PHARMACY POLICY STATEMENT
Marketplace

DRUG NAME	Xolremdi (mavorixafor)
BENEFIT TYPE	Pharmacy
STATUS	Prior Authorization Required

Xolremdi, approved by the FDA in 2024, is a CXC chemokine receptor 4 (CXCR4) antagonist indicated in patients 12 years of age and older with WHIM syndrome (warts, hypogammaglobulinemia, infections and myelokathexis) to increase the number of circulating mature neutrophils and lymphocytes.
WHIM syndrome is a primary immunodeficiency characterized by retention of leukocytes in the bone marrow (myelokathexis), resulting in neutropenia, leukopenia, and sometimes hypogammaglobulinemia or warts, as well as recurrent infections and predisposition to malignancy. It is caused by mutations in *CXCR4,* resulting in hyperactivation of the CXCR4-CXCL12 pathway and myelokathexis.
Xolremdi, the first approved treatment for WHIM syndrome, inhibits the hyperresponsiveness to the CXCR4 ligand CXCL12 to increase mobilization of neutrophils and lymphocytes from the bone marrow into peripheral circulation. This results in improvement in absolute neutrophil counts (ANC), improvement in absolute lymphocyte counts (ALC), and a reduction of infection frequency, severity, and duration.

Xolremdi (mavorixafor) will be considered for coverage when the following criteria are met:

WHIM syndrome (warts, hypogammaglobulinemia, infections and myelokathexis)

For **initial** authorization:
1. Member is at least 12 years of age; AND
2. Medication must be prescribed by or in consultation with an immunologist; AND
3. Member has a diagnosis of WHIM syndrome confirmed by genetic testing that shows mutation of the *CXCR4* gene; AND
4. Member's labs show absolute neutrophil count (ANC) ≤400 cells/μL in the absence of infection; AND
5. Member's chart notes show baseline ALC and infection history.
6. **Dosage allowed/Quantity limit:**
   Weight more than 50 kg: 400 mg orally once daily (4 capsules)
   Weight less than or equal to 50 kg: 300 mg orally once daily (3 capsules)
   QL: 120 capsules per 30 days (1 capsule = 100 mg)

If all the above requirements are met, the medication will be approved for 6 months.

For **reauthorization**:
1. Chart notes must show clinically significant improvement of ANC, ALC, and/or reduced infections since starting Xolremdi.

If all the above requirements are met, the medication will be approved for an additional 12 months.

CareSource considers Xolremdi (mavorixafor) not medically necessary for the treatment of conditions that are not listed in this document. For any other indication, please refer to the Off-Label policy.

DATE	ACTION/DESCRIPTION
05/01/2024	New policy for Xolremdi created.

References:

1. Xolremdi [prescribing information]. X4 Pharmaceuticals, Inc.; 2024.
2. Badolato R, Alsina L, Azar A, et al. Phase 3 randomized trial of mavorixafor, CXCR4 antagonist, in WHIM syndrome. *Blood*. Published online April 21, 2024. doi:10.1182/blood.2023022658
3. Badolato R, Donadieu J; WHIM Research Group. How I treat warts, hypogammaglobulinemia, infections, and myelokathexis syndrome. *Blood*. 2017;130(23):2491-2498. doi:10.1182/blood-2017-02-708552

Effective date: 10/01/2024
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