CASE REPORT

The Effect of Static Ergocycle on Cardiorespiratory Endurance in an Early Ambulatory Duchenne Muscular Dystrophy Patient: A Case Report

Rizky Kusuma Wardhani[1], Fiona Lestari[1]

[1] Department of Physical Medicine and Rehabilitation, Faculty of Medicine Universitas Indonesia, Cipto Mangunkusumo General Hospital Jakarta, Indonesia9

ABSTRACT

Introduction: Duchenne muscular dystrophy (DMD) is a debilitating genetic disorder affecting approximately 1 in 3,600 to 6,000 live male births, caused by mutations in the dystrophin gene. The early ambulatory stage of DMD is typically characterized by difficulty standing, muscle weakness, frequent falls, and impaired motor skills. Previous research suggests that assisted bicycle training may improve patients' quality of life and lung function.

Case Description: A multidisciplinary approach involving medical and rehabilitative interventions is essential to improve quality of life and delay disease progression. The rehabilitation program included physical activities, stretching exercises, and environmental modifications. Spirometry and the 6-Minute Walk Test (6MWT) indicated restrictive lung patterns and reduced exercise capacity. Adherence to corticosteroid treatment, continuous monitoring, and a comprehensive rehabilitation strategy were crucial for maintaining optimal muscle function and enhancing overall quality of life. Pulmonary function evaluation and management are particularly important, given the association of pulmonary complications with mortality in DMD.

Keywords: Duchenne muscular dystrophy, rehabilitation management

Correspondence Detail:

Rizky Kusuma Wardhani

Department of Physical Medicine and Rehabilitation, Faculty of Medicine Universitas Indonesia, Cipto Mangunkusumo General Hospital Jakarta, Indonesia
Email: rizkikusumawardhani@yahoo.co.id

INTRODUCTION

Duchenne muscular dystrophy (DMD) is a severe, progressive disorder that affects 1 in 3,600-6,000 male births. The condition is caused by gene mutations in the dystrophin gene (locus Xp21.2).[1–3] Currently, there is no definitive cure for DMD. However, multidisciplinary approach combining medicine, surgery and

rehabilitation can change the disease's natural progression, thus improving patient's quality of life (QoL) and longevity.[4] Previous research has shown that low-intensity aerobic exercise, particularly assisted bicycle training, may enhance motor function in DMD patient. This case report aims to evaluate the effect of static ergocycle in early ambulatory DMD case to maintain cardiorespiratory function.[5]

CASE PRESENTATION

A 5-year-old boy, suspected to have Duchenne Muscular Dystrophy (DMD), was referred to the Physical Medicine and Rehabilitation Outpatient Clinic. Approximately 7 months ago, he began experiencing several functional limitations, such as needing to take breaks after walking 200-300 meters, crawling to climb stairs, difficulty lifting heavy objects, and slower learning capacity compared to his peers. Since the onset of these symptoms, he has preferred spending most of his time on his phone rather than engaging in outdoor activities.

Physical examination revealed an anterior pelvic tilt, hyperlordotic lumbar posture, pseudohypertrophy of both deltoid and calf muscles, and hypotrophy of all muscles. Range of motion (ROM) was full in both the upper and lower extremities, while manual muscle testing (MMT) indicated weakness in all extremities, more prominent in the proximal regions. The Valley sign and Gowers sign were positive, while the Meryon sign was negative. Gait analysis showed a waddling gait, characterized by initial total foot contact and increased arm swing.

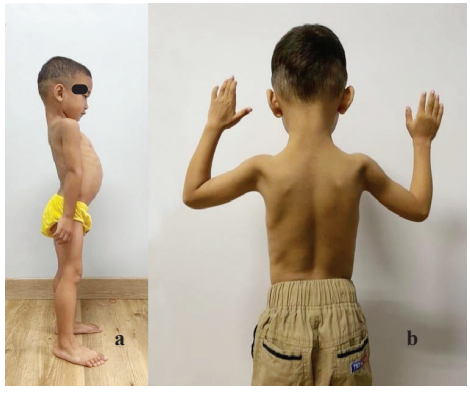

Figure 1.a) Lateral View of Patient Posture. b) Valley Sign.

