MEDICAL MUTUAL®

# Drug Policy

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| **Policy:** | **231204** | **Initial Effective Date: 12/19/2024** |
| **Code(s):** | **HCPCS J3490** | **Annual Review Date: 12/19/2024** |
| **SUBJECT:** | **Zilbrysq ® (zilucoplan)** | **Last Revised Date: 12/19/2024** |

☐Subject to Site of Care

**Prior approval is required for some or all procedure codes listed in this Corporate Drug Policy.**

## OVERVIEW

Zilbrysq is a complement C5 inhibitor indicated for the treatment of generalized myasthenia gravis (gMG) in adult patients who are anti-acetylcholine receptor (AChR) antibody positive.

## POLICY STATEMENT

This policy involves the use of Zilbrysq. Prior authorization is recommended for pharmacy and medical benefit coverage of Zilbrysq. Approval is recommended for those who meet the conditions of coverage in the **Criteria, Dosing (medical benefit requests only), Initial/Extended Approval, Duration of Therapy**, and **Labs/Diagnostics** for the diagnosis provided. **Waste Management** applies for all covered conditions that are administered by a healthcare professional. **Conditions Not Recommended for Approval** are listed following the recommended authorization criteria and Waste Management section. Requests for uses not listed in this policy will be reviewed for evidence of efficacy and for medical necessity on a case-by-case basis.

Because of the specialized skills required for evaluation and diagnosis of patients treated with Zilbrysq as well as the monitoring required for AEs and long-term efficacy, initial approval requires Zilbrysq be prescribed by or in consultation with a physician who specializes in the condition being treated. All approvals for initial therapy are provided for the initial approval duration noted below; if reauthorization is allowed, a response to therapy is required for continuation of therapy unless otherwise noted below.

## RECOMMENDED AUTHORIZATION CRITERIA

Coverage of Zilbrysq is recommended in those who meet the following criteria:

1. **Generalized Myasthenia Gravis**. Approve if the patient meets ONE of the following (A or B):
   A. Initial Therapy. Approve if the patient meets the following (i, ii, iii, iv, v, vi, vii, viii AND ix)
      i. Patient is ≥ 18 years of AGE; AND
      ii. Patient has confirmed anti-acetylcholine receptor antibody-positive generalized myasthenia gravis; AND
      iii. Patient meets both of the following (a and b):

a) Myasthenia Gravis Foundation of America classification of II to IV; AND
b) Myasthenia Gravis Activities of Daily Living (MG-ADL) score of ≥ 6; AND

iv. Patient meets one of the following (a or b):
a) Patient received or is currently receiving pyridostigmine; OR
b) Patient has had inadequate efficacy, a contraindication, or significant intolerance to pyridostigmine; AND

v. Patient meets one of the following (a or b):
a) Patient received or is currently receiving two different immunosuppressant therapies for ≥ 1 year; OR
b) Patient had inadequate efficacy, a contraindication, or significant intolerance to two different immunosuppressant therapies; AND

Note: Examples of immunosuppressant therapies tried include azathioprine, cyclosporine, mycophenolate mofetil, methotrexate, tacrolimus, and cyclophosphamide.

vi. Patient has evidence of unresolved symptoms of generalized myasthenia gravis; AND
Note: Evidence of unresolved symptoms of generalized myasthenia gravis includes difficulty swallowing, difficulty breathing, and a functional disability resulting in the discontinuation of physical activity (e.g., double vision, talking, impairment of mobility); AND

vii. Prescriber has enrolled in the Zilbrysq REMS program; AND

viii. Patient has documentation of completion of meningococcal vaccination (for serogroups A, C, W, and Y, and serogroup B) at least 2 weeks prior to the first dose; AND

ix. The medication is being prescribed by or in consultation with a neurologist.

