Extrapyramidal system and cerebellum

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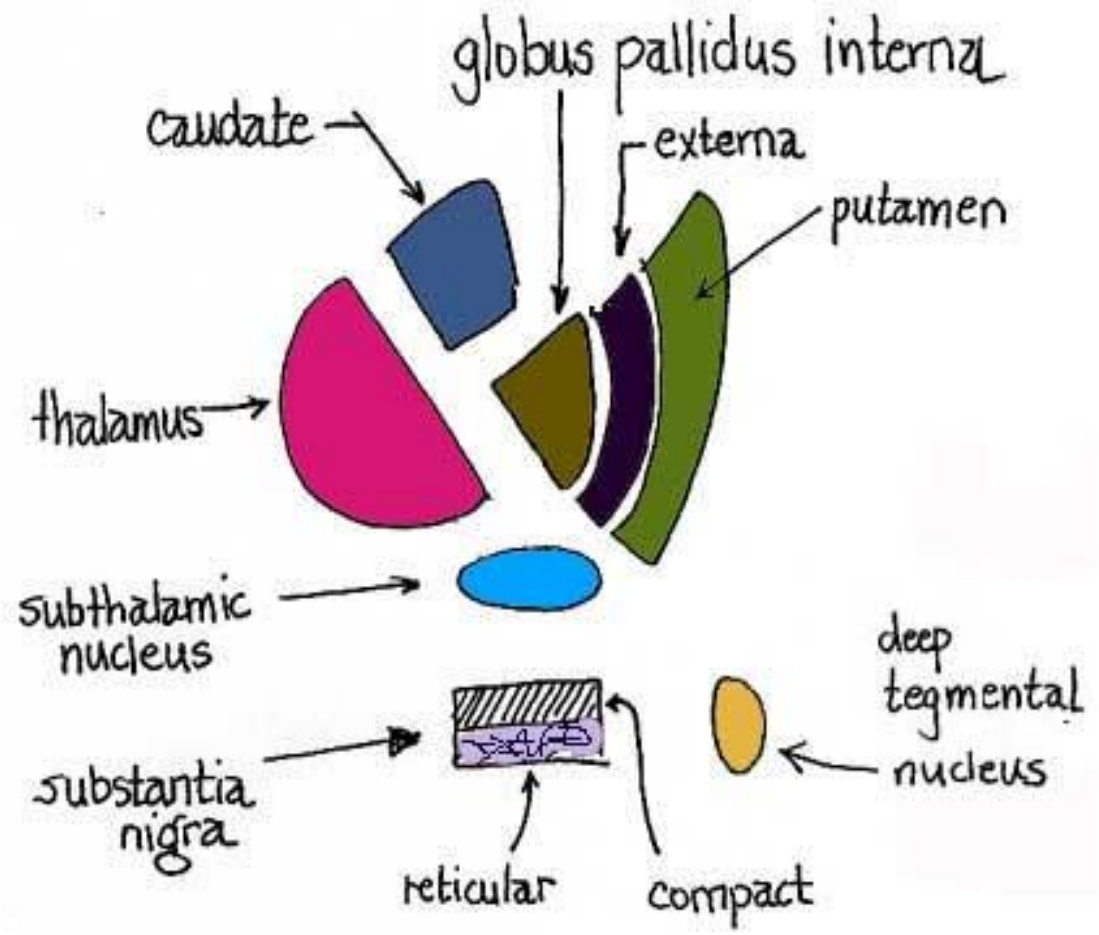
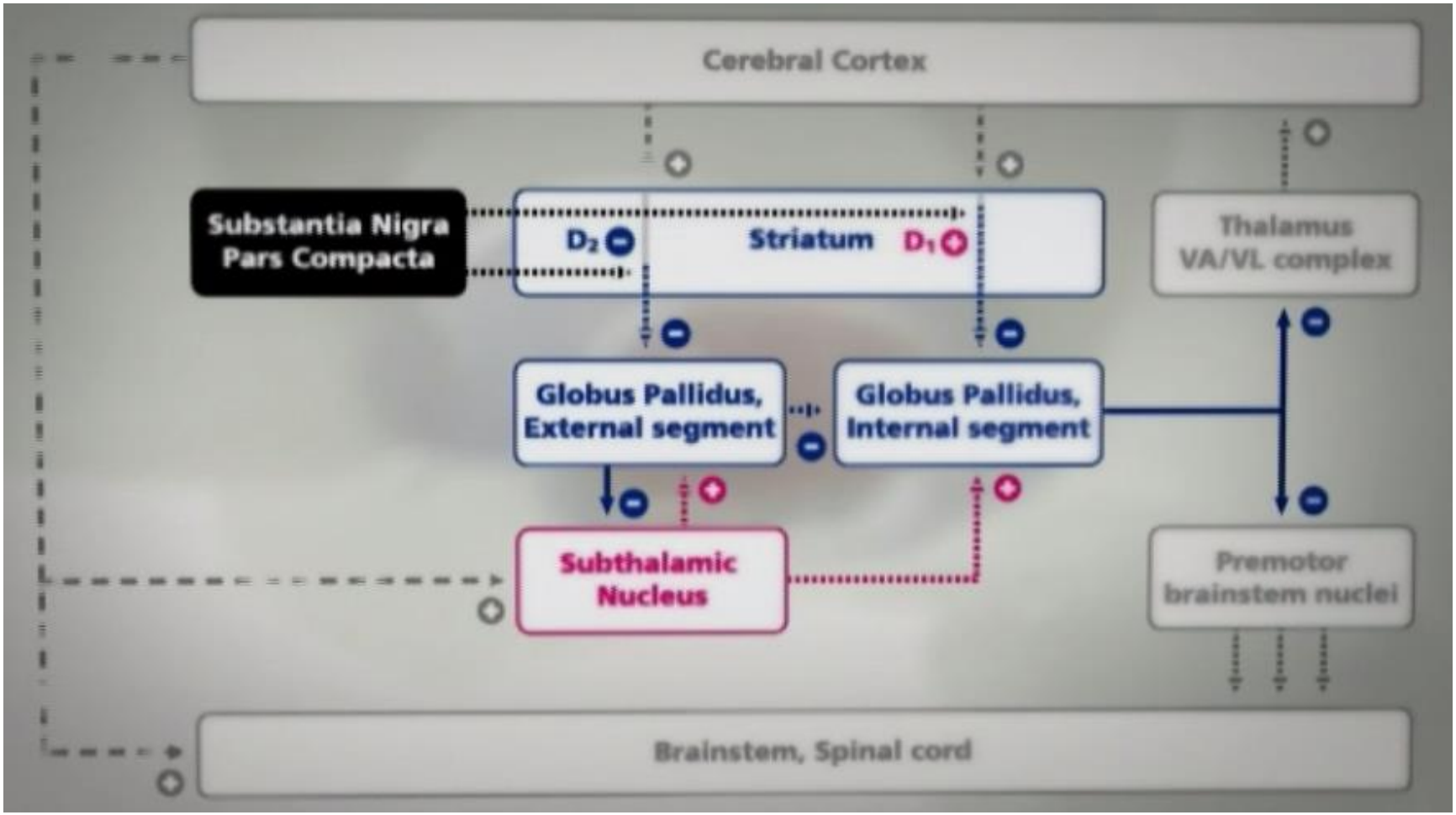
Extrapyramidal system - anatomy



