PEGVALIASE

Generic	Brand	HICL	GCN	Exception/Other
PEGVALIASE-PQPZ	PALYNZIQ	44944		

GUIDELINES FOR USE

INITIAL CRITERIA (NOTE: FOR RENEWAL CRITERIA SEE BELOW)

1. Does the patient have a diagnosis of phenylketonuria and meet **ALL** of the following criteria?
   - The patient is 18 years of age or older
   - The patient has uncontrolled blood phenylalanine concentrations greater than 600 micromol/L on existing management, as confirmed by a measurement in the last 30 days
   - The patient has had a previous trial of Kuvan (sapropterin)
   - The patient is not concurrently receiving Kuvan (sapropterin)

   If yes, **approve for 6 months by GPID for all strengths with the following quantity limits:**
   - **Palynziq 2.5mg/0.5mL (GPID 44791): #1mL (2 syringes) per 7 days.**
   - **Palynziq 10mg/0.5mL (GPID 44792): #0.5mL (1 syringe) per day.**
   - **Palynziq 20mg/mL (GPID 44793): #2mL (2 syringes) per day.**

   APPROVAL TEXT: Renewal requires that the patient has demonstrated a reduction in phenylalanine levels, compared to baseline, by at least 20% or to a level below 600 micromol/L.

   If no, do not approve.
   INITIAL DENIAL TEXT: The guideline named **PEGVALIASE (Palynziq)** requires a diagnosis of phenylketonuria. In addition, the following criteria must be met:
   - The patient is 18 years of age or older
   - The patient has uncontrolled blood phenylalanine concentrations greater than 600 micromol/L on existing management, as confirmed by a measurement in the last 30 days
   - The patient has had a previous trial of Kuvan (sapropterin)
   - The patient is not concurrently receiving Kuvan (sapropterin)

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PEGVALIASE

GUIDELINES FOR USE (CONTINUED)

RENEWAL CRITERIA

1. Does the patient have a diagnosis of phenylketonuria and meet the following criteria?
   - The patient has demonstrated a reduction in phenylalanine levels, compared to baseline, by at least 20% or to a level below 600 micromol/L

   If yes, **approve for 12 months by GPID for all strengths with the following quantity limits:**
   - **Palynziq 2.5mg/0.5mL (GPID 44791): #1mL (2 syringes) per 7 days.**
   - **Palynziq 10mg/0.5mL (GPID 44792): #0.5mL (1 syringe) per day.**
   - **Palynziq 20mg/mL (GPID 44793): #2mL (2 syringes) per day.**

   If no, do not approve.
   RENEWAL DENIAL TEXT: The guideline named **PEGVALIASE (Palynziq)** requires a diagnosis of phenylketonuria. In addition, the following criteria must be met:
   - The patient has demonstrated a reduction in phenylalanine levels, compared to baseline, by at least 20% or to a level below 600 micromol/L

RATIONALE
To ensure appropriate use of Palynziq (pegvaliase) consistent with FDA-approved indication and dosing.

FDA APPROVED INDICATION
Palynziq 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.

DOSAGE AND ADMINISTRATION
Treatment with Palynziq should be managed by a healthcare provider experienced in the management of phenylketonuria. Before initiating treatment, baseline blood phenylalanine concentrations should be obtained. After initiating treatment with Palynziq, blood phenylalanine concentrations should be obtained every 4 weeks until a maintenance dosage is established. After a maintenance dosage is established, periodic blood phenylalanine monitoring is recommended to assess blood phenylalanine control.

For hypersensitivity reactions, premedication may be considered with an H_1-receptor antagonist, H_2-receptor antagonist, and/or antipyretic prior to Palynziq administration based upon individual patient tolerability.

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FDA APPROVED INDICATION (CONTINUED)

DOSAGE AND ADMINISTRATION

Induction:
The recommended initial induction dosage for Palynziq is 2.5 mg subcutaneously once weekly for 4 weeks. The initial dose should be administered under the supervision of a healthcare provider.

Titration:
Palynziq doses should be titrated in a stepwise manner based on tolerability, over at least 5 weeks, to achieve a dosage of 20 mg subcutaneously once daily.

Maintenance:
Therapeutic response may not be achieved until the patient is titrated to an effective maintenance dosage. The lowest effective and tolerated dosage of Palynziq should be used. Palynziq should be maintained at a dosage of 20 mg subcutaneously once daily for at least 24 weeks. Increasing the dosage to a maximum of 40 mg subcutaneously once daily may be considered in patients who have been maintained continuously on 20 mg once daily for at least 24 weeks and who have not achieved either a 20% reduction in blood phenylalanine concentrations from pre-treatment baseline levels or blood phenylalanine concentrations ≤600 micromol/L. Patient tolerability, blood phenylalanine concentrations, and dietary protein and phenylalanine intake should be assessed throughout treatment.

Discontinuation:
Palynziq should be discontinued in patients who have not achieved a response (at least a 20% reduction in blood phenylalanine concentrations from pre-treatment baseline levels or blood phenylalanine concentrations ≤600 micromol/L) after 16 weeks of continuous treatment with the maximum dosage of 40 mg once daily.

Phase of Treatment	Palynziq Dosing Regimen	Duration[a]
Induction	2.5 mg SC once weekly	4 weeks
Titration	2.5 mg SC twice weekly	1 week
	10 mg SC once weekly	1 week
	10 mg SC twice weekly	1 week
	10 mg SC four times per week	1 week
	10 mg SC once daily	1 week
Maintenance[b]	20 mg SC once daily	24 weeks

[a]Additional time may be required prior to each dosage escalation based on patient tolerability.
[b]Treatment should be individualized to the lowest effective and tolerated dosage. Increasing Palynziq to a maximum dosage of 40 mg once daily may be considered in patients who have not achieved a therapeutic response with at least 24 weeks of 20 mg once daily.

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REFERENCES

- Palynziq [prescribing information]. Novato, CA. BioMarin Pharmaceutical, Inc. May 2018.

Library	Commercial	NSA
Yes	Yes	No

Part D Effective: N/A
Commercial Effective: 10/01/18
Created: 08/18
Client Approval: 09/18
P&T Approval: 07/18