

Relative Fatigability of Muscles in Response to Repetitive Nerve Stimulation in Myasthenia Gravis

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Background: Repetitive nerve stimulation (RNS) plays an important role in the diagnosis of myasthenia gravis (MG). Technically, this test can be more easily performed on distal muscles than proximal muscles. On the other hand, proximal muscles have been shown to be more easily fatigued or more sensitive than the distal ones.

Objective: Evaluate the relative fatigability of different muscles in response to RNS in ocular and generalized MG patients.

Material and Method: Two hundred patients with 44 ocular MG and 156 generalized MG were studied. The relative fatigability of each muscle was calculated as percentage of cases with abnormal response. A decrement of 10% or more of the amplitude of the compound muscle action potentials was considered abnormal.

Result: The cases with abnormal response of adductor digiti minimi, anconeus, trapezius, nasalis and orbicularis oculi were 17%, 50%, 55%, 62%, 66% in ocular MG and 53%, 77%, 79%, 85%, 79% in generalized MG respectively. These results showed that in generalized MG, the response in all the muscles was more frequently abnormal and was fatigable in a more comparable degree, whereas in ocular MG, the facial muscles were more often abnormal than the limb muscles.

Conclusion: The abnormal response because of fatigability was more widespread in generalized MG, whereas facial muscles were relatively more sensitive in ocular MG. This finding may be useful for the selection of the muscles to be tested in RNS especially in ocular MG.

Keywords: Myasthenia gravis, Repetitive nerve stimulation, Fatigability

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Repetitive nerve stimulation (RNS) is a reliable and noninvasive electrophysiological test for evaluation of neuromuscular transmission in patients with myasthenia gravis (MG). Technically, this test can be more easily performed on distal muscles than proximal muscles. On the other hand, proximal muscles have been shown to be more easily fatigued or more sensitive than the distal ones correlating with the clinical features of this disease⁽¹⁻³⁾. The purpose of the present study was to evaluate the electrophysiological response and relative fatigability of different muscles in these patients. This information may help choose the

muscles to be tested in RNS for patients with MG of different severity.

Material and Method

Results of RNS in patients with suspected MG in the presented laboratory from January 1999 to December 2003 and the clinical information of these patients were retrospectively reviewed. Only the patients with abnormal RNS were included in the present study. The diagnosis of MG was made by clinical features and response to anticholinesterase drugs. Anticholinesterase medication was withheld for at least 12 hours before testing. The patients were clinically grouped into ocular and generalized MG. A Dantec Keypoint EMG machine was used in the present study. In all the patients, the RNS was performed by

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stimulating supramaximally the radial nerve just above the elbow, facial nerve in front of the ear and accessory nerve at the neck behind the sternocleidomastoid muscle at 3 Hz (0.2 msec duration) with recording electrode on anconeus, orbicularis oculi, nasalis and trapezius muscles respectively. At the early stage of the present study period, the abductor digiti minimi (ADM) was also studied by stimulating the ulnar nerve at the elbow. Ten trains of 10 stimuli each were delivered to each nerve with a brief rest between each successive train. Post exercise stimulation was not performed in these patients. The peak-to-baseline amplitude of compound muscle action potentials (CMAP) was automatically measured and the percentage decrements of amplitude of the fourth CMAP compared to the first were automatically calculated. This rate of stimulation may produce a decremental response of up to 8% in normal muscles⁽⁴⁾. To avoid a false-positive result, decrement of 10% or more is usually considered abnormal⁽⁵⁾. This criterion was used in the present study and each muscle was considered as abnormal if one or more of the trains showed decremental response in each muscle. The relative fatigability of each muscle was calculated as percentage of cases with abnormal response. Accordingly, relative sensitivity of different muscles to the test can be estimated.

Results

There were two hundred patients with 44 ocular and 156 generalized MG. In ocular MG, there were 10 men and 34 women with the age ranging from 17-64 years (mean = 41.8) and 15-68 years (mean = 38.9) respectively. In generalized MG, there were 36 men and 120 women with the age ranging from 16-89 years (mean = 42.5) and 15-79 years (mean = 39.8) respectively. The patients with abnormal response of ADM, anconeus, trapezius, nasalis and orbicularis oculi were 17%, 50%, 55%, 62%, 66% of the cases of ocular MG and 53%, 77%, 79%, 85%, 79% of the cases of generalized MG respectively (Fig. 1).

Discussion

The present study is different from other studies in that only the patients with abnormal response of RNS were included. Therefore, the sensitivity of the test cannot be determined from the present study. It has been shown that RNS is less sensitive in ocular MG than in generalized cases. The sensitivity was about 17-20% in ocular cases and about 55-85% in generalized cases depending on the severity and muscles tested with facial and proximal muscles providing good diagnostic yield⁽⁶⁻⁸⁾. The purpose of the present study was to compare the relative fatigability

