kygevvi™

(doxecitine and doxribtimine)

Powder for Oral Solution

2 g/2 g per packet

Getting Patients Started With KYGEVVI

This resource includes an overview of KYGEVVI acquisition and information that may be required for a prior authorization.

The information provided in this guide is of a general nature and for information purposes only. The responsibility to determine coverage and appropriate coding for a particular patient and/or procedure is always that of the provider or physician.

INDICATION

KYGEVVI is indicated for the treatment of thymidine kinase 2 deficiency (TK2d) in adult and pediatric patients with an age of symptom onset on or before 12 years.

IMPORTANT SAFETY INFORMATION

Increase in Liver Transaminases

Elevated liver transaminase [alanine aminotransferase (ALT) and/or aspartate aminotransferase (AST)] levels were reported in patients treated with KYGEVVI. Obtain baseline liver transaminase (ALT, AST) and total bilirubin levels in patients prior to treatment initiation with KYGEVVI. If signs or symptoms consistent with liver injury are observed, interrupt treatment with KYGEVVI until liver transaminase (ALT, AST) and total bilirubin levels have either returned to baseline or stabilized at a new baseline value. Consider permanently discontinuing KYGEVVI if signs or symptoms consistent with liver injury persist or worsen. Monitor liver transaminases and total bilirubin levels yearly and as clinically indicated.

Please see additional Important Safety Information throughout and the full Prescribing Information.

Acquisition Process Overview

KYGEVVI must be obtained through PANTHERx Rare, the exclusive, authorized specialty pharmacy. If applicable, communicate to your patient and their caregiver that KYGEVVI must be obtained from PANTHERx Rare even if their health insurance has a preferred specialty pharmacy. See the flow chart below for a general overview of the process for getting patients started on KYGEVVI.

HCP completes and sends the KYGEVVI Start Form with a prescription to ONWARD®*

HCP office conducts a benefits investigation to confirm coverage under pharmacy benefit

Collect information that may be required by your patient's health plan before approving KYGEVVI (e.g., copy of your patient's insurance cards, medical history)

ONWARD enrolls the patient in applicable support services once consent is obtained

Inform the patient and their caregiver to expect a call from an ONWARD Care Coordinator† (1-844-669-2731) to discuss the support available

ONWARD triages prescription to PANTHERx Rare for fulfillment

NPI No. 1659762524 | Fax: 1-412-567-6135 | Phone: 1-833-418-7760

PANTHERx Rare verifies insurance coverage

PANTHERx Rare initiates prior authorization (PA) support and will contact your office if PA is not already submitted

HCP office submits a PA to the patient's health plan

Reference the checklist on the next page if a PA is required

Health plan APPROVES treatment with KYGEVVI

Health plan DOES NOT APPROVE treatment with KYGEVVI

HCP office confirms reason for denial and resubmits PA or appeals to health plan

HCP can request PANTHERx Rare support for PA or appeals

Health plan APPROVES treatment with KYGEVVI

If coverage is denied, additional steps may be required

PANTHERx Rare will call the patient to schedule shipment

*The Start Form is used to enroll patients in the ONWARD Patient Support Program for access to important resources and support.
†Please note that when patients receive this call, they may see "ONWARD" on their caller ID.
HCP=healthcare professional; NPI=National Provider Identifier.

Please see Important Safety Information throughout and the full Prescribing Information.

Prior Authorization Checklist

Your patient's health plan may require a PA before KYGEVVI coverage can be approved. A common reason for coverage denial is incomplete or missing information (see 1-5 below) as indicated on the request form. Contact the patient's payer for requirements and clinical coverage guidelines for KYGEVVI, if available. This checklist is provided as an educational resource regarding potential PA requirements for KYGEVVI.

1 Diagnosis Code[1,*]

- ☐ E88.40 Mitochondrial metabolism disorder, unspecified
- ☐ E88.8 Other specified metabolic disorders
- ☐ Z15.89 Genetic susceptibility to other disease
- ☐ E88.49 Other mitochondrial metabolism disorders
- ☐ G71.3 Mitochondrial myopathy, not elsewhere classified
- ☐ Other: _____

*An ICD-10-CM code specific to thymidine kinase 2 deficiency (TK2d) is not available. The diagnosis codes provided are informational and are not intended to be directive or a guarantee of reimbursement. Please consult the most recent version of the ICD-10-CM for a full list of codes that may be applicable.

