motor planning deficits
      - increased response time in choice response tasks relative to controls, even accounting for initial difference in simple response time
  - Dementia syndrome: memory loss, mental changes
  - Vegetative dysfunctions: sexual dysfunction constipation, seborrhoea, fatigability, urinary incontinency,
- **Occurrence:**
  - 0.1-1% of population, 3rd most common neurological disease
  - Occurs after 50; may progress 10-20 y

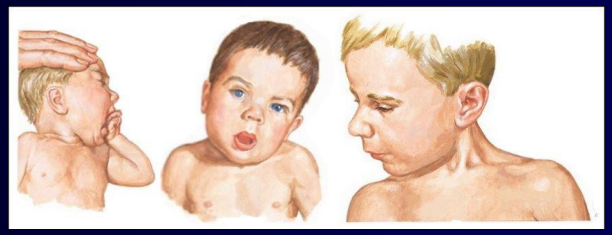
Parkinson' syndrome

- **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



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)
- Tretment:
  - Anticholinergics, botulinum toxin injections

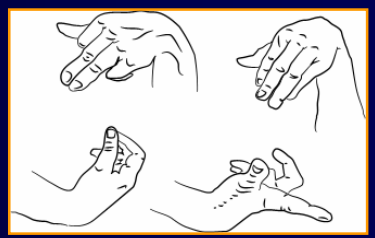


Huntington' disease

- 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 women whose father, an English sailor, carried the gene - > 3000 decendents, 100 with Huntington's disease, 1,100 children with 50% chance of having it!
- Etiology:
  - hereditary AD- transmitted disease (discovered in1993) studying 75 families from Lake Maracaibo
  - defective huntingtin protein (Ch4) trinucleotide repeate mutation; CAG triplet occurs 11-34 times in the normal gene, from 35 to 100 or more times in mutant

Chorea (choreos = dancing)

- Manifestations: distal limbs, head
  - Irregular, fast jerky extra-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:
  - Hungtington'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

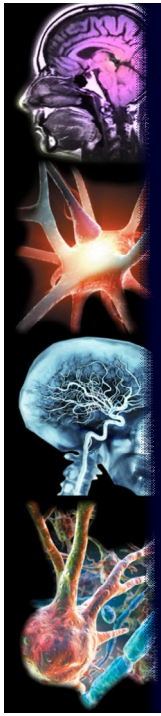


Huntington' disease

- 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

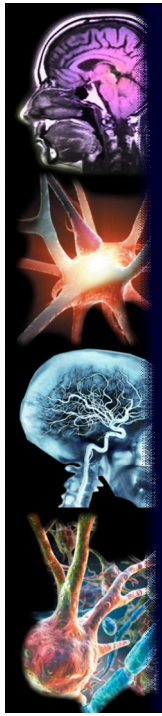
Athetosis (athetos = fidgety)

- **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



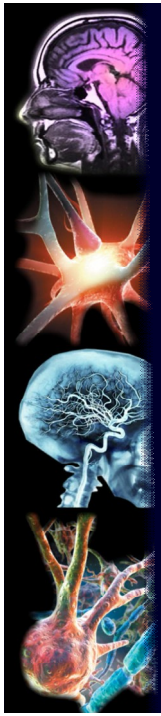
Tremor

- Resting 4-6 Hz (occurs with limb inactivity, chin, hand)
  - Parkinsonism, heavy manual work, emotional distress, midbrain stroke
  - Treatment: dopaminergic agonists
- 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, sympaticus
  - Essential 4-10 Hz
    - 50% inherited familial tremor
    - Treatment: beta-blockers, primidone



Hemiballism

- Manifestation:
  - Sudden, violent, purposeless, excessive, throwing movements, gyrations (ball = throw; ballistic rocket)
  - Limb an trunk involuntary movements
- Causes:
  - Stroke in subthalamic nucleus
- Mechanism:
  - cortical escape from basal ganglia control

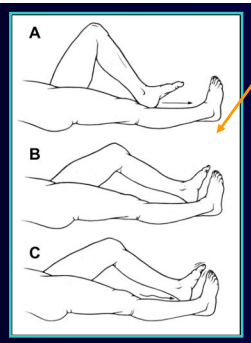


Tardive dyskinesia

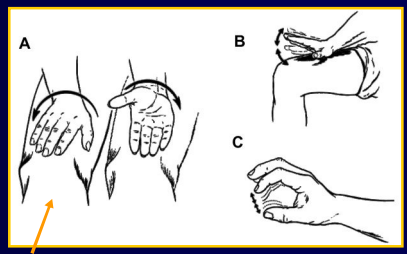
- Etiopathogenesis:
  - Chronic treatment (> 6 weeks) by dopamine antagonists, neuroleptics
  - Hypersensitivity of striatal DA - receptors
- Manifestation:
  - Orofacial repetitive movements
  - Limb an trunk involuntary movements

Tics

- Manifestation:
  - Brief, stereotypic, predictible, suppresible jerks worsening with stress
  - Vocalisations, grimasing,swearing, rising eye brows, gestures, grining
  - Worse with stress
- Mechanism:
  - ? Dopamine excess causing disinhibition of limbic circuit



Test heel – knee. A. normal, B. abnormal



Dysdiadochokinesia:

A. turning of hand
B. tapping, waving
C. thumb- index

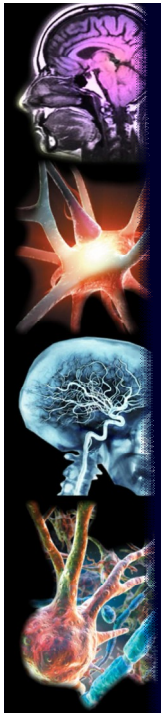
Intentional tremor

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, ponto-cerebellar 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