Rehabilitation in Duchenne Muscular Dystrophy: A Case Report

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ABSTRACT

Background: Duchenne muscular dystrophy (DMD) seems to be the most common X-linked muscular dystrophy condition in children marked by proximal muscle weakness and calf hypertrophy and affected mostly in boys. In more than 20 muscular dystrophies the most prevalent form is Duchenne muscular dystrophy. Each 1/3500 male births, the presence of DMD worldwide, indicating that in the UK alone there are approximately 2400 people living with DMD. Assessing improvements in severity, range of motion and functional mobility, implementation of therapies and guidance on positioning are important key factors.

Clinical Finding: Mostly in proximal muscles around the hips and shoulders, which are those around the trunk of the body, muscle weakness occurs mainly. In the lower extremities, weakness usually begins proximally, and then passes distally. Symptoms typically begin about the

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age of 1-3 years. To see the child run and get up off the floor while searching for signs of DMD is necessary.

**Diagnosis:** A very common physical finding in boys with Duchenne's is Gower's Sign. In order to get up, it requires using their hands to 'climb' their legs. It is because of a weakness in the boy's hip muscles, which was seen in this patient in progressive in nature. The proper examination and evaluation the symptoms the child presented to the physiotherapy clinic played an important in reaching to the diagnosis of the condition.

**Conclusion:** Prevent the child from developing contracture, correct posture of the child and maintain quality of life these are important criteria in for the child suffered from Duchenne muscular dystrophy.

**Keywords:** Gowers sign; rehabilitation; physiotherapy; DMD; pediatric.

## 1. INTRODUCTION

Duchenne muscular dystrophy (DMD) tends to be an unusual musculoskeletal genetic disease that shows early disease signs of progressive muscle weakness. This is a heritable X-linked chronic syndrome that impacts 1 of every 3,500 male live births and was named after the French neurologist Guillaume Benjamin Armand Duchenne around 1860. The incidence of muscular dystrophies was estimated to be between 19.8 and 25.1 per 100,000 individual years [1].

A most popular and serious form of muscular dystrophy in affected boys, occurring at the age of 3-5 years, calf hypertrophy and proximal muscle weakness characterize this condition [2]. With a distinctive and enduring clinical appearance, DMD has a very high mutation rate. In their early to mid-twenties, by the age of twelve, most patients are wheelchair-bound and suffer [3].

Before the age of 5, the first signs of the disease begin. The ability of walk is always lost before puberty, without treatment. While boys are restricted to wheelchairs, their arm and hand functions are increasingly dependent on them, this is important for most everyday activities, including electric wheelchair driving, sleeping, school work, computer and video games activities [4].

In the treatment of people with DMD in the following capabilities, physical therapists play a crucial role: Assessing improvements in severity, range of motion and functional mobility, implementation of therapies and guidance on positioning, stretching, and activity pacing/modification for individuals and their families [5,6].

## 1.1 Patient Information

A 12-year-old patient with the main concern reported difficulty walking to the department. The difficulty of standing, walking and everyday tasks began and progressed steadily at the age of 6 years. His family has a long history of slips, exhaustion, muscle fatigue, and the failure to climb stairs.

## 2. CLINICAL FINDINGS

Patients with proximal weakness, lower and upper extremity weakness, and trouble standing, walking, standing up from a sitting position, and ascending stairs, and hypertrophy of the calf (shown in Fig. 1), contracture of the hamstring muscle, and positive Gower's sign. The history of cranial nerve involvement and muscle aches has not been documented. It was assumed his intelligence quotient had been in the average category.

On observation, the child had a mesomorphic appearance. The hyper-lordotic curve was present and pelvis was anteriorly tilted, weight was shifted forward. When the child is seated, the hyper-lordotic curve disappeared. The ankle was in a state of plantar flexion and slightly inverted (Fig. 2).

On general physical inspection, it has been noticed that superficial, deep and cortical stimuli are intact (At dermatome level). The motor testing was carried and the muscle tone in the bilateral upper limb and lower extremities was typical in grade 0, as per the Modified Ashworth Scale (MAS).
The motion analysis was done passively and it was measured within normal limits by the goniometer. This is seen in table-1. Manual Muscle Testing (MMT) was evaluated, which is seen in Table 2. For activities of everyday life measurement, the Barthel Index Scale was used and the score for the patient was 8.

