Clinical Physiology of the Nervous System I.

Motor disorders

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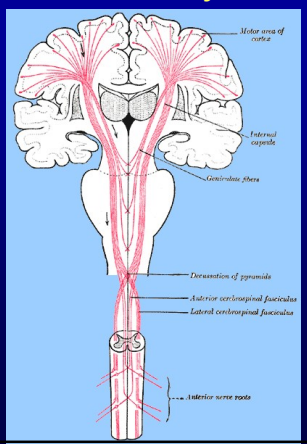
ANATOMY OF THE MOTOR SYSTEM

The motor system



Upper Motor Neurons (UMNs) and Lower Motor Neurons (LMNs)

The terms **UMN** and **LMN** are more helpful in clinical terms of function than anatomic accuracy.

UMNs are all the descending motor fibers coursing through the CNS, that eventually make a synaptic connection to the motor neurons in the PNS.

LMNs are innervated by the upper motor neuron(s).
Cell bodies are in the nuclei of cranial and spinal nerves.
Axon passes through the ventral root.
Motor neuron innervates a collective of muscle fibers, the '*motor unit*'.

3 levels in hierarchy of motor control

1) cerebral cortex
2) brain stem
3) spinal cord (and cranial motor nuclei)
connections are hierarchical (cortex > brainstem > spinal cord) and parallel (cortex > brainstem and cortex > spinal cord)

Motor areas of cerebral cortex - top of hierarchy
three major areas, all in frontal lobe
a) primary motor cortex - executes commands to motoneurons
b) premotor cortex - program, preparation, prox. muscles
c) supplementary motor cortex

All three project directly to spinal cord via the corticospinal tract.

Premotor and supplementary motor cortex (b & c) also project to primary motor cortex and are important in coordinating and planning complex sequences of movement (motor learning).

Motor Cortex

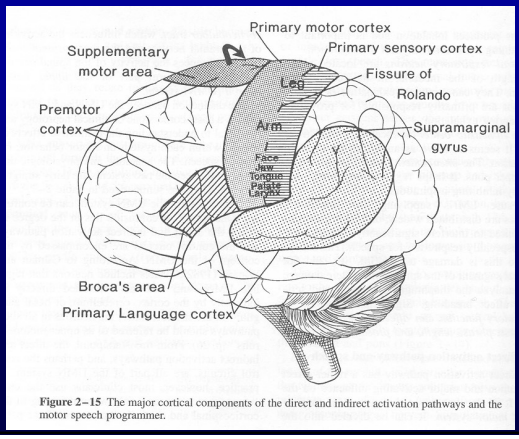
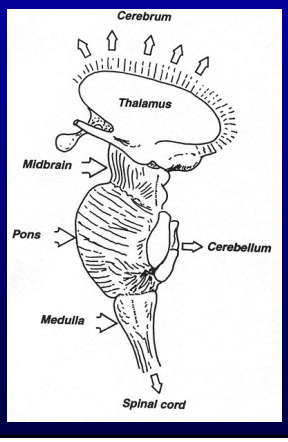


Figure 2–15 The major cortical components of the direct and indirect activation pathways and the motor speech programmer.

Brain stem



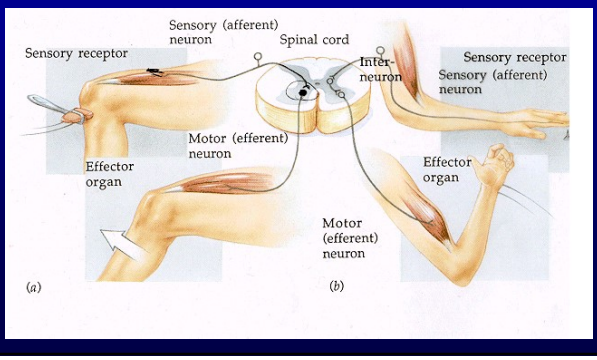
Important nuclei include reticular formation, vestibular nuclei and inferior olivary complex

Axons project and regulate the segmental networks of spinal cord.

Brain stem integrates visual and vestibular information with somatosensory input to modify movements initiated by cortex.

