

Reference number(s)
3219-D

This document applies to the following:

Formulary	Applies
Standard Control (SF)	<input checked="" type="checkbox"/>
Standard Control – Choice (SCCF)	<input checked="" type="checkbox"/>
Preferred Drug Plan Design (PDPD)	<input type="checkbox"/>
Advanced Control Specialty (ACSF)	<input checked="" type="checkbox"/>
Advanced Control Specialty – Choice (ACSCF)	<input checked="" type="checkbox"/>
Managed Medicaid Template (MMT)	<input type="checkbox"/>
Marketplace (MF)	<input type="checkbox"/>
Aetna Small Group Affordable Care Act (SG ACA) Aetna Health Exchange (AHE)	<input type="checkbox"/>
Aetna Individual Lives (IVL)	<input type="checkbox"/>
Value (VF)	<input checked="" type="checkbox"/>

Formulary	Applies
New to Market (NTM)	<input type="checkbox"/>
Standard Formulary Chart (SFC)	<input checked="" type="checkbox"/>
Basic Control Chart Preferred Drug Plan Design (BCC PDPD)	<input type="checkbox"/>
Advanced Control Specialty Formulary Chart (ACSFC)	<input checked="" type="checkbox"/>
Value Formulary Chart (VFC)	<input checked="" type="checkbox"/>
Medical Benefit	<input type="checkbox"/>
Medical Benefit: Advanced Biosimilars First	<input type="checkbox"/>
Medical Benefit: Managed Medicaid (MMMB)	<input type="checkbox"/>
Medicare Part B	<input type="checkbox"/>
Medicare Part B: Advanced Biosimilars First	<input type="checkbox"/>

# Exceptions Criteria

## Phenylketonuria (PKU)

This document informs prescribers of preferred products and provides an exception process for targeted products through prior authorization.

These criteria were developed to align with the following: Standard Control Formulary (SF), Standard Control Choice Formulary (SCCF), Advanced Control Specialty Formulary (ACSF), Advanced Control Specialty – Choice Formulary (ACSCF), Value Formulary (VF), Standard Formulary Chart (SFC), Advanced Control Specialty Formulary Chart (ACSFC), and Value Formulary Chart (VFC).

## Plan Design Summary

This program applies to the phenylketonuria (PKU) products specified in this document. Coverage for targeted products is provided based on clinical circumstances that would exclude the use of the preferred product and may be based on previous use of a product. The coverage review process will ascertain situations where a clinical exception can be made. This program applies to all members requesting treatment with Kuvan. This program applies to members who are new to treatment with Palynziq for the first time.

Each referral is reviewed based on all utilization management (UM) programs implemented for the client.

## Table. Phenylketonuria (PKU)

Medications considered formulary or preferred on your plan may still require a clinical prior authorization review.

	Products
Preferred	<ul style="list-style-type: none"> <li>sapropterin dihydrochloride (generic)</li> </ul>
Target	<ul style="list-style-type: none"> <li>Kuvan (sapropterin dihydrochloride)</li> <li>Palynziq (pegvaliase-pqpz)</li> </ul>

## Exception Criteria

This program applies to members requesting treatment for an indication that is FDA-approved for the preferred product.

### Palynziq

Coverage for Palynziq is provided when any of the following criteria are met:

- Member is currently receiving treatment with Palynziq, excluding when Palynziq is obtained as samples or via manufacturer's patient assistance programs.
- Member has a documented inadequate response or intolerable adverse event with sapropterin dihydrochloride.
- Member has a documented phenylalanine hydroxylase (PAH) deleterious genotype with two null-alleles.

### Kuvan

Coverage for Kuvan is provided when all of the following criteria are met:

- Member has a documented intolerable adverse event with the preferred product.
- The adverse event was not an expected adverse event attributed to the active ingredient as described in the prescribing information.

## References

- Kuvan [package insert]. Novato, CA: BioMarin Pharmaceutical Inc.; August 2024.
- Palynziq [package insert]. Novato, CA: BioMarin Pharmaceutical Inc.; November 2020.
- Sapropterin dihydrochloride [package insert]. Memphis, TN: Northstar RX LLC; September 2023.
- Regier DS, Greene CL. Phenylalanine Hydroxylase Deficiency. GeneReviews [Internet]. Available at <https://www.ncbi.nlm.nih.gov/books/NBK1504/> Updated January 5, 2017. Accessed September 24, 2024.