RAMA UNIVERSITY

www.ramauniversity.ac.in

FACULTY OF NURSING

RAMA UNIVERSITY

MUSCULAR DYSTROPHY

Mr. M.Raghavendran M.Sc(N)
Professor
MSN Dept

MUSCULAR DYSTROPHY

- It is a group of genetically transmitted disease characterized by progressive symmetric wasting of skeletal muscle without evidence of neurologic involvement

Types

- Duchanne (Pseudo Hypertrophic)
- Becker (Benign Pseudo Hypertrophic)
- Lan Douzy – Dejerine
- Erb

Duchanne (Pseudo Hypertrophic)

- *Genetic basis*: X Linked
- *Clinical Manifestations*:

  Onset before age 5

  Progressive weakness of pelvic and shoulder muscles

  Unable to walk after age 12

  Cardiomyopathy

  Respiratory failure in second or third decade

  Mental impairment

Becker (Benign Pseudo Hypertrophic)

- *Genetic basis*: X Linked mutation dystrophin gene
- *Clinical Manifestations*
  - Onset between 5 & 15 yrs
  - Slower course of pelvic and shoulder wasting than duchanne
  - Cardiomyopathy
  - Respiratory failure in fourth or fifth decade

Lan Douzy – Dejerine

- *Genetic basis*: Autosomal Dominant Deletion of 4q5 Chromosome
- *Clinical Manifestations*
  - Onset before age 20
  - Slowly progressive weakness of face
  - Shoulder muscles and foot dorsiflexion and deafness.

Erb

- *Genetic basis*: Autosomal recessive or Dominant
- *Clinical Manifestations*
  - Onset ranges from early childhood to early adulthood
  - Slow progressive weakness of shoulder and hip muscles.

Diagnostic Studies

- Muscle serum enzyme
- Electromyogram
- Muscle fibre biopsy
- ECG abnormalities
- Deficiency of muscle protein dystrophin

Management

- No definitive therapy is available to stop the progressive wasting of muscles.
- Corticosteroid may significantly halt the disease progression for upto 3 yrs.
- An emphasis should be place on teaching the patient and family ROM exercises, Nutrition and signs of progression