

Myasthenia gravis: diagnosis and treatment

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INTRODUCTION

Acquired Myasthenia Gravis (MG) is a very rare disease, with prevalence rates in the US population reaching around 20 per 100,000. Muscle weakness is a symptom of an autoimmune illness, which worsens with exercise and improves with rest. Extrinsic Ocular Muscles (EOMs) involvement is the first symptom in around two-thirds of patients, and it generally progresses to include additional bulbar muscles and limb musculature, resulting in Generalised Myasthenia Gravis (GMG) [1]. About 10% of myasthenia gravis individuals have symptoms restricted to EOMs, resulting in a syndrome known as Ocular Myasthenia Gravis (OMG). Myasthenia gravis appears to be influenced by gender and age. Female to male ratio is around 3:1 under 40 years of age; however, it is nearly equal between 40 years and 50 years of age, as well as throughout puberty. It is more frequent in guys over the age of 50. Childhood Myasthenia Gravis (MG) is rare in Europe and North America, accounting for just 10% to 15% of all MG cases. However, in Asian nations, up to 50% of patients present before the age of 15, with primarily ocular symptoms.

EVALUATION

The majority of MG diagnoses are clinical. In most cases, laboratory tests and techniques assist the clinician in confirming clinical findings.

Serologic tests

Anti-AChR Ab tests are extremely specific and confirm the diagnosis in patients with characteristic clinical symptoms. It is found in four-fifths of generalised MG patients and only half of those with pure ocular MG. Anti-MuSK antibodies will be found in the remaining patients, which will account for around 5% to 10% of the total [2]. Anti-AChR and anti-MuSK antibodies are only found in a few infrequent incidences in the same patient. Anti-LRP4 antibodies will be detected in the 3% to 50% of the remaining individuals who are seronegative to either of these antibodies. Antibodies against striated muscle are found in 30% of MG patients. They're more practical.

Electrophysiologic tests

Patients who are seronegative for antibody testing should be aware of these. The Repeated Nerve Stimulation (RNS) test and single-fiber electromyography are two common MG tests SFEMG. Both tests look for delays in conduction in the NMJ. Before performing these tests, routine nerve conduction investigations are routinely performed to check how well the nerves and muscles are working.

Edrophonium (Tensilon) test

Edrophonium is an acetylcholinesterase inhibitor with a short half-life that enhances ACh availability in the NMJ. This is especially important in cases of ocular MG, where electrophysiologic testing is not possible [3]. It's given intravenously, and the patient's symptoms of ptosis or diplopia are monitored for improvement. For MG diagnosis, it has a sensitivity range of 71% to 95%.

Other laboratory tests

Anti-Nuclear Antibodies (ANA), Rheumatoid Factor (RF), and baseline thyroid functions testing are advised since myasthenia gravis frequently coexists with other autoimmune illnesses.

TREATMENT

Symptomatic treatment

By blocking acetylcholinesterase breakdown, acetylcholinesterase inhibitors raise the amount of ACh at the NMJ. Because of its extended duration of action, pyridostigmine bromide is favoured over neostigmine. Ambenonium chloride can be used to treat bromide intolerance that causes gastrointestinal problems. Patients with MuSK MG may have a poor response to these medicines, necessitating a greater dose.

Immunosuppressive treatment

These are prescribed for people who are still experiencing symptoms after taking pyridostigmine. The first-line immunosuppressive drugs used in the therapy of MG include glucocorticoids (prednisone, prednisolone, and methylprednisolone) and azathioprine. Cyclosporine, methotrexate, mycophenolate, cyclophosphamide, and tacrolimus are examples of second-line agents. When a patient is resistant to therapy, has a contraindication to treatment, or is unable to tolerate first-line medicines, they are utilised [4]. Various monoclonal antibodies have recently been utilised to treat drug-resistant MG, including rituximab and eculizumab, although data from clinical studies on their efficacy has yet to be established.

Intravenous immunoglobulins/plasmapheresis

This is advised to help stabilise a patient before a surgery during the perioperative phase. Due to its quick start of action, it is also the therapy of choice for myasthenic crises and is utilised in instances when immunosuppressive medicines have failed [5].

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