

Market Applicability							
Market	DC	GA	KY	MD	NJ	NY	WA
Applicable	X	X	X	X	X	X	X

Palynziq (pegvaliase-pqpz)

Override(s)	Approval Duration
Prior Authorization	1 year

Medications	Quantity Limit
Palynziq (pegvaliase-pqpz)	May be subject to quantity limit

APPROVAL CRITERIA

Initial requests for Palynziq (pegvaliase-pqpz) may be approved if the following criteria are met:

- I. Individual is 18 years of age or older; **AND**
- II. Individual has a confirmed prescription for an auto-injectable epinephrine agent; **AND**
- III. Individual has a diagnosis of phenylketonuria (PKU); **AND**
- IV. Individual has uncontrolled blood phenylalanine (PHE) concentrations (> 600 micromol/L) on existing management, including but not limited to the following examples:
 - A. Dietary phenylalanine and/or protein restriction;
 - B. Kuvan (sapropterin dihydrochloride) agents.

Requests for continued use of Palynziq (pegvaliase-pqpz) may be approved if the following criteria are met:

- I. Individual is showing signs of continuing improvement, as evidenced by blood PHE level decrease of at least 20% from pre-treatment baseline or a reduction below 600 micromol/L at current dose OR after 16 weeks of treatment as a maximum dose of 40 mg/day.

Note: Palynziq (pegvaliase-pqpz) has a black box warning for anaphylaxis which may occur at any time during treatment. The initial dose must be administered under the supervision of a healthcare provider equipped to manage anaphylaxis and closely observe individual for at least 60 minutes following injection. Concurrent auto-injectable epinephrine must be prescribed and individuals instructed to carry epinephrine with them at all times during treatment. Due to the risk of anaphylaxis, Palynziq is only available through a restricted REMS program.

This policy does not apply to health plans or member categories that do not have pharmacy benefits, nor does it apply to Medicare. Note that market specific restrictions or transition-of-care benefit limitations may apply.

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Additional information and forms for individuals, prescribers, and pharmacists may be found on the Palynziq REMS program website: www.palynziqrems.com.

State Specific Mandates		
State name	Date effective	Mandate details (including specific bill if applicable)
N/A	N/A	N/A

Key References:

1. Clinical Pharmacology [database online]. Tampa, FL: Gold Standard, Inc.: 2019. URL: <http://www.clinicalpharmacology.com>. Updated periodically.
2. DailyMed. Package inserts. U.S. National Library of Medicine, National Institutes of Health website. <http://dailymed.nlm.nih.gov/dailymed/about.cfm>. Accessed: June 7, 2019.
3. DrugPoints® System [electronic version]. Truven Health Analytics, Greenwood Village, CO. Updated periodically.
4. Lexi-Comp ONLINE™ with AHFS™, Hudson, Ohio: Lexi-Comp, Inc.; 2019; Updated periodically.
5. Longo N, Dimmock D, Levy H, et al. Evidence- and consensus-based recommendations for the use of pegvaliase in adults with phenylketonuria. *Genet Med* 2018; doi: 10.1038/s41436-018-0403-z. [Epub ahead of print]. Available at: <https://www.nature.com/articles/s41436-018-0403-z>
6. American College of Medical Genetics and Genomics Therapeutic Committee. Phenylalanine hydroxylase deficiency: diagnosis and management guideline. *Genet Med*. 2014; 16(2):188-200. doi:10.1038/gim.2013.157. Available from: <http://www.nature.com/gim/journal/v16/n2/pdf/gim2013157a.pdf>. Accessed on: June 7, 2019

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