BlueCross BlueShield
Federal Employee Program
Blue Cross Blue Shield Association
750 9th St NW, Suite 900
Washington, D.C. 20001
1-800-624-5060
Fax 1-877-378-4727

5.85.061

Section:	Prescription Drugs	**Effective Date:**	October 1, 2024
Subsection:	Hematological Agents	**Original Policy Date:**	May 31, 2024
Subject:	Xolremdi	**Page:**	1 of 4

Last Review Date: September 6, 2024

Xolremdi

Description

Xolremdi (mavorixafor)

Background
Xolremdi (mavorixafor) is a CXC Chemokine Receptor 4 (CXCR4) antagonist that blocks the binding of the CXCR4 ligand, stromal-derived factor-1α (SDF-1α)/CXC Chemokine Ligand 12 (CXCL12) SDF-1/CXCR4 plays a role in trafficking and homing of leukocytes to and from the bone marrow compartment. Gain of function mutations in the CXCR4 receptor gene that occur in patients with WHIM syndrome (warts, hypogammaglobulinemia, infections and myelokathexis) lead to increased responsiveness to CXCL12 and retention of leukocytes in the bone marrow. Xolremdi inhibits the response to CXCL12 in both wild-type and mutated CXCR4 mobilization of neutrophils and lymphocytes from the bone marrow into peripheral circulation (1).

Regulatory Status
FDA-approved indications: Xolremdi is a CXC chemokine receptor 4 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 (1).

Xolremdi has warnings regarding embryo-fetal toxicity and QTc interval prolongation. Women of reproductive potential should be advised to use effective contraception during treatment with Xolremdi and for three weeks after the final dose. Any modifiable risk factors for QTc

prolongation (e.g., hypokalemia) should be corrected. QTc should be assessed at baseline and monitored during treatment as clinically indicated in patients with risk factors for QTc prolongation (1).

The safety and effectiveness of Xolremdi in pediatric patients less than 12 years of age have not been established (1).

Related policies

Policy

This policy statement applies to clinical review performed for pre-service (Prior Approval, Precertification, Advanced Benefit Determination, etc.) and/or post-service claims.

Xolremdi may be considered **medically necessary** if the conditions indicated below are met.

Xolremdi may be considered **investigational** for all other indications.

Prior-Approval Requirements

Age 12 years of age or older

Diagnosis

Patient must have the following:

WHIM syndrome (warts, hypogammaglobulinemia, infections and myelokathexis)

AND ALL of the following:

a. Patient has a genotype-confirmed variant of CXCR4 consistent with WHIM syndrome
b. Absolute neutrophil count (ANC) ≤ 400 cells/μL
c. Females of reproductive potential **only**: patient will be advised to use effective contraception during treatment with Xolremdi and for 3 weeks after the last dose

Prior – Approval *Renewal* Requirements

Age 12 years of age or older

Diagnosis

Patient must have the following:

WHIM syndrome (warts, hypogammaglobulinemia, infections and myelokathexis)

AND ALL of the following:

a. Patient has had a clinical benefit from therapy (e.g., increased ANC or ALC or a reduction in infections)
b. Females of reproductive potential **only**: patient will be advised to use effective contraception during treatment with Xolremdi and for 3 weeks after the last dose

Policy Guidelines

Pre – PA Allowance

None

Prior - Approval Limits

Quantity 400 mg per day

Duration 12 months

Prior – Approval *Renewal* Limits

Same as above

Rationale

Summary

Xolremdi (mavorixafor) is a CXC chemokine receptor 4 antagonist indicated in patients with WHIM syndrome (warts, hypogammaglobulinemia, infections and myelokathexis) to increase the number of circulating mature neutrophils and lymphocytes. Xolremdi may cause embryo-fetal toxicity and QTc interval prolongation. The safety and effectiveness of Xolremdi in pediatric patients less than 12 years of age have not been established (1).

Prior authorization is required to ensure the safe, clinically appropriate, and cost-effective use of Xolremdi while maintaining optimal therapeutic outcomes.

References

1. Xolremdi [package insert]. Boston MA: X4 Pharmaceuticals, Inc.; April 2024.

Policy History

Date	Action
May 2024	Addition to PA
September 2024	Annual review

Keywords

This policy was approved by the FEP® Pharmacy and Medical Policy Committee on September 6, 2024 and is effective on October 1, 2024.