Spinal Cord

neurons mediate automatic reflexes (e.g., stretch reflex)

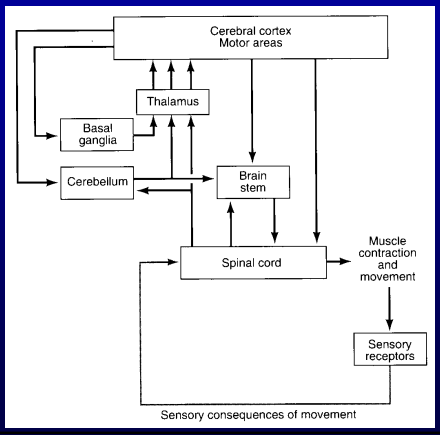


2 important subcortical systems which act on cortex via the thalamus

1) cerebellum
   - receives input from spinal cord
   - projects to both brainstem and thalamus (and onto cortex)
   - improves accuracy of movement (by comparing descending motor commands with information about resulting motor action; thus important in learning)

2) basal ganglia (BG)
   - receives inputs from all cortical areas (not just motor)
   - projects to thalamus and then to areas of cortex involved in motor planning
   - diseases of BG product range of motor abnormalities including hypokinesia and hyperkinesia

Integrated model of motor control



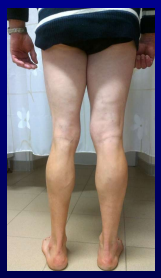
CLINICAL ASPECTS

Complaints typical for damage of the motor system

- Weakness of muscles
- Muscle pain
- Involuntary small contractions (fasciculations), cramps
- Gait disturbance
- Tremor or other involuntary movements
- Imbalance (dissiness)

Physical examination of the motor system (terms to consider for motor disorders)

- Trophism (atrophy, hypertrophy)



Physical examination of the motor system (terms to consider for motor disorders)

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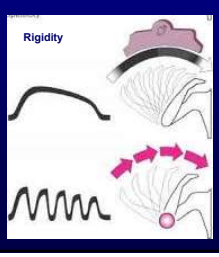

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Hyperkinesia (eg. tremor),
Hypokinesia (Akinesia, Bradykinesia),
Dyskinesia

Physical examination of the motor system (terms to consider for motor disorders)

- *Reflex system*
  - Deep tendon reflexes (biceps, triceps, radius, ulna, patella, Achilles)
  - Pyramidal signs (eg. Hoffmann, Trömner, Babinski)

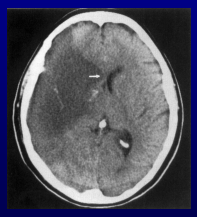
- *Coordination* (limb- and truncal ataxia)
- *Cortical functions* (Apraxia)

- ideomotor apraxia:
disturbed links between motor planning and execution
- Ideator apraxia:
motor plans are missing

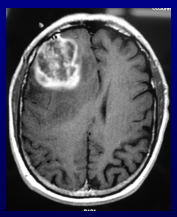
UMN and LMN Lesions

	UMN lesion/syndrome	LMN lesion
Localization	**groups of muscles**	**can restrict to individual muscles**
Muscle tone	**spastic (increased muscle tone)**	**flaccid (decreased tone)**
Reflexes	**exaggerated**	**reduced**
Path. reflexes	**present**	**missing**
Fasciculation	**missing**	**possible (fibrillation by EMG)**
Atrophy	**small, inactivation-induced**	**pronounced**

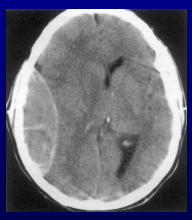
Some of the possible findings in the background of left sided hemiparesis



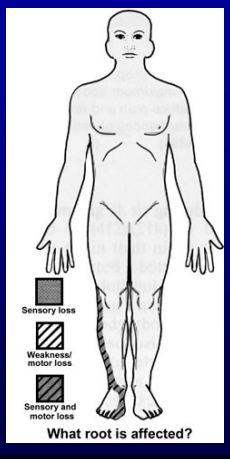
CT: right MCA territory infarct



MRI: right frontal lobe tumor



CT: right epidural haemorrhage



What root is affected?

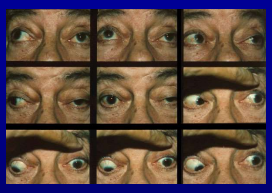
RADICULOPATHY

Pain, sensory, and motor loss.

