Pharmacy Policy Bulletin

**Title:** Pegvaliase-pqpz (Palynziq™)

**Policy #:** Rx.01.210

***Application of pharmacy policy is determined by benefits and contracts. Benefits may vary based on product line, group, or contract. Some medications may be subject to precertification, age, quantity, or formulary restrictions (ie limits on non-preferred drugs). Individual member benefits must be verified.***

***This pharmacy policy document describes the status of pharmaceutical information and/or technology at the time the document was developed. Since that time, new information relating to drug efficacy, interactions, contraindications, dosage, administration routes, safety, or FDA approval may have changed. This Pharmacy Policy will be regularly updated as scientific and medical literature becomes available. This information may include new FDA-approved indications, withdrawals, or other FDA alerts. This type of information is relevant not only when considering whether this policy should be updated, but also when applying it to current requests for coverage.***

***Members are advised to use participating pharmacies in order to receive the highest level of benefits.***

**Intent:**

The intent of this policy is to communicate the medical necessity criteria for **Pegvaliase-pqpz (Palynziq™)** as provided under the member's prescription drug benefit.

**Description:**

**Phenylketonuria** is a disorder affecting the aromatic amino acid, phenylalanine. It results from a deficiency of phenylalanine hydroxylase (PAH) and if untreated is characterized by intellectual disability.

Pegvaliase-pqpz (Palynziq™) is a PEGylated phenylalanine ammonia lyase (PAL) enzyme that converts phenylalanine to ammonia and trans-cinnamic acid. It substitutes for the deficient phenylalanine hydroxylase (PAH) enzyme activity in patients with PKU and reduces blood phenylalanine concentrations.

Pegvaliase-pqpz (Palynziq™) is 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.

**Policy:**

**INITIAL CRITERIA:** Pegvaliase-pqpz (Palynziq™) is approved when BOTH of the following are met:

1. Member is 18 years of age or older, and
2. Member has diagnosis of phenylketonuria with uncontrolled blood phenylalanine concentration of greater than 600 micromol/L on existing management (e.g., medical foods, Kuvan®); and
3. Member will continue to have phenylalanine blood levels measured periodically during therapy

Initial Authorization duration: 6 months

**CONTINUATION CRITERIA:** Pegvaliase-pqpz (Palynziq™) is re-approved when there is documentation of a positive clinical response to Pegvaliase-pqpz (Palynziq™) therapy

Continuation Authorization duration: 2 years

**Black Box Warning as shown in the drug Prescribing Information:**

WARNING: RISK OF ANAPHYLAXIS
See full prescribing information for complete boxed warning.

> Anaphylaxis has been reported after administration of Palynziq and may occur at any time during treatment. Administer the initial dose of Palynziq 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.
Palynziq is available only through a restricted program called the Palynziq REMS.

**Guidelines:**

Refer to the specific manufacturer's prescribing information for administration and dosage details and any applicable Black Box warnings.

**BENEFIT APPLICATION**

Subject to the terms and conditions of the applicable benefit contract, the applicable drug(s) identified in this policy is (are) covered under the prescription drug benefits of the Company's products when the medical necessity criteria listed in this pharmacy policy are met. Any services that are experimental/investigational or cosmetic are benefit contract exclusions for all products of the Company.

**References:**

Pegvaliase-pqpz (Palynziq™) [package insert]. Novato, CA. BioMarin Pharmaceutical, Inc. May 2018. Available from: https://www.accessdata.fda.gov/drugsatfda_docs/label/2018/761079s000lbl.pdf. Accessed September 29, 2021.

Bodamer, O. Overview of Phenylketonuria. UpToDate. Access September 29, 2021.

**Applicable Drugs:**

Inclusion of a drug in this table does not imply coverage. Eligibility, benefits, limitations, exclusions, precertification/referral requirements, provider contracts, and Company policies apply.

| Brand Name | Generic Name |
|---|---|
| Palynziq™ | Pegvaliase-pqpz |

**Cross References:**

Off-Label Use Rx. 01.33

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| **Policy Version Number:** | 4.00 |
| **P&T Approval Date:** | September 23, 2021 |
| **Policy Effective Date:** | January 01, 2022 |
| **Next Required Review Date:** | September 23, 2022 |