

A Case Study: Conventional Physical Therapy and Electrical Stimulation in Duchenne Muscular Dystrophy Patient

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Abstract: Introduction: Duchenne Muscular Dystrophy (DMD) is inherited as an X-linked recessive disorder (Xp21) of muscle characterized by progressive muscle weakness, intellectual impairment, and pseudohypertrophy and wasting of skeletal muscles, smooth and cardiac muscle. Prevalence according to Indian Association of Muscular Dystrophy 30 cases per 1,00,000 in India. Case presentation: 8 years male had difficulty in walking and playing, difficulty in sit to stand, frequently respiratory infection, pain in calf region. Patient had started physiotherapy treatment with electrical stimulation for both lower limb muscles. Management and outcome: In exercise stretching exercise, strengthening exercise, breathing exercise, endurance training, electrical stimulation, hand held dynamometer, North Star ambulant scale. Discussion: Exercise and electrical stimulation were improve in the lower Limb muscles strength & NSAA Score for the lower limb function following intervention was observed in the patient. Conclusion: Exercise and electrical stimulation were used for lower limb muscle strength and functional improvement. So there was improved both the lower limb muscle strength which was examined by hand held dynamometer and functional improvement was examined by North Star ambulant scale. So there was seen improvement in daily activities.

Keywords: Duchenne Muscular Dystrophy, Electrical Stimulation, hand held dynamometer, North Star Ambulatory Assessment, physical therapy

1. Introduction

Duchenne Muscular Dystrophy (DMD) is inherited as an X-linked recessive disorder (Xp21) of muscle characterized by progressive muscle weakness, intellectual impairment, and pseudohypertrophy and wasting of skeletal muscles, smooth and cardiac muscle. It is caused by mutations in the DMD gene on the X chromosome^{1,2,3}, and it has been recently demonstrated that DMD is the result of a defective gene that encodes a protein termed "dystrophin" which leads to the complete absence of the cytoskeletal protein dystrophin in both skeletal and cardiac muscle fibers⁴. The incidence of DMD is approximately 1 in 3500 male birth⁵. The Prevalence of DMD in the general population is reported at about 3 cases per 1,00,000⁶ and according to Indian Association of Muscular Dystrophy 30 cases per 1,00,000 in India⁷.

In the DMD the primary impairment is weakness; the secondary impairments are development of contracture, postural mal alignment, reduced respiratory capacity, easy fatigability and occasional obesity.

Primary impairment - Weakness of muscle is because of muscle cell destruction due to abnormal or missing dystrophin and it affects the muscle cell membrane. An early abnormality during the process of muscle fiber destruction is the breakdown of the muscle fiber plasma membrane. The membrane destruction results in an influx of calcium-rich extracellular fluid and complement components into the muscle fibers. In addition, activation of intracellular proteases and complement occurs, with the ultimate removal of necrotic fibers by macrophages,⁸ these loss of dystrophin results in a weakened cell membrane and it is easily damaged in muscle contraction.¹

The typical progression of weakness is symmetrical from proximal to distal, with marked weakness of the pelvic and shoulder girdle musculature preceding weakness of the trunk and more distal extremity muscles, except the sternocleidomastoids and bowel and bladder function is usually spared. Progression of weakness is slow but persistent. The key muscle groups of lower limb, which are more affected in DMD, are hip extensor weakness which may lead to lumbar lordosis, hip abductor weakness which may lead to waddling gait and knee extensors weakness which may lead to hyperextension of knee and gait deterioration¹⁰.

2. Case Study

8 years male with waddling gait, pseudo hypertrophy of calf muscle and history of fall down during walking and playing, difficulty in sit to stand without support, frequently respiration infection, tiredness during activities, calf pain while stair climbing.

On physical examination the patient had generalized body weakness but lower limb is more affected than upper limb and trunk muscles. Weakness in upper limbs (4/5) and lower limb (3/5) bilaterally. Strength of lower limbs muscles measured by hand held dynamometer. He had difficulty in sit to stand and noticed Gower's sign. His feet were slightly in pronated and plantar flexed. On ambulation he had waddling gait with Hyperlordosis of lumbar spine. He had full range of motion in both upper limbs but reduced range of motion in both lower limbs.

The presenting symptoms were indications for CPK level which was elevated to the normal value in this patient. The patient's family was informed of treatment options of DMD,

who refused stem cell treatment and came for physiotherapy management.