B. Patient is Currently Receiving Zilbrysq. Approve if the patient meets the following (i, ii, iii, AND iv):
i. Patient is ≥ 18 years of age; AND
ii. Patient is continuing to derive benefit from Zilbrysq, according to the prescriber; AND
Note: Examples of derived benefit include reductions in exacerbations of myasthenia gravis; improvements in speech, swallowing, mobility, and respiratory function.
iii. The prescriber and patient have been adherent to the REMS program throughout the course of therapy; AND
iv. The medication is being prescribed by or in consultation with a neurologist.

**Dosing in Generalized Myasthenia Gravis.** *Dosing must meet the following:*
Administer, subcutaneously, the appropriate prefilled syringe once daily based on the patient's actual body weight. For patients less than 56 kg, administer 16.6 mg daily. For patients between 56 kg and less than 77 kg, administer 23 mg daily. For patients 77 kg and above, administer 32.4 mg daily.

**Initial Approval/ Extended Approval.**
**A)** *Initial Approval: 6 months*
**B)** *Extended Approval:1 year*

## CONDITIONS NOT RECOMMENDED FOR APPROVAL

Zilbrysq has not been shown to be effective, or there are limited or preliminary data or potential safety concerns that are not supportive of general approval for the following conditions. (Note: This is not an exhaustive list of Conditions Not Recommended for Approval).

1. **Concomitant Use with Another Complement Inhibitor, a Neonatal Fc Receptor Blocker, or a Rituximab Product.** There is no evidence to support concomitant use of Zilbrysq with another complement inhibitor, a neonatal Fc receptor blocker, or a rituximab product.
   Note: Examples of complement inhibitors are Soliris (eculizumab intravenous infusion) and Ultomiris (ravulizumab-cwvz intravenous infusion or subcutaneous injection).
   Note: Examples of Neonatal Fc receptor blockers are Rystiggo [rozanolixizumab-noli subcutaneous infusion) Vyvgart (efgartigimod alfa-fcab intravenous infusion), and Vyvgart Hytrulo (efgartigimod alfa and hyaluronidase-qvfc subcutaneous injection).

2. Coverage is not recommended for circumstances not listed in the Recommended Authorization Criteria. Criteria will be updated as new published data are available.

**Documentation Requirements:**

The Company reserves the right to request additional documentation as part of its coverage determination process. The Company may deny reimbursement when it has determined that the drug provided or services performed were not medically necessary, investigational or experimental, not within the scope of benefits afforded to the member and/or a pattern of billing or other practice has been found to be either inappropriate or excessive. Additional documentation supporting medical necessity for the services provided must be made available upon request to the Company. Documentation requested may include patient records, test results and/or credentials of the provider ordering or performing a service. The Company also reserves the right to modify, revise, change, apply and interpret this policy at its sole discretion, and the exercise of this discretion shall be final and binding.

## REFERENCES

1. Zilbrysq subcutaneous injection [prescribing information]. Symra, GA: UCB: October 2023.
2. National Institute of Neurological Disorders and Stroke (NINDS). Myasthenia Gravis Fact Sheet. National Institutes of Health (NIH) Publication No. 17-768. Publication last updated: March 2020. Available at: https://www.ninds.nih.gov/sites/default/files/migrate-documents/myasthenia_gravis_e_march_2020_508c.pdf Accessed on October 18, 2023.
3. Sanders DB, Wolfe GI, Benatar M, et al. International consensus guidance for management of myasthenia gravis. *Neurology*. 2016;87:419–425.
4. Howard JF, Bresch S, Genge A, et al on behalf of the RAISE study team. Safety and efficacy of zilucoplan in patients with generalized myasthenia gravis (RAISE): a randomized, double-blind, placebo-controlled, phase 3 study. *Lancet Neurology*. 2023;22:395-406.
5. Narayanaswami P, Sanders DB, Wolfe G, et al. International Consensus Guidance for Management of Myasthenia Gravis: 2020 Update. *Neurology*. 2021;96(3):114-122.

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# Drug Policy

**FOR MEDICAL BENEFIT COVERAGE REQUESTS:**

**Prior approval is required for HCPCS Codes J3490**

| **HCPCS Code(s):** | |
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| J3490 | Unclassified drugs |