# CEREBELLAR DISORDERS

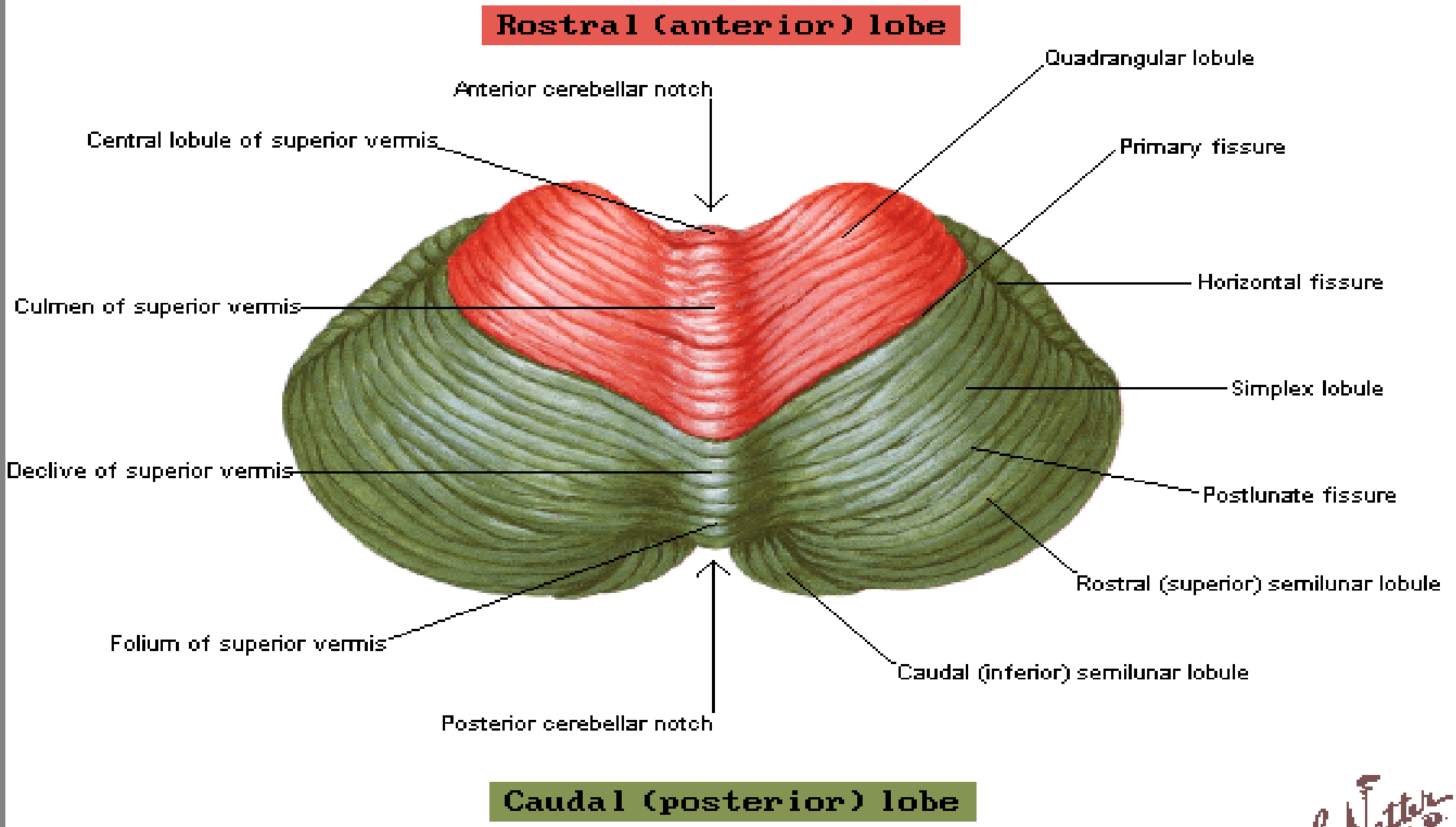
**By Dr./ Alaa Eldin Sedky**
**Lecturer of Neurology**

