UnitedHealthcare Pharmacy
Clinical Pharmacy Programs

Program Number	2025 P 1057-14
Program	Prior Authorization/Notification
Medication	Javygtor™ (sapropterin dihydrochloride)*, Kuvan® (sapropterin dihydrochloride)*
P&T Approval Date	4/2008, 4/2009, 3/2010, 3/2011, 1/2012, 2/2013, 10/2013, 10/2014, 10/2015, 9/2016, 9/2017, 7/2018, 7/2019, 7/2020, 7/2021, 7/2022, 3/2023, 3/2024, 3/2025
Effective Date	6/1/2025

1. Background:

Javygtor and Kuvan are phenylalanine hydroxylase activators indicated to reduce blood phenylalanine (Phe) levels in adult and pediatric patients one month of age and older with hyperphenylalaninemia (HPA) due to tetrahydrobiopterin- (BH4-) responsive Phenylketonuria (PKU). Javygtor and Kuvan are to be used in conjunction with a Phe-restricted diet.

2. Coverage Criteria[a]:

A. Initial Authorization

1. **Javygtor*** and **Kuvan*** will be approved based on **all** of the following criteria:

   a. Diagnosis of phenylketonuria (PKU)

   -AND-

   b. Patient is actively on a Phe-restricted diet

   -AND-

   c. Patient is not receiving the requested medication in combination with Palynziq (pegvaliase-pqpz)

Authorization will be issued for 12 months.

B. Reauthorization

1. **Javygtor*** and **Kuvan*** will be approved based on **all** of the following criteria:

   a. Patient is actively on a Phe-restricted diet

   -AND-

   b. Blood Phe levels continue to remain lower than baseline level

   -AND-

c. Patient is not receiving the requested medication in combination with Palynziq (pegvaliase-pqpz)

Authorization will be issued for 12 months.

[a] State mandates may apply. Any federal regulatory requirements and the member specific benefit plan coverage may also impact coverage criteria. Other policies and utilization management programs may apply.

*Javygtor and Brand Kuvan are typically excluded from coverage. Tried/Failed criteria may be in place. Please refer to plan specifics to determine exclusion status.

3. Additional Clinical Rules:

- Notwithstanding Coverage Criteria, UnitedHealthcare may approve initial and re-authorization based solely on previous claim/medication history, diagnosis codes (ICD-10) and/or claim logic. Use of automated approval and re-approval processes varies by program and/or therapeutic class.
- Supply limits may also be in place.

4. References:

1. Kuvan [package insert]. Novato, CA: BioMarin Pharmaceutical Inc.; August 2024.
2. Javygtor [package insert]. Princeton, NJ: Dr. Reddy's Laboratories, Inc.; October 2024.
3. Javygtor powder for oral solution [package insert]. Princeton, NJ: Dr. Reddy's Laboratories, Inc.; October 2024.
4. Smith WE, Berry SA, Bloom K, et al. Phenylalanine hydroxylase deficiency diagnosis and management: A 2023 evidence-based clinical guideline of the American College of Medical Genetics and Genomics (ACMG). Genet Med. 2025;27(1):101289. doi:10.1016/j.gim.2024.101289

Program	Prior Authorization/Notification – Javygtor and Kuvan (sapropterin dihydrochloride)
Change Control	
10/2013	Removed age criterion.
10/2014	Annual review. Updated references.
10/2015	Annual review. Updated authorization period to 6 mo. Updated reauthorization requirement. Background edit. Updated references.
9/2016	Annual review. Updated references.
9/2017	Annual review with no changes to coverage criteria.
7/2018	Added criteria restricting combination use with Palynziq
12/2018	Administrative change to add statement regarding use of automated processes.
7/2019	Annual review with no changes to coverage criteria. Updated reference.
7/2020	Annual review with no changes to coverage criteria. Updated reference.
7/2021	Annual review with no changes to clinical criteria. Updated re-authorization to 12 months. Added statement that Brand Kuvan is excluded from coverage for the majority of our benefits. Updated background and references.

7/2022	Annual review with no changes to coverage criteria. Added state mandate footnote.
3/2023	Added Javygtor to program. Updated exclusion footnote and references.
3/2024	Annual review. Updated authorization approval duration to 12 months. Updated reference.
3/2025	Annual review. Updated references.