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The Changes in Muscle Strength and Relaxation Time after a Comprehensive Rehabilitation Program for Patients with Myotonic Dystrophy

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Myotonic dystrophy is a muscular disorder characterized by muscle weakness and myotonia. Myotonia manifests with abnormally slow relaxation after strong voluntary contraction of the muscles. In our previous study we reported that quinine sulfate provided therapeutic benefit to myotonia and a home exercise program based on muscle strengthening exercises improved muscle strength. The purpose of this study was to determine the effect of a multi-therapeutic program in patients with myotonic dystrophy. For six months, seven patients with myotonic dystrophy received heat therapy, were given psychologic intervention using relaxation techniques, were trained at home, and were given quinine sulfate. The changes in muscle strength and relaxation time between the post-six-months home exercise program combined with quinine sulfate therapy, and the post-six months multi-therapeutic program, were assessed from the first dorsal interossei, the elbow flexors, and the knee extensors. The results were as follows:

1) The mean muscle strength of the each of the three muscles after the six months multi-therapeutic program was improved but was not significant compared with the post-six-months home exercise program combined with quinine sulfate therapy.

2) The mean relaxation time of each of the three muscles after the six months multi-therapeutic program was significantly reduced compared with the home exercise program combined with quinine sulfate therapy. In conclusion, the multi-therapeutic program undertaken in this study was the better program for the patients with myotonic dystrophy.

Key Words: Myotonic dystrophy, muscle strength, relaxation time, heat therapy, relaxation technique, exercise, quinine sulfate

Myotonic dystrophy is a muscular disorder characterized by muscle weakness and myotonia. Myotonia manifests with abnormally slow relaxation after strong voluntary contraction of the muscles. The patients with myotonic dystrophy, therefore, have difficulty in performing daily activities. The degree of myotonia can be quantified by measuring relaxation time. Relaxation time means the time taken for maximum voluntary contraction to decrease by 100% (Millner-Brown and Miller, 1990). Millner-Brown and colleagues reported that relaxation time measured at the first dorsal interossei of 12 patients with myotonic dystrophy was significantly prolonged. The muscle strengths measured at the first dorsal interossei, the elbow flexors, and the knee extensors of the patients were significantly reduced compared with healthy subjects, and

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amitriptyline therapy (in combination with muscle strengthening exercises) may increase muscle strength and decrease myotonia (Millner-Brown and Miller, 1990).

In another study, Ricker and colleagues showed that the relaxation time of the abductor pollicis muscles of four myotonic dystrophy patients was prolonged (Ricker *et al.* 1977). In our previous study, we reported that the muscle strengths of the first dorsal interossei, the elbow flexors, and the knee extensors of the patients with myotonic dystrophy were significantly reduced, and that the relaxation times of the three muscles of these patients were significantly prolonged compared with healthy subjects (Lee *et al.* 1995). We also showed that quinine sulfate provided therapeutic benefit to myotonia, and that a home exercise program based on muscle strengthening exercises improved muscle strength (Lee *et al.* 1995; Na *et al.* 1996).

The symptom of myotonia is influenced by various factors such as the external temperature, and the physical and psychological conditions of the patients (Engel and Franzini-Armstrong, 1994). We thought, therefore, that heat therapy and psychologic intervention, as well as drug therapy and exercise program, would provide therapeutic benefit to the patients with myotonic dystrophy. The purpose of this study was to determine the effect of heat therapy and psychologic intervention using relaxation techniques for the symptom of myotonia, and to establish a comprehensive rehabilitation program for the patients with myotonic dystrophy.

MATERIALS AND METHODS

The subjects were 7 patients (5 male and 2 female) ranging in age from 17 to 49 years, who were diagnosed with myotonic dystrophy both by clinical manifestations and electrodiagnostic studies. For six months, the seven patients were given one gram of quinine sulfate daily, and were given weight-training at home with weights adapted to their lifting abilities or with no weights at all. And then for one month, the subjects received no therapy in order to eliminate the accumulative effect of drug or exercise.