# )The Cerebellum (gross features

- The cerebellum, or “small brain” is located in the posterior fossa of the skull, separated from the occipital lobes by a dural fold, the tentorium cerebelli.
- It overlies the dorsal surfaces of the pons and medulla oblongata and contributes to the formation of the roof of the fourth ventricle.
- It consists of a midline vermis and two laterally placed hemispheres.
- The cerebellum is divided anatomically by two transverse fissures (anterior and posterolateral) into three lobes: anterior, posterior, and flocculonodular.

# Cerebellum

## Superior Surface

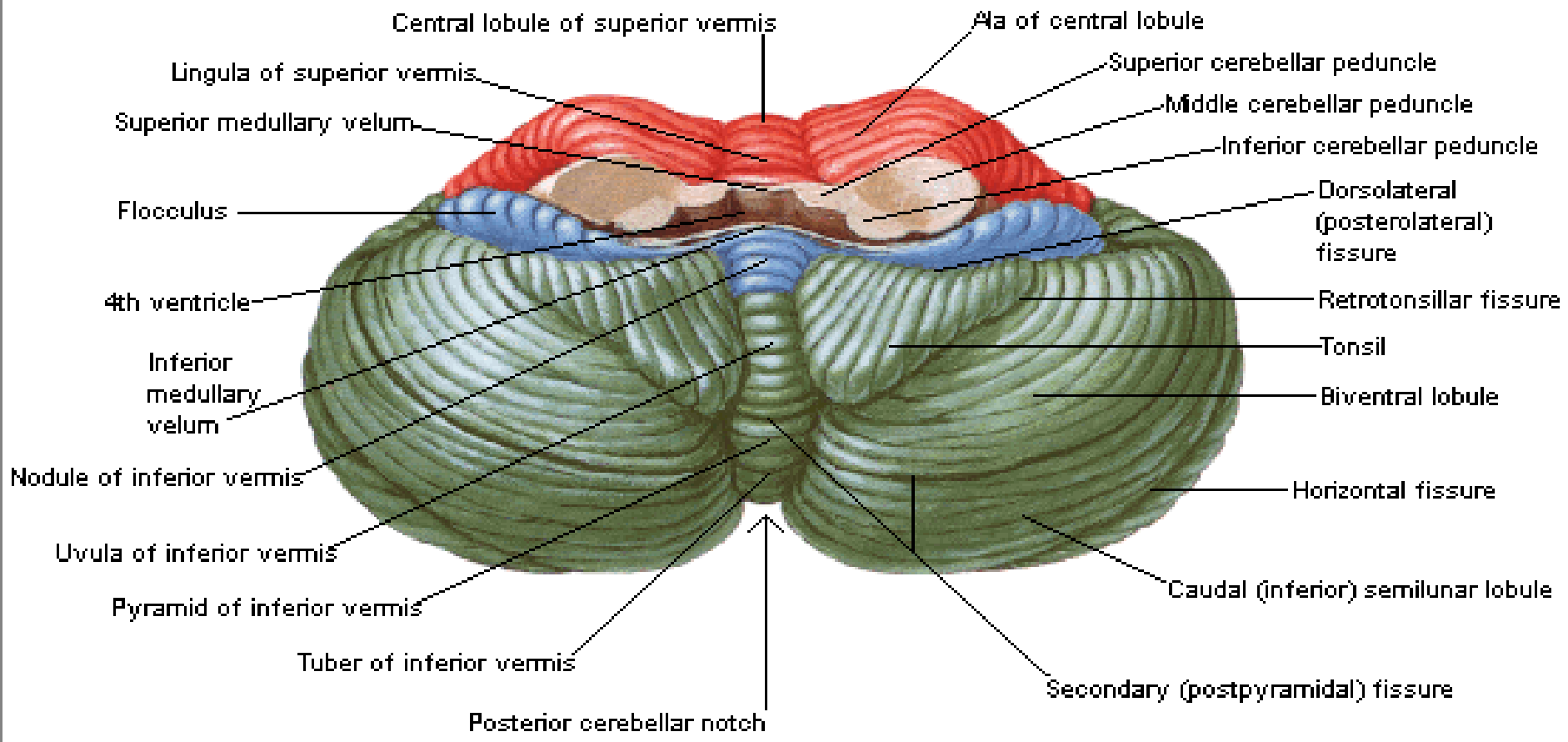


# Cerebellum

## Inferior Surface

**Rostral (anterior) lobe**

**Flocculonodular lobe**




**Caudal (posterior) lobe**

# )The Cerebellum (connections

**The cerebellum is connected to the midbrain, pons, and medulla oblongata by three pairs of peduncles.**

1. **The superior cerebellar peduncle connects the cerebellum with the midbrain.**
2. **The middle cerebellar peduncle connects the pons with the cerebellum.**
3. **The inferior cerebellar peduncle connects the medulla with the cerebellum.**

# The Cerebellum

## Three functional subdivisions (based on fiber connectivity):

1. **The vestibulocerebellum (flocculonodular lobe)** has reciprocal connections with vestibular nuclei & plays a role in control of body equilibrium and eye movement.
2. **The spinocerebellum (the anterior lobe)** has reciprocal connections with the spinal cord and plays a role in control of muscle tone.
3. **The cerebrocerebellum or pontocerebellum (the posterior lobe)** has reciprocal connections with the cerebral cortex and plays a role in planning and initiation of movements, as well as the regulation of discrete limb movements.