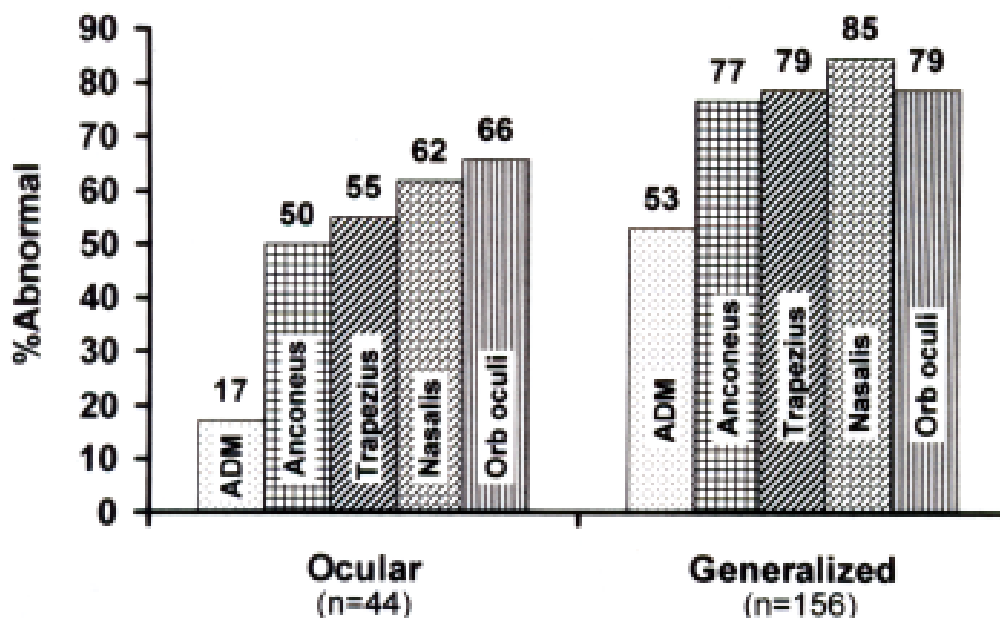


Fig. 1 Relative fatigability of different muscles in response to repetitive nerve stimulation in 200 patients with myasthenia gravis (44 ocular and 156 generalized)

of different muscles tested. These results showed that in generalized MG, the response was more frequently abnormal in all the muscles and was fatigable in a more comparable degree with nasalis being the most sensitive followed by orbicularis oculi, trapezius, and anconeus in equal fatigability. In ocular MG, the orbicularis oculi and nasalis muscles were more often abnormal than the limb muscles. ADM was the least sensitive in either group. This finding confirmed that the fatigability of these muscles in generalized MG was more marked and more widespread compared to that in ocular cases in whom facial and ocular muscles were more fatigable correlating with clinical involvement. This information may be useful for the selection of the muscles to be tested in RNS especially in ocular MG.

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การเปรียบเทียบความล้าของกล้ามเนื้อต่างๆในการตอบสนองต่อการกระตุ้นเส้นประสาทซ้ำๆ ในผู้ป่วยมีัยแอสทีเนียแกรวิส

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การตรวจกล้ามเนื้อด้วยการกระตุ้นเส้นประสาทซ้ำๆ เป็นการตรวจที่สำคัญในการวินิจฉัยผู้ป่วยมีัยแอสทีเนียแกรวิส ในด้านเทคนิคการตรวจนี้ทำได้ง่ายในกล้ามเนื้อส่วนปลายมากกว่าส่วนต้น แต่ในด้านผลการตรวจกล้ามเนื้อส่วนต้นมักจะให้ผลที่ผิดปกติมากกว่า จุดประสงค์ของการศึกษานี้คือ การประเมินความล้าของกล้ามเนื้อต่างๆ ในการตอบสนองต่อการกระตุ้นเส้นประสาทซ้ำๆ ในผู้ป่วยมีัยแอสทีเนียแกรวิสในกลุ่มที่เป็นเฉพาะที่ตาและกลุ่มที่เป็นทั่วตัว ขนาดของคลื่นกล้ามเนื้อที่ลดลงตั้งแต่ร้อยละ 10 ขึ้นไปถือว่าผิดปกติ การเปรียบเทียบความล้าจากอัตราของผู้ป่วยที่ให้ผลผิดปกติในกล้ามเนื้อแต่ละมัด มีผู้ป่วยทั้งหมด 200 คน เป็นเฉพาะที่ตา 44 คนและเป็นทั่วตัว 156 คน ผลการตรวจพบว่า ในผู้ป่วยกลุ่มที่เป็นเฉพาะที่ตา กล้ามเนื้อ adductor digiti minimi, anconeus, trapezius, nasalis และ orbicularis oculi ให้ผลผิดปกติในอัตราร้อยละ 17, 50, 55, 62 และ 66 ของผู้ป่วยตามลำดับ ส่วนในผู้ป่วยกลุ่มที่เป็นทั่วตัว กล้ามเนื้อเหล่านี้ผิดปกติในอัตราร้อยละ 53, 77, 79, 85 และ 79 ของผู้ป่วยตามลำดับ ผลการตรวจนี้แสดงว่า ในกลุ่มที่เป็นทั่วตัว กล้ามเนื้อทั้งหมดที่ตรวจให้ผลผิดปกติบ่อยกว่ากลุ่มที่เป็นเฉพาะที่ตา และในอัตราที่พอๆ กัน ในกล้ามเนื้อส่วนใหญ่ ส่วนในกลุ่มที่เป็นเฉพาะที่ตานั้น กล้ามเนื้อที่หน้าและตาให้ผลผิดปกติบ่อยกว่ากล้ามเนื้อของแขนขา ผลที่ได้นี้อาจมีประโยชน์ในการช่วยเลือกกล้ามเนื้อที่จะตรวจในผู้ป่วยแต่ละกลุ่ม