Myotonia Not Aggravated by Cooling

Force and Relaxation of the Adductor Pollicis in Normal Subjects and in Myotonia as Compared to Paramyotonia

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Summary. The effect of local cooling has been studied in 27 normal subjects, 8 cases of myotonia congenita, 5 of myotonic dystrophy and one of paramyotonia. Using the adductor pollicis we registered the compound muscle action potential, the isometric twitch force and the time to half relaxation, the maximum tetanic force and the time to \( \frac{3}{4} \) relaxation.

1. In normal subjects the twitch force and maximum tetanic force decreased after cooling (Fig. 2). The amplitude of the action potential increased.

2. Myotonia congenita and myotonic dystrophy were not aggravated by cooling. Muscle force was reduced only in the same proportion as in normal subjects (Fig. 2). The myotonic after-contraction was made normal by cooling (Figs. 5 and 6).

3. In paramyotonia initial tonic stiffness with a pronouncedly prolonged twitch relaxation occurred directly after cooling (Fig. 1B). Paradoxical myotonia occurred only after exercise and was accompanied by increasing paresis (Figs. 3 and 8).

The results indicate that exposure to cold has a specific effect on muscle function only in paramyotonia.

Key words: Muscle cooling – Twitch force – Muscle relaxation – Myotonia congenita – Paramyotonia – Myotonic after-contraction.

Zusammenfassung. Der Einfluß lokaler Muskelkühlung wurde an 27 Kontrollpersonen, 8 Patienten mit Myotonia congenita, 5 mit myotoner Dystrophie und 1 mit Paramyotonie untersucht. Wir registrierten am Adductor pollicis das Summenaktionspotential, die isometrische Muskelzuckung und die Zeit bis zur halben Erschaffung, die maximale tetanische Muskelkontraktion und die Zeit bis zur \( \frac{3}{4} \)-Erschaffung.

1. Bei den Kontrollpersonen wurde die Kraft der Einzeltenszuckung und die maximale Kraft nach Kühlung geringer (Abb. 2). Die Amplitude des Aktionspotentials wurde größer.


Die Befunde zeigen, daß nur bei Paramyotonie Kühlung eine spezifische Auswirkung auf die Muskelfunktion hat.

Introduction

Exposure to cold leads to a stiffness of movement and to a muscular paresis in patients with paramyotonia. The weakness develops particularly rapidly with muscular exercise performed after cooling. The paresis remains even after the muscle is warmed again.

The effect of local cooling on myotonia congenita and myotonic dystrophy is not completely clear. Walton and Gardner-Medwin (1974) remark: “Myotonia in all its forms is always made worse by cold and this criterion is insufficient to distinguish paramyotonia from myotonia congenita.” Becker (1973) writes about myotonia congenita: “Exposure to cold temperatures increases the symptoms of myotonia in about half the cases.” Bryant (1973) observed a noticeable increase in stiffness of movement in myotonic goats out of doors during cold weather. In these animals local heat applied by a lamp decreased the duration of the percussion response; local cooling prolonged it.

We tried to determine, in a group of myotonic patients, the influence of cold on muscle force and on active myotonia. For comparison, normal subjects and one patient with paramyotonia were examined. The unexpected result was that active myotonia in patients with myotonia congenita and myotonic dystrophy did not worsen after exposure to cold.

Methods

Isometric Contraction of the Adductor Pollicis by Stimulating the Ulnar Nerve (Slomič et al., 1968; Desmedt, 1973). The muscle action potential (MAP) was registered by subcutaneous belly-tendon electrodes. The isometric contractions were recorded by attaching the thumb to a force transducer (Shinkoh LD/10 KA); the initial tension was adjusted to 100—150 g.

The ulnar nerve was blocked with 4 ml of 2% Scandicain at the elbow. The nerve was stimulated at the wrist with 2 steel needles. The stimulus intensity was definitely supramaximal at any given temperature; stimulus duration 0.1 msec. We used single volleys and tetanic stimulation with a frequency of 50/sec for 1 or 1.5 sec. With this frequency Merton (1954) obtained maximum tetanic contraction using the adductor pollicis. This still lies in the upper range of the physiological discharge frequency of the motor units. In some cases muscle exercise was achieved by stimulating the nerve with 20/sec for 30 or 60 sec.

We measured the intramuscular temperature with a thermo EMG needle. The correct site of the electrode was checked by the shape of the MAP. Temperature unit DISA 14 G 05,