

## In Neurology

### Myasthenia Gravis

**Introduction :** It is the most common disorder affecting the neuromuscular junction (NMJ) of the skeletal muscles.

**Etiology :**

- It occurs in genetically susceptible individuals.
- Precipitating factors include conditions like infections, immunization, surgeries, and drugs.
- The commonly implicated proteins in the NMJ against which autoantibodies are produced include the nicotinic acetylcholine receptors (n-AChR's), muscle-specific kinase (MuSK), and lipoprotein-related protein 4 (LPR4).

**Epidemiology :**

- Prevalence of 20 per 100,000 population.
- A female predominance in those less than 40 years of age.
- A male predominance in those greater than 50 years of age.

**Classification :** Based on the clinical features and the disease severity :-

- **Class I:** Involves any ocular muscle weakness, including weakness of eye closure. All other muscle groups are normal.
- **Class II:** Involves mild weakness of muscles other than ocular muscles. Ocular muscle weakness of any severity may be present.
- **Class IIa:** Involves predominant weakness of the limb, axial muscles, or both. It may also involve the oropharyngeal muscles to a lesser extent.
- **Class IIb:** Involves mostly oropharyngeal, respiratory muscles, or both. It can have the involvement of limb, axial muscles, or both to a lesser extent.
- **Class III:** Involves muscles other than ocular muscles moderately. Ocular muscle weakness of any severity can be present.
- **Class IIIa:** involves the limb, axial muscles, or both predominantly. Oropharyngeal muscles can be involved to a lesser degree.
- **Class IIIb:** Involves oropharyngeal, respiratory muscles, or both predominantly. The limb, axial muscles, or both can have lesser or equal involvement.
- **Class IV:** Involves severe weakness of affected muscles. Ocular muscle weakness of any severity can be present.

- **Class IVa:** Involves limb, axial muscles, or both predominantly. Oropharyngeal muscles can be involved to a lesser degree.
- **Class IVb:** Involves oropharyngeal, respiratory muscles, or both predominantly. The limb, axial muscles, or both can have lesser or equal involvement. It also includes patients requiring feeding tubes without intubation.
- **Class V:** Involves intubation with or without mechanical ventilation, except when employed during routine postoperative management.

### **Clinical Features :**

- Extraocular Muscle Weakness: Diplopia, ptosis, or both.
- Bulbar Muscle Weakness: Difficulty chewing or frequent choking, dysphagia, hoarseness, and dysarthria.
- **Limb Weakness:** Usually involves the proximal muscles more than distal muscles, with the upper limbs more affected than the lower limbs.
- **Myasthenic crisis:** It is due to the involvement of intercostal muscles and diaphragm and is a medical emergency.
- There are no autonomic symptoms like palpitations, bowel, or bladder disturbances.

### **Diagnosis :**

- **Edrophonium test :** It is a short acting acetylcholinesterase inhibitor and with injection, patient gets immediate increases in muscle strength.
- Spirometry to see FEV1 & vital capacity.
- Chest X ray, CT or MRI of chest to rule out thymoma.
- Serum acetylcholine receptor antibodies are present in large majority.
- T3, T4 & TSH levels to rule out associated thyroid disorders.
- ANA, RA factor & antithyroid antibodies may be positive.

### **Treatment :**

- Acetylcholinesterase inhibitor – Pyridostigmine 5times/day.
- Definitive treatment includes immunosuppression by steroids, azathioprine, cyclosporine, rituximab, plasmapheresis & IV immunoglobulin.
- Thymectomy if no response to the above.

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