Loargys (pegzilarginase-nbln)

Effective: January 1, 2027

Guideline Type	☒ Prior Authorization ☐ Non-Formulary ☐ Step-Therapy ☐ Administrative

Applies to:

Public Plans Products

☒ Tufts Health One Care*, (a dual eligible product); Fax 617-673-0956

*The MNG applies to Tufts Health One Care members unless a less restrictive LCD or NCD exists.

Senior Products

☒ Tufts Health Plan Senior Care Options (SCO), (a dual-eligible product); Fax 617-673-0956

☒ Tufts Medicare Preferred HMO, (a Medicare Advantage product); Fax 617-673-0956

☒ Tufts Medicare Preferred PPO, (a Medicare Advantage product); Fax 617-673-0956

Note: While you may not be the provider responsible for obtaining prior authorization, as a condition of payment you will need to ensure that prior authorization has been obtained.

Overview

Arginase 1 deficiency (ARG1-D) is one of eight rare inherited urea cycle disorder subtypes. It is caused by ARG1 gene variants and leads to hyperargininemia, a condition characterized by spasticity, seizures, developmental delay, and intellectual disability. ARG1-D affects approximately 250 individuals in the United States.

The urea cycle (UC) is responsible for the clearance of nitrogen waste that results from protein turnover and nitrogen metabolism. Urea cycle disorders (UCDs) are inherited deficiencies in the enzymes (e.g. arginase 1 [ARG1]) or transporters involved in the UC pathway. When these enzymes are deficient, toxic ammonia compounds cannot be cleared from the body

The accelerated approval was supported by results from the Phase 3 PEACE trial, in which patients ≥2 years of age with ARG1-D were randomized 2:1 to intravenous Loargys or placebo once weekly for 24 weeks. At 24 weeks, Loargys significantly reduced plasma arginine versus placebo. Target arginine levels were achieved in 90% of Loargys-treated patients versus 0% with placebo.

Food and Drug Administration (FDA) - Approved Indications:

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

Clinical Guideline Coverage Criteria

The plan may authorize coverage of Loargys (Pegzilarginase-nbln) for Members when **ALL** of the following criteria are met:

Initial Authorization Criteria

1. Documented hyperargininemia or Arginase-1 deficiency (ARG1-D) diagnosis confirmed by any of the following enzymatic, biochemical, or genetic testing:
    a. Persistently elevated plasma arginine (pArg) levels (>230 µmol/L)
    b. Pathogenic variants in ARG1 gene
    c. Diminished erythrocyte ARG1 activity of a red blood cell (RBC) arginase assay showing < 1% activity

AND

2. The member is ≥2 years of age

AND

3. Documentation the medication will be used in conjunction with a protein-restricted diet

AND

4. Prescribed in consultation with a metabolic disease specialist or neurologist experienced in urea cycle disorders (UCDs)

Reauthorization Criteria

1. The member has a documented response to Loargys with pArg levels of 40 µmol/L–115 µmol/L

Limitations

1. Initial approvals with be authorized for 6 (six) months and reauthorizations will be provided in 12-month intervals.
2. Members new to the plan and stable on the medication will be reviewed against reauthorization criteria.

Codes

The following code(s) require prior authorization:

Table 1: HCPCS Codes

HCPCS Codes	Description
J1818	Injection, pegzilarginase-nbln, 0.05 mg

References

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- National Organization for Rare Disorders (NORD). Arginase-1 deficiency. NORD. April 11, 2023. Accessed April 6, 2026. https://rarediseases.org/rare-diseases/arginase-deficiency/
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Approval And Revision History

September 2026: Joint Medical Policy and Health Care Services UM Committee review (effective 1/1/27)

Subsequent endorsement date(s) and changes made:

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Background, Product and Disclaimer Information

Point32Health prior authorization criteria to be applied to Medicare Advantage plan members is based on guidance from Medicare laws, National Coverage Determinations (NCDs) or Local Coverage Determinations (LCDs). When no guidance is provided, Point32Health uses clinical practice guidance published by relevant medical societies, relevant medical literature, Food and Drug Administration (FDA)-approved package labeling, and drug compendia to develop prior authorization criteria to apply to Medicare Advantage plan members. Medications that require prior authorization generally meet one or more of the following criteria: Drug product has the potential to be used for cosmetic purposes; drug product is not considered as first-line treatment by medically accepted practice guidelines, evidence to support the safety and efficacy of a drug product is poor, or drug product has the potential

to be used for indications outside of the indications approved by the FDA. Prior authorization and use of the coverage criteria within this Medical Necessity Guideline will ensure drug therapy is medically necessary, clinically appropriate, and aligns with evidence-based guidelines. We revise and update Medical Necessity Guidelines annually, or more frequently if new evidence becomes available that suggests revisions.

Treating providers are solely responsible for the medical advice and treatment of Members. The use of this guidelines not a guarantee of payment or a final prediction of how specific claim(s) will be adjudicated. Claims payment is subject to eligibility and benefits on the date of service, coordination of benefits, referral/authorization, utilization management guidelines when applicable, and adherence to plan policies, plan procedures, and claims editing logic.