

Drug Policy

Policy:	Kygevvi™ (doxecitine and doxribtimine powder for oral solution – UCB)	Annual Review Date: 07/16/2026 Last Revised Date: 07/16/2026
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OVERVIEW

Kygevvi is a combination of doxecitine and doxribtimine, both pyrimidine nucleosides, indicated for the treatment of thymidine kinase 2 deficiency (TK2d) in adults and pediatric patients with an age of symptom onset on or before 12 years.¹

POLICY STATEMENT

This policy involves the use of Rhapsido. Prior authorization is recommended for pharmacy benefit coverage of Rhapsido. Approval is recommended for those who meet the conditions of coverage in the **Criteria and Initial/Extended Approval** for the diagnosis provided. **Conditions Not Recommended for Approval** are listed following the recommended authorization criteria. Requests for uses not listed in this policy will be reviewed for evidence of efficacy and for medical necessity on a case-by-case basis.

Because of the specialized skills required for evaluation and diagnosis of patients treated with Rhapsido as well as the monitoring required for adverse events and long-term efficacy, initial approval requires Rhapsido be prescribed by or in consultation with a physician who specializes in the condition being treated. All approvals for initial therapy are provided for the initial approval duration noted below; if reauthorization is allowed, a response to therapy is required for continuation of therapy unless otherwise noted below.

RECOMMENDED AUTHORIZATION CRITERIA

Coverage of Rhapsido is recommended in those who meet the following criteria:

1. **Thymidine Kinase 2 Deficiency (TK2d).** Approve for 1 year if the patient meets ALL of the following (A, B, and C):
 - A) Patient has had a genetic test confirming the diagnosis of TK2d with biallelic pathogenic or likely pathogenic variants in the *TK2* gene*; AND
 - B) According to the prescriber, the patient had onset of symptoms consistent with TK2d at ≤ 12 years of age; AND
Note: Examples of symptoms consistent with TK2d include progressive muscle weakness, hypotonia (i.e., low muscle tone), respiratory insufficiency, loss of motor skills, feeding and/or swallowing difficulties, facial weakness/paralysis, ptosis (i.e., drooping eyelid[s]), ophthalmoparesis (i.e., difficulty moving the eyes), developmental delay and regression, hearing loss, and seizures.
 - C) The medication is prescribed by or in consultation with a neurologist, geneticist, or physician who specializes in metabolic and/or neuromuscular disorders.

Drug Policy

Initial Approval/ Extended Approval.

A) *Initial Approval:* 1 year

B) *Extended Approval:* 1 year

CONDITIONS NOT RECOMMENDED FOR APPROVAL

Rhapsido has not been shown to be effective, or there are limited or preliminary data or potential safety concerns that are not supportive of general approval for the following conditions. (Note: This is not an exhaustive list of Conditions Not Recommended for Approval).

1. Coverage is not recommended for circumstances not listed in the Recommended Authorization Criteria. Criteria will be updated as new published data are available.

***Documentation Requirements:**

The Company reserves the right to request additional documentation as part of its coverage determination process. The Company may deny reimbursement when it has determined that the drug provided or services performed were not medically necessary, investigational, or experimental, not within the scope of benefits afforded to the member and/or a pattern of billing or other practice has been found to be either inappropriate or excessive. Additional documentation supporting medical necessity for the services provided must be made available upon request to the Company.

Documentation requested may include patient records, test results and/or credentials of the provider ordering or performing a service. The Company also reserves the right to modify, revise, change, apply and interpret this policy at its sole discretion, and the exercise of this discretion shall be final and binding.

REFERENCES

1. Kygevvii™ powder for oral solution [prescribing information]. Smyrna, GA: UCB; November 2025.
2. National Organization for Rare Disorders. Thymidine kinase 2 deficiency. Available at: <https://rarediseases.org/rare-diseases/thymidine-kinase-2-deficiency/>. Updated March 4, 2025. Accessed on December 1, 2025.
3. Wang J, El-Hattab AW, Wong LJC. TK2-Related Mitochondrial DNA Maintenance Defect, Myopathic Form. 2012 Dec 6 [Updated 2018 Jul 26]. In: Adam MP, Feldman J, Mirzaa GM, et al., editors. GeneReviews® [Internet]. Seattle (WA): University of Washington, Seattle; 1993-2025.
4. Hirano M, Garone C, Haas R, et al. Survival analyses in patients with thymidine kinase 2 deficiency aged ≤12 years at symptom onset who received pyrimidine nucleos(t)ide therapy. Presented at: Muscular Dystrophy Association (MDA) Clinical & Scientific Conference; 2025.