

UnitedHealthcare Pharmacy
Clinical Pharmacy Programs

Program Number	2025 P 1057-15
Program	Prior Authorization/Notification
Medication	Javygtor™ (sapropterin dihydrochloride)*, Kuvan® (sapropterin dihydrochloride)*, sapropterin dihydrochloride, Zelvysia (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, 11/2025
Effective Date	2/1/2026

1. Background:

Javygtor, Kuvan, and Zelvysia 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, Kuvan, and Zelvysia are to be used in conjunction with a Phe-restricted diet.

2. Coverage Criteria^a:

<p>A. <u>Initial Authorization</u></p> <p>1. Javygtor*, Kuvan*, sapropterin dihydrochloride, or Zelvysia will be approved based on all of the following criteria:</p> <p style="padding-left: 40px;">a. Diagnosis of phenylketonuria (PKU)</p> <p style="text-align: center;">-AND-</p> <p style="padding-left: 40px;">b. Patient is actively on a Phe-restricted diet</p> <p style="text-align: center;">-AND-</p> <p style="padding-left: 40px;">c. Patient is not receiving the requested medication in combination with Palynziq (pegvaliase-pqpz) or Sephience (sepiapterin)</p> <p>Authorization will be issued for 12 months.</p> <p>B. <u>Reauthorization</u></p> <p>1. Javygtor*, Kuvan*, sapropterin dihydrochloride, or Zelvysia will be approved based on all of the following criteria:</p> <p style="padding-left: 40px;">a. Patient is actively on a Phe-restricted diet</p> <p style="text-align: center;">-AND-</p> <p style="padding-left: 40px;">b. Blood Phe levels continue to remain lower than baseline level</p>

-AND-

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

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. Zelvysia [package insert]. Piscataway, NJ: Aucta Pharmaceuticals, Inc.; April 2025.
5. 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

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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.
11/2025	Added Zelvysia to program. Added Sephience to combination use criteria. Updated references.