ALASKA MEDICAID
Prior Authorization Criteria

Palynziq™ (pegvaliase-pqpz)

FDA INDICATIONS AND USAGE[1]

Palynziq™ is a phenylalanine-metabolizing enzyme indicated to reduce blood phenylalanine concentrations in adult patients with phenylketonuria who have uncontrolled blood phenylalanine concentrations greater than 600 micromol/L on existing management.

APPROVAL CRITERIA[1,2]

1. Patient is 18 years of age or older **AND;**
2. Patient has a confirmed diagnosis of phenylketonuria **AND;**
3. Prescribed by or in consultation with metabolic specialist and both prescriber and patient are enrolled in REMS program **AND;**
4. Baseline phenylalanine levels have been documented **AND;**
5. Patient is actively on a phenylalanine restricted diet **AND;**
6. Patient has uncontrolled blood phenylalanine concentrations >600 micromol/L on existing management (I.E. Kuvan agents) **AND;**
7. Phenylalanine levels are being monitored and recorded through therapy **AND;**
8. Patient has been prescribed an auto-injectable epinephrine device and has been trained on proper use.

DENIAL CRITERIA[1,2]

1. Patient is less than 18 years of age **OR**;
2. Patient does not have a confirmed diagnosis of phenylketonuria **OR;**
3. Not being prescribed by or in consultation with metabolic specialist and both prescriber and patient are not enrolled in REMS program **OR;**
4. Baseline phenylalanine levels have not been documented **OR;**
5. Patient is not actively on a phenylalanine restricted diet **OR;**
6. Patient does not have uncontrolled blood phenylalanine concentrations >600 micromol/L on existing management (I.E. Kuvan agents) **OR;**
7. Patient is concomitantly using Kuvan agents with Palynziq™ **OR;**
8. Phenylalanine levels are not being monitored and recorded through therapy **OR;**
9. Patient has not been prescribed an auto-injectable epinephrine device and has not been trained on proper use.

CAUTIONS[1]

- Black Box Warning of anaphylaxis has been reported after administration of Palynziq™ and may occur at any time during treatment.
- Administration of the initial dose of Palynziq™ should be under the supervision of a healthcare provider equipped to manage anaphylaxis, and closely observe patients for at

least 60 minutes following injection. Prior to self-injection, confirm patient competency with self-administration, and patient's and observer's (if applicable) ability to recognize signs and symptoms of anaphylaxis and to administer auto-injectable epinephrine, if needed.

- Prescribe auto-injectable epinephrine. Prior to first dose, instruct the patient and observer (if applicable) on its appropriate use. Instruct the patient to seek immediate medical care upon its use. Instruct patients to carry auto-injectable epinephrine with them at all times during Palynziq™ treatment.
- Obtain blood phenylalanine concentrations every 4 weeks until a maintenance dosage is established, then periodically monitor blood phenylalanine concentrations.

DURATION OF APPROVAL

- Initial Approval: 3 months
- Reauthorization: up to 6 months if the patient has shown continued improvement measured by at least a 20% reduction from the pre-treatment baseline or a reduction below 600 micromol/L and the patient has not had any toxicities or adverse reactions to the drug.

QUANTITY LIMITS

- Up to 40mg per day (2 syringes max daily dose)
- 60 syringes per month

REFERENCES / FOOTNOTES:

1. Palynziq™ [Package Insert]. BioMarin Pharmaceutical Inc., Novato, CA. 2018. Available at: https://www.palynziq.com/prescribinginformation.pdf. Accessed on: December 7, 2018.
2. Vockley J, Andersson HC, Antshel KM, et al. Phenylalanine hydroxylase deficiency: diagnosis and management guideline. Genet Med. 2014 Feb;16(2):188-200 Available at: https://www.ncbi.nlm.nih.gov/pubmed/24385074. Accessed on December 7, 2018.