Figure 2. Gowers Sign.

Spirometry results indicated restrictive lung disease, with a forced expiratory volume (FEV1) of 0.44 L (58% of predicted), forced vital capacity (FVC) of 0.45 L (51% of predicted), a FEV1/FVC ratio of 98%, and weak peak cough flow (115 L/min). The 6- Minute Walk Test (6MWT) was terminated prematurely at 5 minutes 15 seconds due to fatigue, covering a distance of 210 meters (48% of predicted), with normal vital signs and no falls. His Brooke Scale score was 1 point, Vignos Scale score was 3 points, and his ambulation function classification system for DMD (AFCSD) was Level II. The Muscular Dystrophy Functional Rating Scale showed the following: 81% for the mobility domain, 75% for the basic ADL domain, 100% for the arm function domain, and 95% for the impairment domain.

Creatine Kinase (CK) levels were elevated at 9,872 U/L. Electromyography (EMG) of the right tibial anterior muscle revealed a myogenic lesion with chronic denervation. Musculoskeletal ultrasonography of both upper and lower extremities showed decreased muscle fiber with dominant hyperechoic homogeneous fibrosis, suggestive of DMD.

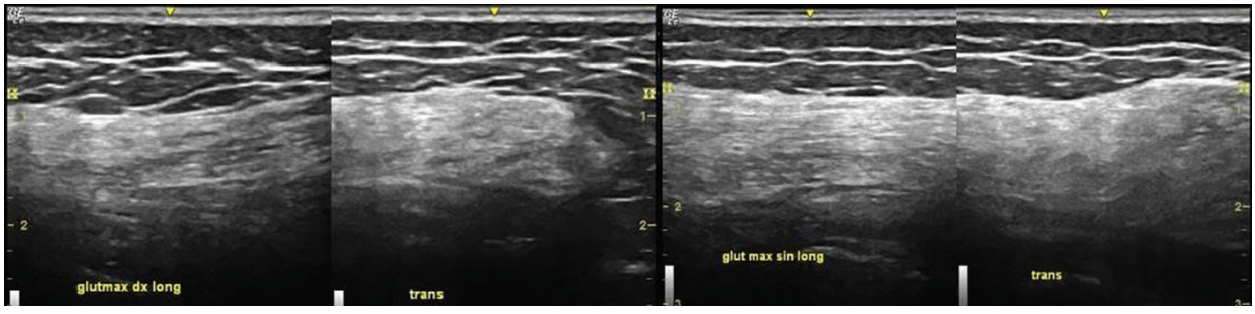

Figure 3. Ultrasonography of gluteus maximus muscle on a) left side and b) right side. Note that both pictures depict minimal muscle fibre and hyperechoic features with homogeneity.

He was placed on a regimen consisting of intermittent Methylprednisolone at 0.75 mg/kg/day, divided into two doses for 10 days, followed by 10 days off, along with omeprazole and Vitamin D as prescribed by his pediatrician. The patient's rehabilitation program includes the following:

- Mild to moderate intensity aerobic exercise (static ergocycle: 3 x 10 minutes, with a 10-minute break between each session, 5 times per week or as tolerated by the patient)
- Mild to moderate intensity isometric strengthening exercises (hip extensors, knee extensors, and dorsiflexors: 3 x 10 repetitions, with a 6-second hold each, 3 times per week or as tolerated by the patient)
- Functional upper extremity exercises (ball throwing activity: 3 x 10 repetitions, 3 times per week or as tolerated by the patient)
- Passive stretching (hip, knee, and ankle joints: 3 x 10 repetitions, with a 10-second hold each, 7 days per week)
- Incentive spirometry (5 x 5 repetitions, with a 3-second hold each, 7 days per week)
- Active air stacking exercise (3 x 5 repetitions, 7 days per week)
- Voluntary coughing 3 times per day