# :Clinical feature of cerebeller disorders

**Head:**

- Nodding, abnormal postures (tilting)

**Eye:**

- Nystagmus , ocular dysmetria.

**Speech:**

- Dysartheria in the form of staccato, scanning, explosive, speech

**Upper limbs:**

- Intentional kinetic tremors (tremors on reaching the target).
- Decomposition of movements.
- Dysmetria, dysdiadocokinesia ,jerkiness of movement.
- Hypotonia, rebound phenomenon.

# Clinical feature of cerebeller disorders:

## Trunk:

- Titubation , trunkal instability

## Gait:

- Ataxic gait: ( wide base, drunken) ,
- Deviated to one side (in unilateral cerebellar lesions)

## Hypotonia:

- Generalized , more in U.L.

# Clinical Classification Of Ataxia

I. Congenital ataxia
II. Acute/subacute onset ataxia
III. Slowly progressive ataxia
IV. Intermittent ataxia

# I. Congenital ataxia

1. Ataxic cerebral palsy
2. Hereditary congenital ataxias

# II. Acute/ subacute onset ataxia

1. Infarction / haemorrhage
2. Demyelination (MS - ADEM)
3. Post-infectious cerebellar ataxia
4. Paraneoplastic
5. Toxins
6. Abscess/tumour

# III. Slowly progressive ataxia

1. Early onset hereditary degenerative ataxia (<25 yrs)
2. Late onset hereditary degenerative ataxia (>25 yrs)
3. Sporadic idiopathic cerebellar degeneration

# III. Slowly progressive ataxia (cont.)

4. Tumour
5. Foramen magnum compression
6. Alcoholic cerebellar ataxia
7. Drugs, e.g. phenytoin
8. Prion disease
9. Metabolic ataxias
10. Vitamin E deficiency

# IV. Intermittent ataxia

1. Drugs / toxins
2. Multiple sclerosis
3. Transient ischaemic attacks
4. Foramen magnum compression
5. Metabolic ataxias
6. Periodic ataxias (hereditary)