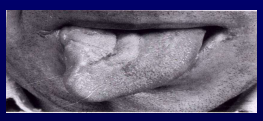
Referable to a dermatome and weakness in muscles innervated by the same root. E.g. in spondylosis

C-5 root damage: atrophy of the periscapular and deltoid muscles

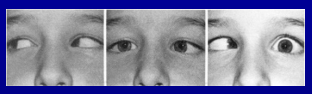
Peripheral Nerve Disorders - Cranial nerves



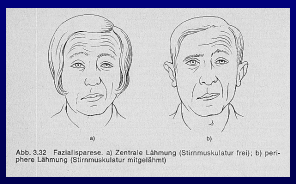
Left oculomotor paresis



Right peripheral
Hypoglossic nerve lesion



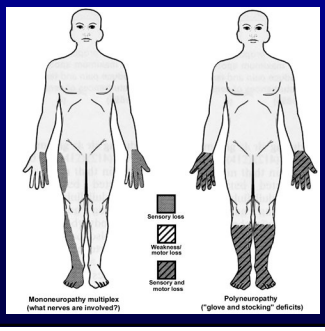
Left abducent paresis



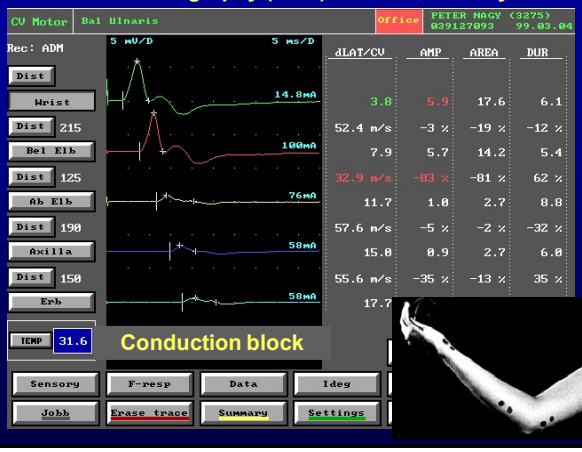
Central facial palsy Peripheral facial palsy

Peripheral Nerve Disorders

Mononeuropathies: sensory and motor deficit referable to the territory of one specific nerve (eg. median, ulnar, peroneal)
Polyneuropathies: distally prominent sensory and motor symptoms (glove and stocking distribution). Motor or sensory system, demyelinating or axonal damage may be more predominant.



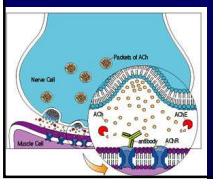
Electroneurography (ENG): cubital tunnel sy



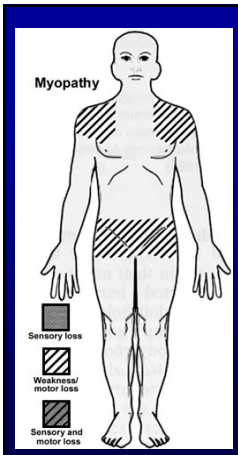
NEUROMUSCULAR JUNCTION DYSFUNCTION
Mysthenia gravis

Autoimmune disease (antigen: AChR, MUSK, titin …)
Congenital (rare, NMJ proteins)

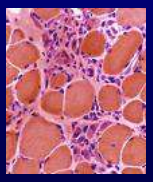
- Muscle weakness, worsens during the day and after exercise
- Main subtypes
  - Bulbar (dysarthria, dysphonia, dysphagia)
  - Ocular (ptosis, diplopia)
  - Generalized (proximal musces, respiratory muscles)
  - MYASTHENIC CRISIS!!!



MUSCLE DISEASES



Affects specific muscles, usually proximal muscles giving weakness with no sensory loss.
Myopathies may be inherited and then termed dystrophies.
Other common causes of myopathy are inflammation (polymyositis), endocrine abnormalities or drugs/toxins.



