MEDICAL MUTUAL®

Drug Policy

Policy:	Xolremdi™ (mavorixafor)	Annual Review Date: 06/20/2024 Last Revised Date: 06/20/2024

OVERVIEW

Xolremdi, a CXC chemokine receptor 4 (CXCR4) antagonist, is indicated for the treatment of **WHIM syndrome** (warts, hypogammaglobulinemia, infections and myelokathexis) to increase the number of circulating mature neutrophils and lymphocytes in adults and children ≥ 12 years of age.[1]

POLICY STATEMENT

This policy involves the use of Xolremdi. Prior authorization is recommended for pharmacy benefit coverage of Xolremdi. Approval is recommended for those who meet the conditions of coverage in the **Criteria and Initial/Extended Approval** for the diagnosis provided. **Conditions Not Recommended for Approval** are listed following the recommended authorization criteria. 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 Xolremdi as well as the monitoring required for adverse events and long-term efficacy, initial approval requires Xolremdi 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 Xolremdi is recommended in those who meet the following criteria:

FDA-Approved Indication

1. **WHIM syndrome.** Approve Xolremdi for the duration noted if the patient meets ONE of the following (A or B):
   - **A)** Initial Therapy. Approve if the patient meets ALL of the following (i, ii, iii, and iv):
     - **i.** Patient is ≥ 12 years of age; AND
     - **ii.** Genetic testing confirms pathogenic and or likely pathogenic variants in the CXCR4 gene; [Documentation required] AND
     - **iii.** Patient meets ONE of the following (a or b):
       - **a)** At baseline, patient had an absolute neutrophil count ≤ 400 cells/µL; [Documentation required] OR
       - **b)** At baseline, patient had a white blood cell count ≤ 400 cells/µL; [Documentation required] AND
     - **iv.** The medication is prescribed by or in consultation with an immunologist, hematologist or dermatologist.
   - **B)** Patient is Currently Receiving Xolremdi. Provide extended approval if all of the following are met:
     - **i.** Patient is ≥ 12 years of age; AND

ii. The medication is prescribed by or in consultation with an immunologist, hematologist or dermatologist; AND
iii. if, according to the prescriber, the patient is continuing to derive benefit from Xolremdi as determined by the most recent objective measurement.

Note: Examples of objective measurements of a response to Xolremdi therapy are reduced frequency, duration, or severity of infections, less frequent treatment with antibiotics, fewer warts, or improved or stabilized clinical signs/symptoms of WHIM syndrome (e.g., absolute neutrophil count, white blood cell count, and absolute lymphocyte count).

Initial Approval/ Extended Approval.
A) *Initial Approval:* 3 months
B) *Extended Approval:* 1 year

CONDITIONS NOT RECOMMENDED FOR APPROVAL

Xolremdi 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. Coverage is not recommended for patients taking Plerixafor.

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. Xolremdi™ oral capsules [prescribing information]. Boston, MA: X4 Pharmaceuticals; June 2024.
2. Mavorixafor. *Lexi-Drugs*. UpToDate Lexidrug. UpToDate Inc. https://online.lexi.com. Accessed June 7, 2024.