

Myasthenia Gravis Activities of Daily Living Scale (MG-ADL)^{1,2}

This Practice Aid has been provided as a quick reference to help learners apply the information to their daily practice and care of patients.

Grade	0	1	2	3	Score
Talking	Normal	Intermittent slurring or nasal speech	Constant slurring or nasal, but can be understood	Difficult to understand speech	
Chewing	Normal	Fatigue with solid food	Fatigue with soft food	Gastric tube	
Swallowing	Normal	Rare episode of choking	Frequent choking necessitating changes in diet	Gastric tube	
Breathing	Normal	Shortness of breath with exertion	Shortness of breath at rest	Ventilator dependence	
Impairment of ability to brush teeth or comb hair	None	Extra effort, but no rest periods needed	Rest periods needed	Cannot do one of these functions	
Impairment of ability to arise from chair	None	Mild, sometimes uses arms	Moderate, always uses arms	Severe, requires assistance	
Double vision	None	Occurs, but not daily	Daily, but not constant	Constant	
Eyelid droop	None	Occurs, but not daily	Daily, but not constant	Constant	
Total Score					

1. Wolfe GI et al. *Neurology*. 1999;52:1487-1489.
 2. Muppidi S et al. *Muscle Nerve*. 2011;44:727-731.

Access the activity, "Taking Stock of Recent Clinical Research in Generalized Myasthenia Gravis: What Is the Potential Impact of New Therapies on Disease Management?" at www.peerview.com/RMU40.

Summary of International Consensus Guidance Statements for Myasthenia Gravis Management¹

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Symptomatic and Immunosuppressive (IS) Treatment

Pyridostigmine should be part of the initial treatment in most patients

Corticosteroids or IS therapy should be used in all patients with MG who have not met treatment goals after an adequate trial of pyridostigmine

- Nonsteroidal IS agents should be used alone when corticosteroids are contraindicated/refused
- Nonsteroidal IS agents that can be used include azathioprine, cyclosporine, mycophenolate mofetil, methotrexate, and tacrolimus
- Nonsteroidal IS agents should be used initially in conjunction with corticosteroids when the risk of steroid side effects is high based on medical comorbidities

IVIg and PLEX

PLEX and IVIg are appropriately used as short-term treatments:

- In patients with life-threatening signs such as respiratory insufficiency or dysphagia
- In preparation for surgery in patients with significant bulbar dysfunction
- When a rapid response to treatment is needed
- When other treatments are insufficiently effective
- Prior to beginning corticosteroids if deemed necessary to prevent or minimize exacerbations

Choice between PLEX and IVIg depends on individual patient factors and on the availability of each

IVIg and PLEX probably equally effective in treatment of severe generalized MG

Efficacy of IVIg less certain in milder MG or in ocular MG

PLEX may be more effective than IVIg in MuSK MG

Thymectomy

With rare exceptions, all patients with MG and thymoma should undergo surgery to remove the tumor; all thymus tissue should be removed along with the tumor

In nonthymomatous MG, thymectomy is performed as an option to potentially avoid or minimize the dose or duration of immunotherapy, or if patients fail to respond to an initial trial of immunotherapy or have intolerable side effects from that therapy

Refractory MG

Patients with refractory MG should be referred to a physician or a center with expertise in management of MG

In addition to the previously mentioned IS agents, the following therapies may also be used:

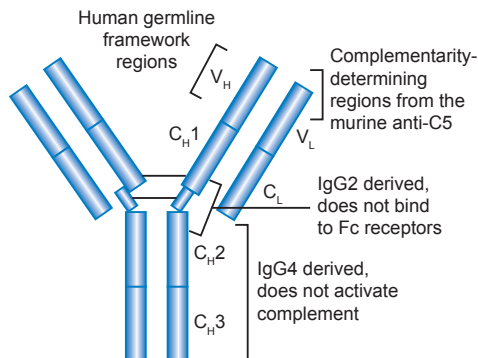
- Chronic IVIg
- Cyclophosphamide
- Chronic PLEX
- Rituximab

IVIg: intravenous immunoglobulin; MG: myasthenia gravis; MuSK: muscle-specific tyrosine kinase; PLEX: plasma exchange.
1. Sanders DB et al. *Neurology*. 2016;87:419-425.

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Eculizumab: Dosing and Safety Considerations¹

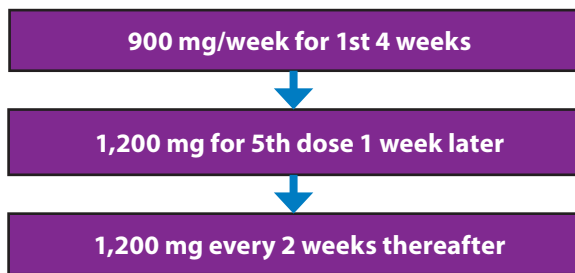
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Eculizumab

- Humanized mAb that specifically binds with high affinity to human terminal complement protein C5
- Inhibits enzymatic cleavage to proteins C5a and C5b, preventing C5a-induced chemotaxis of proinflammatory cells and formation of C5b-induced membrane attack complex
- Approved for treatment of adult patients with generalized MG who are anti-AChR antibody positive

Recommended Dosing Regimen



Dosage Adjustments in Case of PLEX

Most Recent Eculizumab Dose	Supplemental Eculizumab Dose With Each PLEX Session	Timing of Supplemental Eculizumab Dose
≥600 mg	600 mg per each PLEX session	Within 60 min after each PLEX

Safety Considerations

- Most frequently reported AE in generalized MG trial (≥10%): musculoskeletal pain
- Life-threatening and fatal meningococcal infections have occurred in patients treated with eculizumab and may become rapidly life-threatening or fatal if not recognized and treated early

- Comply with the most current ACIP recommendations for meningococcal vaccination in patients with complement deficiencies
- Immunize patients with meningococcal vaccines at least 2 weeks prior to administering the first dose of eculizumab, unless the risks of delaying eculizumab therapy outweigh the risks of developing a meningococcal infection
- Monitor patients for early signs of meningococcal infections, and evaluate immediately if infection is suspected

AChR: acetylcholine receptor; ACIP: Advisory Committee on Immunization Practices; C_H: constant heavy region; C_L: constant light region; IgG: immunoglobulin G; mAb: monoclonal antibody; MG: myasthenia gravis; PLEX: plasma exchange; V_H: variable heavy region; V_L: variable light region.

1. Soliris (eculizumab) Prescribing Information. http://www.alexion.com/Documents/Soliris_USPI.aspx. Accessed February 27, 2018.

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