Effective Date:08/01/2024
Reviewed: 5/2024

SPECIALTY GUIDELINE MANAGEMENT

ZILBRYSQ (zilucoplan)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indication
Zilbrysq is indicated for the treatment of generalized myasthenia gravis (gMG) in adult patients who are anti-acetylcholine receptor (AChR) antibody positive.

All other indications are considered experimental/investigational and not medically necessary.

II. DOCUMENTATION

Submission of the following information is necessary to initiate the prior authorization review:

A. For initial requests chart notes, medical records, or claims history documenting:
   1. Positive anti-acetylcholine receptor (AChR) antibody test
   2. Clinical classification of myasthenia gravis score
   3. MG activities of daily living score
   4. Previous medications tried, including response to therapy. If therapy is not advisable, documentation of clinical reasons to avoid therapy.

B. For continuation requests: Chart notes or medical record documentation supporting positive clinical response.

III. CRITERIA FOR INITIAL APPROVAL

Generalized myasthenia gravis (gMG)
Authorization of 6 months may be granted for the treatment of generalized myasthenia gravis (gMG) when all of the following criteria are met:

1. Member is ≥ 18 years of age
2. Prescribed by or in consultation with a neurologist
3. Diagnosis of gMG with a positive test for Anti-acetylcholine receptor (AChR) antibodies
4. Myasthenia Gravis Foundation of America (MGFA) clinical classification II to IV at the start of therapy
5. MG activities of daily living (MG-ADL) total score ≥6
6. Meets one of the following:

   a. Member has had an inadequate response or intolerable adverse event to at least two immunosuppressive therapies over the course of at least 12 months (e.g., azathioprine, corticosteroids, cyclosporine, methotrexate, mycophenolate, tacrolimus)
   b. Member has had an inadequate response or intolerable adverse event to at least one immunosuppressive therapy and intravenous immunoglobulin (IVIG) over the course of at least 12 months
   c. Member has a documented clinical reason to avoid therapy with immunosuppressive agents and IVIG
7. The member has an inadequate response or contraindication to one of the following:
   a. efgartigimod (Vyvgart) OR efgartigimod-hyaluronidase (Vyvgart Hytrulo)
   b. rozanolixizumab (Rystiggo)
8. Will not be used in combination with Soliris (eculizumab), Ultomiris (ravulizumab), Uplizna(inebilizumab), Vyvgart/Vyvgart Hytrulo (efgartigimod), Rituxan (rituximab), Rystiggo (rozanolixizumab) or chronic IVIG
9. The dose does not exceed the following based on actual body weight (current weight provided):
   a. Less than 56kg: 16.6mg daily
   b. 56kg to less than 77kg: 23mg daily
   c. 77 kg and above: 32.4mg daily

IV. CONTINUATION OF THERAPY

Authorization of 6 months may be granted for continued treatment in members requesting reauthorization when there is no evidence of unacceptable toxicity or disease progression while on the current regimen and member demonstrates a positive response to therapy (e.g., improvement in MG-ADL score, MG Manual Muscle Test (MMT), MG Composite).

V. QUANTITY LIMIT

Zilbrysq 16.6mg/0.416ml, 23mg/0.574ml, 32.4mg/0.81ml: 1 syringe per day or 28 syringes per 28 days

VI. REFERENCES

1. Zilbrysq [package insert]. Smyrna, GA: UCB, Inc.; March 2024.
2. Sanders D, Wolfe G, Benatar M et al. International consensus guidance for management of myasthenia gravis. *Neurology*. 2021; 96 (3) 114-122.
3. Howard JF, et al. Safety and efficacy of zilucoplan in patients with generalised myasthenia gravis (RAISE): a randomised, double-blind, placebo-controlled, phase 3 study. Lancet Neurol. 2023;22(5):395-406.
4. Barnett C, Herbelin L, Dimachkie MM, Barohn RJ. Measuring Clinical Treatment Response in Myasthenia Gravis. Neurol Clin. 2018 May;36(2):339-353.