

# Myasthenia Gravis

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## Myasthenia Gravis

### MG: Associated Autoimmune Dx

- ◇ Hypothyroidism #1
  - ◇ 2.2 to 16.9%
- ◇ Rheumatoid Arthritis #2
  - ◇ Up to 10.3%
- ◇ Other Autoimmune Disorders
  - ◇ Sjogren's, sarcoidosis, scleroderma, polymyositis, etc.
  - ◇ Lambert Eaton Myasthenic Syndrome

### MYASTHENIA IS:

- ◇ A Neuromuscular Junction Disease
- ◇ AN AUTOIMMUNE DISEASE that targets the Ach receptors

## Purely Ocular Myasthenia

- **Initial presentation of MG in up to 70%**
  - Ocular precedes clinically generalized MG in 50-70 % of patients.
  - **Usually generalizes within 2 years of onset of ocular symptoms**
- ◇ If very symptomatic: how aggressive and how should we treat Ocular MG ?

### **WHY GRAVIS ? BECAUSE PT CAN DIE ....**

- **OMINOUS SYMPTOMS ARE:**
  - **Hoarseness** –problems with respiratory system can progress to pt 's **inability to breathe.**
  - **Swallowing difficulty:** can lead to patient choking with food causing aspiration and death

## MG: Clinical Features

- o Ocular Myasthenia: Ptosis and Diplopia
  - o Presenting feature in 50 to 60%
  - o 90% of MG patient will ultimately manifest ocular symptoms
  - o 85% of “Pure Ocular” patients will generalize usually within 2 years.
  - o “Pure ocular” over 2 years: Only has a 10% chance of progression.
  - o Characteristic signs helpful on examination

## MG: Bulbar

- Bulbar symptoms and Signs
  - o Paucity of facial wrinkles/expression
  - o Poor eyelid closure
  - o Difficulty puckering/whistling
  - o Decreased horizontal excursion on smile.
  - o Masseter weakness – cannot chew
  - o Weakness/cramping neck extensors
  - o Dysarthria/hypernasality of speech
  - o Dysphagia

## MG: generalized + bulbar

- Generalized Weakness
  - o Prominent fatigability of extremities
  - o May be predominantly proximal.
  - o RESPIRATORY COMPROMISE
    - Highest degree of vigilance
    - Always better to intubate too early than too late.

## Clues Diagnosis in office

### o History

Variability and late in day history

Ptosis and diplopia -ocular

Problems with chewing, hoarseness, swallowing, breathing, neck weakness, extremity weakness- general

Diseases that may be associated:

Thyroid

Collagen vascular

Pernicious Anemia

- o **Exam**

- o **Ocular**

- Lids : check for ptosis and signs of fatigue and poor orbicularis function

- Pupils : check for symmetry and good reaction

- EOM's : check alignment and fatigue induced change

Exophthalmometry : **Check for absence of proptosis : association with thyroid eye disease.**

- o **Exam**

- o **General**

- Neck: Flexion and extension weakness

- Extremities: fatigue

### **Ice Test for ptosis**

**Laboratory testing -** MG antibodies, thyroid testing

**Electrophysiology -**

- o EMG for repetitive stimulation decrement

- o Single fiber of frontalis very helpful and very sensitive for jitter

**Imaging**

- o Thymus- for thymoma

- o +/- Brain/Orbit if you are not certain

Tensilon not really necessary in our opinion

**Treatment**

**Symptomatic**

**For ocular symptoms and mild general symptoms**

**Pyridostigmine or Mestinon orally**

## **Immunosuppression**

**Steroids**

**Cellcept**

**Imuran**

**Other**

**See below**

## **Rescue therapy**

**IVIg**

**Plasmaphoresis**

## **Surgery**

**Thymectomy**

## **Whats new**

**Monoclonal Antibody agents**