



kygevvi™

[doxycitine and doxribtimine]

Powder for Oral Solution

2 g/2 g per packet

KYGEVVI is the first and only FDA-approved treatment for **thymidine kinase 2 deficiency (TK2d)** in adults and pediatric patients with an age of symptom onset on or before 12 years<sup>1</sup>

# Dosing Guide

## 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.

Please see the full [Important Safety Information](#) on [page 5](#).  
Please see the full [Prescribing Information](#).

# KYGEVVI dosage<sup>1</sup>

Obtain baseline liver transaminase (alanine aminotransferase [ALT] and aspartate aminotransferase [AST]) and total bilirubin levels in patients prior to treatment initiation with KYGEVVI.

The recommended dosage of KYGEVVI is based on the patient's weight. Titrate to the next dosage level based on tolerability after a minimum of 2 weeks at the current dosage level.

Administer KYGEVVI orally in 3 equally divided doses approximately 6 hours apart (plus or minus 2 hours) with food.

## Recommended starting, intermediate, and maintenance dosage of KYGEVVI

KYGEVVI Dosage Level	KYGEVVI Dosage (mg/kg/day)
<b>Starting</b>	260 mg/kg/day (consisting of 130 mg doxecitine and 130 mg doxribtimine)
<b>Intermediate</b>	520 mg/kg/day (consisting of 260 mg doxecitine and 260 mg doxribtimine)
<b>Maintenance</b>	800 mg/kg/day (consisting of 400 mg doxecitine and 400 mg doxribtimine)



Since dosage is **based on a patient's weight**, reassess the dose based on their potential for change in weight.<sup>1</sup>



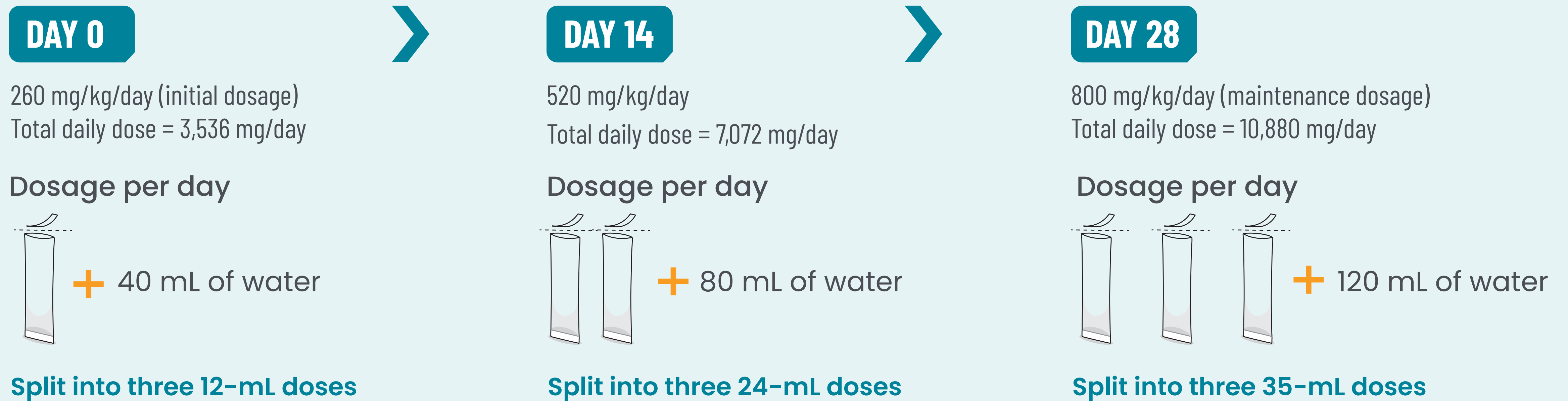
KYGEVVI is titrated, based on tolerability, up to the recommended maintenance daily dose of **800 mg/kg** split into 3 equal doses.<sup>1</sup>

Please see the full [Important Safety Information](#) on page 5.  
Please see the full [Prescribing Information](#).

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# KYGEVVI recommended **titration schedule**<sup>1</sup>

## Example titration for a 30-lb (13.6 kg) patient



### IMPORTANT SAFETY INFORMATION

#### Adverse Reactions

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

Access a preparation and dosing table based on total daily dosage range from the KYGEVVI full Prescribing Information [here](#).

Please see the full [Important Safety Information](#) on page 5.  
Please see the full [Prescribing Information](#).

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# Getting started with KYGEVVI

KYGEVVI is a powder for solution taken orally or via feeding tube 3 times each day about every 6 hours (plus or minus 2 hours) with food. KYGEVVI should be prepared and administered by adults only.<sup>1</sup>

When prescribed KYGEVVI, a patient will be provided with:<sup>1</sup>



Carton(s) of 30 KYGEVVI single-use powder packets.

ZX2000 convenience kit, which contains 1 dosing cup, 1 mixing bottle, 2 oral syringes, 2 spare seals, and KYGEVVI Instructions for Use.

Each single-use packet contains 4 g of KYGEVVI. Use 40 mL of room temperature water per packet to prepare the oral solution. Refer to the Instructions for Use for preparation and administration instructions.

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Please see the full Important Safety Information on page 5.  
Please see the full Prescribing Information and Instructions for Use.



ONWARD® is a single point of entry for patients to receive **personalized support**, tools, and resources throughout their treatment journey with KYGEVVI. Learn about how to enroll your patients in ONWARD at [kygevvihcp.com](https://kygevvihcp.com).

## 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.

### 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.

### Adverse Reactions

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

**Please see the full Prescribing Information.**

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TK2d is a progressive, genetic, and **life-threatening** mitochondrial disease<sup>2</sup>

**KYGEVVI is the first and only FDA-approved treatment for TK2d in adults and pediatric patients with symptom onset  $\leq$ 12 years of age<sup>1</sup>**

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.<sup>1</sup>



**Studies found improved survival time from treatment initiation with KYGEVVI is possible<sup>1</sup>**

## **IMPORTANT SAFETY INFORMATION**

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**Please see the full Important Safety Information on page 5.**

**References:** **1.** KYGEVVI [Prescribing Information]. Smyrna, GA: UCB, Inc. **2.** Garone C, Taylor RW, Nascimento A, et al. Retrospective natural history of thymidine kinase 2 deficiency. *J Med Genet.* 2018;55(8):515-521. doi:10.1136/jmedgenet-2017-105012



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