Movement disorders

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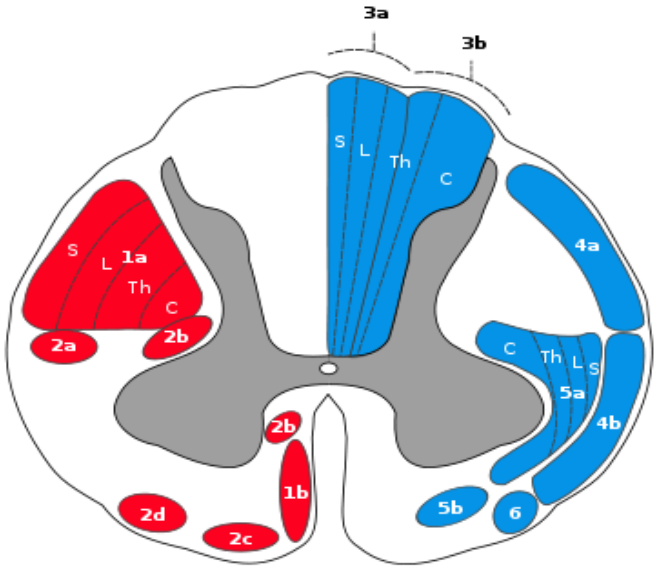
Extrapyramidal system

- Structures involved in the management of the motor function
- Influences the regulatory motor circulus in the spinal cord, brain stem, cerebellum, cortex
- It consists of basal ganglia- nuclei of grey matter in the depths of the hemispheres
- nucleus caudatus, putamen, globus pallidus, nucleus subthalamicus, substantia nigra
- Connection to stem structures and cortex
- Transmiters
- dopamine, acetylcholine, GABA, glutamate

Extrapyramidal system



Motor and decending (efferent) pathways (left, red)	Sensory and ascending (afferent) pathways (right, blue)
1. Pyramidal Tracts	**3. Dorsal Column Medial Lemniscus System**
1a. Lateral corticospinal tract	3a. Gracile fasciculus
1b. Anterior corticospinal tract	3b. Cuneate fasciculus
2. Extrapyramidal Tracts	**4. Spinocerebellar Tracts**
2a. Rubrospinal tract	4a. Posterior spinocerebellar tract
2b. Reticulospinal tract	4b. Anterior spinocerebellar tract
2c. Vestibulospinal tract	**5. Anterolateral System**
2d. Olivospinal tract	5a. Lateral spinothalamic tract
	5b. Anterior spinothalamic tract
Somatotopy Abbreviations:	6. Spino-olivary fibers
S: Sacral, **L: Lumbar**	
Th: Thoracic, **C: Cervical**	

Extrapyramidal system

- Control muscle tone
- Creating synkinesis
-
- Create and control automatic movements
-
- Control of mimic and pantomimic

Movement disorders

- A disorder which impairs the regulation of voluntary motor activity without directly affecting strength, sensation or cerebellar function

- Sometimes also known as “extrapyramidal disorders”
- Many neurologists restrict the term “extrapyramidal” to refer only to **Parkinsonism**

Extrapyramidal syndromes
Movement disorders, basal ganglia disorders

- **Parkinson syndrome**
  Hypokinetic-rigide
  Hypokinetic-hypertonic
  nigrostriate system
-
- **Abnormal-involuntary movements**
  Hyperkinetic – dyskinesis, dystonia
- Tremor, chorea, dystonia, myoclonus, tic

Abnormal and involuntary movements

- **Dystonia**-muscle tone discoordination
- Involuntary contraction of muscles
- Trunk muscles - twisting movement around own axis while walking

- **Dyskinesis-involuntary movements**
  Chorea-fast, free effort uncontrollable jerky movements, especially at UE
- **Athetosis** – slow, tonic, twisting serpentine movements-dis n. caudatus

Types of abnormal movement

- Chorea
- Ballismus
- Athetosis
- Myoclonus
- Dystonia
- Tics
- Tremor

Huntington disease

- Prevalence, genetics
- 4-8/100 000
- HD gene - short arm of chr. 4 (4p16.3)
- Autosomal dominant transfer
- Huntingtin protein is missing
- Function is unclear
- A role in cytoskeletal anchoring or transport of mitochondria

Huntington disease

- Progresive hereditary disorder

- **Chorea**+ dementia+personality disorder
- Ncl. caudatus + **putamen** most affected
- Decrese of GABA and enkephalin→
  gl. pallidus not inhibited= chorea
- Increase Dopamine
- Other parts of brain also involved-subcortical
- glutamic acid decarboxylase
  N-methyl-d-aspartat reduced

Signs and symptoms

- Appear 30-50 yrs (5-70)
- Chorea+dementia+ personality changes
- Onset is gradual
- Clumsiness, dropping of objects, neglect of duties, hyperkinesis
- Chorea-progressive from fingers to limbs
- Sway of trunk, falls down

Signs and symptoms

- Apatia, dementia
- Irresponsible behavior, agresivity
- Prefrontal syndrome

- Later on dystonia and rigidity
- Vegetative dysfunction
- Insomnia, loss of weight
- Lasting 15 years and more

Laboratory data

- CT, MR: enlarged ventricles (butterfly appearance of the lateral ventricles), or striatal hyperintensity on TW2 MRI
- Atrophy
- Genetic examination

