Muscular Dystrophy

Muscular Dystrophy :-

- It is an Autosomal Dominant disorder.
- Muscular dystrophies are characterized by progressive skeletal muscle weakness.
- Defects in muscle proteins, and the death of muscle cells and tissue

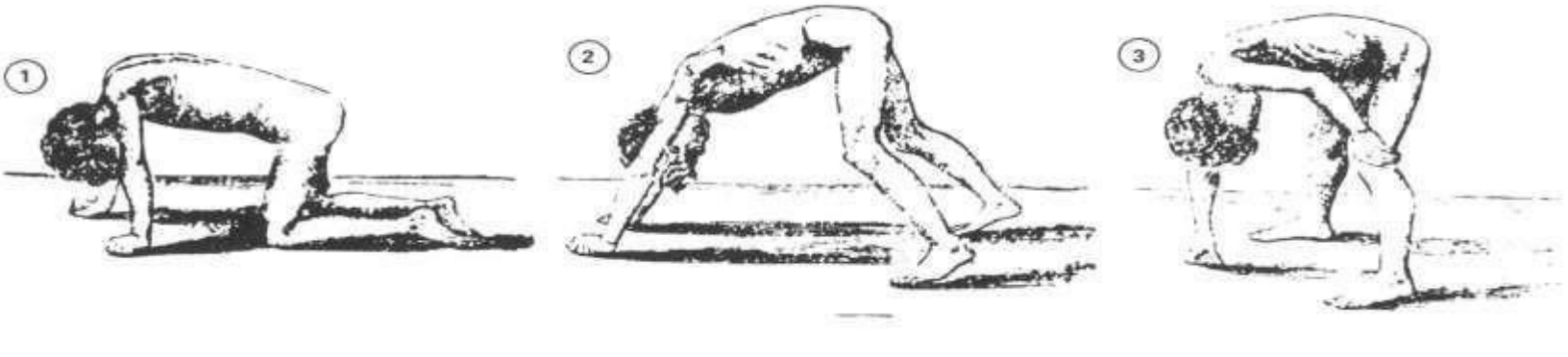
- French neurologist Guillaume Duchenne has first found this disease -which now carries his name—Duchenne muscular dystrophy.

Clinical Features :-

- Progressive muscularwasting
- Poor balance
- Atrophy
- Scoliosis (curvature of the spine and the back)
- Inability to walk.
- Frequent falls
- Waddling gait.
- Calf deformation.
- Limited range of movement .
- Respiratory difficulty
- Muscle spasms.
- Gowers' sign[10]

Gower’s Sign :-

- **Gowers' sign** is a medical sign that indicates weakness of the proximal muscles, namely those of the lower limb. The sign describes a patient that has to use his hands and arms to "walk" up his own body from a squatting position due to lack of hip and thigh muscle strength



Causes Of MD :-

- Muscular Dystrophy is genetically inherited,
- However, mutations of the dystrophin gene and nutritional deficits are responsible for the disease.
- The main cause of the disease is due to the lack of Muscle proteins like
- Dystrophin and Dystrophin Associated Protein Complex.

Types :-

- There are Around 30 Types of Muscular Dystrophies out of
- Duchenne Muscular Dystrophy.
- Becker's Muscular Dystrophy .
- Limb-girdle Muscular dystrophy
- Myotonic Muscular dystrophy

are Common

Diagnosis :-

- The diagnosis of muscular dystrophy is based on the results of
- Muscle biopsy,
- Increased creatine phosphokinase (CpK3),
- Electromyography,
- Electrocardiography
- DNA analysis.