# Friedrich's Ataxia

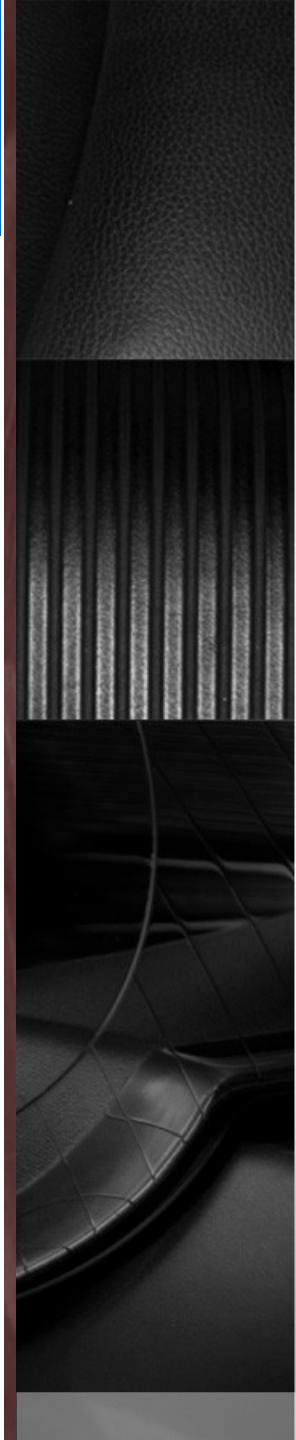
# Pathogenesis

## Degeneration of :

- Dorsal column.
- Spinocerebellar tracts.
- Pyramidal tracts.

## Loss of:

- Dorsal root ganglion cells.
- Large myelinated fibres in peripheral nerves.



# Clinical picture

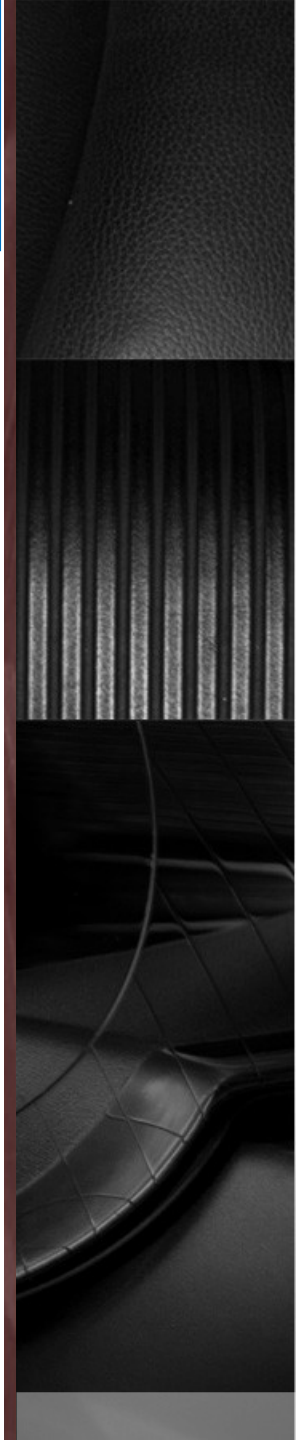
- Age <25ys old (8-26).
- All cases have :

A. <5ys of onset:

✓ Progressive ataxia (Gait-Limb-Speech → late)