Finally, for the next six months, the patients received a multi-therapeutic program (heat therapy, psychologic intervention using relaxation techniques, daily training at home with weights adapted to their lifting abilities or without any tools at all, and one gram of quinine sulfate daily). The patients' muscle strengths and relaxation times before therapy, six-months after the home exercise program combined with quinine sulfate therapy, and six-months after the multi-therapeutic program, were assessed from the first dorsal interossei, the elbow flexors, and the knee extensors.

Heat therapy

Hot pack and ultrasound were applied to the hand, the elbow, and the knee joint area of the patients three times a week for 20 minutes per session during the 6 months. We used commercial hot packs for application. The canvas cases (packs) were heated in the water inside a thermostatically controlled cabinet (71.1℃). The canvas cases (packs) were wrapped in 6 to 8 layers of towels before the application. The intensity of ultrasound was 3 watts per cm^2 with a total output of 30 watts, and was applied with a moving applicator.

Psychologic intervention using relaxaton techniques

The patients received psychologic intervention using relaxation techniques-introduced by a psychologist experienced in relaxation techniques-twice a week for 20 to 30 minutes per session during the 6 months. The patients were instructed to contract and relax all of the major muscle groups progressively from the feet to the facial muscles, leading eventually to a completely relaxed state. During the relaxation therapy, which involved substantial dialogue between patient and therapist, patients frequently referred to personal experiences of tension, anxiety and/or stress within their daily lives. If these experiences were judged to be affecting the effectiveness of in-session training, cognitive-behavioral interven-

tions were undertaken to resolve the related issues.

Home exercise program

Each patient trained daily at home with weights adapted to his/her lifting abilities or without any tools at all for six months. The intensity of exercise was adjusted to each patient's ability: a weight was selected with which the patient could perform a maximum of 10 repetitions. The patients performed isometric exercises using a tennis ball and a grip exerciser for strengthening of the hand muscles including the first dorsal interossei. For the elbow flexors and extensors, and the knee flexors and extensors, isotonic exercises with the dumbells and sand bags were performed. Squatting exercises for strengthening the entire lower extremity muscles and heel - toe rising exercises for strengthening the ankle dorsiflexors and plantar flexors were performed. The weights of the dumbells and the sand bags were adjusted to each patient's ability and ranged from 2 kg to 10 kg. The weight was progressively increased if the patient could do more than 10 repetitions per set. The initial number of repetitions with weights was 60 per day and was increased progressively.

Measurement of muscle strength and relaxation time

The muscle strength and relaxation time of the first dorsal interossei, the elbow flexors, and the knee extensors were measured by a strain gauge; axis muscle tester[a].

First dorsal interossei: The patient was comfortably seated. The forearm of the patient was strapped to the examining table and the hand and fingers were securely fixed with tapes. After the sensor of the force transducer of the axis muscle tester was attached to the distal part of the second finger, three second maximum voluntary contractions (second finger abduction) were performed. Muscle strength was quantified using the maximum force generated in the three seconds, with the aid of a loud oral support. The greater of the two maximum force measurements was recorded as muscle strength in pounds (lbs) (Fig. 1). Relaxation time was recorded as the time taken for the maximum voluntary contraction to decrease by 100% (Fig. 1).

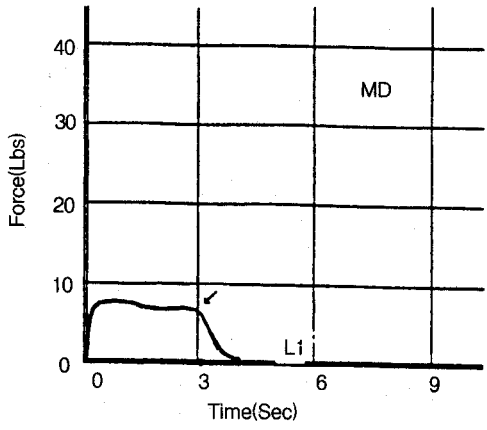

Fig. 1. *Muscle strength and relaxation time of the patient with myotonic dystrophy(MD). Arrow indicates starting point of relaxation.*

Elbow flexors: The patient was comfortably seated, with the elbow held in a specially designed device to ensure that the angle between the upper arm and the forearm was maintained at 90 degrees. Patient's upper arm was strapped to a wooden frame, which formed a part of the elbow support. After the sensor was attached to the ventral aspect of the distal forearm, three seconds of maximum voluntary contractions (elbow flexion) were performed. Muscle strength was quantified using the maximum force generated in the three seconds, with the aid of a loud oral support. The greater of the two maximum force measurements was recorded as muscle strength in pounds (lbs) (Fig. 1). Relaxation time was quantified using the time taken for the maximum voluntary contraction to decrease by 100% (Fig. 1).

