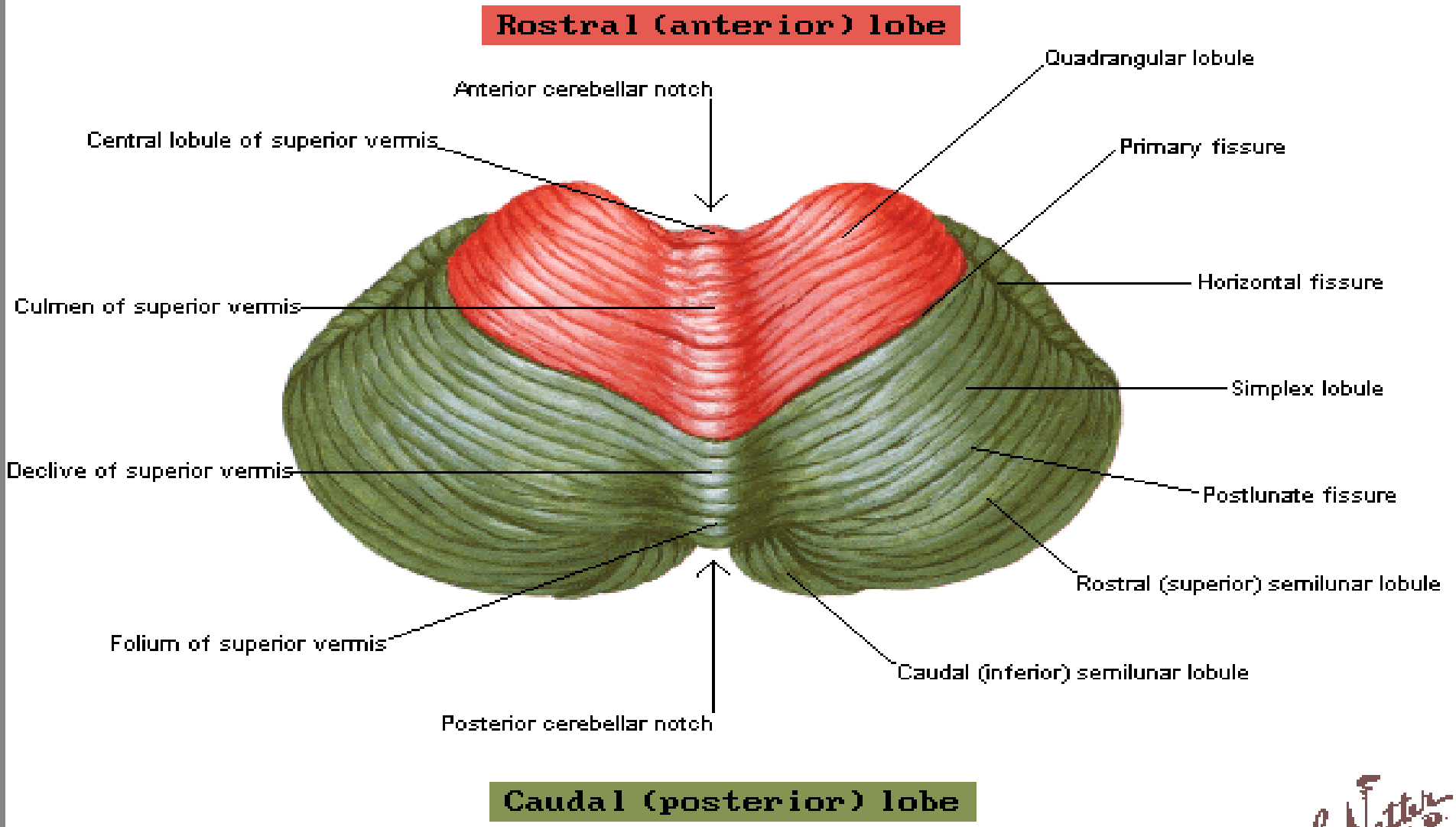
Cerebeller Disorders

Cerebellum

Superior Surface

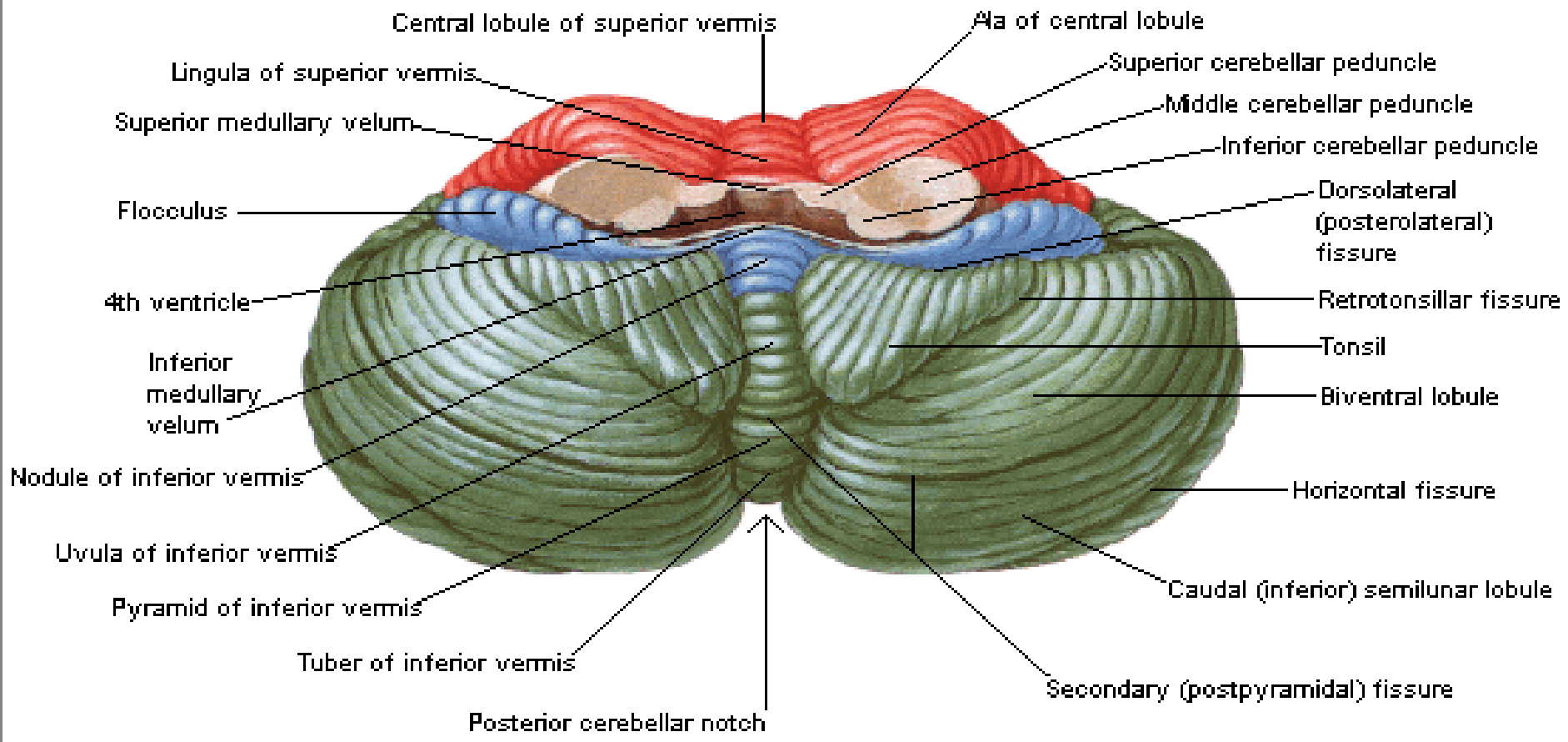


Cerebellum

Inferior Surface

Rostral (anterior) lobe

Flocculonodular lobe



Caudal (posterior) lobe

Anatomical & functional review

- (3) lobes