✓ Lower limb areflexia

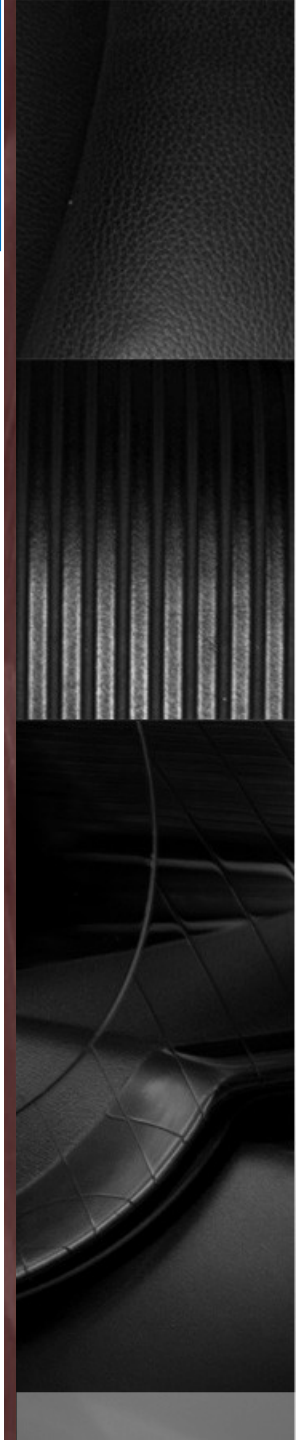
✓ Extensor planter response



# Clinical picture

B. **> 5ys of onset**

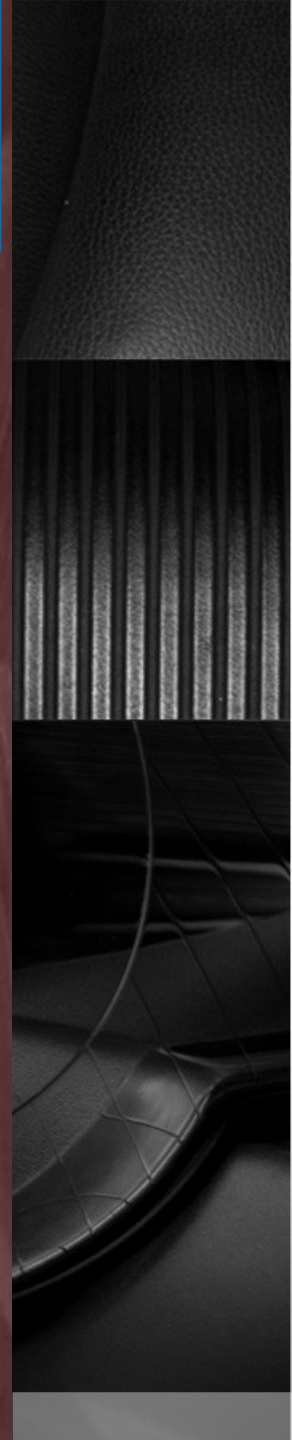
- ✓ **Leg weakness.**
- ✓ **Lost deep sensation in lower limbs.**
- ✓ **Generalized areflexia.**



# Clinical picture

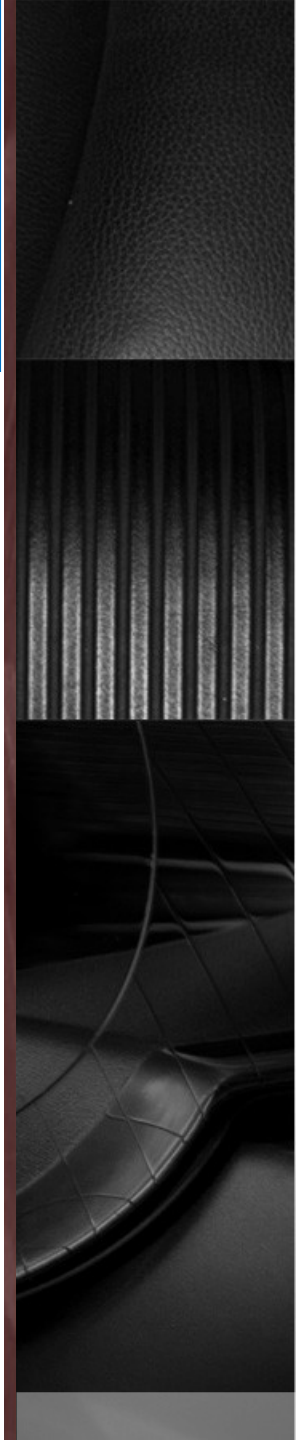
**Variable features:**

- **Scoliosis in 80% (severe in 10%)**
- **Cardiac in 60% (hypertrophic or dilated cardiomyopathy, T-wave abnormalities on ECG).**
- **Optic atrophy in 25% (severe in 5%)**
- **Nystagmus in 20%**
- **Deafness in 10%**
- **DM in 10%**
- **Pes cavus**
- **Mild cognitive impairment.**



