Market Applicability				
Market	**GA**	**MD**	**NJ**	**NY**
Applicable	X	X	X	X

Palynziq (pegvaliase-pqpz)

Override(s)	Approval Duration
Prior Authorization	Initial and continuation: 1 year

Medications	Quantity Limit
Palynziq (pegvaliase-pqpz) 20 mg/mL prefilled syringe	1 syringe per day*

*Initial requests for up to two 20 mg syringes per day may be approved if the following criteria are met:

I. Individual has been on treatment with 20 mg per day for at least 24 weeks; **AND**
II. Individual has not achieved a blood PHE level below 600 micromol/L.

*Initial requests for up to three 20 mg syringes per day may be approved if the following criteria are met:

I. Individual has been on treatment with 40 mg per day for at least 16 weeks; **AND**
II. Individual has not achieved a blood PHE level below 600 micromol/L.

Requests for continued use of 2 or 3 syringes per day may be approved if increased dosing resulted in reduction and maintenance of blood PHE levels below 600 micromol/L.

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. Documentation is provided that 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:

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.

Market Applicability				
Market	**GA**	**MD**	**NJ**	**NY**
Applicable	X	X	X	X

I. Documentation is provided that there is positive response to therapy as evidenced by a reduction and maintenance of blood PHE levels below 600 micromol/L; **OR**
II. Individual is showing signs of continuing improvement but has not completed titration to maximum tolerated dose.

Palynziq (pegvaliase-pqpz) may not be approved for the following:

I. All other indications not included above; **OR**
II. Individual is using in combination with Kuvan (sapropterin dihydrochloride).

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

Key References:

1. Clinical Pharmacology [database online]. Tampa, FL: Gold Standard, Inc.: 2020. 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, 2020.
3. DrugPoints® System [electronic version]. Truven Health Analytics, Greenwood Village, CO. Updated periodically.
4. Lexi-Comp ONLINE™ with AHFS™, Hudson, Ohio: Lexi-Comp, Inc.; 2020; 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

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Market Applicability				
Market	**GA**	**MD**	**NJ**	**NY**
Applicable	X	X	X	X

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, 2020.

Federal and state laws or requirements, contract language, and Plan utilization management programs or polices may take precedence over the application of this clinical criteria.

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