- Embryo-genetically… 3 phenogenetic regions
- (3) functions
- (3) pairs of nuclei
- (3) peduncles
- (3) arterial blood supply

Clinical feature of cerebeller disorders:

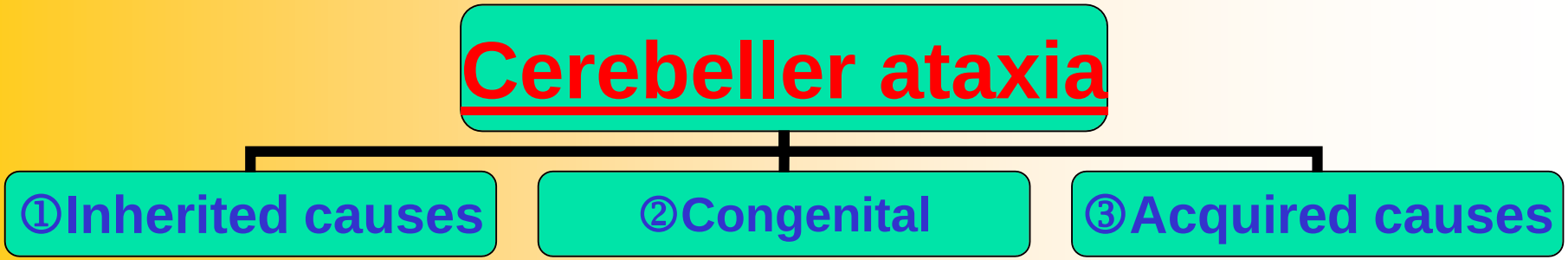
- Head:
- Eye:
- Speech:
- Upper limbs:
- Trunk:
- Lower limbs:
- Gait:
- Hypotonia:

Other unusual features

- Asthenia & fatigability- barognosia
- Macrographia
- Catalepsy
- Cerebeller mutism
- Cerebeller fits

- Remember all by>>> HANDS tremors

Etiology of cerebeller ataxia



I. **With known metabolic defect.**
II. **without known metabolic defect:**
    I. **Early onset ataxia**
    II. **Late onset ataxia**

- **Mostly reversible causes**
- **Frequently.. Acute onset**
- **Often treatable if early recognized**

Acquired causes

- Vascular
- Neoplasm
- Infection
- Inflammatory & autoimmune
- Metabolic
- Drugs & toxins
- Post traumatic
- Developmental

Inherited ataxia with known metabolic defect

- Ataxia with hyperammonaemia
- Ataxia with aminoacidurias
- Ataxia with disorders of pyruvate and lactate metabolism
- Ataxia telangiectasia(with defective DNA repaire)
- Leucodystrophies
- mucolipidosis

Inherited ataxia with unknown Aetiology of early onset.

- All are Aut.R
- Age of onset < 25ys

- **A- Friedreich's ataxia:**

- The Commonest early onset ataxia
- Present > 50% of early onset ataxias

C/P of Friedrich's ataxia

- Aut.R
- Age of onset 8-15 ys.
- C/F reflects sites of pathological changes in
  - Spino-cerebeller tracts & purkinje cells of cerebellum
  - Pyramidal tract
  - Peripheral nerves
  - Posterior column
  - Optic tract

Other associated features

- Nerve deafness in > 10%
- Scoliosis…frequent,pes cavus, equinovarus
- Cardiomyopathy in > 2/3 of cases
- Dementia … mild degree
- DM in > 10% of cases
- Sphincteric dysfunction
- Most patients become unable to walk within 15 ys after the onset.

Other examples of early onset ataxias

Holmes ataxia

- Cerebeller ataxia plus..
- MR
- Dementia
- Hypogonadotrophic hypogonadism
- Choreoathetosis
- Retinopathy
- deafness

Other examples of early onset ataxias

Ramsy hunt syndrome

- **Cerebeller ataxia**
- **Myoclonus**
- **Tonic clonic seizures**

Behr's ataxia

- **Ataxia**
- **Optic atrophy**
- **Spasticity**
- **MR**

Late onset inherited ataxia of unknown etiology

- (familial spino-cerebeller ataxia)
- All are Aut.D
- Age of onset usually > 25ys

A- type I

- Including Machado-josef disease
- Age of onset commonly 30-40 ys.& up to 60ys.
- Gait ataxia…the frequent presentation
- Optic atrophy
- Distal wasting
- Ophthalmoplegia
- Dementia
- Extrapyramidal features
- Speech disorders of cerebeller and pseudobulbar features

B-type II

- Age of onset 20-35 ys.
- Cerebeller ataxia
- Pigmentary retinopathy
- Visual failure
- Dementia
- Supranuclear ophthalmoplegia
- Extensor planter response

C- Type III

- Pure cerebeller syndrome
- Age > 50ys.

D- idiopathic late onset ataxia ‘Marie’s ataxia’

- Mean age for onset 55 ys.
- Proressive gait ataxia…the prominent feature
- Relative preserved coordination of UL.
- Mild dysartheria
- Dementia
- Idiopathic

Investigations of a case with cerebellere ataxia

- Neuroanatomical imaging
  - CT-brain and MRI
- Neurophysiological studies:
  - -EEG
  - -VEP
  - -BAEP
  - -SSEP
  - EMG & NCS

Investigations..continuo

- Neurophysiological imaging
  - PET & SPECT
- Metabolic screen

  to search for the aetiology of acquired ataxia

Management

- In aquired ataxia
- In inherited ataxias
  - No definite measures
  - Physiotherapy
  - Management of associated medical disorders
  - Supportive measures and symptomatic
  - Recently introduced benificial medications
    - INH - TRH- Physostigmine- propranolol- vit. E- Amantadine- clonazepam