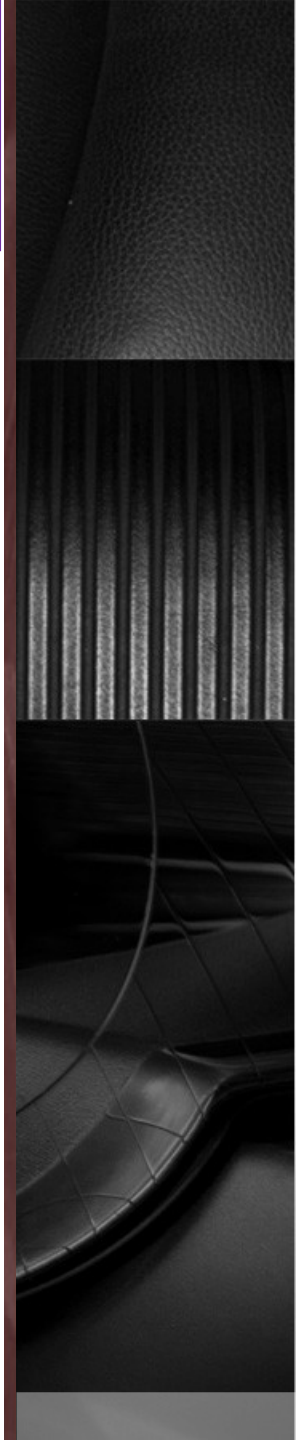
# Features against the diagnosis of FA

- Congenital onset.
- Marked dementia.
- Ophthalmoplegia.
- Parkinsonism or dystonia.
- Marked cerebellar atrophy at early stage.



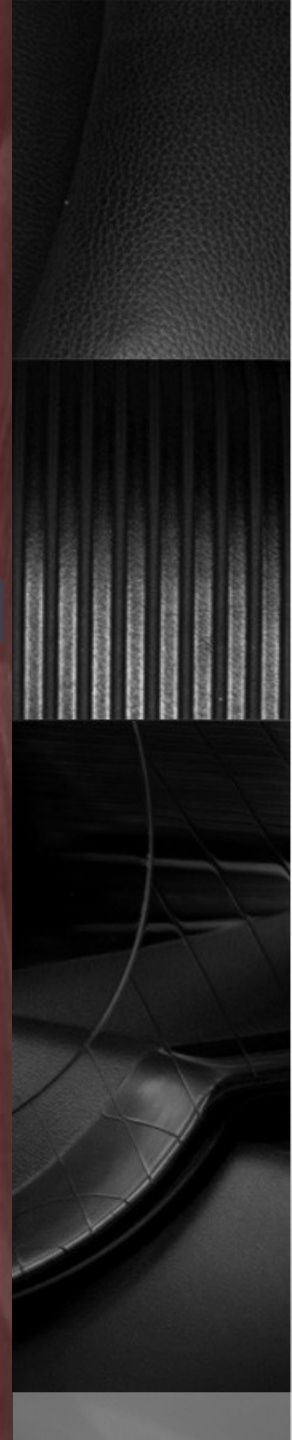
# Investigations

1. **Neuroimaging:**
   - **Early → normal.**
   - **Late → mild atrophy of vermis, medulla and cervical cord.**
2. **NCS.**
3. **VEP.**
4. **Genetic.**



