CEREBELLAR DISORDERS IN CHILDREN

Smuts
2011
Movement

Cerebellum
- Control of movement patterns
- Motor learning
- Judge speed, force and direction
- Coordinator of information
- Receives information from
  - Muscle spindles
  - Labyrinth, eyes, parietal cortex
  - Joints
  - Pressure receptors

Extra pyramidal system
- Control and execution of movements
- Includes:
  - Basal ganglia
  - Thalamus
  - Subthalamic nuclei
  - Substantia nigra
  - Red nucleus
  - Brainstem reticular formation

Fluent movement
Start and stop of movement
CEREBRAL CORTEX
BRAINSTEM
(descending motor pathways)

DEEP CEREBELLAR
NUCLEI

PURKINJE
CELLS

MOSSY FIBERS
CLIMBING FIBERS

FUNCTION
COMPARES MOTOR OUTPUT OF
CEREBRAL CORTEX WITH
PROPRIOCEPTIVE INPUT FROM
PERIPHERAL NERVOUS SYSTEM

CEREBRAL
CORTEX
(proprioceptive
input)

MUSCLES
TENDONS
JOINTS
(proprioceptive
Input)

VESTIBULAR
NUCLEI
(input concerning
equilibrium)
Functional anatomical organization of the cerebellum

- Vestibulocerebellum
- Spinocerebellum
- Pontocerebellum
Spinocerebellum

To medial descending systems

To lateral descending systems

Cerebrocerebellum

To motor and premotor cortices

Vestibulocerebellum

To vestibular nuclei

Motor execution

Motor planning

Balance and eye movements
Functional anatomical organization of the cerebellum

- **Vestibulocerebellum**
  - Adjustment of axial muscle tone
  - Eye movements
  - Coordination of the head and eyes

- **Spinocerebellum**
  - Execution of movement
  - Regulates muscle tone

- **Pontocerebellum**
  - Precision in control of rapid limb movement
Cerebellar signs

- Non specific symptoms
  - Headache
  - Nausea
  - Vomiting
  - Gait difficulty
  - Vertigo
Cerebellar signs

- Midline cerebellar disease
  - Abnormal gait
  - Abnormal posture of the head
  - Ocular motor dysfunction
    - Nystagmus
    - Ocular dysmetria
Cerebellar signs

- Lateral cerebellar disease
  - Hypotonia
  - Dysarthria
  - Limb ataxia
  - Intention tremor
- Oculomotor disorder
  - Opsoclonus
  - Ocular bobbing
Ataxia - 3 broad categories

- Acute ataxia
- Chronic non-progressive
- Chronic progressive
Acute ataxia

- Sudden onset
- Can’t walk
- Extremely clumsy
- Can’t feed due to tremor
- Dysarthria
- Nystagmus unusual
- Look for signs of infections e.g. chickenpox
- History of possible intoxication
- If signs are symmetrical, no raised ICP, and no focal signs, usually benign
Acute ataxia: Aetiology

- **Infections**
  - Cerebellar abscess
  - Viral cerebellitis
  - Bacterial

- **Metabolic:**
  - Organic acidurias
  - Leigh’s encephalopathies
  - Hypoglycaemia
  - Hyperammonaemia

- **Toxins**
  - Alcohol
  - Phenytoin
  - Phenobarbitone,
  - Lead
  - Glue
  - Vit A

- **Posterior fossa tumour**

- **Vascular**
  - Haemorrhage
  - Embolism
  - AVM

- **Pseudo-ataxia**
Chronic non progressive ataxia

- Ataxic/Hypotonic CP
- Often a congenital malformation of the cerebellum
Aetiology: Chronic non-progressive ataxia

- **Perinatal insults**
  - Birth asphyxia
  - Metabolic
  - Intra ventricular haemorrhage
  - Meningitis

- **Congenital malformations**
  - Primary cerebellar hypoplasia
  - Hydrocephalus

- **Foetal alcohol syndrome**

- **Joubert syndrome**

- **Cerebellar/ kidney associations**

- **Postnatal acquired**
  - Hypoxia
  - Hypoglycaemia
  - Chronic phenytoin
  - Thiamine deficiency
  - Trauma
Chronic progressive ataxia

- Lesion in cerebellum with loss of:
  - Purkinje cells
  - Cerebellar nuclei
  - Afferent or efferent pathways
    - Olivary atrophy
    - Spinocerebellar degeneration
    - Post column demyelination
    - Peripheral nerve lesion
Ataxia telangiectasia

- Progressive ataxia (1-4 years)
- Abnormal eye movements - oculomotor apraxia
- Telangiectasia (3 years - adolescence)
- Cutaneous manifestations
- High risk for malignancies
- Abnormality in cellular and humoral immunity
- Elevated alpha feto protein
Friedreich’s ataxia

- Onset before 20 years
- AR
- Progressive ataxia - gait difficulties, speech problems
- No nystagmus
- Weakness
- Positive Babinski but absent ankle and knee reflexes - involvement of the corticospinal tract
- Loss of position and vibration sense
- Positive Romberg test - involvement of the posterior columns
- Bladder dysfunction
- Involvement of cranial nerves
- Scoliosis
- Cardiomyopathy
- Diabetes mellitus