

kygevv[™]

(doxecitine and doxribtimine)

Powder for Oral Solution

2 g/2 g per packet

INSTRUCTIONS FOR USE KYGEVVI[™] (KY-JEH-vee)

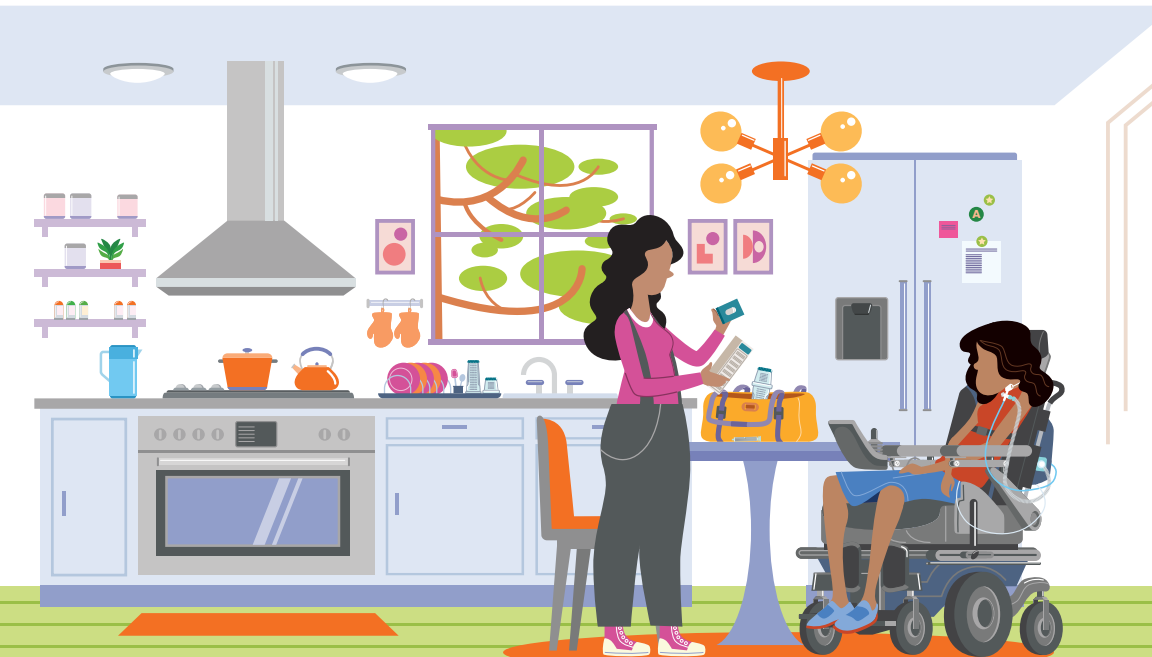


TABLE OF CONTENTS

Instructions for use

Important information	5
-----------------------------	---

Before you start

Supplies for preparing and taking or giving KYGEVVI	6
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Important information

What you need to know before preparing and taking or giving KYGEVVI ...	10
Storing KYGEVVI packets and oral solution	11

Preparing your one-day supply of KYGEVVI

Get supplies ready.....	12
Measure water and add powder from packets.....	14
Mix and inspect medicine.....	16

Dosing methods

How to measure your individual dose.....	18
--	----

Individual doses 50 mL or more

Measure and take or give your individual dose.....	20
--	----

Individual doses less than 50 mL

Measure and take or give your individual dose.....	22
--	----

Between individual doses

Clean up after first and second individual dose.....	26
--	----

End-of-day clean up

Pour out and clean up after third individual dose.....	28
--	----

Dosing cup maintenance

Replacing the seal if misplaced or damaged.....	30
---	----

Contact information.....

	32
--	----

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OVERVIEW

KYGEVVI is a prescription medicine used for the treatment of **thymidine kinase 2 deficiency (TK2d)** in adults and pediatric patients with a symptom onset on or before 12 years of age.

This document includes the **Instructions for Use (IFU)** for this treatment.

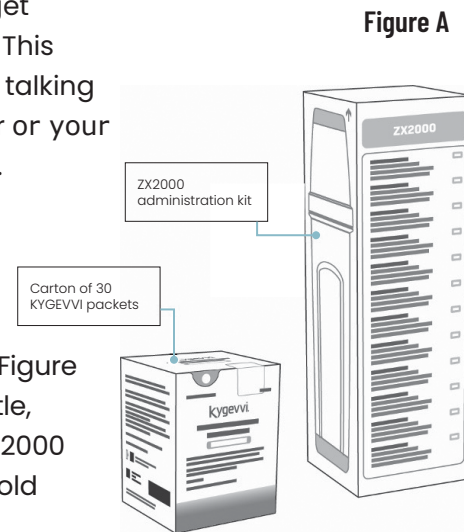
INSTRUCTIONS FOR USE

Important information

This Instructions for Use contains information on how to prepare a one-day supply of KYGEVVI, and take or give an individual dose of KYGEVVI.

