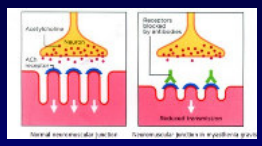
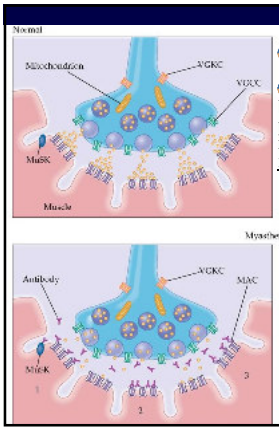
Myasthenia gravis



- Definition: autoimmune disease with circulating antibodies against Ach-R (Acetylcholine Receptor), or other antigens of postsynaptic membrane (titin, MUsK-enzyme,...)
- Clinical symptoms: progredient muscle weakness during daily activities, with evening acces, repair after rest time
- Onset: small muscles – eye (diplopia), pharynx (dysphagia), soft palate (rhinolalia), general ...

MG- epidemiology

- **Incidence**: 14,8 / mil. inhabitants
- Prevalence: 191 /mil. inhab. /SR 1.1.2007/
- **Disease onset**: mostly: 30. year (F), 60.-70. year (M)
- Sex rate F:M= 1,7:1
- No hereditary cases, familial increased susceptibility for autoimmune disease (HLA)



- Acquired autoimmune disease
- **Ab+Ach-Receptor => functional block and destruction=> decrease postsynaptic actional potential => insuficient muscle contraction => muscle weakness**

MG- manifestation

- Clinical symptoms: progredient muscle weakness during daily activities, with evening acces, repair after rest time
- Onset: small muscles – eye (diplopia), pharynx (dysphagia), soft palate (rhinolalia), general ...
- Thymus abnormalities - ¾ of MG patients
- **85% - hyperplasia**
  15% - thymoma (benign / malign)

MG- disease course

- **Subjective**: abnormal muscle fatigue, weakness - paresis, recovery after resting
- **Objective**:
- repetitive muscle activity provocates weakness- ptosis, diplopia, rhinolalia, dysphagia, dysarthria, dysphonia
- weak of jawing, mimic paresis
- neck dicrease
- short breathing
- tendon reflexes- presented or slight decrease

MGFA (Foundation of America)- Clinical symptoms scaling
(Osserman´s classification)

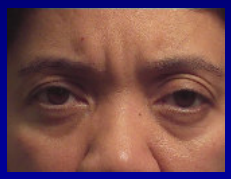
- I. Ocular form MG
- II. Ocular + slight generalised MG (limbs /bulbar)
- III: Moderate weakness of ocular + extraocular muscles (limbs, respiratory, bulbar muscles)
- IV: Severe weakness of ocular + extraocular muscles
- V: Respiratory failure, suported ventilation

MG- diagnosis

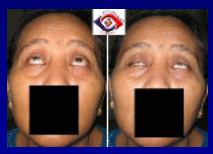
1. **History**
2. **EMG**
3. **Lab:** serum antibodies anti-Ach-R (75% positivity)
4. **Clinical tests**: Simpson´s test- vertical gaze
   Seeman´s test- dysarthria
   Gorelick´s test
   pharmacol. Tensilon test: iv.
   amp. inhib. AchE
5. **X-rays chest, mediastinal CT, MRI**

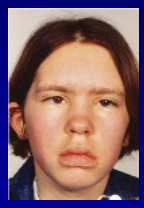
Static and dynamic (repetitive) tests

- Demasking of latent MG or enhancing of present muscle weakness
- **Simpson´s test –** slight ptosis – patient is looking upward 1 minute- more severe ptosis

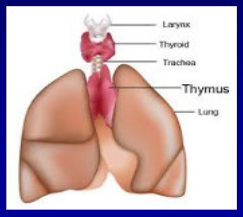


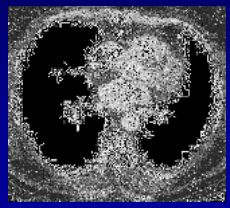
- **Gorelick´s sign** – bilateral asymetrical ptosis: patient is looking up, finger elevation of eyelid opposite side, on contralateral side wa can see total decrease
- (pathognomic for MG)





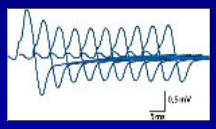
Thymoma

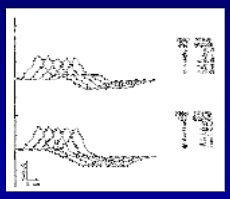




MG- EMG

- EMG - repetitive stimulation, low freq.stimulation - 3Hz
  Abnormality: **gradual decrement of response amplitude**
- min.15%, max. in the 2.- 4. response, normalization of EMG after Tensilone inj.





MG- therapy

Currently - no deaths, previously - 30% mortality

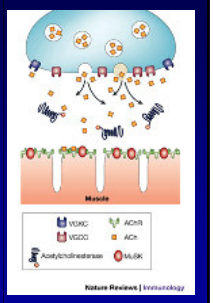
1. **Pharmacological**: IS + symptomatic th
2. **Surgical** – thymectomy

Pharmacological therapy:

- Immunosupression: **Prednison, Azathioprine,Cyclosporine A**
- Plasma exchange/ or IVIG
- inhib.ChE **(Pyridostigmine, Mestinon)**

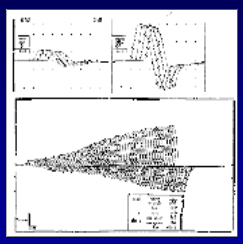
LEMS, Lambert- Eaton myasthenic syndrom

- is a rare autoimmune disorder
- muscle weakness of the limb
- Antibodies against presynaptic voltage-gated calcium channels, and likely other nerve terminal proteins
- Prevalence: 3.4 cases/million
- Around 60% of LEMS -have an underlying malignancy (small cell lung cancer)
- paraneoplastic syndrome
- KP: fatigue, weakness of proximal mm., inferior extremities, spared eye and bulbar mm.
- autonomic difficulties: dry mouth, low lacrimation, orthostatic collapses, impotentia



LEMS

High frequency repetitive EMG (30 Hz) => gradual increase of AP amplitude



Therapy:

steroids, Azathioprine, plasma exchange