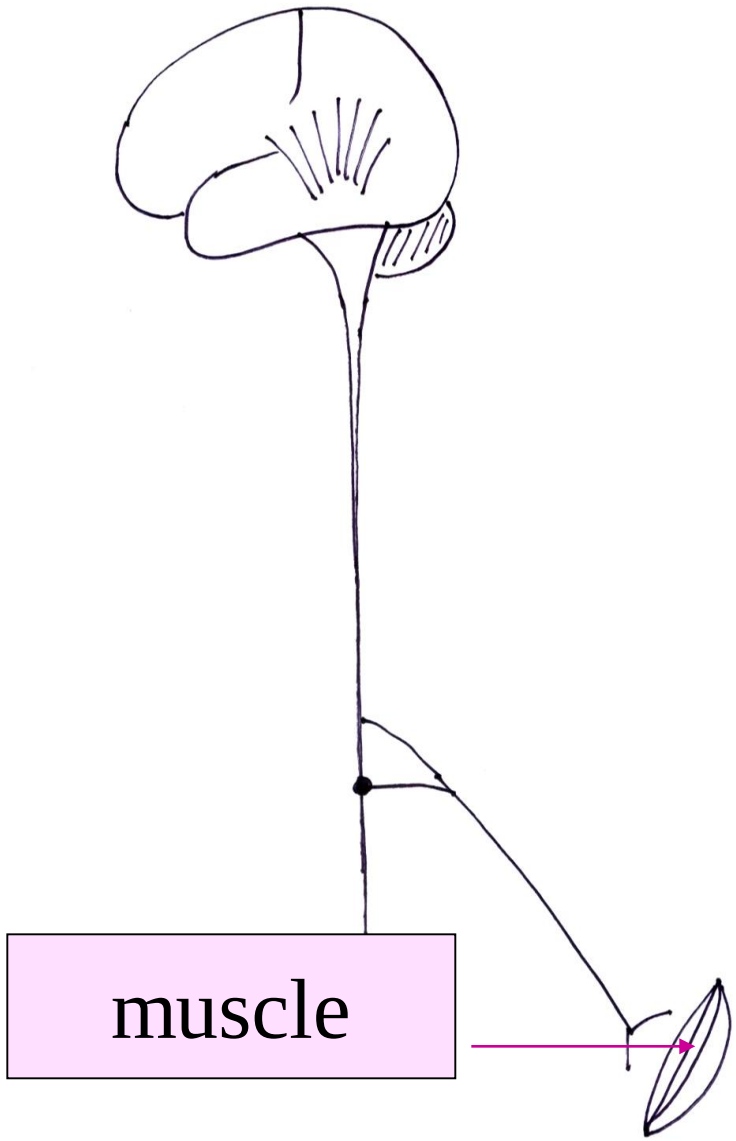
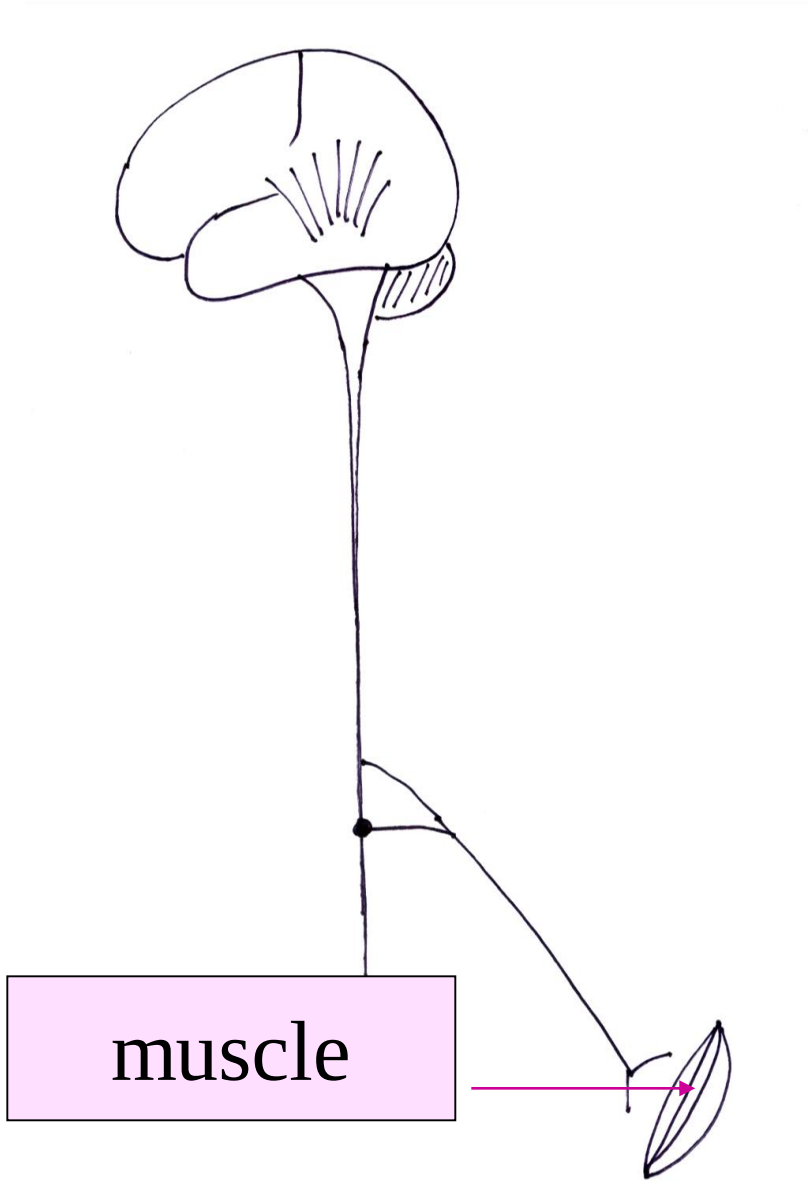


