

Effective Date 06/07/2019
Reviewed: 6/2019, 6/2020, 10/2020, 4/2021, 3/2022, 5/2023, 4/2024, 5/2025
Scope: Medicaid

Palynziq (pegvaliase-pqpz)

POLICY

I. CRITERIA FOR APPROVAL

An authorization of 3 months may be granted when all the following criteria are met:

- A. Patient is ≥ 18 years of age
- B. Patient has a diagnosis of phenylketonuria (PKU)
- C. Prescriber must be certified with Palynziq REMS program
- D. Patient currently uncontrolled on existing management and has a blood phenylalanine concentration greater than 600 micromol/L (labs to be provided)
- E. Patient has failed therapy with sapropterin (Kuvan)
- F. Patient must have prescription of auto-injectable epinephrine on hand at all times
- G. Patient has failed therapy with dietary restriction of phenylalanine
- H. Palynziq is not being used in combination with sapropterin (Kuvan)

II. CONTINUATION OF THERAPY

- A. Current blood phenylalanine concentration lab results must be provided; **AND**
- B. Palynziq is not being used in combination with sapropterin (Kuvan); **AND**
- C. Patient has been titrated to 20mg daily over 3 months, or lowest effective and tolerated dose; **OR**
- D. If requesting new daily dose of 40mg: Documentation that member has been adherent to 20mg daily for at least 24 weeks and requires increase to 40mg once daily because the patient has not achieved blood Phe control (blood phenylalanine concentration less than or equal to 600 micromol/L); **OR**
- E. If requesting new daily dose of 60mg: Documentation that member has been adherent to 40mg daily for at least 16 weeks and requires increase to 60mg once daily because the patient has not achieved blood Phe control (blood phenylalanine concentration less than or equal to 600 micromol/L); **OR**
- F. If requesting maintenance daily dose of 20 mg, 40mg or 60mg: documentation of blood phenylalanine concentration less than or equal to 600 micromol/L

III. QUANTITY LIMIT

- Palynziq 2.5mg/0.5 ml: 8 syringes per 28 days (daily dose of 0.143 ml)
- Palynziq 10mg/0.5 ml: 30 syringes per 30 days (daily dose of 0.5 ml)
- Palynziq 20mg/ml: 60 syringes per 30 days (daily dose of 2 ml), with post-exception limit for 90 syringes per 30 days (daily dose of 3 ml) if 60 mg dose approved.

IV. COVERAGE DURATION

- Initial 3 months
- Renewal:
 - Dose of 20mg once daily: 6 months
 - Dose of 40mg or 60mg once daily : 4 months

V. DOSING

Treatment	Palynziq Dosage	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
	10mg four times weekly	1 week
	10mg once a daily	1 week
Maintenance	20mg once daily	24 weeks
	40mg once daily	16 weeks
Maximum	60mg once daily	16 weeks

VI. REFERENCES

Palynziq [package insert]. Novato, CA: BioMarin Pharmaceutical Inc.; December 2022.