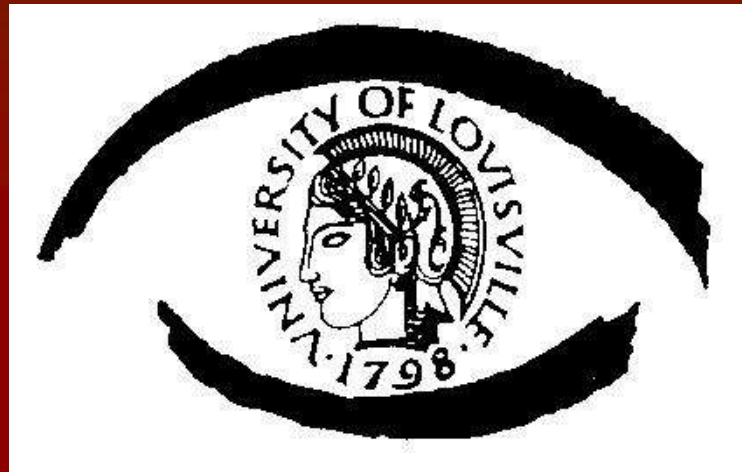


# Grand Rounds



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# Subjective

CC: “ R eye ptosis and double vision x 6 months”

HPI: 55 year-old AAM presented c/o diplopia and ptosis OD x 6 months. Onset was gradual . Symptoms have been progressively getting worse over the last several months. Diplopia resolves when one eye is closed. Denies headaches, ocular pain and irritation. No history of trauma or cerebrovascular accidents .

POH: EOM surgery OS 18 yrs ago

PMH: Hyperthyroidism (s/p radioactive I<sup>131</sup> therapy 18 yrs ago), type II DM

MEDS: Levothyroxine, Januvia

All: NKDA

ROS: c/o increased generalized fatigue

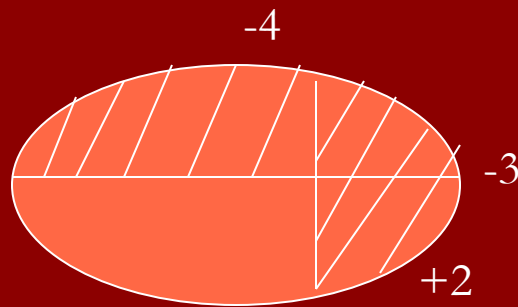
# Objective

VA(sc)  $\left\{ \begin{array}{l} 20/20^{-2} \\ 20/20 \end{array} \right.$

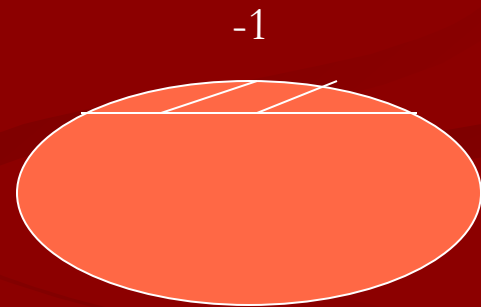
P  $\left\{ \begin{array}{l} 3 \rightarrow 2 \text{ brisk OU} \\ 3 \rightarrow 2 \text{ } \emptyset \text{ RAPD} \end{array} \right.$

T  $\left\{ \begin{array}{l} 20 \\ 19 \end{array} \right.$

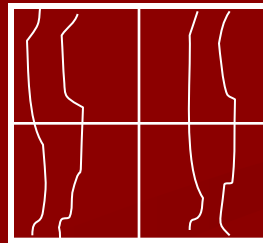
## Motility:



25 PD RHoT  
10 PD XT

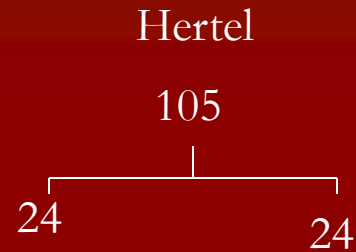
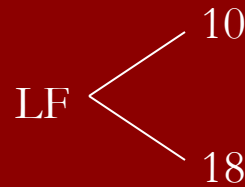