There has been no thinning, muscle twitching, muscle tone twitching, all developmental milestone appeared till the age.

Neurologic examination was done where dermatomes and myotomes was evaluated and there were no diminished or absent sensation, nerve roots were intact. Reflexes were intact.

Egen classification scale score was -13. North Star ambulatory assessment score was – 4/34.

3. HAND FUNCTION ASSESSMENT

Hand function were assessed for grasp and grip the following findings were obtained which are tabulated.

Grasp  a) spherical  No  b) cylindrical  No  c) hook  No
Grip  a) pulp to pulp  No  b) tip to tip  No  c) three jaw chuck  No  d) lateral prehension  No

Table 1. Range of motion

| Joint movement       | Pre-Range of motion | Post-Range of motion |
|----------------------|---------------------|----------------------|
| Shoulder Flexion     | 0-90°               | 0-105°               |
| Shoulder Extension   | 0-10°               | 0-18°                |
| Shoulder Abduction   | 0-70°               | 0-90°                |
| Elbow Flexion        | 0-100°              | 0-115°               |
| Elbow Extension      | 0°                  | 0°                   |
| Hip Flexion          | 0-10°               | 0-20°                |
| Hip Extension        | 0°                  | 10°                  |
| Hip Abduction        | 0-15°               | 0-35°                |
| Hip Adduction        | 0-5°                | 0-10°                |
| Knee Flexion         | 0-90°               | 0-100°               |
| Knee Extension       | 0°                  | 0°                   |
| Ankle Dorsiflexion   | 0°                  | 5°                   |
| Ankle Plantarflexion | 0-40°               | 0-45°                |

Table 2. Manual muscle testing

| Joint movement    | Pre-rehab Grade | Post-rehab Grade |
|-------------------|-----------------|------------------|
| Shoulder Flexion  | 3               | 4+               |
| Shoulder Extension| 3               | 4                |
| Shoulder Abduction| 3               | 4                |
| Elbow Flexion     | 4               | 4                |
| Elbow Extension   | 4               | 4+               |
| Hip Flexion       | 3               | 4                |
| Hip Extension     | 3               | 4                |
| Hip Abduction     | 3               | 4                |
| Hip Adduction     | 2               | 3                |
| Knee Flexion      | 3               | 4+               |
| Knee Extension    | 3               | 4+               |
| Ankle Dorsiflexion| 2               | 3                |
| Ankle Plantarflexion| 3             | 3+               |
Fig. 1. Show the calf hypertrophy

Fig. 2. Proximal muscle weakness of upper and lower limb

Fig. 3. Standing with support
3.1 Intervention

The medications were given and physical therapy also started, we took the patient’s follow-up regularly.

3.2 Description

3.2.1 Phase 1 (1-6 Days)

Initially the bed mobility exercise was started in that the bed side sitting, bed side standing was taught to the patient and parents. Standing in optimum positions used pillows either splints to help the child sit, then lie in optimal positions, shoulder flexion and extension, shoulder abduction and adduction, shoulder internal and external rotation, elbow flexion and extension, and wrist flexion and extension exercises were included in the upper extremity ROM training program. Each ROM exercise was done by subjects 5 or 10 times 3 times a day [5]. Hand gripping exercises were also started to improve the strength in upper extremity. Passive lower limb mobility exercise- ankle toe movement, heel slide, hip abduction-adduction. For any particular joint or muscle group, such as, passive stretching to avoid or reduce contractures was performed at least 3 times a day. Tendo Achilles stretching, hamstring stretching, adductor stretching, piriformis stretching was performed for 5 repetition for 30 sec stretching. In order to preserve respiratory function and avoid a decrease in total lung volume and forced vital capacity, inspiratory muscle training was implemented. Early respiratory preparation has been shown to promote quality of life and decrease the risk of potential respiratory complications [3,7].

3.2.2 Phase 2 (6-12 Days)

The above all exercises were continued to perform. PNF stretching may be added. The strengthening exercises was started with the help of weight cuff hip abduction, adduction, straight leg raise, heel slide, dynamic quads etc. the use of anti-swan neck orthosis for preventing the swan neck deformity.

