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

Plan	☒ MassHealth ☐Commercial/Exchange	**Program Type**	☒ Prior Authorization ☐ Quantity Limit ☐ Step Therapy
Benefit	☒ Pharmacy Benefit ☐ Medical Benefit (NLX)		
Specialty Limitations	This medication has been designated a specialty medication and must be filled at a contracted pharmacy.		
Contact Information	**Specialty Medications**		
	All Plans	Phone: 866-814-5506	Fax: 866-249-6155
	Non-Specialty Medications		
	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
	Medical Specialty Medications (NLX)		
	All Plans	Phone: 844-345-2803	Fax: 844-851-0882
Exceptions	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 ≥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

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