HIGHMARK WHOLECARE

Updated: 02/2025
PARP Approved: 03/2025

Prior Authorization Criteria
Myasthenia Gravis Medications

All requests for Soliris (eculizumab), Ultomiris (ravulizumab-cwvz), Vyvgart (efgartigimod alfa-fcab), Vyvgart Hytrulo (efgartigimod alfa and hyaluronidase-qvfc), Zilbrysq (zilucoplan) and Rystiggo (rozanolixizumab-noli) require a prior authorization and will be screened for medical necessity and appropriateness using the criteria listed below.

****For all requests for complement Inhibitors for diagnoses other than Myasthenia Gravis please refer to policy PH-206.74-MD-PA C5b and C3 Complement Inhibitors ****

Coverage may be provided with a diagnosis of **generalized Myasthenia Gravis (gMG)** and the following criteria is met:

- Medication is prescribed by, or in consultation with, a neurologist
- Documentation of a positive serologic test for one of the following:
    - anti-acetylcholine antibodies
    - anti-muscle specific tyrosine kinase (MUSK) - **Rystiggo only**
- Documentation the member meets the following Myasthenia Gravis Foundation of America Clinical Classification Class
    - Vyvgart (efgartigimod alfa-fcab), Vyvgart Hytrulo (efgartigimod alfa and hyaluronidase-qvfc), Zilbrysq (zilucoplan), Soliris (eculizumab), or Ultomiris (ravulizumab-cwvz)
        - II to IV
    - Rystiggo (rozanolixizumab-noli)
        - II to IVa
- Documentation the member has a Myasthenia Gravis-Specific Activities of Daily Living (MG-ADL) total score of one of the following:
    - Zilbrysa (zilucoplan), Soliris (eculizumab), or Ultomiris (ravulizumab-cwvz)
        - ≥6
    - Vyvgart (efgartigimod alfa-fcab) or Vyvgart Hytrulo (efgartigimod alfa and hyaluronidase-qvfc)
        - ≥5
    - Rystiggo (rozanolixizumab-noli)
        - ≥ 3 (with at least 3 points from non-ocular symptoms)
- Documentation of a baseline Quantitative Myasthenia Gravis (QMG) scale score
- Laboratory testing demonstrating IgG levels of the following:
    - Vyvgart (efgartigimod alfa-fcab) or Vyvgart Hytrulo (efgartigimod alfa and hyaluronidase-qvfc)
        - at least 6g/L
    - Rystiggo (rozanolixizumab-noli)
        - at least 5.5 g/L
- Documentation of at least one of the following:
    - Failed treatment over 1 year or more with 2 or more immunosuppressive therapies either in combination or as monotherapy (e.g. azathioprine, cyclophosphamide, methotrexate)

    - Failed treatment over 1 year or more with at least 1 immunosuppressive therapy while on chronic plasmapheresis or plasma exchange (PE)
- The requested dose and frequency is in accordance with FDA-approved labeling, nationally recognized compendia, and/or evidence-based practice guidelines
- Must be age-appropriate according to FDA-approved labeling, nationally recognized compendia, or evidence-based practice guidelines
- If requesting Soliris, must have documentation of inadequate response, contraindication, or intolerance to Ultomiris
- The requested agent must not be used in combination with another Myasthenia Gravis medication listed in this policy [e.g. Soliris (eculizumab), Ultomiris (ravulizumab-cwvz), Vyvgart (efgartigimod alfa-fcab), Vyvgart Hytrulo (efgartigimod alfa and hyaluronidase-qvfc), Zilbrysq (zilucoplan), and Rystiggo (rozanolixizumab-noli)]
- **Initial Duration of Approval:** 6 months
- **Reauthorization criteria**
    - First reauthorization criteria (member on therapy for 0 to 6 months)
        - Documentation from the provider that the member had a positive clinical response and tolerates therapy supported by at least one of the following:
            - A 2 point improvement in the member's total MG-ADL score
            - A 3 or more point improvement in QMG total score
    - Subsequent reauthorization criteria (member on therapy ≥ 6 months)
        - Documentation from the prescriber indicating stabilization or improvement in condition.
- **Reauthorization Duration of Approval:** 12 months

Coverage may be provided for any non-FDA labeled indication if it is determined that the use is a medically accepted indication supported by nationally recognized pharmacy compendia or peer-reviewed medical literature for treatment of the diagnosis(es) for which it is prescribed. These requests will be reviewed on a case by case basis to determine medical necessity.

When criteria are not met, the request will be forwarded to a Medical Director for review. The physician reviewer must override criteria when, in their professional judgment, the requested medication is medically necessary.

