Pharmacy Medical Necessity Guidelines: Palynziq™ (pegvaliase-pqpz)

Effective: June 15, 2020

Prior Authorization Required	√	Type of Review – Care Management	
Not Covered		Type of Review – Clinical Review	√
Pharmacy (RX) or Medical (MED) Benefit	RX	Department to Review	RXUM

These pharmacy medical necessity guidelines apply to the following:

Commercial Products

☒ Tufts Health Plan Commercial products – large group plans
☒ Tufts Health Plan Commercial products – small group and individual plans
☒ Tufts Health Freedom Plan products – large group plans
☒ Tufts Health Freedom Plan products – small group plans
- CareLink℠ – Refer to CareLink Procedures, Services and Items Requiring Prior Authorization

Tufts Health Public Plans Products

☒ Tufts Health Direct – A Massachusetts Qualified Health Plan (QHP) (a commercial product)
☒ Tufts Health Together – MassHealth MCO Plan and Accountable Care Partnership Plans
☒ Tufts Health RITogether – A Rhode Island Medicaid Plan

Fax Numbers:

RXUM: 617.673.0988

Note: This guideline does not apply to Medicare Members (includes dual eligible Members).

OVERVIEW

FOOD AND DRUG ADMINISTRATION (FDA)-APPROVED INDICATIONS

Palynziq (pegvaliase-pqpz) 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.

The recommended initial induction for Palynziq (pegvaliase-pqpz) is 2.5 mg subcutaneously once weekly for four weeks. The first dose should be administered under the supervision of a healthcare provider.

The dose of Palynziq (pegvaliase-pqpz) should be titrated in a step-wise manner, based on tolerability, over at least five weeks (2.5 mg twice weekly for one week, 10 mg once weekly for one week, 10 mg twice weekly for one week, 10 mg four times per week for one week, then 10 mg once daily for one week), to achieve a maintenance dose of 20 mg subcutaneously once daily. Therapeutic response may not be achieved until the patient is titrated to an effective maintenance. Use the lowest effective and tolerated dose of Palynziq (pegvaliase-pqpz).

COVERAGE GUIDELINES

The plan may authorize coverage of Palynziq (pegvaliase-pqpz) for Members, when all of the following criteria are met:

Initial Therapy

1. Documented diagnosis of phenylketonuria

AND

2. The prescribing physician is a metabolic disease specialist

AND

3. The Member is at least 18 years of age

AND

4. Documentation of a baseline phenylalanine level of at least 600 umol/L

Reauthorization Criteria

1. Documented diagnosis of phenylketonuria

AND

2. The prescribing physician is a metabolic disease specialist

AND

3. The Member is at least 18 years of age

AND

4. Documentation the Member has received at least 33 weeks of Palynziq

AND

5. Documentation the Member has experienced a therapeutic response as defined by one of the following:
    a. At least a 20% reduction in blood phenylalanine concentration from pre-treatment baseline
    b. Blood phenylalanine concentration less than or equal to 600 umol/L

LIMITATIONS

- Initial coverage of Palynziq (pegvaliase-pqpz) will be authorized for 33 weeks. Reauthorization of Palynziq (pegvaliase-pqpz) will be provided in 12-month intervals.
- Members new to the plan stable on Palynziq (pegvaliase-pqpz) should be reviewed against Initial Therapy if they have received less than 33 weeks of therapy. Members new to the plan stable on Palynziq (pegvaliase-pqpz) should be reviewed against Reauthorization Criteria if they have received at least 33 weeks of therapy.
- Palynziq (pegvalise-pgnz) will not be authorized in combination with Kuvan (sapropterin).
- The following quantity limitation applies:
    a. Palynziq (pegvalise-pgnz) 20 mg/mL syringe: 1 unit per day
    b. Requests for quantities above the quantity limitation will be reviewed against the Drugs With Quantity Limitations Medical Necessity Guideline.

CODES

None

REFERENCES

1. Blau N, van Spronsen FJ, Levy H. Phenylketonuria. Lancet. 2010; 376:1417-27.
2. 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.
3. Kuvan (sapropterin) [prescribing information]. Novato, CA: BioMarin Pharmaceutical Inc.; 2019 February.
4. Palynziq (pegvaliase-pqpz) [prescribing information]. Novato, CA: BioMarin Pharmaceutical Inc.; 2018 May.
5. National Institutes of Health. U.S. National Library of Medicine Genetics Home Reference. Phenylketonuria. URL: https://ghr.nlm.nih.gov/condition/phenylketonuria. Available from Internet. Accessed 2018 October 22.
6. 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(1):27-38.
7. Vockley J, Andersson HC, Antshel KM et al. Phenylalanine hydroxylase deficiency: diagnosis and management guideline. Genet Med. 2014; 16(2):188-200.

APPROVAL HISTORY

November 13, 2018: Reviewed by Pharmacy & Therapeutics Committee

Subsequent endorsement date(s) and changes made:

1. September 10, 2019: No changes
2. June 9, 2020: Administrative update to move duration of approval rules into the Limitations as "Initial coverage of Palynziq (pegvaliase-pqpz) will be authorized for 33 weeks. Reauthorization of Palynziq (pegvaliase-pqpz) will be provided in 12-month intervals" and "Members new to the plan stable on Palynziq (pegvaliase-pqpz) should be reviewed against Initial Therapy if they have received less than 33 weeks of therapy. Members new to the plan stable on Palynziq (pegvaliase-pqpz) should be reviewed against Reauthorization Criteria if they have received at least 33 weeks of therapy."

BACKGROUND, PRODUCT AND DISCLAIMER INFORMATION

Pharmacy Medical Necessity Guidelines have been developed for determining coverage for plan benefits and are published to provide a better understanding of the basis upon which coverage decisions are made. The plan makes coverage decisions on a case-by-case basis considering the individual member's health care needs. Pharmacy Medical Necessity Guidelines are developed for selected therapeutic classes or drugs found to be safe, but proven to be effective in a limited, defined population of patients or clinical circumstances. They include concise clinical coverage criteria based on current literature review, consultation with practicing physicians in the service area who are medical experts in the particular field, FDA and other government agency policies, and standards adopted by national accreditation organizations. The plan revises and updates Pharmacy Medical

Necessity Guidelines annually, or more frequently if new evidence becomes available that suggests needed revisions.

For self-insured plans, coverage may vary depending on the terms of the benefit document. If a discrepancy exists between a Pharmacy Medical Necessity Guideline and a self-insured Member's benefit document, the provisions of the benefit document will govern.

Treating providers are solely responsible for the medical advice and treatment of members. The use of this policy is not a guarantee of payment or a final prediction of how specific claim(s) will be adjudicated. Claims payment is subject to member eligibility and benefits on the date of service, coordination of benefits, referral/authorization and utilization management guidelines when applicable, and adherence to plan policies and procedures and claims editing logic.