MEDICATION POLICY:
Loargys (pegzilarginase-nbln)

Applicable Drugs: Loargys (pegzilarginase-nbln)

Preferred: N/A

Non-preferred: N/A

Date of Origin: 7/29/2026

Date Last Reviewed / Revised: 7/29/2026

Prior Authorization Criteria

(May be considered medically necessary when criteria I through V are met)

I. Documented diagnosis of hyperargininemia due to arginase 1 deficiency (ARG1-D) and meets criteria A through C:
   A. Documented ARG1-D diagnosis as confirmed by one of the following:
      i. Genetic testing confirms biallelic pathogenic variants in the ARG-1 gene.
      ii. Reduced RBC arginase enzyme activity (i.e., <1% of normal in red blood cell extracts).
   B. Elevated plasma arginine levels (i.e., greater than or equal to 250 µmol/L).
   C. The requested medication will be used as an adjunct to a protein-restricted diet.

II. Minimum age requirement: greater than or equal to 2 years old.

III. Treatment must be prescribed by or in consultation with a geneticist, neurologist, or metabolic disease specialist.

IV. Request is for a medication with the appropriate FDA labeling, or its use is supported by current clinical practice guidelines.

V. Refer to the plan document for the list of preferred products. If the requested agent is not listed as a preferred product, must have a documented failure, intolerance, or contraindication to a preferred product(s).

Exclusion Criteria

- History of liver or hematopoietic transplant
- Known active infection with human immunodeficiency virus (HIV), hepatitis B, or hepatitis C

Other Criteria

- N/A

Quantity / Days Supply Restrictions

- Loargys is available in 2 mg/0.4 mL and 5 mg/mL in a single-dose vial.

- Dose of Loargys is calculated based on patient's actual body weight.
- Dosing will vary based on patient weight and plasma arginine level. Do not exceed 0.2 mg/kg once weekly.

APPROVAL LENGTH

- **Authorization:** 6 months
- **Re-Authorization:** 12 months, with an updated letter of medical necessity or progress notes showing current medical necessity criteria are met and that the medication is effective, such as plasma arginine levels between 40 µmol/L and 115 µmol/L. Provider attestation that the member has been adherent to a protein-restricted diet and will continue this diet during treatment with Loargys.

APPENDIX

Loargys Dosing Information:

- Initial dose: 0.1 mg/kg IV once weekly using actual body weight
- Dose Adjustments
    - If two consecutive weekly pre-dose levels are outside target range:
        - < 50 micromolar: increase dose by 0.05 mg/kg/week
        - > 150 micromolar: decrease dose by 0.05 mg/kg/week
- Maximum dose: 0.2 mg/kg once weekly
- After 8 weeks of IV therapy, patients may transition to subcutaneous administration at the same weekly dose.

REFERENCES

1. Loargys (pegzilarginase-nbln) injection, for intravenous or subcutaneous use [prescribing information]. Immedica Pharma AB; 2026. Accessed July 27, 2026. https://www.accessdata.fda.gov/drugsatfda_docs/label/2026/761211Orig1s000lbl.pdf
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. Bin Sawad A, Jackimiec J, Bechter M, et al. Epidemiology, methods of diagnosis, and clinical management of patients with arginase 1 deficiency (ARG1-D): A systematic review. Mol Genet Metab. 2022;137(1-2):153-163. doi:10.1016/j.ymgme.2022.08.005
5. Arginase-1 Deficiency. Accessed July 29, 2026. https://rarediseases.org/rare-diseases/arginase-deficiency/
6. 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

DISCLAIMER: Medication Policies are developed to help ensure safe, effective and appropriate use of selected medications. They offer a guide to coverage and are not intended to dictate to providers how to practice medicine. Refer to Plan for individual adoption of specific Medication Policies. Providers are expected to exercise their medical judgement in providing the most appropriate care for their patients.