Attachments

Attachment 1. Myasthenia Gravis Activities of Daily Living (MG-ADL) profile

Attachment 2. Quantitative Myasthenia Gravis scale (QMG) scale

Attachment 3. Myasthenia Gravis Foundation of America Clinical Classification

Attachment 1. MG Activities of Daily Living (MG-ADL) profile

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 a 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 _____

Attachment 2. Quantitative Myasthenia gravis scale

Test Items Weakness	None	Mild	Moderate	Severe
Double vision on lateral gaze **right** or **left** (circle one)	61	11–60	1–10	spontaneous
Ptosis (upward gaze)	61	11–60	1–10	spontaneous
Facial muscles	normal lid	complete, weak, some resistance	complete, without resistance	incomplete
Swallowing 4 oz. water (½ cup)	normal	Minimal coughing or throat clearing	severe coughing/ choking or nasal regurgitation	cannot swallow (test not attempted)
Speech following counting aloud from 1 to 50 (onset of dysarthria)	none at #50	dysarthria at #30–49	dysarthria at #10–29	dysarthria at #9
Right arm outstretched (90° sitting)	240	90–239	10–89	0–9
Left arm outstretched (90° sitting)	240	90–239	10–89	0–9
Vital capacity (% predicted)	≥80%	65–79%	50–64%	<50%
Right hand grip (kg)				
male	≥45	15–44	5–14	0–4
female	≥30	10–29	5–9	0–4
Left hand grip (kg)				
male	≥35	15–34	5–14	0–4
female	≥25	10–24	5–9	0–4
Head lifted (45° supine)	120	30–119	1–29	0
Right leg outstretched (45° supine)	100	31–99	1–30	0
Left leg outstretched (45° supine)	100	31–99	1–30	0

[a]Total QMG score range 0–39.

Attachment 3. Myasthenia Gravis Foundation of America Clinical Classification

Class	Clinical symptoms
I	Any ocular weakness
II	Mild Weakness. May also have ocular muscle weakness of any severity
II A	Predominantly affecting limb, axial muscles, or both. May also have lesser involvement of oropharyngeal, respiratory muscles or both
II B	Predominantly affecting orорpharyngeal, respiratory muscles, or both. May also have lesser or equal involvement of limb, axial muscles or both
III	Moderate weakness affecting other than ocular muscles. May also have ocular muscle weakness of any severity
III A	Predominantly affecting limb, axial muscles, or both. May also have lesser involvement of oropharyngeal, respiratory muscles or both
III B	Predominantly affecting orорpharyngeal, respiratory muscles, or both. May also have lesser or equal involvement of limb, axial muscles or both
IV	Severe weakness affecting other than ocular muscles. May also have ocular muscle weakness of any severity
IV A	Predominantly affecting limb, axial muscles, or both. May also have lesser involvement of oropharyngeal, respiratory muscles or both
IV B	Predominantly affecting orорpharyngeal, respiratory muscles, or both. May also have lesser or equal involvement of limb, axial muscles or both
V	Defined by intubation, with or without mechanical ventilation, except when employed during routine postoperative management

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MYASTHENIA GRAVIS MEDICATIONS PRIOR AUTHORIZATION FORM

Please complete and fax all requested information below including any progress notes, laboratory test results, or chart documentation as applicable to Highmark Wholecare Pharmacy Services. **FAX:** (888) 245-2049
If needed, you may call to speak to a Pharmacy Services Representative. **PHONE**: (800) 392-1147 Mon – Fri 8:30am to 5:00pm

PROVIDER INFORMATION

Requesting Provider:	Provider NPI:
Provider Specialty:	Office Contact:
State license #:	Office NPI:
Office Address:	Office Phone:
	Office Fax:

MEMBER INFORMATION

Member Name:	DOB:	
Member ID:	Member weight:	Height:

REQUESTED DRUG INFORMATION

Medication:	Strength:	
Directions:	Quantity:	Refills:
Is the member currently receiving requested medication? ☐ Yes ☐ No	Date Medication Initiated:	

Billing Information

This medication will be billed: ☐ at a pharmacy **OR** ☐ medically, JCODE:_____

Place of Service: ☐ Hospital ☐ Provider's office ☐ Member's home ☐ Other

Place of Service Information

Name:	NPI:
Address:	Phone:

MEDICAL HISTORY (Complete for ALL requests)

Diagnosis:	ICD Code:

Is there documentation of a positive serologic test for anti-acetylcholine antibodies? ☐ Yes ☐ No
Is there documentation of a positive serological test for anti-muscle specific tyrosine kinase (MUSK) ☐ Yes ☐ No

Is there documentation the member meets the Myasthenia Gravis Foundation of America Clinical Classification II to IV generalized myasthenia gravis? ☐ Yes ☐ No

What are the member's baseline Myasthenia Gravis-Specific Activities of Daily Living (MG-ADL) score, Quantitative Myasthenia Gravis (QMG) scale score and IgG levels? **Please list:** MG-ADL: QMG: IgG:

Has the member tried and failed treatment over 1 year or more with 2 or more immunosuppressive therapies either in combination or as monotherapy or tried and failed treatment over 1 year or more with at least 1 immunosuppressive therapy while on chronic plasmapheresis or plasma exchange (PE)? ☐ Yes- Please list in medication section below. ☐ No

Is the requested agent being used in combination with another Myasthenia Gravis medication? ☐ Yes, please list which one:
☐ No

CURRENT or PREVIOUS THERAPY

Medication Name	Strength/ Frequency	Dates of Therapy	Status (Discontinued & Why/Current)

REAUTHORIZATION

Is there documentation the member had a positive clinical response by an improvement in MG-ADL or QMG score? Please list improvement score: MG-ADL: QMG:

Has the member experienced an improvement with treatment? ☐ Yes ☐ No

SUPPORTING INFORMATION or CLINICAL RATIONALE

Prescribing Provider Signature	Date