## Diplopia



# Objective

Lacrimal/Orbit/Lids: ptosis OD and eyelid retraction OS



SLE: mild nasal conjunctival injection OU, otherwise unremarkable

DFE: unremarkable OU



Marked ptosis OD and lid retraction OS

# Motility



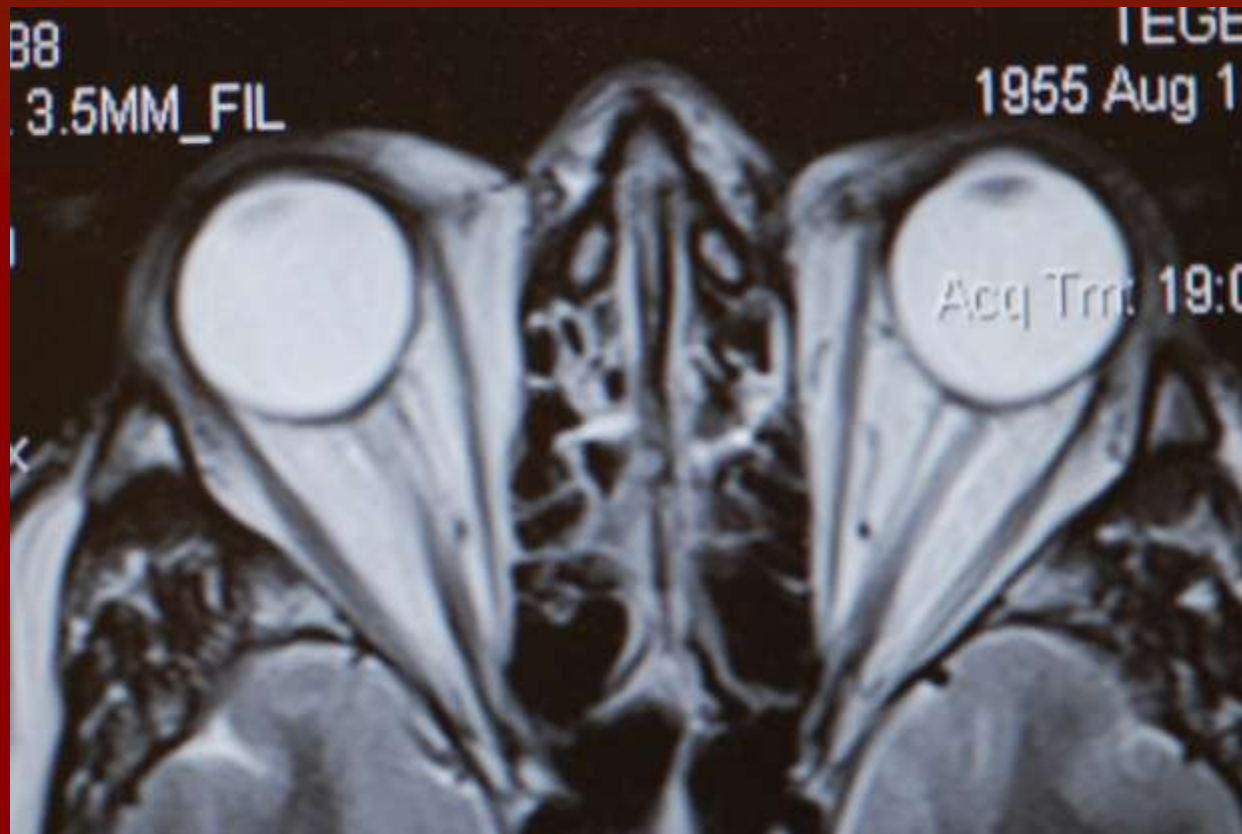
# Assessment

55 yo AAM with RHoT, XT, limited motility in supraduction and adduction, diplopia and ptosis OD.