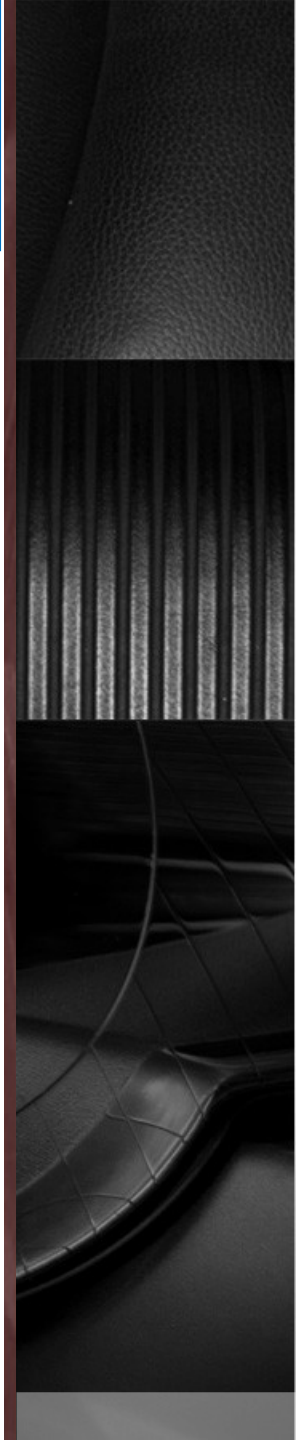
# Late onset hereditary cerebellar ataxia

- AD.
- Age of onset >25ys old.
- Slowly progressive ataxia of gait and limbs.
- Dysarthria.
- Nystagmus.



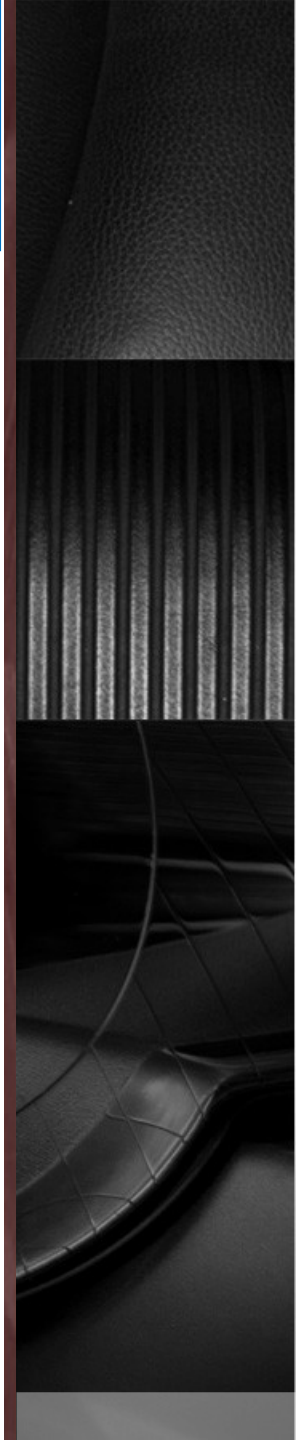
# Late onset hereditary cerebellar ataxia

- **Type □:**
  - **Optic atrophy**
  - **Ophthalmoplegia**
  - **Pyramidal and extrapyramidal features.**
  - **And or distal wasting (amyotrophy)**



# Late onset hereditary cerebellar ataxia

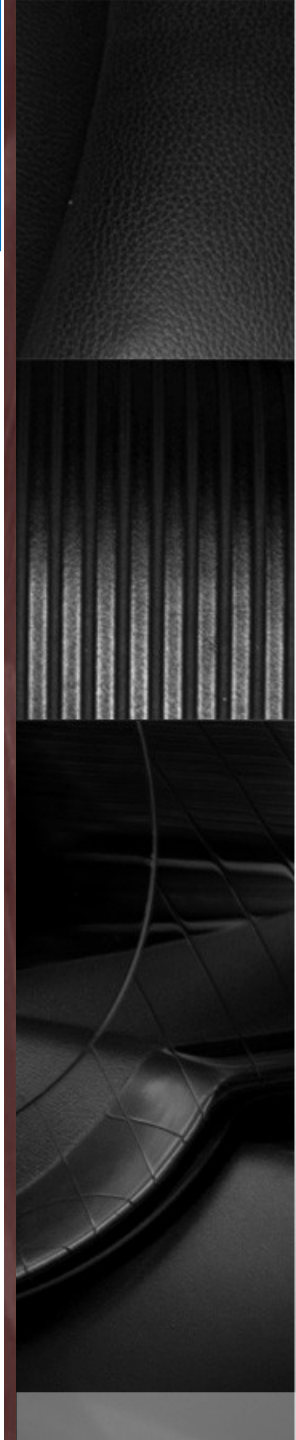
- **Type □□ :**
  - **Pigmentary retinal degeneration**
  - **Ophthalmoplegia**
  - **Pyramidal features**
  - **Dementia**



