MEDICATION POLICY:
Kygevvi (doxecitine and doxribtimine)

Applicable Drugs: Kygevvi (doxecitine and doxribtimine)

Preferred: N/A

Non-preferred: N/A

Date of Origin: 10/20/2026

Date Last Reviewed / Revised: 10/20/2026

PRIOR AUTHORIZATION CRITERIA

(May be considered medically necessary when criteria I through IV are met)

I. Documented diagnosis of Thymidine Kinase 2 deficiency (TK2d) and meets the following criteria (A through C):

   A. Presence of a known pathological mutation in the thymidine kinase 2 (TK2) gene with phenotypical manifestations.

      i. Phenotypical manifestations include but are not limited to ptosis, progressive external ophthalmoplegia (PEO), facial or cervical weakness, dysphagia, and early diaphragmatic involvement requiring mechanical ventilation.

   B. Documentation of baseline motor development milestones, feeding and/or respiratory support requirements, or other objective measures of function including but not limited to 6-minute walk test (6MWT) distance (in meters), Motor Function Measure [MFM] 20 or MFM 32, Egen Klassifikation, North Star Ambulatory Assessment, Children's Hospital of Philadelphia Infant Test of Neuromuscular Disorders [CHOP INTEND], or Hammersmith Functional Motor Scale-Expanded [HFMSE]

   C. Documented age of symptom onset on or before 12 years.

II. Prescribed by or in consultation with a neurologist or specialist experienced in managing mitochondrial myopathies.

III. Request is for a medication with the appropriate FDA labeling, or its use is supported by current clinical practice guidelines.

IV. Refer to the plan document for the list of preferred products. If the requested agent is not listed as a preferred product, must have a documented failure, intolerance, or contraindication to a preferred product(s).

EXCLUSION CRITERIA

- Age of symptom onset after 12 years.

OTHER CRITERIA

- N/A

Quantity / Days Supply Restrictions*

- Quantity limits are dependent on patient weight and number of doses and are limited to a maximum of a 30-day supply per fill.
- Use Appendix tables 1 and 2 to calculate the appropriate daily dose and total number of Kygevvi packets required per day.

Approval Length

- **Authorization:** 6 months.
- **Re-Authorization:** 12 months. An updated letter of medical necessity or progress notes showing current medical necessity criteria are met, updated patient weight is documented, and the medication is effective in improving patient symptoms or maintaining condition such as no longer requiring mechanical ventilation or improvement from baseline motor development milestones as described in criterion I.B.

Appendix

Table 1: Recommended Dosage Levels

Dosage Level	Total Daily Dose	Component Doses
Starting	260 mg/kg/day	130 mg doxecitine and 130 mg doxribtimine
Intermediate	520 mg/kg/day	260 mg doxecitine and 260 mg doxribtimine
Maintenance	800 mg/kg/day	400 mg doxecitine and 400 mg doxribtimine
Administration: All doses are given by mouth or via enteral feeding tube in 3 equally divided doses with food. **Titration:** Titration to the next dosage level is based on tolerability after a minimum of 2 weeks at the current dosage level.		

Table 2: Recommended Dosage - Daily-Dose Ranges and Packet Requirements

Total Daily Dose (mg/day)	Total Number of KYGEVVI Packets for Reconstitution[1]
750–4,049	1 packet
4,050–8,249	2 packets
8,250–12,749	3 packets
12,750–17,249	4 packets

17,250–21,749	5 packets
21,750–24,749	6 packets
24,750–29,249	7 packets
29,250–33,749	8 packets
33,750–38,249	9 packets
38,250–42,749	10 packets
42,750–47,249	11 packets
47,250–51,749	12 packets
51,750–54,749	13 packets
54,750–59,249	14 packets
59,250–63,749	15 packets
63,750–68,249	16 packets

[1] Each single use packet contains 4 grams of KYGEVVI as a white to off-white powder (2 grams doxecitine and 2 grams doxribtimine) in a carton of 30 packets.

REFERENCES

1. Kygevvi. Prescribing information. UCB, Inc; 2025. Accessed October 7, 2026. https://www.accessdata.fda.gov/drugsatfda_docs/label/2025/219792s000lbl.pdf
2. Wang LT, Jhuang HY. Clinical development in primary mitochondrial diseases at a translational inflection point: lessons, mechanisms, and emerging therapeutic strategies. *Biomed Pharmacother.* 2026;201:119699. doi:10.1016/j.biopha.2026.119699

DISCLAIMER: Medication Policies are developed to help ensure safe, effective and appropriate use of selected medications. They offer a guide to coverage and are not intended to dictate to providers how to practice medicine. Refer to Plan for individual adoption of specific Medication Policies. Providers are expected to exercise their medical judgement in providing the most appropriate care for their patients.