

# EVALUATION OF REPETITIVE STIMULATION TEST (RST) IN 30 PATIENTS WITH MYASTHENIA GRAVIS, WHO WERE PREVIOUSLY CONFIRMED BY CLINICAL SIGN AND TENSILON TEST 1996-1999

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**Abstract** - Jolly Test (RST) is the most commonly used electrodiagnostic test to assess the defect of neuromuscular transmission, which is reported to be positive in the diffuse and restricted ocular forms 60-95% and 14-50%, respectively. In a cross-sectional study, to determine the efficacy of repetitive stimulation test in myasthenia gravis, we evaluated the results in 30 cases who were hospitalized in Imam Khomeini Hospital during 1996-1999. Patients were first selected clinically and then confirmed by Tensilon test.

Various clinical types including generalized and restricted ocular forms with different severity and duration were entered in this study. Considering the fact that the positiveness of the test is enhanced by assessment of more muscle groups, we evaluated decremental response in the facial, proximal and distal muscles of limbs. 90% of patients had the generalized form of the disease, whereas ocular myasthenia gravis was seen only in 10% of the cases. 74% of females and 73% of males showed positive response (overall: 73.3%). No significant association was found between the positive response, and age and sex. Peaks of incidences of the disease for the males were in fourth and sixth decades and for the females in third decades.

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**Key Words:** Jolly test (RST), myasthenia gravis

## INTRODUCTION

Myasthenia gravis is an acquired autoimmune disease associated with a fault in neuromuscular transmission, resulting in muscle weakness (1,2). Its prevalence was reported to be 14 to 125 cases per million (3,4). Before age 40, the disease is three times more common in women but at older ages both sexes are affected equally. However, it has been recently found to be more common after the age 50 in men. The age of onset shows a bimodal pattern with 2nd and 3rd decades in women and 6th and 8th in men. Ocular

involvement is the initial sign in 50%, but ultimately ocular weakness occurs in up to 90% of the cases (5,6,8). Weakness gets aggravated with increase in body temperature and gets improved with cold (7,8,9). In 16% of the cases the symptoms of the disease remain restricted to ocular muscles. Serum antibodies against nicotinic acetylcholine receptors is positive in 70-80% of generalized and 50% of ocular types (10,11,12). According to Martegazza, the prognosis is worse in those associated with thymoma and those above 40 (13). Muscle biopsy reveals decrease of acetylcholine receptors and widening of synaptic cleft (14,15,16,17). Tensilon test, muscular fatigability, Mary-Walker phenomenon (18), ice test (19,20) and stapedius reflex (21,22), measurement of striated muscle and acetylcholine receptor antibodies as well as sleep and electrodiagnostic tests are used in diagnosis of the disease.

Jolly test (repetitive stimulation with 2-5 Hz) is among the simplest, most commonly used, noninvasive and necessary electrophysiological method for evaluation of neuromuscular transmission. Repetitive stimulation test is reported to be positive (decrement more than 10% in amplitude, the drop in amplitude is graduated and very often takes the form of the letter U) in 60-95% of generalized myasthenia and 14-50% of restricted ocular type (23,24). Evaluation of distal and proximal muscles, increasing the temperature and testing under ischemic condition (double step technique) increase the chance of positive response (25,26). It is better to stimulate each muscle twice with an interval of 30 seconds.

## MATERIALS AND METHODS

Aims of the study were:

A) Evaluation of the response in repetitive stimulation test in myasthenia gravis;

and B) association between the response and sex, age, initial symptoms, severity of the disease, and thymic pathology.

In this cross sectional study among 47 patients, admitted in the neurology ward of Imam Khomeini Hospital, 30 cases with positive Tensilon test, clinical criteria and without any evidence of respiratory distress were entered into the study. The test was performed 12 hours after discontinuation of anticholinesterase.

