

Loargys (pegzilarginase-nbln) for SC Administration
Effective 10/01/2026

Plan	☐ MassHealth UPPL ☒ Commercial/Exchange		Program Type	☒ Prior Authorization
Benefit	☒ Pharmacy Benefit ☐ Medical Benefit			☐ Quantity Limit ☐ Step Therapy
Specialty Limitations	N/A			
Contact Information	Medical Benefit Pharmacy Benefit	Phone: 833-895-2611 Phone: 800-711-4555	Fax: 888-656-6671 Fax: 844-403-1029	
Exceptions	N/A			

Overview

Loargys (pegzilarginase-nbln) is an arginine specific enzyme indicated for the treatment of hyperargininemia in adult and pediatric patients 2 years of age and older with Arginase 1 Deficiency (ARG1-D), in conjunction with dietary protein restriction.

Coverage Guidelines

If member is new to the plan (as evidenced by coverage effective date of less than or equal to 90 days), submission of medical records documenting that the member is currently receiving treatment with the requested drug, excluding when the product is obtained as samples or via manufacturer's patient assistance programs

OR

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

- Confirmed diagnosis of Arginase-1 Deficiency (ARG1-D) variants in *ARG1* on molecular genetic testing, as demonstrated by at least ONE of the following features of the disease:
 - Identification of biallelic pathogenic (or likely pathogenic) variants in *ARG1* on molecular genetic testing
 - Reduced RBC arginase enzyme activity (i.e., < 1% of normal in red blood cell extracts)
- Member will maintain a protein-restricted diet (i.e., will maintain the current level of protein consumption, including natural protein and essential amino acid (EAA) supplementation)
- Member has history of hyperargininemia (e.g., two to three times the upper limit of normal)
- Member has not experienced a hyperammonemic episode within the six weeks prior to the start of therapy (defined as an event in which the member has an ammonia level $\geq 100 \mu\text{M}$ with one or more symptoms related to hyperammonemia requiring hospitalization or emergency room management)
- Member plasma arginine concentration will be monitored as clinically indicated
- Member has not previously received a liver transplant
- Submission of medical records (e.g., chart notes) documenting that the member has received at least eight weeks of therapy with Loargys IV and is transitioning to subcutaneous administration

Continuation of Therapy

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

1. Request is to continue subcutaneous administration of Loargys
2. Absence of unacceptable toxicity from requested medication (e.g., severe hypersensitivity reactions, anaphylaxis, etc.)
3. Member has demonstrated a beneficial response to therapy compared to pretreatment baseline, as evidenced by stabilization or improvement in plasma arginine levels and/or complications of disease manifestations

Limitations

1. Initial approvals and reauthorizations will be granted for 12 months

References

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2. Burlina A, Ardisson A, Battini R, et al. Arginase 1 deficiency: a treatable form of spastic paraplegia. *Neurol Sci.* 2025;46(9):4219-4228. doi:10.1007/s10072-025-08153-3
3. Catsburg C, Anderson S, Upadhyaya N, Bechter M. Arginase 1 Deficiency: using genetic databases as a tool to establish global prevalence. *Orphanet J Rare Dis.* 2022;17(1):94. Published 2022 Mar 2. doi:10.1186/s13023-022-02226-8
4. Guyatt G, Oxman AD, Vist GE, et al. GRADE: An emerging consensus on rating quality of evidence and strength of recommendations. *BMJ.* 2008;336:924-926.
5. 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
6. McNutt M, Rutsch F, Russo RS, et al. Long-term efficacy and tolerability of pegzilarginase in arginase 1 deficiency: Results of two international multicenter open-label extension studies. *J Inherit Metab Dis.* 2025;48(4):e70066. doi:10.1002/jimd.70066
7. Loargys (pegzilarginase-nbln) [prescribing information]. Chicago, IL: Immedica Pharma US Inc.; February 2026.
8. 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
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Review History

07/15/2026 – Created and reviewed at July P&T. Effective 10/01/2026.