Knee extensors: The patient was seated on an examining table with attached hand grips. The pelvis was securely fixed with a seat belt strap. The hip and knee joint angles were held at 90 degrees. After the sensor was attached to the dorsal aspect of the distal leg, three seconds of maximum voluntary contractions (knee extension) were performed. Muscle strength was quantified using the maximum

force generated in the three seconds, with the aid of a loud oral support. The greater of the two maximum force measurements was recorded as muscle strength in pounds (lbs) (Fig. 1). Relaxation time was quantified using the time taken for the maximum voluntary contraction to decrease by 100% (Fig. 1).

Statistics

The Wilcoxon signed-ranks test was used to compare the changes in muscle strength and relaxation time between pretherapy and the home exercise program combined with quinine sulfate and the heat therapy combined with the psychologic intervention. If the calculated p-values were less than 0.05, the changes were considered statistically significant.

RESULTS

Muscle strength

Table 1 compares the mean (± SD) muscle strength of 7 patients between pretherapy and the post-six-months home exercise program combined with quinine sulfate therapy and the post-six-months multi-therapeutic program; 1) The mean muscle strength of the each of the three muscles after the six-months multi-therapeutic program was improved but was not significant compared with the post-six-months home exercise program combined with quinine sulfate therapy (Table 1). 2) The mean muscle strength of the each of three muscles after the six-months home exercise program combined with quinine sulfate therapy was significantly improved compared with pretherapy ($p<0.05$, Table 1).

Relaxation time

Table 2 compares the mean (± SD) relaxation time of 7 patients between pretherapy and the post-six-months home exercise program combined with quinine sulfate therapy and the post-six months multi-therapeutic program; ① The mean relaxation times of each

Table 1. Comparison of effects on muscle strength(1bs.)

	Pretherapy(n=7)		Q+E(n=7)		Q+E+H+P(n=7)	
	Right	Left	Right	Left	Right	Left
1st DI	4.9± 1.8	4.6± 2.2	6.0± 1.4*	5.7± 1.2*	6.1± 1.7	6.0± 1.3
Elbow flexor	22.6± 9.7	21.9±10.9	32.6±11.0*	31.7±11.7*	34.1±10.0	31.9±11.5
Knee extensor	44.3±34.8	45.3±30.7	62.3±35.2*	58.3±29.6*	63.0±32.4	58.4±28.3

Values are means±S.D.(1bs). *: $p<0.05$
1st DI: First dorsal interossei
Q: Quinine sulfate, E: Home exercise program, H: Heat therapy
P: Psychologic intervention using relaxation techniques

Table 2. Comparison of effects on relaxation time(sec.)

	Pretherapy(n=7)		Q+E(n=7)		Q+E+H+P(n=7)	
	Right	Left	Right	Left	Right	Left
1st DI	0.93±0.17	0.80±0.12	0.55±0.16*	0.51±0.11*	0.46±0.17*	0.45±0.13*
Elbow flexor	1.18±0.47	1.15±0.54	0.82±0.46*	0.91±0.54*	0.71±0.32*	0.79±0.42*
Knee extensor	1.49±0.65	1.38±0.53	0.68±0.22*	0.93±0.33*	0.57±0.21*	0.76±0.34*

Values are means±S.D.(sec). *: $p<0.05$
1st DI: First dorsal interossei
Q: Quinine sulfate, E: Home exercise program, H: Heat therapy
P: Psychologic intervention using relaxation techniques

of the three muscles after the six months multi-therapeutic program was significantly reduced compared with the post-six-months home exercise program combined with quinine sulfate therapy ($p<0.05$, Table 2). ② The mean relaxation times of each of the three muscles after the six-months home exercise program combined with quinine sulfate therapy was significantly reduced compared with pretherapy ($p<0.05$, Table 2).

