INTRODUCTION

Myasthenia Gravis (MG), is a slowly progressive neuromuscular disorder which is caused by an autoimmune disorder characterized by fluctuating pathological weaknesses with remissions and exacerbations involving one or several skeletal muscle groups. Fatigable muscle weakness is the clinical hallmark of MG.\(^1\)\(^,\)\(^2\) mainly caused by antibodies in the acetylcholine receptor (AChR) at the post-synaptic site of the neuromuscular junction. MG affects between 5 – 12 people per 100,000, with two peaks, one between 20 and 40 year old women; and the other between 60 and 80 year old men.\(^3\) One of the most serious symptoms of myasthenia is weakness of the tongue and posterior pharyngeal muscles that produces dysphagia, which continues to be a major source of morbidity in MG.\(^3\)

Dysphagia or oropharyngeal weakness is the presenting symptom in 15-40% of MG patients.\(^4\) The occurrence of dysphagia, along

ABSTRACT

**Objective:** To evaluate whether Shaker Exercise can improve dysphagia in MG patients.

**Methods:** Pre-Post test design. Ten patients with a confirmed diagnosis of MG (Osserman class IIB) were recruited for this study. All patients did Shaker exercise an hour after taking their medication, with holding time and repetition which increased gradually for 6 weeks. Examinations using Fiber Optic Endoscopic Evaluation of Swallowing (FEES) and Eating Assessment Tool (EAT-10) were conducted in between.

**Results:** All patients completed 6 weeks of exercise with increased holding time (pre 14±6.11; post 27.5±4.25) and repetition (pre 12.7±6.9; post 20.7±7.75). Only one patient experienced discomfort in their neck muscles. Pre intervention FEES Examination showed that all ten patients have residue (100%), two have spillage (20%), three have penetration risk (30%) and two have aspiration risk (20%). Post intervention showed no residue in seven patients, while three showed decreased residue (70%; 30%; 100%). There was no spillage, penetration risk or aspiration risk in all patients. Pre (11.1±9.386) and post (2.2±2.573) intervention EAT-10 scores show significant improvement (p=0.002).

**Conclusion:** Shaker Exercise can be used safely to improve dysphagia in Myasthenia Gravis patients.

**Keywords:** Myasthenia Gravis, Shaker Exercise, Dysphagia
with respiratory involvement, continues to be a significant source of both morbidity and mortality in MG. Aspiration and pneumonia have been shown to be a significant risk factor for a potentially life threatening myasthenic crisis.\(^2\)

Swallowing is an event that requires an elaborate coordination of neuromuscular activity. The main component of the UES is the cricopharyngeus muscle. During swallowing, this muscle relaxes and is pulled open by the contraction of the suprahoid muscles. The suprahoid muscles involved in this movement include the geniohyoid, thyrohyoid, and digastric muscles.\(^3\) These muscles are striated muscle that can be affected by Myasthenia Gravis.\(^3\) Neurological conditions resulting from Myasthenia Gravis cause dysfunction of the upper esophageal sphincter opening. This causes a barrier to food-bolus flow from the pharynx, leading to post-swallow residue and aspiration.

Muscle weakness will cause deconditioning which leads to muscle waste and so on. This condition can be managed by exercise prescription. The specific role of exercise prescription in the management of MG is limited; however some research has been conducted into the effects of exercise prescription in other slowly progressive neuromuscular disorders (NMD) with similar symptoms. The principles behind exercise prescription in the management of NMD include maintenance of muscle strength within the limitations of the disease process, thus minimising atrophy.\(^7\)

Over the past 25 years, a number of laboratories around the world have studied and defined the factors responsible for the opening of the upper esophageal sphincter (UES). The upward-forward movement of the hyoid pulls the upper sphincter open after the cricopharyngeal muscle relaxes, enabling the upward-forward movement of the larynx. The Shaker Exercise based on that physiology, involves isometric and isokinetic neck exercises aimed at strengthening the suprahoid muscles including the geniohyoid, thyrohyoid, and digastric muscles.\(^8\) The Shaker exercise requires less muscle effort than traditional therapy, which often involved a series of exercises like mendelsohn, supraglottic or super supraglottic. Therefore, Shaker Exercise might be preferable for Myasthenia Gravis patients. Research on this exercise shows improvement in the duration and the width of the UES opening in normal elderly patients. Other clinical trials show that Shaker exercise significantly reduces post-swallow aspiration to a greater degree than the traditional therapy.\(^8\)

The aim of this study is to evaluate whether Shaker Exercise can improve dysphagia in MG patients. We hypothesized that Myasthenia Gravis patients will improve in terms of residue and to decrease aspiration risk.

