Reference number(s)
7401-A

Standard Medicare Part B Management Loargys

Products Referenced by this Document

Drugs that are listed in the following table include both brand and generic and all dosage forms and strengths unless otherwise stated. Over-the-counter (OTC) products are not included unless otherwise stated.

Brand Name	Generic Name
Loargys	pegzilarginase-nbln

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 Indications[1]

Loargys is indicated for the treatment of hyperargininemia in adult and pediatric patients two years of age and older with Arginase 1 Deficiency (ARG1-D), in conjunction with dietary protein restriction.

This indication is approved under accelerated approval based on reduction of plasma arginine. Continued approval for this indication may be contingent upon verification and description of clinical benefit in a confirmatory trial.

All other indications will be assessed on an individual basis. Submissions for indications other than those listed in this criteria should be accompanied by supporting evidence from Medicare approved compendia.

Documentation

The following documentation must be available, upon request, for all submissions:

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Initial Requests

- Lab results documenting elevated plasma arginine levels (i.e., greater than or equal to 250 micromol/L).
- Chart notes, medical records, or lab results documenting one of the following:
  - Pathogenic (or likely pathogenic) variant in the ARG1 gene.
  - Enzyme assay demonstrating a deficiency of arginase enzyme activity (i.e., <1% of normal) in erythrocytes.

Continuation Requests

- Lab results documenting pre-dose plasma arginine levels.

Coverage Criteria

Arginase 1 Deficiency (ARG1-D)[1-4]

Authorization of 12 months may be granted for treatment of ARG1-D when all of the following criteria are met:

- The member is 2 years of age or older.
- The member has elevated plasma arginine levels (i.e., greater than or equal to 250 micromol/L) prior to initiating therapy with the requested medication.
- The diagnosis of ARG1-D is confirmed by one of the following:
  - Pathogenic (or likely pathogenic) variant in the ARG1 gene
  - Enzyme assay demonstrating a deficiency of arginase enzyme activity (i.e., <1% of normal) in erythrocytes
- The requested medication will be used in conjunction with dietary protein restriction.
- Baseline and subsequent pre-dose plasma arginine levels will be collected and monitored as outlined in the manufacturer's prescribing information.
- The dose of the requested medication will be adjusted as outlined in the manufacturer's prescribing information if the member's pre-dose plasma arginine levels fall outside of the therapeutic range (i.e., 50 micromol/L to 150 micromol/L).
- Initial and subsequent doses of the requested medication will not exceed 0.2 mg/kg once weekly.

Continuation of Therapy

All members (including new members) requesting authorization for continuation of therapy must be currently receiving therapy with the requested agent.

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Authorization for 12 months may be granted when all of the following criteria are met:

- The member is currently receiving therapy with the requested medication.
- The requested medication is being used to treat an indication in the coverage criteria section.
- Either of the following criteria apply:
    - The member has achieved a pre-dose plasma arginine level between 50 micromol/L and 150 micromol/L, or
    - The member has not achieved a pre-dose plasma arginine level between 50 micromol/L and 150 micromol/L, and the dose will be adjusted as outlined in the manufacturer's prescribing information.
- The requested dose does not exceed 0.2 mg/kg once weekly.

Summary of Evidence

The contents of this policy were created after examining the following resources:

- The prescribing information for Loargys.
- The available compendium
    - National Comprehensive Cancer Network (NCCN) Drugs and Biologics Compendium
    - Micromedex DrugDex
    - American Hospital Formulary Service- Drug Information (AHFS-DI)
    - Lexi-Drugs
    - Clinical Pharmacology
- Efficacy and safety of pegzilarginase in arginase 1 deficiency (PEACE): a phase 3, randomized, double-blind, placebo-controlled, multi-centre trial.
- Arginase Deficiency – Gene Reviews.
- Suggested guidelines for the diagnosis and management of urea cycle disorders: First revision.

After reviewing the information in the above resources, the FDA-approved indications listed in the prescribing information for Loargys are covered.

Explanation of Rationale

Support for FDA-approved indications can be found in the manufacturer's prescribing information.

References

1. Loargys [package insert]. Chicago, IL: Immedica Pharma US, Inc.; February 2026.

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2. Russo RS, Gasperini S, Bubb G, et al. Efficacy and safety of pegzilarginase in arginase 1 deficiency (PEACE): a phase 3, randomized, double-blind, placebo-controlled, multi-centre trial. EClinicalMedicine. 2024;68:102405. Published 2024 Jan 12. doi:10.1016/j.eclinm.2023.102405.
3. Sun A, Crombez EA, Wong D. Arginase Deficiency. 2004 Oct 21 [Updated 2020 May 28]. In: Adam MP, Bick S, Mirzaa GM, et al., editors. GeneReviews® [Internet]. Seattle (WA): University of Washington, Seattle; 1993-2026. Available from: https://www.ncbi.nlm.nih.gov/books/NBK1159/.
4. Häberle J, Burlina A, Chakrapani A, et al. Suggested guidelines for the diagnosis and management of urea cycle disorders: First revision. J Inherit Metab Dis. 2019;42(6):1192-1230. doi:10.1002/jimd.12100.

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