Treatment :-

- There is no cure for any form of muscular dystrophy.
- In Duchenne muscular dystrophy, corticosteriods may slow muscle destruction.
- In myotonic muscular dystrophy, phenytoin, procainamide, or quinine can treat delayed muscle relaxation.
- Gene Transplation Researches are going on

Physiotherapy Management :-

- Duchenne musculardystrophy is often divided into 3 stages
- Early stage
- Transitional Stage
- Late / Non Ambulatory Stage

EARLY STAGE :-

Weakness of

Hip :- Extensors (Gluteus Maximus),
Abductors(G.Medius) Adductors become weak

Ankle Dorsiflexors become weak

Abdominals , Neck Flexors ,Shoulder Abductors
,Shoulder Elevators and depressors are also involved

Compensations of Early stage :-

- Increased Lumbar Lordosis
- Lack of heel strike
- Increased hip flexion during swing to clear foot
- Foot may be Pronated and Everted
- Cadence is decreased (Speed)

Transitional Stage :-

- Progression of Muscles Listed in Early stage
- With more marked Increased weakness in

  Quadriceps & Ankle Evertors.

- **Compensations:-**
- Base of support widens.
- More increased Falls
- Knee Buckling(Quads weakness) Causes more falls

Tightness Develops in muscles like

- Illio Tibial Band and Tensor facia lata.
- Hip Flexors
- Hamstrings
- Gastrosoleus
- Posterior Tibialis

Functional Losses :-

1. Inactivity of Elevation against gravity
2. Inability to rise from floor
3. Inability in stair Climbing
4. Difficulty from rising of a chair

LATE OR NON –AMBULATORY STAGE :-

- Upper limb weakness becomes more prominent

Elbow extension weaker than Flexion

Fore arm supination is weaker than pronation

Wrist & Finger extension weaker than Flexion

Scoliosis is seen Caused due to (posterior pelvic tilt)

Compensations:-

1. Leaning for stability
2. Contralateral trunk leaning to compensate upper extremity function
3. Using Mouth to grab fingers to compensate UE function

RESPIRATORY INVOLVEMENT:-

- Respiratory insufficiency is major cause of death in 90% of DMD patients
- Causes of Respiratory Problems
- Less inspiration due to muscle weakness
- Decreased Lung Expansion
- Decreased Coughing ability
- Restricted Chest wall Mobility
- Impact of Spinal Deformity

CARDIAC INVOLVEMENT :-

- Cardiac muscle is affected by Dystrophic process.

- Myocardial fibrosis may occur primarily involving walls of left ventricle.
- Spectrum of abnormalities with Cardiac involvement includes

- AV Block
- Atrial paralysis
- Atrial Fibrillation
- Ventricular arrhythmias
- Conduction Defects
- Reduced Ejection Fraction

GOALS OF Physiotherapy Management :-

- **Long Term Goals:-**

1. To Prevent Deformity
2. To Maximise & maintain Strength of muscles
3. To Maximise & maintain Respiratory status
4. To maintain Ambulation as long as possible
5. To maintain highest posssible level of Functional independence
6. Using Adaptive Equipment and Orthotics as needed

SHORT TERM GOALS :-

- To increase or maintain Rangeof motion of joints
- To increase or maintain Strength & Endurance.
- To pramote Optimal body alignment & symmetry
- To maintain sitting ability
- To provide an active respiratory programme.
- To strengthen or maintain respiratory muscle endurance
- To establish and monitor Home programmes
- To promote Relaxation & comfort

ASSESSMENT :-

- Range of Motion
- Assesment in all positions & Transition between positions
- MMT (Manual Muscle Testing)
- Vital capacity Analysis (Spirometry)
- Patterns of Breathing
- Gait Assesment
- Assesment of ADL acivities (Functional Activity scale)
- Physical Environment & Accesibility

PT – Management :-

- **Early stage:-**
  Education of family , prevention of deformity
  Maximizing Strength & Functional capabilities
  Intervention to maintain Ambulation.