The patient receives physical therapy and occupation therapy.

In this case, to found out the strength of lower limbs muscles before and after physical therapy. Muscles were Gluteus maximus, Gluteus medius and quadriceps bilaterally.

On the initial assessment, I was examined that the strength of gluteus maximus on right side 3 kg and left side 5kgs, gluteus medius right side 3 kgs and left side 7 kgs and quadriceps on right side 4.5 kgs and left side 4 kgs. In functional assessment NSAA score was 16.

3. Treatment Plan

- 1) Stretching exercise (To prevent contracture)¹²
 - Positioning, splinting (AFO) at night, stretching of hip flexors, knee flexors and plantar flexors.
- 2) Strengthening exercise (To improve the strength)¹²
 - Concentric muscle contraction (ROM exe). Hip extensor, knee extensor, Dorsiflexors muscles were strength
- 3) Breathing exercise (To maintain strength of respiratory muscles)¹²
 - Diaphragmatic breathing exercise 5-7 rep
- 4) Endurance training (To maintain the maximal functional level)¹³
 - Walking/cycling once in a day for 1-20 min
- 5) Electrical stimulation^{14,15}
 - Quadriceps femoris – supine lying
 - Gluteus maximus – prone lying
 - Gluteus medius- side lying
 - Placement of electrodes: Muscles to stimulated Quadriceps femoris, gluteus Maximus and gluteus medius for 10 minutes for each muscle, total 30 minutes, 5 sessions a week for 12 weeks.

4. Discussion

The purpose of the study was to evaluate the effectiveness of conventional and ES in DMD participants. ES plus conventional therapy was given in 40 sessions. The outcome measures were Hand Held Dynamometer (HHD) for lower limb muscles strength and North Star Ambulatory Assessment score (NSAA) for functional activities of lower limb.

In this study a significant improvement in the lower Limb muscles strength & NSAA Score for the lower limb function following intervention was observed in the patient.

After intervention strength was improved. I was examined that the strength of gluteus maximus on right side 6.5 kgs and left side 4.5kgs, gluteus medius right side 6.5 kgs and left side 9.5 kgs and quadriceps on right side 9 kgs and left side 8.5 kgs. In functional assessment NSAA score was 20.

The present results show that muscles of children suffering from Duchenne muscular dystrophy can be influenced by

electrical stimulation, provided that the stimulation is applied at a time when the children are not yet severely disabled. This beneficial effect of stimulation could be due (a) to slower deterioration of the existing diseased muscle fibers, (b) to a better and more rapid growth of regenerating fibers that are known to be present in muscles from Duchenne muscular dystrophy children, particularly in the younger age groups, and (c) to hypertrophy of either healthy fibers or affected fibers.

Muscle activity induced by electrical stimulation is in many respects unnatural and has often been viewed with some reservation. Two fundamental differences exist between voluntary elicited contractions and those induced by electrical stimulation of muscle. During voluntary or reflex movements motor units are activated asynchronously and a strict hierarchical order of recruitment is always maintained. During this hierarchical recruitment of motor units the smallest motor units are activated first followed by contractions of larger units. The force of contraction is graded, in general by increase in 1) the number of motor units' recruited (spatial summation); 2) the frequency of nerve impulses (temporal summation). Therefore during voluntary movement the largest motor units are least active and are used only during maximal effort. Type 1 muscle fibers are recruited are first and later type 2. It may leads to fatigue, rapidly in type 2b and most slowly in type 1.

In involuntary contraction, motor unit are recruited from smallest to largest as requirements for force are increased. In stimulated contraction, recruitments tend to occur from largest to smallest as the stimulation strength is gradually increased. Electrostimulation was often considered to recruit motor units in the opposite order from voluntary drive, contrary to Hennemann's "size principle". The principle states that slow motor units, associated with small-diameter motoneurons axons, are active before fast motor units, which are associated with larger-diameter axons. These principle based on 2 commonly agreed upon findings: (1) the axons of the larger motor units have a lower resistance to current and conduct action potentials at faster rates than the axons of the smaller motor units, and (2) few data demonstrate increased fatigue with EMS versus voluntary activation.

5. Conclusion

Stretching Exercise, strengthening exercise, breathing exercise, endurance training, electrical stimulation for gluteus maximus, gluteus medius and quadriceps femoris muscles are helpful for improving lower muscle strength which was examined by hand held dynamometer and lower limb functions was examined by North Star ambulant scale.

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