Myopathy patterns

- **Proximal (limb-girdle)**
  a. Onset at birth
    i. Congenital dystrophy (LGMD-Limb girdle muscular dystrophy)
    ii. Congenital myopathy (all types): central core, myotubular, nemaline, CFTD (fiber disproportion), myofibrillar
    iii. Glycogen storage ds
    iv. Lipid storage ds
  b. Childhood or adult onset
    . Muscular dystrophy (dystrophinopathy, FSH-fascioscapulohumeral, ED-Emery-Dreiffus, Bethlem, LGMD, PROMM-proximal myotonic myopathy)
    i. Congenital myopathy (all types)
    ii. Inflammatory
    iii. Toxic
    iv. Metabolic
    v. Endocrine
- **Distal**
  . Late adult 1 (Welander)
  a. Late adult 2 (Markesbury/Udd)
  b. Early adult 1 (Nonaka)-IBM2, distal myopathy with rimmed vacuoles (DMRV)-GNE mutations
  c. Early adult 2 (Miyoshi)
  d. Early adult 3 (Laing)
  e. Myofibrillar (Desmin)
  f. Myotonic dystrophy
  g. FSH, ED and other scapuloperoneal patterns
  h. OPMD-oculopharyngial dystrophy
  i. IBM-Inclusion body myopathy
  j. Debrancher, AMD-acid maltase dystrophy
  k. Nemaline, Central core, Centronuclear
- **Proximal arm/distal leg (scapuloperoneal)**
  . FSH
  a. ED
  b. Many LGMD
  c. Some congenital Myopathies
  d. AMD
- **Proximal leg/distal arm (IBM pattern)**
  - IBM
  - Some LGMD
  - Late adult distal MD1 (Welander)
- **Ptosis**
  . Myotonic dystrophy
  a. Centronuclear

b. Nemaline
c. Central core
d. Myofibrillar
e. **With weakness of extraocular muscles**- OPMD, mitochondrial

- **Head drop**
  . Isolated neck Extensor Myopathy
  a. PM/DM- polymyositis,dermatomyositis
  b. IBM
  c. Carnitine deficiency
  d. FSH
  e. DM
  f. Congenital myopathy
  g. Hyperparathyroidism
  h. CMS/MG- congenital myasthenic syndrome/myasthenia gravis
  i. ALS

EMG FINDINGS IN MYOPATHIES

Myopathies with fibrillations
Congenital: centronuclear, nemaline, myofibrillar (desmin), some CFTD
Dystrophy: dystrophinopathies, FSH, some LGMD, most distal dystrophies, myotonic
Inflammatory: all
Toxic: acute alcoholic myopathy, lipid lowering drugs
Metabolic: acid maltase, other GSD especially after attack, HKPP, Paramyotonia, K-sensitive myotonia, critical illness

Myopathies with myotonic discharges
Myotonic dystrophy
Paramyotonia congenita
Myotonia congenita
Hyperkalemic periodic paralysis
Potassium sensitive myotonia
Centronuclear myopathy
Myofibrillar (desmin)
Inflammatory myopathy
Hypothyroid myopathy
Lipid-lowering drugs (CLAM)
Acid maltase deficiency
Amyloid

Myopathies with mixed MUP
Chronic dystrophies
Chronic inflammatory neuropathies
Myopathies associated with neuropathy or MND
Infantile acid maltase
Debrancher or brancher deficiency

Mitochondrial myopathy
Connective tissue diseases
Amyloid

Myopathies with distal weakness
Centronuclear myopathy
Nemaline myopathy
Central core myopathy
Myofibrillar (desmin)
Myotonic dystrophy
Distal dystrophy
IBM
Polymyositis
Amyloid