- **Transitional stage :-**

  Strechings to muscles

  Lower Extremities –

  Illiotibial bands , Tensor facia lata
  Hipflexors, Hamstrings, gastrosoleus , posterior Tibialis

  Upper Extremities :-

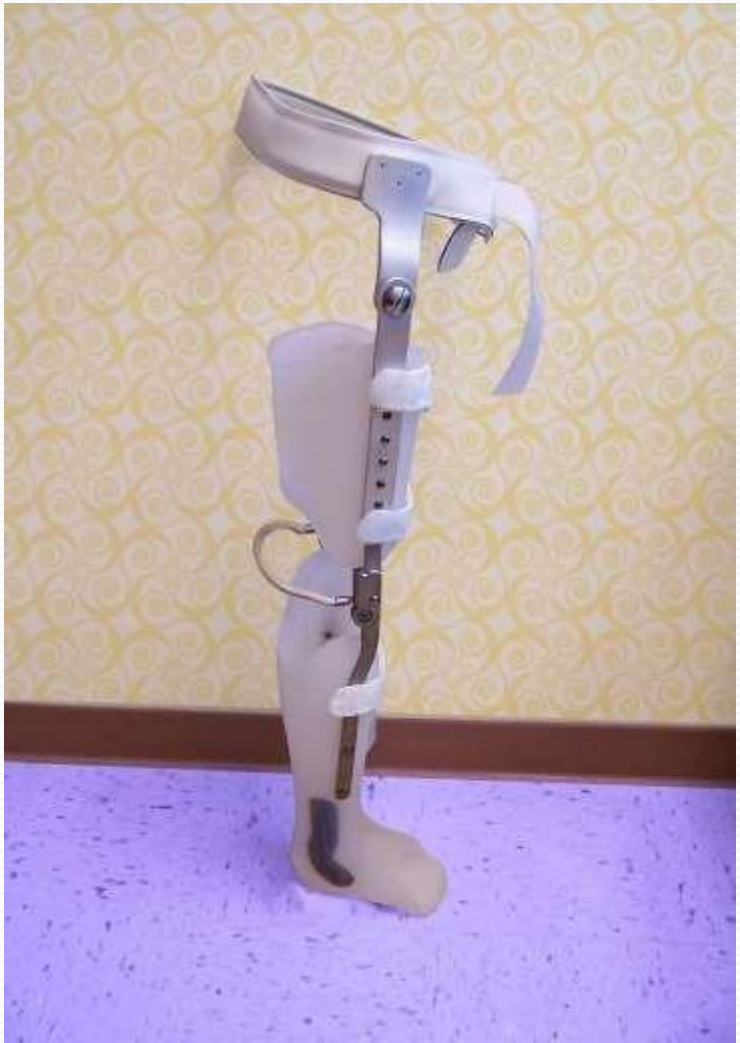
  Elbow Flexors ,Fore arm pronators , Wrist and finger flexors

- Passive streching must be done and it is best achived by standing
- PNF Techniques (hold/relax)
- Joint Mobilization – Patella,elbow,anterior and posterior movements of tibia on femur
- Myofacial release
- Moist Heat – to increase comfort & plasticity of tissue increases but excessive heat should ne avoided it can damage tissue.
- Positioning – Prone Lying and Wheel chair Positioning should be trained

Late Stage :-

- Continuation of Programme in transitional stage
- Long periods of Rest and Immobility are avoided
- Position and support for fuction
- Swimming (Hydrotherapy)
- Trike Riding (Not Uphill)
- Promotion of Orthotics in case of deformities HKAFO
- Gait Training in Parlell bars
- Transitions from one position to other
- (Sitting to standing-Standing to sitting)
- Spinal Bracing To avoid /Correct Scoliosis.

HKAFO



Spinal Brace

Respiratory Management:-

Inspiratory Breathing / Segmental breathing .

- to strengthen Diaphragm
- For lung Expansion and chest wall mobility
- For efficient breathing

GPB- Glossopharangeal breathing

(means of pistoning air into the lungs to volumes greater than can be achieved by the person's breathing muscles (greater than maximum inspiratory capacity). The technique involves the use of the glottis)

Postural drainage as necessary by percussions & oscillations

Periodic review of bronchial hygeine at home

Surgeries :-

- Spinal surgeries like

Segmental Stabilization of spine is achived by spinal surgeries to prevent the abnormal curvature of spine

Thank you