DD<sub>x</sub>

1. Third Nerve Palsy
2. Myasthenia Gravis
3. Thyroid-associated Ophthalmopathy

# Imaging



MRI was unremarkable except mild increase in intraorbital fat content



Ice Test: ice was applied over OD for 5 min that resulted in significant improvement of ptosis and moderate improvement in motility

# Diagnosis

55 yo AAM with:

1. Myasthenia Gravis
2. Thyroid-associated lid retraction OS

# Discussion

## Myasthenia Gravis (MG)

- Autoimmune disorder characterized by weakness and fatigability of skeletal muscles
- Hallmark of MG is fluctuating muscle weakness
- Affects women > men
- Bimodal distribution with peaks in 2<sup>nd</sup> and 3<sup>rd</sup> decade affecting mostly women and 6<sup>th</sup> and 7<sup>th</sup> affecting predominately men

# Clinical Presentation

- Fluctuating muscle weakness mostly occurs in ocular, bulbar, limb and respiratory muscles
- Weakness worsens later in the day or after exercise
- Two clinical forms of MG recognized: ocular (OMG) and generalized (GMG)
- Isolated ocular symptoms of ptosis and diplopia are presenting sign of disease in >50% of patients
- Only 15% of patients remain having isolated ocular problems

# Ocular Myasthenia Gravis

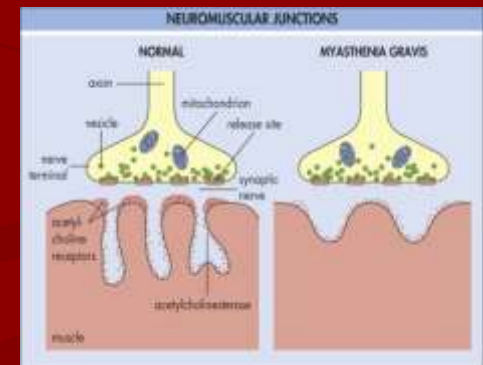
- Characterized by ptosis and oculomotor paresis
- Ptosis
  - often unilateral and asymmetric
  - occasionally alternates from side to side
  - can fluctuate throughout examination
- Diplopia
  - binocular
  - can fluctuate in the degree or direction of gaze
  - can virtually mimic any motility disorder

# Generalized Myasthenia Gravis

- Involvement of muscles of mastication results in weakness with prolonged chewing
- Dysarthria and dysphagia occur with oropharyngeal muscle involvement and can cause aspiration
- Neck extensor and flexor muscles commonly affected and can produce “drooped head syndrome”
- Respiratory muscle involvement cause the most serious symptoms that can lead to respiratory failure called “myasthenic crisis”

# Pathophysiology

- Pathogenicity mediated by antibodies (Ab) attacking the acetylcholine receptor (AChR), fixing complement, and reducing the number of AChRs over time
- Ab originate in hyperplastic germinal center in the thymus where myoid cells expressing AChR are found
- AChR Ab present in 80%-90% of patients
- 60-70% of AChR Ab positive patients have thymic hyperplasia and 10-12% have thymoma
- 40-70% of AChR negative MG patients have Ab directed against the muscle-specific receptor tyrosine kinase (MuSK)



# Diagnosis

- Bedside test: ice-pack test and edrophonium (Tensilon) test
- Serologic tests: AChR-Ab and MuSK-Ab
- Electrophysiologic tests: repetitive nerve stimulation (RNS) studies and single-fiber electromyography (SFEMG)
- Imaging: CT Chest

# Treatment

- Symptomatic treatments (anticholinesterase agents)
- Chronic immunomodulating treatments (glucocorticoids and other immunosuppressive drugs)
- Rapid immunomodulating treatments (plasma exchange and intravenous immune globulin)
- Surgical treatment (thymectomy, eyelid, strabismus)

# The effect of prednisone on the progression from ocular to generalized myasthenia gravis

Monsul et al. *Journal of Neurologic Science* 2004 Feb 15;217(2):131-3.

- 50-85% of patients with OMG progress to GMG with 90% progressing in 3 year from onset
- Objective: determine whether treatment with oral prednisone initiated and completed within 2 years from the onset of ocular symptoms would affect the progression of ocular myasthenia to generalized myasthenia gravis (GMG)
- Methods: retrospective study, 56 patients included in review, 27 patients in the prednisone-treated and 29 patients in the untreated groups

- Results: At 2 years, 3 of 27 (11%) patients in prednisone-treated group progressed to generalized myasthenia and 10 of 29 (34%),  $p=0.04$
- Conclusion: early introduction of prednisone therapy appears to be important in delaying the onset of GMG, but larger studies needed to confirm this recommendation

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Thank You