Cerebellar disorders

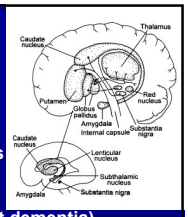
- one of two important subcortical processing loops involved in motor control
- receives input from spinal cord and projects to both brainstem and cortex (the latter via the thalamus)
- has three functional division with distinct anatomical connections to the brain and spinal cord

1) vestibulocerebellum - involved in balance and eye movements (flocculonodular lobe), lesions result in inability to use vestibular information to adjust movement

2) spinocerebellum - modulates muscle tone and motor execution (adjusts ongoing movements) (central part of anterior and posterior lobes), lesions produce hypotonia and ataxia

3) cerebrocerebellum - involved in motor planning (lateral part of anterior and posterior lobes), lesions produce delays in initiating and terminating movement, terminal tremor at end of movement, i.e. intention tremor, disorders in temporal or spatial coordination

Disorders of the Basal "Ganglia"

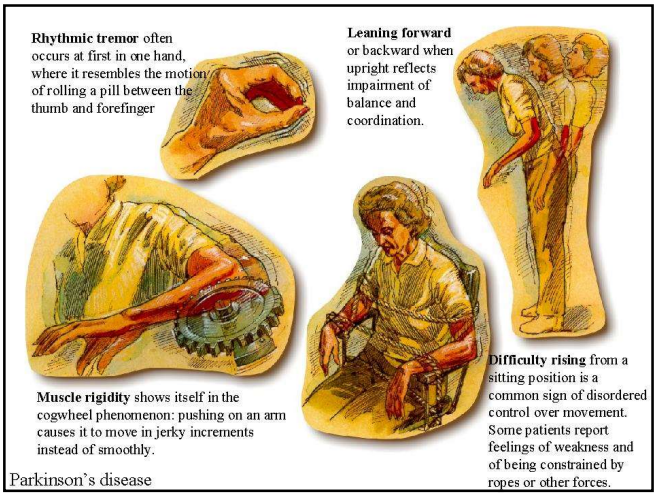


basal ganglia: (misnomer; actually nuclei)
second of two important subcortical processing loops involved in motor control receives inputs from all cortical areas (not just motor; e.g. 100% of Huntington's and >30% of Parkinson's patients exhibit dementia)

projects to thalamus and then to areas of cortex involved in motor planning functional grouping of 5 extensively interconnected nuclei
1-3) corpus striatum - in telencephalon
1) caudate (with putamen forms striatum)
2) putamen - (with caudate forms striatum)
3) globus pallidus (internal and external segments)
4) subthalamic nucleus - in diencephalon
5) substantia nigra (pars reticulata and compacta) - in mesencephalon

Parkinson's Disease

- first described by Londoner James Parkinson in 1817 who termed it *paralysis agitans,*
- disorder characterized by resting rhythmic tremor, akinesia, bradykinesia, rigidity, cogwheel phenomenon and postural reflex impairment,
- most common disorder of BG affecting 500K in the US
- onset of symptoms as early as 40 years but typically 60 years
- results from idiopathic loss of dopaminergic neurons 80% of which are in Substantia Nigra pars compacta
- etiology is unclear, partly genetic (α-synuclein, parkin), partly environmental factors (toxins) thus idiopathic



Huntington's chorea

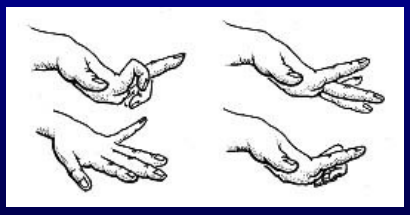
described in 1872 by George Huntington

affects 1 in 20,000
results from selective cell death in striatum
genetic disorder, inherited, AD, 4p16.3, huntingtin, CAG repeat, polyglutamate (1993)

characteristics:
1) onset of symptoms typically in 4th to 5th decade > post- reproductive age
2) chorea (involuntary, rapid, jerky movements)
3) progressive dementia
4) untreatable; death 15-20 (median 16.2) years after onset of symptoms (death results usually from infectious complications of immobility which characterizes terminal phase of disease)

Athetosis

[Gr. *athetos* not fixed + *-osis*] a form of dyskinesia marked by ceaseless occurrence of slow, sinuous, writhing movements, especially severe in the hands, and performed involuntarily.



Positions of fingers in movements of athetosis.

Ballism, Hemiballism

violent flailing movements (bilateral, unilateral)
unilateral ballism results from vascular lesion within the subthalmic nucleus