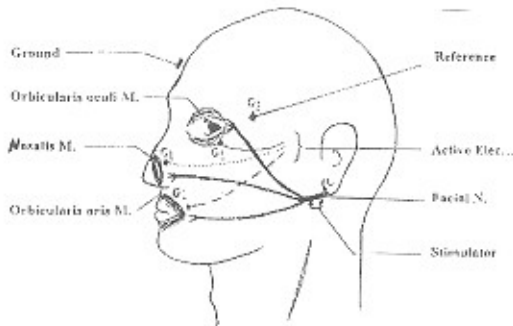


Fig. 1. Position of electrodes on facial muscles

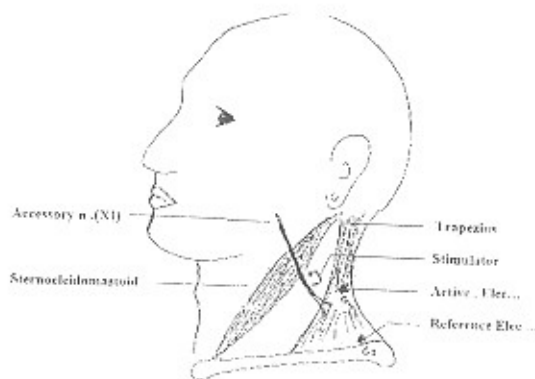


Fig. 2. Position of electrodes on trapezius

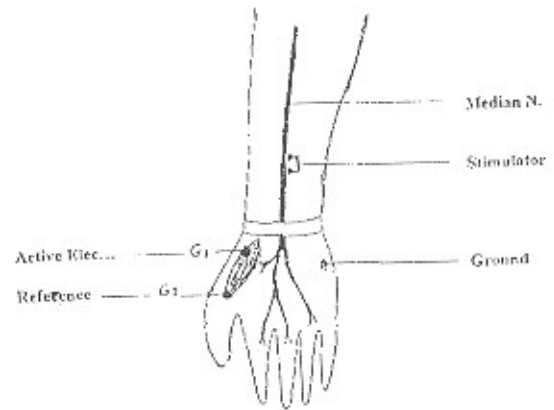


Fig. 3. Position of electrodes of abd. pollicis. brevis

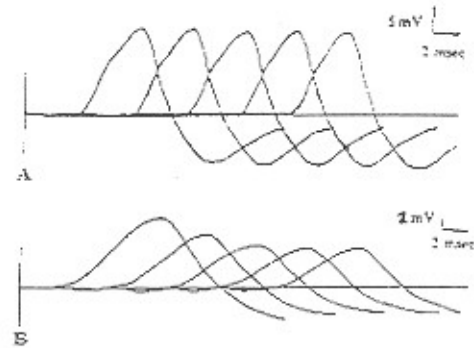


Fig. 4. A: Normal response , B: Decremental response

Table 1. Clinical feature in 30 patients with myasthenia gravis in Imam Khomeini Hospital during 1996-1999

Type	Gender		Total
	Male	Female	
Generalized	9	18	27 (90%)
Ocular	2	1	3 (10%)
Total	11 (36.3%)	19 (63.3%)	30 (100%)

Stimulation was done repetitively three times per second, and each muscle tested twice with 30-60 seconds intervals. The test is defined positive if the

**Table 2.** Age of onset in 30 patients with myasthenia gravis in Imam Khomeini Hospital during 1996-1999

Group	Male		Female		Total	
	Frequency	%	Frequency	%	Frequency	%
10-19	1	9.0	3	15.7	4	13.3
20-29	2	19.0	9	47.6	11	36.8
30-39	3	27.0	3	15.7	6	20.0
40-49	0	0.0	3	15.7	3	10.0
50-59	3	27.0	1	5.3	4	13.3
60-69	1	9.0	0	0.0	1	3.3
70-79	1	9.0	0	0.0	1	3.3
Total	11	100	19	100	30	100

**Table 3.** Severity of disease in 30 patients with myasthenia gravis in Imam Khomeini Hospital during 1996-1999

Severity	Male		Female		Total	
	Frequency	%	Frequency	%	Frequency	%
Mild	3	27.2	2	10.5	5	16.6
Moderate	2	18.2	15	79.0	17	56.7
Severe	6	54.5	2	10.5	8	26.7
Total	11	100	19	100	30	100

**Table 4.** Pathology of thymus in 30 patients with myasthenia gravis in Imam Khomeini Hospital during 1996-1999

Pathology	Male		Female		Total	
	Frequency	%	Frequency	%	Frequency	%
Normal	0	0.0	2	13.3	2	11.2
Thymoma	1	33.3	6	40.0	7	38.8
Hyperplasia	2	66.7	7	46.7	9	50.0
Total	3	100	15	100	18	100

**Table 5.** Percent of positive Jolly test in 30 patients with myasthenia gravis in Imam Khomeini Hospital during 1996-1999

Muscle	Frequency of tested muscle	Frequency of positiveness	% of positiveness
Nasalis	31	17	54.8
Orbicularis oculi	23	8	34.8
Orbicularis oris	24	4	16.7
Trapezius	20	6	30.0
Abd. Pollicis Brevis	25	9	36.0
Hypothenar	7	0	0.0

**Table 6.** Percent of positive Jolly test in different groups of muscles in 30 patients with myasthenia gravis in Imam Khomeini Hospital during 1996-1999

