

**Palynziq (pegvaliase-pqpz)**  
**Effective 04/01/2019**

<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 (NLX)		
<b>Specialty Limitations</b>	This medication has been designated a specialty medication and must be filled at a contracted pharmacy		
<b>Contact Information</b>	<b>Specialty Medications</b>		
	All Plans	Phone: 866-814-5506	Fax: 866-249-6155
	<b>Non-Specialty Medications</b>		
	MassHealth	Phone: 877-433-7643	Fax: 866-255-7569
	Commercial	Phone: 800-294-5979	Fax: 888-836-0730
	Exchange	Phone: 855-582-2022	Fax: 855-245-2134
	<b>Medical Specialty Medications (NLX)</b>		
	All Plans	Phone: 844-345-2803	Fax: 844-851-0882
<b>Exceptions</b>	N/A		

### Overview

Palynziq (pegvaliase-pqpz) is a phenylalanine-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

Member must meet **ALL** the following criteria and documentation has been submitted:

- Member has a diagnosis of uncontrolled PKU on current therapy
- Member is at least 18 years of age
- Member has a baseline phenylalanine level of  $\geq 600$  micromol/L despite current therapy
- Member has had an inadequate response or adverse reaction or a contraindication to Kuvan
- Member has been educated to recognize signs and symptoms of anaphylaxis, has been prescribed auto-injectable epinephrine and instructed on its appropriate use.

### Continuation of Therapy

Reauthorization may be approved upon receipt of documentation evidencing one of the following:

1. 20% reduction from baseline in blood phenylalanine
2. Blood phenylalanine concentrations are 600 micromol/L or less

### Limitations

1. Initial approvals may be granted for 33 weeks to allow for induction and titration to 20mg
2. Reauthorizations will be granted for 12 months

## References

1. Palynziq (pegvaliase-pqpz) [prescribing information]. Novato, CA: BioMarin Pharmaceutical Inc; May 2018.
2. Blau N, van Spronsen FJ, Levy HL. Phenylketonuria. *Lancet* 2010; 376:1417.
3. 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.
4. Bernegger C, Blau N. High frequency of tetrahydrobiopterin-responsiveness among hyperphenylalaninemias: a study of 1,919 patients observed from 1988 to 2002. *Mol Genet Metab* 2002; 77:304.
5. 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.
6. 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.
7. Kuvan (sapropterin) [prescribing information]. Novato, CA: BioMarin Pharmaceutical Inc.; 2016 August.

## Review History

02/20/19 – Reviewed.

