Mass General Brigham
Health Plan

# Xolremdi (mavorixafor)
# Effective 11/01/2024

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| **Plan** | ☐ MassHealth UPPL<br>☒Commercial/Exchange | **Program Type** | ☒ Prior Authorization<br>☐ Quantity Limit<br>☐ Step Therapy |
| **Benefit** | ☒ Pharmacy Benefit<br>☐ Medical Benefit | | |
| **Specialty Limitations** | This medication has been designated specialty and must be filled at a contracted specialty pharmacy. | | |
| **Contact Information** | **Medical and Specialty Medications**<br>All Plans \| Phone: 877-519-1908 \| Fax: 855-540-3693 | | |
| | **Non-Specialty Medications**<br>All Plans \| Phone: 800-711-4555 \| Fax: 844-403-1029 | | |
| **Exceptions** | N/A | | |

**Overview**
Xolremdi (mavorixafor) 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.

**Coverage Guidelines**
Authorization may be reviewed for members new to the plan within the last 90 days who are currently receiving treatment with the requested medication excluding when the product is obtained as samples or via manufacturer's patient assistance programs.
**OR**
Authorization may be granted for members when ALL the following criteria are met:

1. Member has a diagnosis of WHIM (warts, hypogammaglobulinemia, infections, and myelokathexis) syndrome
2. Member is 12 years of age or older
3. Documentation member has a genotype-confirmed variant of CXCR4
4. Xolremdi is prescribed by or in consultation with an immunologist, hematologist, dermatologist, or infectious disease specialist
5. Member has an absolute neutrophil count (ANC) ≤ 400 cells/microliter

**Continuation of Therapy**
Requests for reauthorization will be approved when all of the following criteria are met:

1. Documentation the member has had a positive clinical response to therapy (e.g., improvement in absolute neutrophil count (ANC) or absolute lymphocyte count (ALC); reduced infection frequency, duration, or severity; decreased number of warts; improvement in symptoms)

**Limitations**

1. Initial requests will be approved for 6 months.
2. Reauthorization requests will be approved for 12 months.

**References**


1. Badolato R, Alsina L, Azar A, et al. Phase 3 randomized trial of mavorixafor, CXCR4 antagonist, in WHIM syndrome. *Blood*. 2024. doi:10.1182/blood.2023022658
2. Badolato R, Donadieu J; WHIM Research Group. How I treat warts, hypogammaglobulinemia, infections, and myelokathexis syndrome. *Blood*. 2017;130(2):2491-2498. doi:10.1182/blood-2017-02-708552
3. Bonilla FA, Khan DA, Ballas ZK, et al. Practice parameter for the diagnosis and management of primary immunodeficiency. *J Allergy Clin Immunol*. 2015;136(5):1186-205.e2078. doi:10.1016/j.jaci.2015.04.049.
4. Efficacy and Safety Study of Mavorixafor in Participants With Warts, Hypogammaglobulinemia, Infections, and Myelokathexis (WHIM) Syndrome. ClinicalTrials.gov identifier: NCT03995108. Updated 05/02/2024. Accessed 06/17/2024. https://clinicaltrials.gov/study/NCT03995108
5. European Society of Society for Immunodeficiencies, Pan-American Group for Immunodeficiency. Diagnostic criteria for WHIM (warts-hypogammaglobulinemia-infections-myelokathexis) syndrome. November 2019. Accessed June 2024. https://esid.org/Working-Parties/Registry-Working-Party/Diagnosis-criteria
6. Food and Drug Administration (FDA). Xolremdi integrated review. FDA. August 31, 2023. Accessed June 17, 2024. https://www.accessdata.fda.gov/drugsatfda_docs/nda/2024/218709Orig1s000IntegratedR.pdf
7. Geier CB, Ellison M, Cruz R, Pawar S, et al. Disease progression of WHIM syndrome in an international cohort
8. of 66 pediatric and adult patients. *J Clin Immunol*. 2022;42:1748-1765. doi: 10.1007/s10875-022-01312-7.
9. Heusinkveld LE, Majumdar S, Gao JL, McDermott DH. WHIM syndrome: from pathogenesis towards personalized medicine and cure. *J Clin Immunol*. 2019;39(6):532-556. doi:10.1007/s10875-019-00665-w.
10. Heusinkveld LE, Yim E, Yang A, Azani AB, Liu Q, et al. Pathogenesis, diagnosis and therapeutic strategies in WHIM syndrome immunodeficiency. *Expert Opin Orphan Drugs*. 2017;5(1):813-25. doi:10.1080/21678707.2017.1375403
11. McDermott DH, Velez D, Cho E, Cowen EW, et al. A phase III randomized crossover trial of plerixafor versus G-CSF for treatment of WHIM syndrome. *J Clin Invest*. 2023;133(19):e164918. doi: 10.1172/JCI164918.
12. Orphanet. Orphan Drugs Database. Updated May 28, 2024. Accessed June 17, 2024. https://www.orpha.net/en/drug
13. Perez EE, Orange JS, Bonilla F, Chinen J, et al. Update on the use of immunoglobulin in human disease: a review of evidence. *J Allergy Clin Immunol.* 2017;139:S1-46.
14. Xolremdi [package insert]. X4 Pharmaceuticals, Inc. April 2024.


**Review History**

09/11/2024 – Reviewed at September P&T. Effective 11/01/2024.