Disorders of muscle



- motor weakness
- wasting
- proximal
- bilateral, symmetrical
- tendon reflexes normal/↓

- NO sensory loss

Disorders of muscle (“Myopathy”)

- Acquired
- Inherited - Genetically determined

Acquired myopathies

Acquired myopathies

- inflammatory - ‘myositis’
  - pain, tenderness, swelling, warmth
  - ↑ ESR, CRP
- metabolic/ endocrine

Acquired myopathies

- inflammatory - ‘myositis’
  - infections - viral, bacterial, parasitic
  - autoimmune - polymyositis/ dermatomyositis
- metabolic/ endocrine
  - hypokalaemia, hypocalcaemia
  - Cushing’s, thyroid dis, …..
  - alcohol
  - drugs - steroids, …..

- myositis due to infection
  - viral
  - staphylococcal, streptococcal
  - gas gangrene
  - TB
  - parasitic - trichinosis, cysticercosis, hydatid disease, toxoplasmosis

Polymyositis

- autoimmune inflammatory myopathy
  - proximal weakness with wasting
  - muscle pain & tenderness - less prominent
  - bulbar, respiratory involvement
  - arthralgia, Raynaud's, …
- dermatomyositis - PM+ skin changes
- PM/DM associated with
  - other connective tissue diseases
  - internal malignancy

Inherited myopathies

Inherited myopathies

- muscular dystrophies
  - Duchenne, limb girdle, facio-scapulo-humeral
- myotonias
  - dystrophia myotonica, myotonia congenita
- periodic paralysis - ‘channelopathies’
  - hypo/ hyper/ normo - kalaemic
- mitochondrial myopathies
- inherited metabolic myopathies

Muscular dystrophies

Duchenne muscular dystrophy

- X- linked recessive - males affected
- loss of muscle protein ‘dystrophin’
- onset - 1st few years, disabled by 10 yrs
- proximal weakness, Gower’s sign
- ‘pseudo-hypertrophy’ of calf muscles
- cardiac muscle involvement
- death due to aspiration pneumonia

Dystrophia myotonica

- myotonia
- ‘distal’ weakness
- frontal balding, ptosis, wasting- face, sternomastoids
- cardiac muscle involvement
- endocrine - diabetes, hypogonadism

- AD, trinucleotide repeat expansion
- onset - 20-40 yrs

Periodic paralysis

- ion channel disorders - ‘channelopathies’

- onset - childhood, young adults
- recurrent episodes of generalised weakness
- precipitated by - heavy CHO meal, exercise

- hypokalaemic -
- hyperkalaemic -
- normokalaemic -

Investigation of muscle disease

Investigation of muscle disease

- muscle enzymes - CPK, aldolase
  - ↑↑ in PM/DM, Duchenne
- EMG
  - myopathy, myotonia
- muscle biopsy
- genetic testing
- other - autoimmune, inflammatory, infection, …

Treatment

- specific treatment
  - PM/DM - steroids, immunosuppressants
  - myotonia - phenytoin
- physiotherapy
- genetic counselling

Remember

- ‘proximal myopathy’
- Acquired/ Inherited
  - inflammatory, metabolic
  - Duchenne
  - dystrophia myotonica
  - periodic paralysis
- Ix - CPK, EMG, biopsy