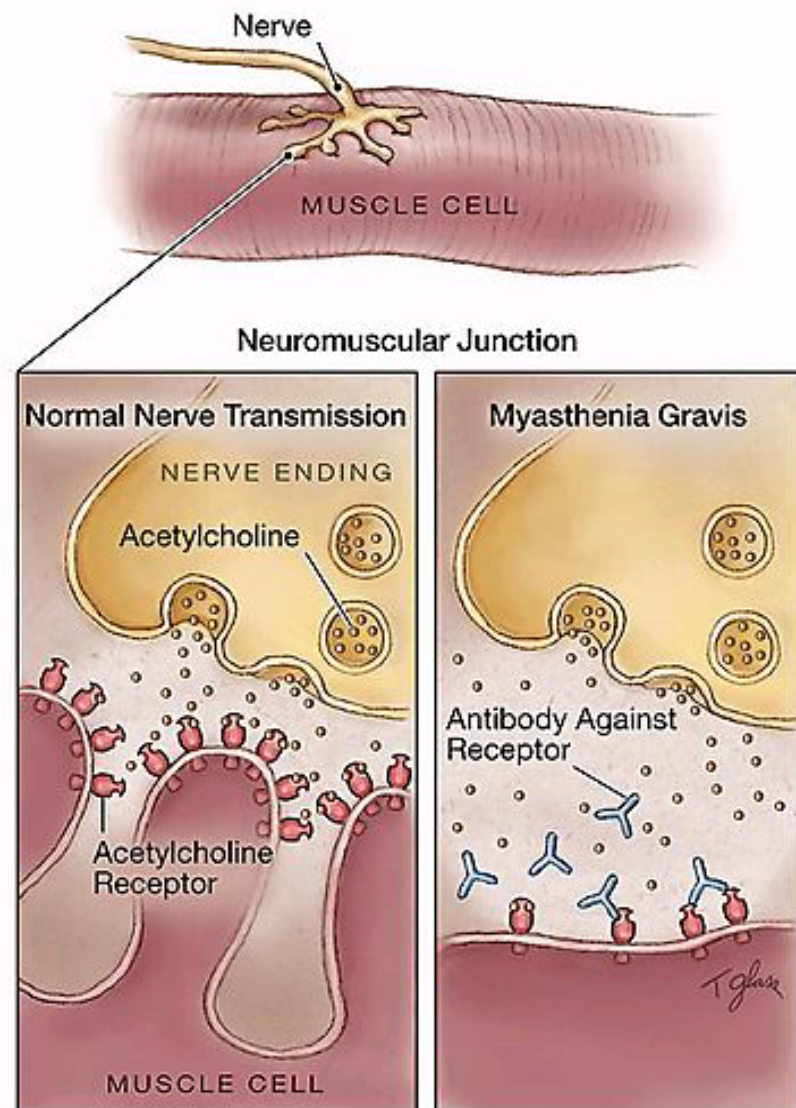


OMG – a part of MG

- *ocular sign is a warning sign of MG* -

- An **autoimmune disease**
 - producing an antibody against acetylcholine receptor
 - as a consequence, resulting in
 - inhibition of ACh releasing
 - dysfunction of neuro-muscular junction
- An **antibody-mediated disorder**
 - similar to other autoimmune disorders
 - therefore it can be **treated by immunosuppression**



Types of MG syndrome - based on AB property

- 1./ Classic form of myasthenia gravis (MG) POST synaptic defect
- 2./ Congenital MG POST synaptic defect
- 3./ Lambert – Eaton syndrome (LEMS) PRE-synaptic defect

Similarities of MG forms

- clinical signs – same
- consequence – disorders of NMJ

Differences of MG forms

- different AGs and ABs
- different pathomechanism
- numerous types of IS therapies

BASIC therapy

thymectomy	thymoma +/- MG	<i>in all cases- except for ocular signs</i>
pyridostigmin (Mestinon) <i>(inhibition of degrad. of Acho)</i>	opening therapy every, new cases of MG	daily dose 2x30mg /day– --5x60mg /day
STEROID + IS <i>(methylpredn., Prednisolon)</i>	basic therapy + AZA <i>prevention of MG crisis</i>	10-25 mg/ on alternate days - 10mg/d -up to: 60-80mg <i>High dose therapy not proposed!</i>

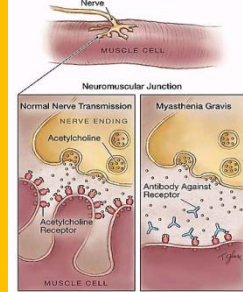
IMMUNOSUPPRESSION (IS)

AZATHIOPRIN (AZA) (Imuran) <i>-inhibition of RNA-DNA synthesis</i> <i>- T-cells functions)</i>	steroid +AZATHIOPRIN combined <ul style="list-style-type: none"> <i>• one hand, steroid has more side effects than AZA</i> <i>• steroid can be reduced gradually in the same time</i> <i>• AZA dose remains the same</i>
MYCOPHENOLAT MOFETIL (CELLCEPT) <i>(selective inhibition the proliferation of lymphocta</i>	<i>indication:</i> <ul style="list-style-type: none"> <i>• steroid non-respondance MG cases</i> <i>• AZA non-respondance or intolerance</i> <i>combine with:</i> Cyclosporin+ corticosteroid
METHOTREXAT	<i>indication:</i> in case of non-respondance for AZA, Cellcept
CYCLOPHOSPHAMID <i>(with serious side effects)</i>	indicated : <i>Non response or non-compliance for: steroid+AZA, Methotrexat, Cyclosporin, Cellcept</i>
CYCLOSPORIN <i>(with serious side effects)</i>	<i>(inhibition effect of T cell, by inhibition of calcineurin- signalling)</i> <i>indication:</i> AZA no efficiency or intolerance
LEUKOCYTA antigen against at antibody (AB)	RITUXIMAB – B-cells inhibitor anti-CD4 – T cells inhibitor <i>indication:</i> resistance to all types of IS

IMMUNOMODULATED therapy

PLASMAPHERESIS (PE) <i>(removing AB from serum)</i>	<i>indications:</i> <ul style="list-style-type: none"> <i>• onset of immunosuppression</i> <i>• in acute myasthenic crisis or near crisis</i> <i>• preoperative treatment</i>
iv. IMMUNGLOBULIN (iv-IVIG)	<i>indications:</i> <ol style="list-style-type: none"> <i>1. acute phase of MG with immediate progression</i> <i>2. as a preoperative treatment</i> <i>3. during the IS therapy to decrease the side effect</i> <i>4. in serious MG cases</i> IF: non - response for steroid or IS

Suggestions of Neurological Advisory Board of Ministry of Health of Hungary



1./ **Beginning with cholinesterase- inhibitor**

2./ **Thymectomy:**

- absolute indication- in case of thymoma
- not suggested in case of OMG !

3./ **Steroid indication:**

- starting low dose, increase gradually
- long term steroid therapy + AZA-/or other IS/

4./ **Immunosuppression therapy indicates:**

- in all progressive MG cases
- symptoms persist in spite of applications of the cholinesterase-inhibitor therapy
- when the patient's every days life is limited by MG symptoms

5./ **Plasmapheresis or IVIG:**

- acute myasthenic exacerbation

Dr. Rózsa Csilla, Dr. Diószeghy Péter, prof. Dr. Komoly Sámuel:

*Clinical guiding principles for the treatment of autoimmune diseases
with myasthenia gravis and disorders of neuromuscular transmission.*

Hungarian Professional Board of Neurology, 2007