DISCUSSION

Muscle weakness and myotonia, which are the characteristic manifestations of myotonic dystrophy, have different pathophysiology: Atrophic fibers, internal nuclei, and occasional necrotic fibers are not infrequent in muscle biopsies from patients with myotonic dystrophy; this muscle fiber degeneration leads to progressive weakness (Brumback *et al.* 1981). Bryant suggests that a reduction of chloride permeability of the muscle membrane could be the basis for myotonia (Bryant, 1962; Bryant and Morales-Aguilera, 1971). Engel hypothesized that increased sodium concentration inside the cell caused by non-physiological opening of the sodium channel could be the pathophysiology (Engel and Franzini-Armstrong, 1994). Myotonic dystrophy is a disorder inherited by autosomal dominant trait and frequently shows various systemic manifestations. No drug is currently available for clinical use that alters the permeability of the myotonic muscle membrane to chloride or sodium. However, quinine sulfate, diphenylhydantoin, procainamide, carbamazepine, n-propyl-ajmaline, and amitriptyline are sometimes useful in alleviating myotonia (Birnberger *et al.* 1975; Adams and Victor, 1977; Millner-Brown and Miller, 1990). In our previous study, quinine sulfate was effective for myotonia (Lee *et al.* 1995). Quinine increases the refractory period of the muscle so that the response to tetanic stimulation is diminished. The excitability of the motor end-plate region decreases so that responses to repetitive nerve stimulation and to acetylcholine are reduced (Gilman *et al.* 1985). A home exercise program based on muscle strengthening exercises showed significant improvements in muscle strength for patients with myotonic dystrophy. Muscle strength of the first dorsal interossei was significantly improved by isometric exercises. Muscle strengths of the elbow flexors and the knee extensors were significantly improved by isotonic exercises. In an isometric exercise, strength gain occurs mainly at the angle at which the exercise is done (Lindh, 1979). DeLorm suggested that isotonic exercises using progressive resistance with ten repetitions at 25%, 50%, 75%, and 100% of the maximum capability increases muscle strength (DeLorme and Watkins, 1948). The body itself can be used as a resistance by moving the body against gravity in such techniques as push-ups, sit-ups, and chin-ups (DeLisa and Gans, 1993). Squatting exercises and heel - toe rising exercises performed in this study, therefore, could be beneficial for the strengthening of the lower extremity muscles - especially the ankle dorsiflexors and the plantar flexors.

Hot packs and ultrasound are therapeutic heating agents widely used for many therapeutic purposes. Hot packs elevate the temperature of the skin and tissues within 1~2 cm from the surface. But tissues below the adipose tissue are minimally affected by such superficial heating agents as hot packs. Ultrasound elevates the temperature of the tissues below the adipose tissue such as the muscles, ligaments, and tendons. Biophysical effect of temperature elevation can be summarized as follows: First, the neuromuscular effect; Resolution of muscle spasm and analgesic effect can be seen. Second, the connective tissue effect; temperature elevation alters viscoelastic properties of connective tissues. So the elasticity of the connective tissues increases. Third, the metabolic reactions; Chemical activities in the cells and the metabolic rate increases twofold to threefold for each 10℃ rise. Increased chemical reactions have a positive effect on human function, and increased oxygen uptake helps tissue healing. Fourth, the vascular effect; Vasodilation of heat-exposed skin occurs (Michlovitz, 1990).

Relaxation refers to the lengthening of the skeletal muscle fibers, while tension refers to

the contraction or shortening of the muscle fibers. Relaxation therapy may be used to decrease anxiety, to promote sleep, to reduce pain or the perception of pain, to reduce or prevent the physiological and psychological effects of stress, to alleviate muscle tension, and to decrease blood pressure (Zahourek, 1988). We concluded that the heat therapy and the psychologic intervention using relaxation techniques practiced in this study were effective in relaxing the muscles of the patients with myotonic dystrophy and in reducing their relaxation time. In conclusion, heat therapy and psychologic intervention using relaxation techniques combined with a home exercise program and a quinine sulfate therapy showed significant improvements in the muscle strength and the shortening of relaxation time (decreased myotonia) for the patients with myotonic dystrophy. We propose that the comprehensive rehabilitation program undertaken in this study provide therapeutic benefit to the patients with myotonic dystrophy.

SUPPLIERS

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