Group of muscles	Frequency of tested muscle	Frequency of positiveness	% of positiveness
Facial	78	29	37.2
Proximal	20	6	30.0
Distal	43	11	26.5

**Table 7.** Results of Jolly test in 30 patients with myasthenia gravis in Imam Khomeini Hospital during 1996-1999

Type of Myasthenia	Positive		Negative		Total
	Frequency	%	Frequency	%	
Generalized	21	77.8	6	22.2	27
Ocular	1	33.3	2	66.7	3
Total	22	73.3	8	26.7	30

**Table 8.** Relationship between positive Jolly test and sex in 30 patients with myasthenia gravis in Imam Khomeini Hospital during 1996-1999

Gender	Results	Positive		Negative		Total
		Frequency	%	Frequency	%	
Male		8	73.0	3	27.0	11
Female		14	74.0	5	26.0	19
Total		22	73.3	8	26.7	30

**Table 9.** Relationship between positive Jolly test and presenting symptoms in 30 patients with myasthenia gravis in Imam Khomeini Hospital during 1996-1999

Presenting symptoms	Results	Positive		Negative		Total
		Frequency	%	Frequency	%	
Diplopia and ptosis		12	80.0	3	20.0	15
Dysphagia		7	87.5	1	12.5	8
Weakness of extremity		3	43.0	4	57.0	7

**Table 10.** Relationship between positive Jolly test and pathology of thymus in 30 patients with myasthenia gravis in Imam Khomeini Hospital during 1996-1999

Pathology of thymus	Results	Positive		Negative		Total
		Frequency	%	Frequency	%	
Normal		1	50.0	1	50.0	2
Hyperplasia		6	67.0	3	33.0	9
Thymoma		5	71.4	2	28.6	7

amplitude of 4th or 5th response shows more than 10% decrement in comparison to the first potential. Regarding the fact that the test is not positive in all muscles of the body, we evaluated at least 3 groups of the muscles (face and distal and proximal of limbs). The points of stimulation and recording of potentials have been shown in figures 1, 2 and 3.

The P-value for tables were calculated using the Fisher exact tests. All statistical analyses were done using the SAS system and S-Plus.

## RESULTS

In half of the patients (15 out of 30), the initial symptoms were ocular. Involvement of the bulbar muscles and weakness of extremities were the next most frequent presentation (8 and 7 cases, respectively). 10% had ocular myasthenia and the remainder had the generalized form of the disease. The disease was more common (63.3%) in the females (Table 1). The peak age was 3rd decade in females and 4 and 6 decades in males (Table 2). Regarding the severity of illness, most patients were ranked as moderate (56.7%) (Table 3). Only 18 cases of our patients underwent thymectomy at this center. The histopathological findings have been thymic hyperplasia (9 out of 18), thymoma (7 out of 18) and normal pattern in two patients (Table 4). Jolly

test was positive in nasalis, abductor pollicis, orbicularis oculi and trapezius in 55%, 36%, 35% and 30%, respectively (Table 5). Positive response was obtained in 37% of facial muscles, 30% proximal and 26.5% distal muscles (Table 6). 77.7% of generalized myasthenia and 33.3% of ocular ones had positive response (overall 73.3%) (Table 7). Positive Jolly test was the same in both sexes (Table 8) but significant difference was detected regarding the presenting symptoms (Table 9).

## DISCUSSION

Although Jolly test has been regarded as the most popular test to confirm the diagnosis of myasthenia gravis, but it has a very low sensitivity and yields too many false negative results when compared with clinical examination and tensilon test. The sensitivity of the test has been reported to be 60-95% and 14-50% in generalized and ocular MG, respectively. Thus our study confirms the above finding, (77% and 33%, respectively).

In this study half of the patients showed ocular involvement as a presenting symptom which is the same as previous reports (4). Positive results were dependent on the pattern of onset, so that patients with onset of

ptosis, diplopia, bulbar palsy and limb weakness showed positive results in 80, 87.5 and 43%, respectively. Therefore, in cases in whom myasthenia initiates from limbs, they are expected to show lower positive response than cases who present with ocular or bulbar symptoms. An interesting finding in our study is that testing of nasalis muscle on both sides increases the chance of positive response, thus we suggest this muscle should be tested bilaterally in all types of the disease. Finally, our study showed that the peak incidence of MG in the females is between 20-30 years, compared to the males with a peak incidence in 30-40 and 50-60 years, indicating a bimodal age distribution of the disease.

The P-value for tables 8, 9 and 10 were  $>0.15$ , indicating no statically significant relationship between variables in these tables (9,10,19).

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