2 Clinical Information

Provide relevant information and supporting documentation regarding age of symptom onset, including chart notes and lab tests.

Patient's age: _____ Age at symptom onset: _____

Baseline functional status: _____

_____

Comorbidities (if any): _____

Confirmed genetic variant(s) in *TK2* gene: ☐ Yes ☐ No ☐ Not known Date of assessment: _____

Include results of additional tests that may have been conducted to confirm TK2d diagnosis. These tests may not be required and can include blood tests, muscle biopsy, brain or skeletal muscle MRI, and electromyography tests.

3 Disease Management History

Include information on medical equipment or devices (e.g., adaptive eating utensils, back braces, breathing support devices, wheelchair/walker) and medications the patient uses to manage symptoms and address muscle weakness.

_____

_____

4 Prescribed Dose[2]

- ☐ **Initiate with starting dose and titrate up to maintenance dose:** 260 mg/kg/day divided equally TID approximately 6 hours apart (plus or minus 2 hours) with food for a minimum of 2 weeks, then increase to 520 mg/kg/day divided equally TID for a minimum of 2 weeks, then increase to 800 mg/kg/day divided equally TID thereafter
- ☐ **Maintenance dose only:** 800 mg/kg/day divided equally TID approximately 6 hours apart (plus or minus 2 hours) with food
- ☐ Other: _____

Route of administration: ☐ Oral ☐ Feeding tube

Dispense days supply: _____ days Refills: ☐ Refill for 1 year ☐ Other: _____

5 Reauthorization

If the patient has already been approved for KYGEVVI under this plan, provide documentation demonstrating continued clinical benefit for the patient.

FDA=US Food and Drug Administration; ICD-10-CM=International Classification of Diseases, 10th Revision, Clinical Modification; MRI=magnetic resonance imaging; PA=prior authorization; TID=three times a day; *TK2*=thymidine kinase 2 gene.

IMPORTANT SAFETY INFORMATION (cont'd)

Gastrointestinal Adverse Reactions

Diarrhea and vomiting leading to hospitalization, dose reduction, and permanent discontinuation were reported in patients treated with KYGEVVI. Based on the severity of the diarrhea and/or vomiting, reduce the dosage of KYGEVVI or interrupt treatment until diarrhea and/or vomiting improves or returns to baseline. Consider restarting KYGEVVI at the last tolerated dose, and increase the dose as tolerated. For persistent or recurring diarrhea and/or vomiting, consider discontinuing KYGEVVI permanently and provide supportive care with electrolyte repletion as clinically indicated.

Please see additional Important Safety Information throughout and the full Prescribing Information.

Patient Support for KYGEVVI

ONWARD

Patient Support

If you have questions about enrolling your patients who are receiving KYGEVVI in the ONWARD® Patient Support Program, please visit ucbONWARD.com to access resources for healthcare professionals.

ONWARD is provided as a service of UCB and is intended to support the appropriate use of UCB medicines. ONWARD may be amended or canceled at any time without notice. Some program and eligibility restrictions may apply.

If you have questions or for more information, please contact your Mito Case Manager

IMPORTANT SAFETY INFORMATION (cont'd)

Adverse Reactions

The most common adverse reactions (incidence ≥5%) are diarrhea, abdominal pain (including abdominal pain upper), vomiting, alanine aminotransferase increased (ALT), and aspartate aminotransferase increased (AST).

Please see the full Prescribing Information.

For more information about KYGEVVI, visit KYGEVVIhcp.com.

For additional information, contact UCBCares® at 1-844-599-CARE (2273).

References: 1. Centers for Medicare & Medicaid Services. ICD-10-CM tabular list of diseases and injuries. Available at: https://www.cms.gov/medicare/coding-billing/icd-10-codes. Accessed November 19, 2025. **2.** KYGEVVI [prescribing information]. Smyrna, GA: UCB, Inc.

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