- Ncl. caudatus
- Ncl. lentiformis (putamen + gl. pallidus)
- Corpus striatum - ncl. caudatus a putamen
- Ncl. Accumbens
- Ncl. Basalis Meynerti
- Ncl. Subthalamicus
- Substantia nigra – pars compacta / pars reticularis
- Ncl. ruber
- Pedunculopontine nucleus

Extrapyramidal syndromes

- Hypokinetic syndrome
- Hyperkinetic syndrome


Cerebral Cortex
Substantia Nigra
Pars Compacta
D2
Striatum
D1
Thalamus
VA/VL complex
Globus Pallidus,
External segment
Globus Pallidus,
Internal segment
Subthalamic
Nucleus
Premotor
brainstem nuclei
Brainstem, Spinal cord

Hypokinetic syndrome

- Bradykinesia – pathological slowness of movements
- Hypokinesia – lower amplitude of movements
- Akinesia – problem with movement initiation
- Rigidity – increased muscle toneýšenie svalového tonusu (compared to spasticity resistance is the same during whole range of passive movement – „lead pipe phenomenon / cogwheel phenomenon“)

Parkinsonian syndrome

- Bradykinesia (hypokinesia, akinesia) + at least one of the following:
  - Rigidity
  - Resting tremor

Parkinsonian syndrome - aetiology

- Parkinson‘s disease 80%
- Neurodegenerative disorders 10%
  - Atypical parkinsonism
    - Progressive supranuclear palsy
    - Corticobasal degeneration
    - Multiple system atrophy
    - Dementia with Lewy Bodies
- Secondary 10%
  - Drug-induced, vascular, toxic, posttraumatic, postencephalitic

Hyperkinetic syndrome

- **Tremor**
- **Chorea**
- **Balismus**
- **Dystonia**
- **Myoclonus**
- **Tics**

Tremor

- Rytmical oscillation of a body part around a joint
- Alternating contractions of agonist and antagonist
- Dif.dg. – rytmical myoclonus, dystonic tremor