**METHODS**

Ten patients with a confirmed diagnosis of MG were recruited for this study. The inclusion criteria was (1) Moderate-severe generalized myasthenia classified by Osserman classification (Osserman class IIB), (2) 18 - 60 years old. The exclusion criteria were (1) myasthenic crisis, (2) other systemic conditions which make exercise contraindicated, and (3) thymoma. All patients conducted the exercise program at home using the Shaker method for 6 weeks. The exercise doses were modified due to the fatigable characteristics of Myasthenia Gravis. All patients had prostigmine as their routine medication therapy and the exercise was conducted 1-2 hours after oral administration. They also attended weekly evaluations for a general check up and evaluation of exercise dosage.

**Description of Exercise**

The traditional Shaker Exercise involves isometric and isokinetic neck exercises. The isometric strengthening portion involves three consecutive head lifts for 60 seconds, with a 60-second rest period between each head lift. The isokinetic strengthening portion involves 30 consecutive head lifts without holding. We modified the Shaker Exercise isometric holding time and isotonic repetition because of Myasthenia Gravis fatigable muscles. The Shaker Exercise consisted of some head
lifts in the supine position with a 1-min rest between lifts. These sustained head-raising exercises were followed by some consecutive repetition of head raising in the same supine position. The initial minutes of head lifts and consecutive repetition were determined from exercise testing. The test was repeated once a week for six weeks. For both sustained and repetitive head raising, patients were instructed to raise the head high enough to be able to see their toes without raising their shoulders. The exercise test was stopped if the patient (1) was no longer able to lift their head as high as before (for isotonic), or maintain the height of the head lift (for isometric); (2) their shoulders begin to raise along with their head; and (3) the patient complains of fatigue or discomfort. The amount of holding minutes and consecutive repetition before those events were taken into account as data.

**Outcome Measurement**

Objective examination for dysphagia taken from FEES procedure. The interpretation of FEES are spillage, residue, penetration and aspiration. Spillage is a condition that causes food bolus in the pharyngeal region before the swallowing process can begin. Residue is the condition of residual bolus left in the hypopharyngeal region after the swallowing process. Penetration is taken into account if there’s bolus dropped into the laryngeal vestibulum. Aspiration means there’s bolus going through the vocal cord.

The EAT-10 is a self administered survey for the subjective assessment of dysphagia. It can be utilized as a clinical instrument to document the initial dysphagia severity and monitor the treatment response in people with a wide array of swallowing disorders including Myasthenia Gravis. The instrument has displayed good consistency, test-retest reproducibility, and validity. An elevated EAT-10 score indicates a higher self perception of dysphagia. Normative data suggests a score of 3 or more is abnormal.

**Statistical Analysis**

Patient characteristics were determined by univariate analysis. The outcomes were compared before and after exercise using a non parametric method due to the small sample size (Friedman test).

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**Pathway**

MG patient diagnosed by neurologist

**INCLUSION**

- FEES (without drugs)
- EAT -10

Refered to Dysphagia Team RSHS

**Shaker Exercise**

6 weeks
(an hour after take medicine)

**Evaluation**

- FEES (without drugs)
- EAT -10

Data Analysis

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Figure 1. Study Pathway
RESULTS

All ten patients completed 6 weeks of exercise. Their characteristic are shown in table 1. There was only one who experienced discomfort in their neck muscles. All of them have increased holding time and repetition in Shaker Exercise dosage. Pre intervention FEES showed all patients have residue, two have spillage, three have penetration risk and two have aspiration risk. Post intervention showed no residue in seven patients while three showed decreased residue, there were no spillage, penetration risk and aspiration risk in all patients (Figure 2). Pre and post intervention EAT-10 scores show significant improvement (Table 3).

