HIGHMARK WHOLECARE

Updated: 10/2025
PARP Approved: 12/2025

Prior Authorization Criteria

Phenylketonuria Medications

All requests for Phenylketonuria Medications require a prior authorization and will be screened for medical necessity and appropriateness using the criteria listed below.

Phenylketonuria Medications include Kuvan (sapropterin dihydrochloride), Javygtor (sapropterin dihydrochloride), Palynziq (pegvaliase-PQPZ), Zelvysia (sapropterin dihydrochloride) and Sephience (sepiapterin). New products with this classification will require the same documentation.

Phenylketonuria Medications Prior Authorization Criteria:

For all requests the following criteria must be met in addition to the diagnosis specific criteria below:

- Diagnosis of phenylketonuria (PKU)
- The requested dose and frequency is in accordance with FDA-approved labeling, nationally recognized compendia, and/or evidence-based practice guidelines
- Must be age-appropriate according to FDA-approved labeling, nationally recognized compendia, or evidence-based practice guidelines
- Member must have documentation of failure to Phe restricted diet as monotherapy
- Member is not receiving Kuvan (sapropterin dihydrochloride), Javygtor (sapropterin dihydrochloride), Palynziq (pegvaliase-pqpz), Zelvysia (sapropterin dihydrochloride) or Sephience (sepiapterin) in combination with each other
- The medication is prescribed by or in consultation with a metabolic disease specialist (or a specialist who focuses in the treatment of metabolic diseases)
- Must be used in conjunction with a Phe-restricted diet, including dietary protein and Phe restriction

For all requests for Kuvan (sapropterin dihydrochloride) or Javygtor (sapropterin dihydrochloride), Zelvysia (sapropterin dihydrochloride) or Sephience (sepiapterin) all of the following criteria must be met:

- Member who are neonates through 12 years of age must have Phe levels greater than or equal to 6mg/dL (360 micromol/L)
- Members who are 12 years of age or older must have Phe levels greater than or equal to 10 mg/dL (600 micromol/L)
- **Initial Duration of Approval:** 8 weeks
- **Reauthorization criteria**
    - Documentation of baseline (pre-treatment) blood Phe levels.
    - Documentation of Phe levels decreased by 20% or greater from baseline or Phe levels between 120 and 600 micromol/L.
- **Reauthorization Duration of approval:** 12 months

For all requests for Palynziq (pegvaliase-PQPZ), all of the following criteria must be met:

- Member must have Phe levels greater than 10mg/dL (600micromol/L)

- Must provide documentation showing the member has tried and failed, had an intolerance or contraindication, or has a genotype that is known to be non-responsive to Kuvan, Javygtor or Zelvysia (prior authorization required, in conjunction with a phenylalanine-restricted diet).
- **Initial Duration of Approval:** 16 weeks
- **Reauthorization criteria**
    - Documentation of baseline (pre-treatment) blood Phe levels.
    - Documentation of Phe levels decreased by 20% or greater from baseline or Phe levels between 120 and 600 micromol/L.
- **Reauthorization Duration of approval:** 12 months

Coverage may be provided for any non-FDA labeled indication if it is determined that the use is a medically accepted indication supported by nationally recognized pharmacy compendia or peer-reviewed medical literature for treatment of the diagnosis(es) for which it is prescribed. These requests will be reviewed on a case by case basis to determine medical necessity.

When criteria are not met, the request will be forwarded to a Medical Director for review. The physician reviewer must override criteria when, in their professional judgment, the requested medication is medically necessary.

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PHENYLKETONURIA MEDICATIONS
PRIOR AUTHORIZATION FORM

Please complete and fax all requested information below including any progress notes, laboratory test results, or chart documentation as applicable to Highmark Wholecare Pharmacy Services. **FAX:** (888) 245-2049
If needed, you may call to speak to a Pharmacy Services Representative. **PHONE**: (800) 392-1147 Mon – Fri 8:30am to 5:00pm

PROVIDER INFORMATION

Requesting Provider:	Provider NPI:
Provider Specialty:	Office Contact:
State license #:	Office NPI:
Office Address:	Office Phone:
	Office Fax:

MEMBER INFORMATION

Member Name:	DOB:	
Member ID:	Member weight:	Height:

REQUESTED DRUG INFORMATION

Medication:	Strength:	
Directions:	Quantity:	Refills:
Is the member currently receiving requested medication? ☐ Yes ☐ No	Date Medication Initiated:	

Billing Information

This medication will be billed: ☐ at a pharmacy **OR** ☐ medically, JCODE:_____

Place of Service: ☐ Hospital ☐ Provider's office ☐ Member's home ☐ Other

Place of Service Information

Name:	NPI:
Address:	Phone:

MEDICAL HISTORY (Complete for ALL requests)

Diagnosis:	ICD Code:

Has the member failed a Phe restricted diet as monotherapy (please attach clinical documentation)? ☐ Yes ☐ No

Is the member receiving any Phenylketonuria medications in conjunction with each other? ☐ Yes ☐ No

If requesting Palynziq, does the member have a genotype known to be non-responsive to Kuvan, Javygtor or Zelvysia? ☐ Yes ☐ No

Will the member be taking the requested medication in conjunction with a Phe-restricted diet? ☐ Yes ☐ No

REFERENCE VALUES

LAB	Baseline (pre-treatment) Value	Units (circle one)	Date	Post-Therapy Value (Reauthorization Only)	Units (circle one)	Date
Blood Phenylalanine (Phe) Levels		mg/dL or micromole/L			mg/dL or micromole/L	

CURRENT or PREVIOUS THERAPY

Medication Name	Strength/ Frequency	Dates of Therapy	Status (Discontinued & Why/Current)

REAUTHORIZATION

Has the member experienced an improvement with treatment? ☐ Yes ☐ No

Has the member's Phe levels decreased by 20% or greater from baseline? ☐ Yes, please include documentation ☐ No

SUPPORTING INFORMATION or CLINICAL RATIONALE

Prescribing Provider Signature	Date