

**Palynziq (pegvaliase-pqpz)**  
**Effective 03/01/2025**

<b>Plan</b>	<input type="checkbox"/> MassHealth UPPL <input checked="" type="checkbox"/> Commercial/Exchange	<b>Program Type</b>	<input checked="" type="checkbox"/> Prior Authorization <input type="checkbox"/> Quantity Limit <input type="checkbox"/> Step Therapy
<b>Benefit</b>	<input checked="" type="checkbox"/> Pharmacy Benefit <input type="checkbox"/> Medical Benefit		
<b>Specialty Limitations</b>	This medication has been designated specialty and must be filled at a contracted specialty pharmacy.		
<b>Contact Information</b>	<b>Medical and Specialty Medications</b>		
	All Plans	Phone: 877-519-1908	Fax: 855-540-3693
<b>Exceptions</b>	<b>Non-Specialty Medications</b>		
	All Plans	Phone: 800-711-4555	Fax: 844-403-1029
<b>Exceptions</b>	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  $\geq 600$  micromol/L
4. Member has had an inadequate response or adverse reaction or a contraindication to 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  $\leq 600$  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

### 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; November 2020.
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.

