Acta Scientific Ophthalmology (ISSN: 2582-3191)

Volume 5 Issue 10 October 2022

Short Communication

# Tests to Diagnose Ocular Myasthenia Gravis

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**Received:** August 08, 2022

**Published:** September 07, 2022



- Myasthenia gravis is an autoimmune, neuromuscular junction disorder causing fatiguability and weakness of the muscles [1].
- Ocular myasthenia gravis (OMG) is where weakness is isolated to the periocular muscles, namely, extraocular muscles, levator palpebrae superioris and orbicularis oculi [2].
- Extraocular muscles (EOMs) are more commonly affected as they contain twitch fibers.
- These twitch fibres develop tension faster and have a higher frequency of synaptic firing than limb muscles.
- They are more susceptible to fatigue.
- Tonic muscle fibers are needed to maintain the gaze in any direction.
- This type of fiber has fewer acetyl choline receptors, making them prone to receptor loss and damage [3].
- Ptosis and diplopia are usually the presenting signs in these patients [4].
- There is a stronger predilection in males [2,3].

## Sleep test [3]

- Involves the patient sleeping or resting with their eyes closed for 30 minutes.
- Prior to the test, ocular motility and the amount of ptosis present in the eyelids are documented.
- Diagnosis of myasthenia is established when the ptosis or ophthalmoparesis disappears after a 30 minute period of sleep.
- There is a reappearance of myasthenic signs over the next 30 seconds to 5 minutes after the test.
- Rest Test: Shorter version of sleep test where patient gently closes the eyelids for two to five minutes and the ocular motility and ptosis is measured before and after the test.

## Ice pack test [2]

- An icepack is kept over the patient's closed eyelids for a period of 2 min (for ptosis) to 5 min (for ophthalmoparesis).
- The ocular motility deficits and ptosis should be measured before and after the test.
- A positive test is when there is an elevation of the upper eyelid by atleast 2 mm following application of ice.
- Mechanism of action: Cooling reduces acetylcholine esterase activity which increases the amount of available acetyl choline (ACh) available at the neuromuscular junction.
- This promotes the efficiency of ACh in eliciting depolarization at the motor end plate [5].

## Pharmacological tests

### Edrophonium test [2]

- Edrophonium (Tensilon) is a rapidly acting and quickly hydrolyzed anticholinesterase drug.
- They cause an increase in ACh in the synapse and cause maximum saturation of limited available receptor population in myasthenia.

- Ptosis, ocular motility defects must be assessed before and after administering it.
- It is given as 1 to 2mg IV dose along with saline flush and observed for idiosyncratic reaction.

### Neostigmine test [1]

- Neostigmine is a longer acting anticholinesterase inhibitor.
- Alternative to edrophonium.

### Other investigations

### Acetylcholine receptor antibodies [2]

- Elevated AChR-Ab titres are required for confirming the diagnosis.
- Positive in 30-77% of patients with OMG.
- Normal titres do not exclude the disease.

### Electrodiagnostic tests

### Repetitive nerve stimulation [6]

- The nerve to be studied is electrically stimulated 6-10 times at 2 or 3 Hz (slow rate) with a supramaximal stimulus and the compound muscle action potential (CMAP) is recorded with surface electrodes.
- The results were considered abnormal if the amplitude in response to the fifth stimulus compared with the first stimulus showed a decrement of more than ten percent with adequate baseline stability.
- This decremental response is seen in only 33% of patients with purely OMG [7].
- A decremental response to RNS is not specific and may also be seen in Lambert-Eaton myasthenic syndrome, motor neuron diseases, and myopathies.

### Single fibre electromyography [1]

- Most sensitive diagnostic test for detecting abnormal neuromuscular transmission.
- Individual muscle fiber action potentials generated by the same motor neuron are recorded by a specialized concentric needle with a 25 µm diameter recording surface and a 500 Hz high-frequency filter.
- Sensitivity of 85 to 100% for OMG when used on frontalis or orbicularis oculi.
- Sensitivity of 91% in generalised MG.

Additional thyroid function tests to rule out thyroid disorder [8].

### Imaging

- CT Chest- To look for thymic hyperplasia/thymoma [9].

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