# Late onset hereditary cerebellar ataxia

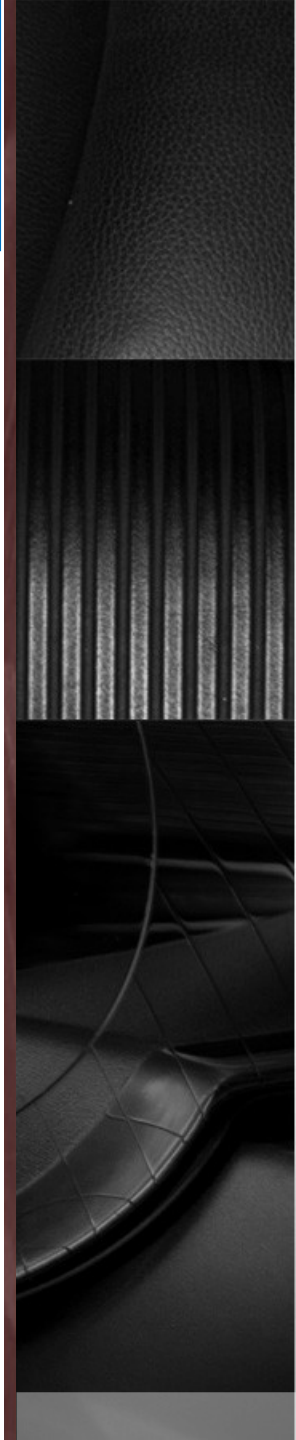
- **Type □□□ :**

**Pure cerebellar ataxia**



# Late onset idiopathic cerebellar ataxia

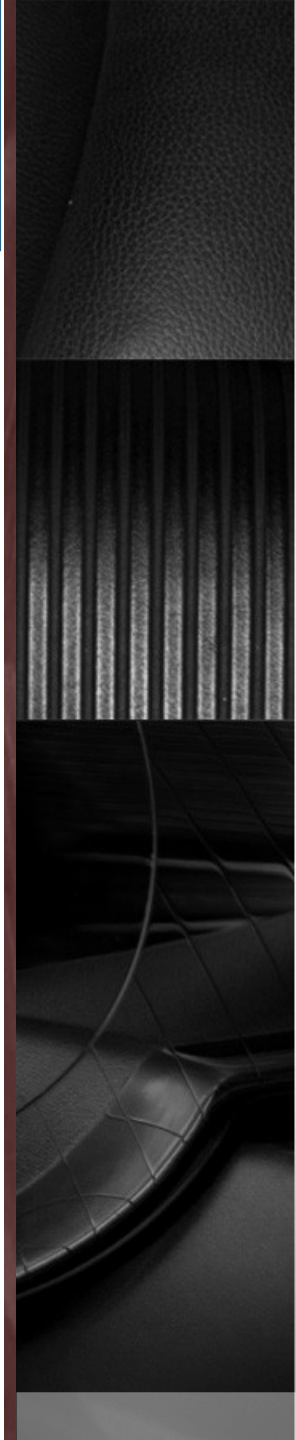
- More common than hereditary.
- Mean age of onset about 55 ys old.
- No family history.
- No optic atrophy or retinopathy.
- Ophthalmoplegia less common.



# Management of cerebellar ataxia

A. Acquired ataxia
B. Hereditary and idiopathic ataxia:

- Physiotherapy
- Symptomatic treatment e.g DM, HTN, cardiac
- Surgical




THANK YOU
اللهم
اجعلنا من الذين إذا
أحسنوا استبشروا ،
وإذا أساءوا استغفروا