

Palynziq® (pegvaliase-pqpz)

When requesting Palynziq® (pegvaliase-pqpz), the individual requiring treatment must be diagnosed with an FDA-approved indication and meet the specific coverage guidelines and applicable safety criteria for the covered indication.

FDA-approved indication

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 on existing management.

Approved Off-label Compendial use(s)

None

Coverage Guidelines

Initial authorization:

The individual must meet **ALL** of the following criteria for approval:

- Is at least 18 years of age
- Has uncontrolled blood phenylalanine concentration despite existing treatment(s) (i.e., phenylalanine restricted diet and/or prior treatment with sapropterin dihydrochloride)
- Has blood phenylalanine concentration greater than 600 micromol/L
- An initial dose of Palynziq will be administered under the supervision of a healthcare provider who is equipped to manage anaphylaxis
- An auto-injectable epinephrine will be prescribed

Approval duration: 40 weeks

Reauthorization:

The individual must meet **ALL** of the following criteria for renewal:

- Is at least 18 years of age
- Has achieved at least 20% reduction in blood phenylalanine concentration from pre-treatment baseline OR has current blood phenylalanine concentration level less than or equal to 600 micromol/L

Approval duration: 12 months

Dosing Recommendations

- The recommended initial induction dosage for Palynziq is 2.5 mg subcutaneously once weekly for 4 weeks.

- Palynziq dosage should be titrated in a step-wise manner, based on tolerability, over at least 5 weeks, to achieve a dosage of 20 mg subcutaneously once daily.
- Palynziq dosage should be increased to a maximum of 40 mg subcutaneously once daily in patients who have been on 20 mg once daily continuously for at least 24 weeks and have not achieved either a 20% reduction in blood phenylalanine concentration from pre-treatment baseline.
- Palynziq should be discontinued in patients who have not achieved at least 20% reduction in blood phenylalanine concentration from pre-treatment baseline or blood phenylalanine concentration less than or equal to 600 micromol/L after 16 weeks of continuous treatment with the maximum dosage of 40 mg once daily.

References

1. Palynziq prescribing information. Novato, CA: BioMarin Pharmaceutical Inc. May 2018.
2. Vockley J, Andersson HC, Antshel KM, et al. (2014). Phenylalanine hydroxylase deficiency: diagnosis and management guideline. *American College of Medical Genetics and Genomics*. . Advance online publication. doi:10.1038/gim.2013.157.