Classification

- According to body part affected – most frequently arms
- According to position where tremor is observed
  - Resting (parkinsonian)
  - Postural (essential, physiological, drug-induced)
  - Kinetic/intentional (cerebellar)
- According to frequency
- According to amplitude

History

1. Anatomical localization?
2. Task-specific?
3. Age of onset?
4. Disease course? (progressive – PD, static – ET, sudden onset and/or sudden remission – functional?)
5. Factors that improve or worsen tremor? – alcohol, fatigue, anxiety, stress, mental tasks, …
6. Drug history
7. Bradykinesia/rigidity in history
8. Metabolic diseases? – hyperthyroidism,…
9. Family history

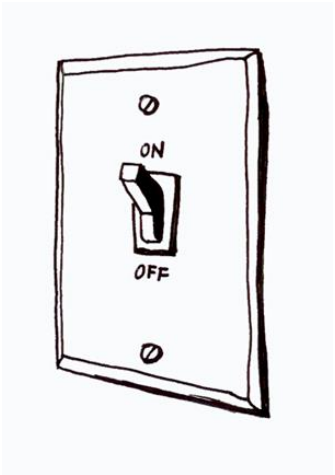
Clinical examination

- Anatomical location – hands, arms, head, chin, legs, trunk
- Symetrical/asymetrical
- Position – resting, postural, kinetic, intentional (finger-nose-finger)
- Frequency (low/high), amplitude (big/small)
- Activation maneuvers
- Other extrapyramidal signs – rigidity, dystonia, …
- Oculomotor abnormalities
- Signs of neuropathy
- Gait abnormalities

Activation maneuvers

- Cognitive task – naming months backwards, counting (increase of supressed organic tremor/ decrease or character change of functional)
- Archimedes spiral
- Drinking a cup of water
- Writing

Patophysiology of tremor



Helmich 2013

Chorea

- Disorder characterised by involuntary brief jerky or twisting movements, which appear at random in the affected body part
- Important is the random nature and unpredictability of the following movement.
- Movements are brief and brisk, but can be also longer and more twisting (choreoathetosis), they are most pronounced at distal body segments

Chorea

- Worsening during speech, movements, emotions
- E.g. „milking maid handshake“, can‘t keep the tounge protruded, dancing gait (st. Vitus dance)
- Distribution
  - Focal
  - Segmental
  - Hemichorea
  - Generalized

Chorea - aetiology

- Neurodegenerative – Huntington‘s disease
- Metabolic, toxic and drug-induced (hepatal encephalopathy, CO poisoning, Mn, tardive dyskinesias after neuroleptics,…)
- Secondary due to BG lessions (trauma, stroke, tumors)
- Autoimunne (assoc. with antibodies against streptococcus – chorea minor assoc. with rheumatic fever, chorea gravidarum)

Ballism

- Variant of chorea with large amplitude involuntary movements of proximal parts of the limbs
- Usually due to lesions of ncl.subthalamicus Luysi or putamen
- Most frequently hemiballism after contralateral lesion of the subthalamic nucleus

Dystonia

- Dystonia is defined as a movement disorder characterized by sustained or intermittent muscle contractions causing abnormal, often repetitive, movements, postures, or both.
- Dystonic movements are typically patterned and twisting, and may be tremulous.
- Dystonia is often initiated or worsened by voluntary action and associated with overflow muscle activation.

Albanese et al. 2013

Dystonia

- Fixed dystonia
- Mobile dystonia
- Dystonic myoclonus
- Dystonic tremor

- Dif.dg. Of dystonie
  - Chorea
  - Myoclonus
  - Tics
  - Stereotypies

Axis I: clinical characteristics

I. Age at onset
   I. Infancy (birth to 2 years)
   II. Childhood (3–12 years)
   III. Adolescence (13–20 years)
   IV. Early adulthood (21–40 years)
   V. Late adulthood (>40 years)
II. Body distribution
   I. Focal
   II. Segmental
   III. Multifocal
   IV. Generalized (with or without leg involvement)
   V. Hemidystonia
III. Temporal pattern
   I. Disease course
      I. Static
      II. Progressive
   II. Variability
      I. Persistent
      II. Action-specific
      III. Diurnal
      IV. Paroxysmal
IV. Associated features
   I. Isolated dystonia or combined with another movement disorder
   II. Occurence of other neurological or systemic manifestations

Axis II: Etiology

I. Nervous system pathology
   I. Evidence of degeneration
   II. Evidence of structural (often static) lesion
   III. No evidence of degeneration or structural lesion
II. Inherited or acquired
   I. Inherited
      I. AD, AR, X-linked, mitochondrial
   II. Acquired
      I. Perinatal brain injury
      II. Infection
      III. Drug-induced
      IV. Toxic
      V. Vascular
      VI. Neoplastic
      VII. Posttraumatic
      VIII. Psychogenic
   III. Idiopathic
      I. Sporadic
      II. Familial