The functional reach activity was started. In sitting, lifting one leg at a time weight transfer through the arm behind and the sideways to stimulate balance reaction. Reach outs sideways and forward to control the trunk. Standing static and dynamic equilibrium exercises was concentrated for balance. We have prescribed orthosis, it is important for rest or stretch ankle-foot orthosis (AFOs), to prevent and minimize progressive plantar flexion contractures [1,8].

3.2.3 Phase 3 (12-18 Days)

Knee-ankle-foot orthosis (KAFOs) for early non ambulatory and late ambulatory times, limited ambulation for medicinal cause, and the avoidance of deformity and atrophy. Resting knee ankle-foot orthosis (KAFOs) worn at night and during the day will help prevent or slow the progression of equinus contractures and are comfortable to wear for the rest of its life. We began with KAFO patient standing, assisted standing, minimal ambulation for therapeutic purposes. Patient with support gradually we increase the period of standing initially it was 30sec. then progress to 2 min and walking was initiated, initially 10steps then progress to walk in hall [9].

We began training with bicycle exercise equipment for their arms and legs, including KPT Cyclo for 30-min sessions of 15 minutes for arm and 15 minutes for leg exercise. He was told to ride at a steady speed throughout each practice session and to complete 700-1000 revolutions with both their legs and arms without being tired [10].

3.4 Home Program

The patient was told to continue the exercise programme and ergonomic advice, to maintain correct posture during rest and work.

4. RESULTS

The exercises and bed mobilization improved the patient’s quality of life. The physiotherapy sessions were held for approximately a month. The patient was reassessed once and a prognosis was made. Mat exercises actually reach up to hands and he starts to stand early with at least 10 seconds of assistance and does not lead to early exhaustion. Patient was encouraged for walking. Preparation for walking was made, as walking is the best way to prevent blood clots. Patient was also advised to maintain a good posture to reduce additional stress. As it focuses on the functional impact of a disease and consequent treatment on a patient, as perceived by the patient, we have also planned to develop a patient health-related quality of life.
5. DISCUSSION

Duchenne muscular dystrophy is the most prevalent muscle syndrome in India and even the world. Genetic changes in the dystrophin gene make it impossible for the body to produce the protein dystrophin, which is essential for muscle contraction. Damage to muscles occurs once the muscle contracts, and is healed but protein-deficient, leading to a muscle that is still weak [11,12].

At the initial evaluation, all individuals in that center got regular education, including job instructions, risk of loss of range of motion, use of orthotics, and importance of the home stretching service. Maintaining ambulation, avoiding scoliosis, delaying the development of respiratory problems and prolonging life are the most important objectives of the DMD recovery program for children [5,13].

A selection of fitness plans for the lower extremities and other recovery measures, and enhancement in the quality of treatment and increased physiotherapy understanding of the family and supportive use of medications, ensure ambulatory care for most children with DMD before late childhood or early adolescence, are necessary to retain the strength of the lower limb muscle and maintain the ability to move. Other factors that may contribute to the result must be considered. The findings of this study show that plantar flexion contracture is progressing and that physical function is deteriorating [11,14]. Strong ankle foot orthotics, personalized to a convenient end range, and worn for 8 hours a day are common guidelines for retaining range of motion at this center (usually overnight), passive ankle stretching for 90 seconds once a day, plus lateral and self-stretching as required [8,15]. Few of the related studies were reviewed [16,17].

Physical activity in boys with Duchenne Muscular Dystrophy: the procedure of the no use idisuse trial. This study is the first clinical trial investigating the impact on muscle strength and functional abilities of low-intensity physical exercise in boys with DMD. It would be a step toward closing the existing void in our understanding of the value of physical training for these boys, as well as improving our understanding of the forms of physical training that can be prescribed.

Until the late outpatient or early non-ambulatory age, upper extremity muscle weakness, associated limitations and reliance on everyday life tasks and typically, hand-arm roles are usually neglected.

6. CONCLUSION

The patient should be taught breathing techniques to prevent respiratory issues. Reinforcing upper and lower limb muscles and improving their daily life duties of the patient. For children suffering from Duchenne muscular dystrophy, preventing the children from experiencing contracture, proper posture of the child and preserving quality of life are important criteria.

7. LIMITATIONS

Planning of further physiotherapy sessions was done but due to the arrival of Covid pandemic patient's further treatment sessions could not take place.

PATIENT CONSENT

Proper consent was taken from patient for writing case report.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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