HIGHMARK HEALTH OPTIONS

Updated: 10/2025
DMMA Approved: 11/2025

Request for Prior Authorization for Phenylketonuria Medications
Website Form – www.highmarkhealthoptions.com
Submit request via: Fax - 1-855-476-4158

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 Prior Authorization Criteria:

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.

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

- Member must have Phe levels greater than 6mg/dL (360 mm/L) for neonates through 12 years of age.
- Phe levels must be greater than 10mg/dL (600 mmol/L) on average after the age of 12.
- **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 (600mm/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 or Javygtor (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.

Drugs are authorized in generic form unless the branded product is on the preferred drug list or the prescriber has indicated in writing that the branded product is medically necessary. If only the branded product is on the preferred drug list, the generic form will be considered non-preferred and shall not require the prescriber to indicate in writing that the branded product 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 Health Options Pharmacy Services. **FAX: (855) 476-4158**
If needed, you may call to speak to a Pharmacy Services Representative. **PHONE**: (844) 325-6251 Mon – Fri 8:00 am to 7:00 pm

PROVIDER INFORMATION

Requesting Provider:	NPI:
Provider Specialty:	Office Contact:
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:	

Is this medication being used for a chronic or long-term condition for which the medication may be necessary for the life of the patient? ☐ Yes ☐ No

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)

Please provide pre-treatment blood Phe levels (please also attach clinical documentation): _____
Has the member tried and failed a Phe restricted diet(please attach clinical documentation)? ☐ Yes ☐ No
If requesting Palynziq, does the member have a genotype known to be non-responsive to Kuvan? ☐ Yes ☐ No

CURRENT or PREVIOUS THERAPY

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

REAUTHORIZATION

Has Phe levels decreased by 20% or greater from baseline? ☐ Yes ☐ No
Please provide pre-treatment and post-treatment blood Phe levels (please also attach clinical documentation):
Pre-treatment: _____ Post-treatment: _____

SUPPORTING INFORMATION or CLINICAL RATIONALE

Prescribing Provider Signature	Date