Furthermore, optimization of the home environment was recommended, such as using a seated toilet or a commode chair. After 3 months of intervention, the 6-Minute Walk Test (6MWT) was repeated, resulting in a distance of 185 meters (41.0% of predicted from Rizky's Formula), with the test being stopped at 5 minutes 5 seconds due to exhaustion. He also completed the 10-Meter Walk Test (10mWT), with a time of 24 seconds. After 4 months of intervention, his subsequent 6MWT showed a distance of 210 meters (47.5% of predicted from Rizky's Formula), during which he completed the full 6-minute course without stopping or falling. However, the patient continued to report experiencing multiple falls during daily activities, which made him unwilling to participate in outdoor activities.

DISCUSSION

The patient's age falls within the typical range for the onset of Duchenne Muscular Dystrophy (DMD), which is between 2 and 6 years old. The mean age of elevated creatine kinase (CK) levels is approximately 4.7 years, according to existing research, which explains the significant elevation of the patient's CK levels when he was 4 years old.[1,6]

Although genetic testing and muscle biopsy are considered the gold standards for diagnosis, these tests could not be performed due to financial limitations. As an alternative, Nerve Conduction Studies, Electromyography (EMG), and Musculoskeletal Ultrasonography were conducted, all of which supported DMD as the working diagnosis. EMG results revealed low-amplitude, short-duration polyphasic motor unit action potentials (MUAPs) and increased recruitment, or "early" recruitment, observed with slight effort. Complex repetitive discharges and abnormal spontaneous rest activity were also present, indicating membrane instability.[6]

The patient was prescribed intermittent corticosteroid therapy, specifically prednisone, which has been shown to help maintain muscle strength and prolong independent ambulation by approximately 2 years. The dose and potential side effects are closely monitored, as corticosteroids can predispose patients to compression fractures, and DMD itself impairs bone metabolism, leading to reduced bone mineralization.6 Routine spine radiography is also recommended, a s scoliosis may develop in patients taking glucocorticoids, particularly after discontinuation of the medication. Annual follow-up assessments will be necessary as the patient progresses towards a non-ambulant state. Echocardiography is planned, as DMD can affect the cardiac muscles, leading to progressive dilated cardiomyopathy, congestive heart failure, cardiac insufficiency, conduction abnormalities, and an increased risk of sudden early death.[4]

The effects of exercise on muscle degeneration in dystrophinopathies are not fully understood, with both supporting and opposing evidence.[8] It is important to note that eccentric muscle exercises and high-resistance strength training should be avoided, as they can be harmful due to muscle fragility, metabolic abnormalities induced during exercise, and reduced exercise capacity. On the other hand, submaximal aerobic exercise or activity is recommended, particularly in the early stages of the disease, as it can promote muscle regeneration and reduce oxidative stress. Static cycling, with a duration ranging from 15 to 30 minutes at an intensity of 60% of the heart rate,

repeated 3-5 times per week, has been shown to produce beneficial effects.[5,9,10]

The results of the 6-Minute Walk Test (6MWT) are as follows: 210 meters at initial contact, 185 meters at 3 months after intervention, and 210 meters at 4 months after intervention; these correspond to 47.5%, 41.0%, and 47.5% of the predicted distance from Rizky's Formula (441.58 meters), respectively. The decrease in distance at the 3-month follow-up may be attributed to fatigue and overexertion, even though the patient theoretically could have continued further. This is supported by the fact that the 6MWT result after 4 months of intervention was the same as the initial 6MWT result, which was 210 meters. These findings suggest that the patient was able to maintain his cardiorespiratory endurance with the rehabilitation program prescribed in this case report.

CONCLUSION

Static ergocycle is a safe aerobic exercise option for ambulatory DMD patients. It can be incorporated as a recreational activity, allowing patients to maintain their cardiorespiratory endurance, as demonstrated by the 6MWT results.

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