Table 1. Characteristics of Participants at Baseline

<table>
<thead>
<tr>
<th>Variable</th>
<th>n</th>
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<tbody>
<tr>
<td>Gender</td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>3</td>
</tr>
<tr>
<td>Female</td>
<td>7</td>
</tr>
<tr>
<td>Age (year)</td>
<td></td>
</tr>
<tr>
<td>Mean</td>
<td>39.2</td>
</tr>
<tr>
<td>Median</td>
<td>43</td>
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<tr>
<td>SD</td>
<td>15.8</td>
</tr>
<tr>
<td>Range</td>
<td>18-57</td>
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<tr>
<td>Duration of illness (year)</td>
<td></td>
</tr>
<tr>
<td>Mean</td>
<td>2.7</td>
</tr>
<tr>
<td>Median</td>
<td>2.9</td>
</tr>
<tr>
<td>SD</td>
<td>1.5</td>
</tr>
<tr>
<td>Range</td>
<td>1-5</td>
</tr>
</tbody>
</table>

Table 2. EAT-10 and Exercise Performance Before and After Rehabilitation

<table>
<thead>
<tr>
<th></th>
<th>Pre Mean (SD)</th>
<th>Post Mean (SD)</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>EAT-10</td>
<td>11.1 (9.4)</td>
<td>2.2 (2.6)</td>
<td>0.002</td>
</tr>
<tr>
<td>Shaker Isometric (second)</td>
<td>14 (6.1)</td>
<td>27.5 (4.2)</td>
<td>0.002</td>
</tr>
<tr>
<td>Shaker Isotonic (repetition)</td>
<td>12.7 (6.9)</td>
<td>20.7 (7.7)</td>
<td>0.002</td>
</tr>
</tbody>
</table>

Figure 2. FEES Before and After Rehabilitation
DISCUSSION

This study, to conduct exercise for the suprathyroid muscles in Myasthenia Gravis patients, is the first as far as we know. The ten patients who participated in this study had already had regular pharmacology therapy for more than 6 months. Most of them had been taking the medicine for years. Baseline characteristics showed classic patterns of MG epidemiology.3

The baseline FEES result showed residue in all patients regardless of their dysphagia complaint. Residue remaining in the pharynx region will lead to an increased risk of post swallow aspiration.9 FEES after an exercise period showed significant improvement in residue outcomes, most of the patients no longer had residue in post exercise FEES. Three patients still have residue remaining, which is probably due to the severity and longer duration of their illness. The EAT-10 questionnaire showed significant improvement after 6 weeks of exercise. Most of them have an EAT-10 score of 3 or below (the cut off point for an abnormal score).10 This is a promising finding regarding a Myasthenia Gravis patients’ quality of life.

Research investigating the benefits of exercise for people with MG is limited. Few studies suggested that lower limb strengthening and low impact aerobic exercise programmes may be beneficial to people with mild to moderate MG.7 This study shows a similar conclusion for the swallowing muscles.

Traditional Shaker Exercise is aimed at strengthening the suprathyroid muscles including the geniohyoid, thyrohyoid, and digastric muscles. Our study modified this because of the pathophysiological processes of MG. The initial holding time and repetition was determined by patient tolerance as described above (description of exercise section).6,8 Initial holding time and repetition (Table 2) suggests that pharmacology treatment did not prevent muscle strength deterioration because all patients have a low baseline exercise tolerance (low holding time and repetition). At the end of six weeks, there’s significant improvement in holding time (isometric) and repetition (isotonic). This data shows muscle strength can be improved with exercise without making the disease worse.7,11

Physiologically, six weeks of strengthening exercise will increase neural adaptation in muscle recruitment that leads to an increase in muscle strength. However, none of the patients were able to achieve this with the traditional Shaker Exercise dosage in six weeks (Table 2). The data shows that all the patients have a low intensity load. This finding suggests that the dysphagia improvement is more likely to be caused by peripheral adaptation of endurance rather than strengthening adaptation.12 The physiological effects, such as an increased number and density of mitochondria, increase in skeletal muscle mass and size of capillary beds, and improved lactate degradation may improve the muscles ability to cope with fatigue due to the pathophysiological processes of MG.13, 14 Further research is needed to determine the optimal exercise duration for Myasthenia Gravis patients.

CONCLUSION

Shaker Exercise can be used safely by Myasthenia Gravis patients in order to improve dysphagia. It improves a patients quality of life and it appears to reduce the risk of aspiration by reducing post swallow residue. Further research is needed to investigate the optimal exercise dosage and long term maintenance effect.

REFERENCES


