

Palynziq (pegvaliase-pqpz)
Effective 03/01/2026

Plan	<input type="checkbox"/> MassHealth UPPL <input checked="" type="checkbox"/> Commercial/Exchange	Program Type	<input checked="" type="checkbox"/> Prior Authorization <input type="checkbox"/> Quantity Limit <input type="checkbox"/> Step Therapy
Benefit	<input checked="" type="checkbox"/> Pharmacy Benefit <input type="checkbox"/> Medical Benefit		
Specialty Limitations	These medications have been designated specialty and must be filled at a contracted specialty pharmacy.		
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

Palynziq (pegvaliase-pqpz) is a phenylalanine (Phe)-metabolizing enzyme indicated to reduce blood phenylalanine concentrations in adult patients with phenylketonuria (PKU) who have uncontrolled blood phenylalanine concentrations greater than 600 micromol/L on existing therapy management.

Coverage Guidelines

Authorization may be granted for members new to the plan within the past 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 all of the following criteria are met:

1. Member has a diagnosis of phenylketonuria (PKU)
2. Member is 18 years of age or older
3. Member has a baseline phenylalanine level of ≥ 600 micromol/L
4. Member has had an inadequate response or adverse reaction or a contraindication to sapropterin (Kuvan)

Continuation of Therapy

Requests for reauthorization will be approved when the member meets ONE of the following:

1. $\geq 20\%$ reduction from baseline in blood phenylalanine
2. Blood phenylalanine concentrations are ≤ 600 micromol/L

Limitations

1. Initial approvals will be granted for 9 months to allow for induction and titration to 20mg
2. Reauthorizations will be granted for 12 months
3. The following quantity limits apply:

Drug Name and Dosage Form	Quantity Limit
Palynziq 2.5 mg/0.5 mL prefilled syringe	8 syringes per 28 days
Palynziq 10 mg/0.5 mL prefilled syringe	30 syringes per 30 days
Palynziq 20 mg/mL prefilled syringe	90 syringes per 30 days

References

1. Bernegger C, Blau N. High frequency of tetrahydrobiopterin-responsiveness among hyperphenylalaninurias: a study of 1,919 patients observed from 1988 to 2002. *Mol Genet Metab* 2002; 77:304.
2. Blau N, van Spronsen FJ, Levy HL. Phenylketonuria. *Lancet* 2010; 376:1417.
3. Palynziq (pegvaliase-pqpz) [prescribing information]. Novato, CA: BioMarin Pharmaceutical Inc; October 2025.
4. Harding CO, Amato RS, Stuy M et al. Pegvaliase for the treatment of phenylketonuria: a pivotal, double-blind randomized discontinuation phase 3 clinical trial. *Mol Genet Metab*. 2018; 124(1):20-6.
5. van Wegberg AMJ, MacDonald A, Ahring K, et al. The complete European guidelines on phenylketonuria: diagnosis and treatment. *Orphanet J Rare Dis* 2017; 12:162.
6. Kuvan (sapropterin) [prescribing information]. Novato, CA: BioMarin Pharmaceutical Inc.; 2016 August.
7. Thomas J, Levy H, Amato S, et al. Pegvaliase for the treatment of phenylketonuria: Results of a long-term phase 3 clinical trial program (PRISM). *Mol Genet Metab* 2018; 124:27.

Review History

02/20/2019 – Reviewed.

12/11/2024 – Reviewed at December P&T. Updated initial criteria to remove “on current therapy” from diagnosis and lab requirements. Removed requirement regarding anaphylaxis education for member. Updated initial approval period from 33 weeks to 9 months. Effective 03/01/2025.

12/10/2025 – Reviewed at December P&T. Administrative update – added quantity limits to the Limitations section. Effective 03/01/2026.