Albanese et al. 2013

Clinical characteristics

- Body distribution
  - Focal – only 1 muscle group or body segment
  - Segmental – 2 neighbouring muscle groups or body segments (e.g. head + neck)
  - Multifocal – 2 non-neighbouring muscle groups or body segments (e.g. right arm + left leg)
  - Hemidystonia
  - Generalized – affection of the trunk + at least 2 other body segments – with or without leg involvement

Age of onset

I. Infancy (<2 years)

II. Childhood (3-12 years)

III. Adolescence (13-20 years)

IV. Early adulthood (21-40 years)

V. Late adulthood (>40 years)

Temporal pattern

I. Disease course
    I. Static
    II. Progressive
II. Variability
    I. Persistent symptoms
    II. Action-specific
    III. Diurnal fluctuations
    IV. Paroxysmal dystonia

Associated features

I. Dystonia isolated or combined with other extrapyramidal symptom
    I. Isolated dystonia
    II. Combined dystonia
II. Other neurological or systemic symptoms

Myoclonus

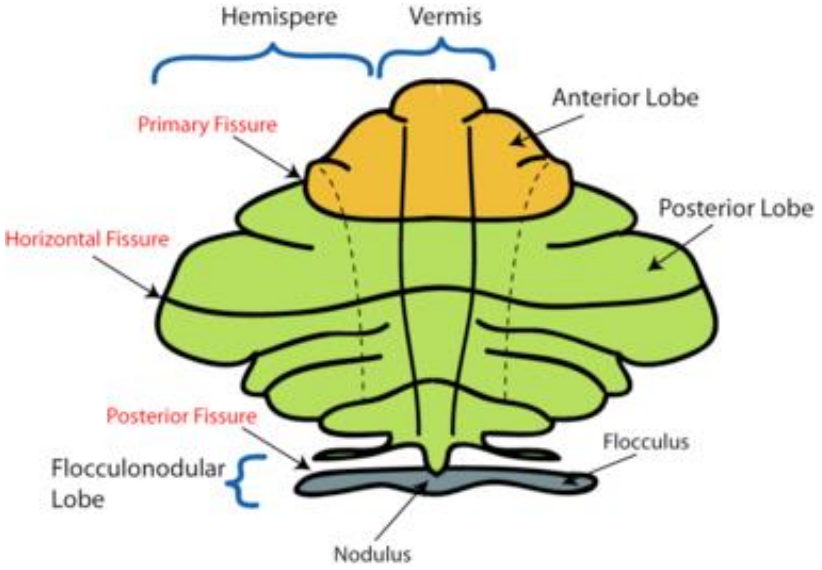
- Very brief involuntary jerky movements caused by intermitent muscle contractions or relaxations
- Regular / irregular
- Positive / negative (asterixis)
- Focal, segmental, multifocal, generalized
- Stimulus sensitive to sound, touch?
- Cortical / subcortical / spinal
- Dif.dg. Tremor, chorea, epilepsy, fasciculations, psychogenic

Tics

- Fast (clonic) or slow (tonic) irregular repetitive stereotyped movements or vocalizations
- Preceded by growing inner tension and urge to perform the tic and followed be relief after performing the tic
- Partially voluntarily supressible
- Simple motor tics
- Complex motor tics
- Simple vocal tics
- Complex motor tics

Cerebellar anatomy

- Archicerebellum
  - Oldest part
  - Flocculonodular lobe
  - Balance, eye movements
- Palleocerebellum
  - Ant. a post. part of vermis
  - Modulates sequential movements
- Neocerebellum
  - Youngest part
  - Hemispheres and central part of vermis
  - Fine motor and speech control



Clinical features of cerebellar dysfunction

- Ataxia (appendicular/axial)
  - Dysmetria (hyper) – disturbance of aiming
  - Dysdiadochokinesis – disturbance of fast alternating movements
  - Dyssynergy – loss of coordination between involved muscle groups
- Rebound
- Failure/disturbance of inhibition

Clinical features of cerebellar dysfunction

- Tremor (kinetic/intentional)
- Titubations and oscillations around the axis on standing
- Hypotonia
- Dysarthria, Ataxic (scanning) speech
- Dysphagia
- Ocular movement abnormalities (nystagmus, slow saccades, saccadic dysmetria, occular flutter, opsoclonus, skew deviation)
- Cognition – slow thinking, problems in motor learning

- Palleocerebellar syndrome
  - Ataxia of stance and gait (archicerebellum)
  - Trunk assynergz (axial muscles)
  - Falls (frequent, usually backwards)
- Neocerebellar syndrome
  - Hypermetria
  - Adiadochokinesis
  - Assynergy of fine movements
  - Intentional tremor
  - Pasivity (lower muscle tone)