- **Treatment**
- Not known
- Symptomatic: antidpresants,antipsychotics

Syndenham chorea
St.Vitus dance, chorea minor

- Disease of childhood (5-15) 2/100 000
- Autoimunne disorder
- Complication of previous infection of A beta hemolytic streptococcus
- Incidence had fallen dramatically
- Acute/subacute onset, tics, dystonia
- May have behavioural problems, usually benign disease, remits spontaneously, complete recovery
- No specific treatment, PNC, IVIG, plasmaferesis

Senile chorea

- Older than 60
- Vascular encefalopathy
- Putamen, ncl subthalamicus
- Hemichorea, hemibalismus
- No mental problems
- Might be a variant of HD
- No therapeutic measures are neded
- Remision

Chorea gravidarum

- Chorea of any cause that begins in pregnancy
- May represent recurrence of Sydenham's chorea
- Most commonly associated with anti-phospholipid sy, +/- SLE
- Usually resolves spontaneously

Myoclonus

- Myoclonus
  “sudden, brief, shock-like involuntary movements”
- Positive myoclonus
- May be caused by active muscle contraction

- Negative
- May be caused by inhibition of ongoing muscle activity
  eg. Asterixis

Myoclonus

- **Focal, segmental, generalised**
- Generalised - widespread throughout body
- Focal / segmental – restricted to particular part of body
- Rare isolated events, many in each minute

- Usually stimulus-sensitive (noise, movement, visual threat, touch, light

Myoclonus

- Can arise from lesions anywhere in the CNS
- Cortical – sensomotor cortex
- Subcortical-brainstem, thalamus
- Segmental (e.g.oculo-palato-pharyngeal) or generalised (reticular myoclonus)
- Spinal
- Oculo-palato-pharyngeal myoclonus:
  lesion in Guillain-Mollaret triangle
- which arises due to any lesion that interrupts pathway between n.dentatus,ruber, inferior, oliva

Etiology

- **Physiologic** - nocturnal (usually on falling sleep jerk, hiccups)
- **Essential-** Occurs in the absence of other abnormality Benign and sometimes inherited familial, sporadic- is nonprogressive
- **Epileptic** - demonstrable cortical source

- **Symptomatic** widespread encephalopathies- viral, degenerative,metabolic, toxic, posthypoxic
- Opsoclonus-myoclonus (encephalopathy in infants, postviral sy, paraneoplastic sy)

Etiology

- Secondary to disease process
- Neurodegenerative eg. Wilson's disease
- Infectious - CJD, viral encephalitis
- Toxic - penicillin, antidepressants
- Metabolic - anoxic brain damage, hypoglycemia,
- hepatic failure " asterixis",renal failure hyponatremia…

- **TREATMENT**
- Anticonvulsans- clonazepam,valproic acid
- 5-hydroxytryptophan

Tics

- Recurrent, sterotyped abnormal movements
- Simple brief jerks to a complex pattern of rapid, coordinated, involuntary movements
- May be shortly suppressed voluntarily or with distraction- then even worse
- Voluntary suppression leads to anxiety and a build-up of internal unrest
- Worsen under stress
- Vocal tics are typical

Gilles de la Tourette syndrome

- Onset
- 2-15 years
- Begin in the face and neck
- Spread to limbs
- Explosive sounds (barking, throat clearing, foul utterances- coprolalia)
- TS - plus
  Attention Deficit Hyperactivity Disorder - ADHD
  Obsedative compulsion disorder

M. le Dr GILLES DE LA TOURETTE,
Médecin des Hôpitaux de Paris, directeur en chef
du service médical de l'Exposition de 1900.
Cliché E. Pirou.

Prevalence

- Estimated 0,05-3% general population
- Many very mild cases (Mozart)
- Many resolve spontaneously by adult life
- No specific morphologic changes in the brain on necropsy
- No mental retardation, high inteligence, memory

- **Treatment**
- In mild cases no treatment
- Clonidin, clonazepam, dopamine antagonists and depleters- more effective- more adverse events

Dystonia

- After parkinsonism most commonly
- Dystonia -condition in which the patient assumes a sustained, abnormal posture or limb position
- Due to co-contraction of agonist and antagonist muscles in the part of body

Dystonia

- **Symptomatic** (90%)
- **Idiopathic**: torsion dystonia

- focal→segmental→generalized
- In advanced disease- contractions become constant

- **Prevalence** 30/100 000
- Focal 50%
- Segmental dystonia 30%

Idiopatic torsion dystonia

- Begins between ages 5-15
- In legs and arms on walking
- Bizzare stepping or bowing gait
- Later spasms in neck and face, difficulty in speech
- Progression extremely variable
- Mental activity remains normal

Adult onset dystonia

- Writer´s cramp: Dystonic posturing of arm when hand used to perform specific tasks e.g. writing, playing piano
- Torticollis: Tendency of neck to twist to one side
- Blepharospam: involuntary forceful closure of eyes
- Oromandibular dystonia
- Lingual dystonia
- Spastic dysphonia

Pathology of dystonia

- Not known, no morphologic changes
- Problaby biochemical abnormalities in BG,genetically determied (noradrenalin, serotonin)

Blepharospasm

- Contraction of the mm.orbicularis oculi
- Sometimes the closure is forcefull, could be intermittent
- Worse by walking and by bright light
- Cocontration of the lower facial muscles (Meige syndrome)
- Begins after age 50
- Botulotoxin injection more than 80% effect

Writer´s cramp

- Limited to one limb (usually dominant)
- Pt should learn to write with nondominant hand
- Botulotoxin is efficient

Symptomatic dystonia

- Wilson disease
- Encephalitis lethargica
- Hallervorden-Spatz disease
- Traumatic hemidystonia (or infarction)
- Tardive dystonia
- Perinatal trauma
- Brain tumours