DRUG QUANTITY MANAGEMENT POLICY – PER RX

POLICY: Phenylketonuria – Palynziq Drug Quantity Management Policy – Per Rx
- Palynziq® (pegvaliase-pqpz subcutaneous injection – BioMarin)

REVIEW DATE: 08/31/2022

OVERVIEW
Palynziq is indicated to reduce blood phenylalanine concentrations in adult patients with **phenylketonuria** (PKU) who have uncontrolled blood phenylalanine concentrations greater than 600 micromol/L (µmol/L) on existing management.[1] Treatment with Palynziq should be managed by a healthcare provider experienced in the management of PKU. Baseline blood phenylalanine concentrations should be obtained before initiating treatment.

Dosing
Dosing of Palynzq is individualized. During titration and maintenance of Palynziq treatment, patients may experience blood phenylalanine concentrations below 30 micromol/L. For blood phenylalanine concentrations below 30 micromol/L, the dosage of Palynziq may be reduced and/or dietary protein and phenylalanine intake may be modified to maintain blood phenylalanine concentrations within a clinically acceptable range and above 30 micromol/L. In the Phase III PRISM-2 open-label extension study (n = 261), the maintenance dose of Palynziq could be adjusted between 5 mg/day and 60 mg/day based on investigator-determined efficacy and tolerability.[2]

Table 1. Palynziq Dose Titration.[1]

Treatment	Palynziq Dose	Duration*
Induction	2.5 mg once weekly	4 weeks
Titration	2.5 mg twice weekly	1 week
	10 mg once weekly	1 week
	10 mg twice weekly	1 week
	10 mg four times per week	1 week
	10 mg QD	1 week
Maintenance	20 mg QD	24 weeks
	40 mg QD	16 weeks
Maximum	60 mg QD	16 weeks

* Additional time may be required prior to each dosage escalation based on patient tolerability; QD – Once daily.

Availability
Palynziq is available in the following strengths: 2.5 mg/0.5 mL, 10 mg/0.5 mL, and 20 mg/1mL syringes.[1] The product is provided in a 1 mL glass syringe with a 26 gauge, 0.5 inch needle. Each carton contains 1 or 10 trays with single-dose prefilled syringe(s).

POLICY STATEMENT
This Drug Quantity Management program has been developed to manage potential dose escalation of Palynziq and to provide a sufficient quantity for approvable indications. If the Drug Quantity Management rule is not met for the requested medication at the point of service, coverage will be determined by the Criteria below. All approvals are provided for 1 year.

Automation: None.

Drug Quantity Limits

Product	Strength and Form	Retail Maximum Quantity per Rx	Home Delivery Maximum Quantity per Rx
Palynziq® (pegvaliase-pqpz subcutaneous injection)	2.5 mg/0.5 mL syringe	8 syringes	8 syringes
	10 mg/0.5 mL syringe	30 syringes	90 syringes
	20 mg/1 mL syringe	60 syringes	180 syringes

CRITERIA

Palynziq 2.5 mg/0.5 mL syringe

1. If the patient has blood phenylalanine concentrations below 30 micromol/L, approve 60 syringes per dispensing at retail and 180 syringes per dispensing at home delivery.

Palynziq 20 mg/1 mL syringe

1. If the patient requires a maintenance dose of 60 mg/day, approve 90 syringes per dispensing at retail and 270 syringes per dispensing at home delivery.

Palynziq 10 mg/0.5 mL syringe

No overrides recommended.

Note: For patients who are receiving ≥ 20 mg/day, refer the patient to the 20 mg/1 mL syringe.

REFERENCES

1. Palynziq injection [prescribing information]. Novato, CA: BioMarin; November 2020.
2. Thomas J, Levy H, Amato S, et al; PRISM investigators. Pegvaliase for the treatment of phenylketonuria: results of a long-term phase 3 clinical trial program (PRISM). *Mol Genet Metab*. 2018 May;124(1):27-38.