



AETNA BETTER HEALTH®
Coverage Policy/Guideline

Name: Kygevvi

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Effective Date: 7/9/2026

Last Review Date: 6/8/2026

Applies to: Illinois
 Florida Kids

New Jersey
 Pennsylvania Kids

Maryland
 Virginia

Intent:

The intent of this policy/guideline is to provide information to the prescribing practitioner outlining the coverage criteria for Kygevvi under the patient's prescription drug benefit.

Description:

FDA-Approved Indication

KYGEVVI is 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.

All other indications are considered experimental/investigational and not medically necessary.

Applicable Drug List:

Kygevvi

Policy/Guideline:

Criteria for Initial Approval:

Submission of the following information is necessary to initiate the prior authorization review:

Initial Requests

Chart notes or medical record documentation documenting ALL of the following:

- Genetic confirmation of pathogenic TK2 variants supporting diagnosis.
- Absence of other genetic disease, polygenic disease, or inborn errors of metabolism.
- Symptom onset on or before 12 years of age.
- Baseline liver transaminases (ALT, AST) and total bilirubin levels prior to treatment initiation.

Continuation Requests

Chart notes and/or medical records documenting a response to therapy.

Prescriber Specialties

This medication must be prescribed by or in consultation with a pulmonologist or a physician who specializes in the treatment of metabolic or neuromuscular disorders.

Coverage Criteria

Thymidine Kinase 2 Deficiency

Authorization of 12 months may be granted for treatment of thymidine kinase 2 deficiency when ALL of the following criteria are met:

- Member has genetic confirmation of pathogenic TK2 variants.
- Member has absence of other genetic disease, polygenic disease, or concurrent inborn error of metabolism.



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- Member has experienced symptom onset on or before 12 years of age.
- Baseline liver transaminases (ALT, AST) and total bilirubin levels prior to treatment initiation.
- Member does not have a history of liver disease or liver function tests (ALT, AST, or total bilirubin) greater than or equal two times the upper limit of normal at baseline per the laboratory administering the test.
- Member does not have renal insufficiency requiring dialysis.
- Member does not have severe end-organ hypo-perfusion syndrome secondary to cardiac failure resulting in lactic acidosis.

Continuation of Therapy

Authorization of 12 months may be granted for members requesting continuation of therapy when the member has demonstrated a response to therapy [e.g., improvement or stabilization in motor function (e.g., 6-minute walk test [6MWT], Motor Function Measure [MFM] 20 or MFM 30), pulmonary function, ability to meet growth and developmental milestones].

Approval Duration and Quantity Restrictions:

Initial and Renewal Approval: 12 months

Quantity Level Limit: Reference Formulary for drug specific quantity level limits

References:

1. Kygevvi [package insert]. Smyrna, GA: UCB INC.; November 2025.
2. Hernandez-Voth A, Sayas Catalan J, Corral Blanco M, Castaño Mendez A, Martin MA, De Fuenmayor Fernandez de la Hoz C, Villena Garrido V, Dominguez-Gonzalez C. Deoxynucleoside therapy for respiratory involvement in adult patients with thymidine kinase 2-deficient myopathy. *BMJ Open Respir Res.* 2020 Nov;7(1):e000774. doi: 10.1136/bmjresp-2020-000774. PMID: 33246973; PMCID: PMC7703425.
3. Domínguez-González C, Chiang C, Colson AO, Rebollo Mesa I, Baixauli E, Quan J, VanMeter S, Hirano M. Pyrimidine Nucleos(t)ide Therapy in Patients With Thymidine Kinase 2 Deficiency: A Multicenter Retrospective Chart Review Study. *Neurology.* 2025 Sep 23;105(6):e213908. doi: 10.1212/WNL.0000000000213908. Epub 2025 Sep 5. PMID: 40911819; PMCID: PMC12413740.
4. ClinicalTrials.gov [Internet]. Bethesda (MD): National Library of Medicine (US). 2000 Feb 29 – . Identifier NCT05017818, A Retrospective Study of Subjects With Thymidine Kinase 2 Deficiency Treated With the Combination of Pyrimidine Nucleos(t)ides as Well as Untreated Subjects to Collect Vital Status Data and Supporting Information; 2021 Jul 23 [cited 2025 Nov 7]; [about 4 screens]. Available from: <https://clinicaltrials.gov/study/NCT05017818> [clinicaltrials.gov]
5. UCB BIOSCIENCES, Inc. (2025, September 12). *Doxecitine and Doxribtimine-Expanded Access* (NCT06590493). ClinicalTrials.gov. U.S. National Library of Medicine. <https://clinicaltrials.gov/study/NCT06590493> [clinicaltrials.gov]