Read this Instructions for Use before taking or giving KYGEVVI and each time you get a refill. There may be new information. This information does not take the place of talking to your healthcare provider about your or your child's medical condition or treatment.

When you are prescribed KYGEVVI for the first time, you will be provided with the carton(s) of 30 KYGEVVI packets and the ZX2000 administration kit (see Figure A). Only use the dosing cup, mixing bottle, and oral syringes provided with your ZX2000 administration kit. **Do not** use a household measuring cup or measuring spoon.

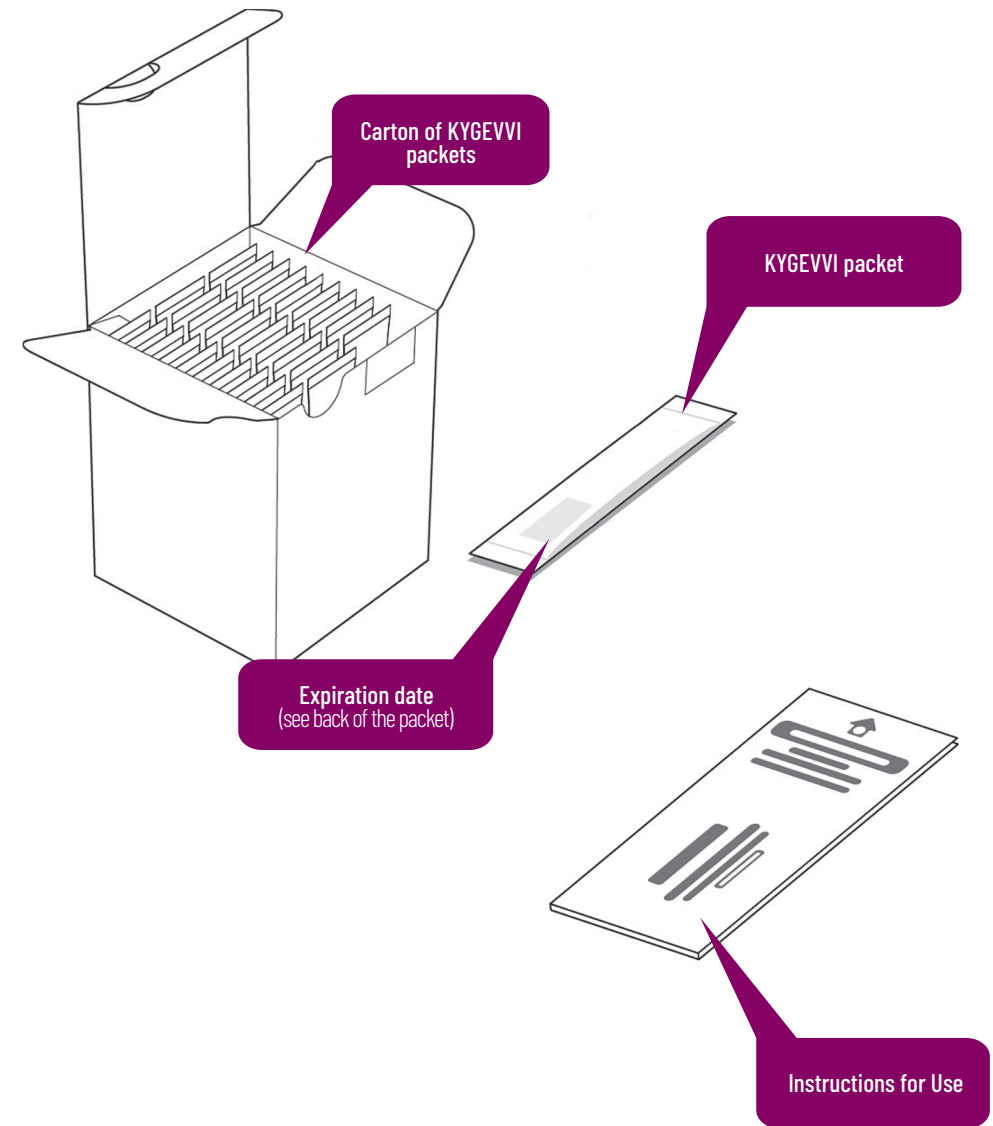
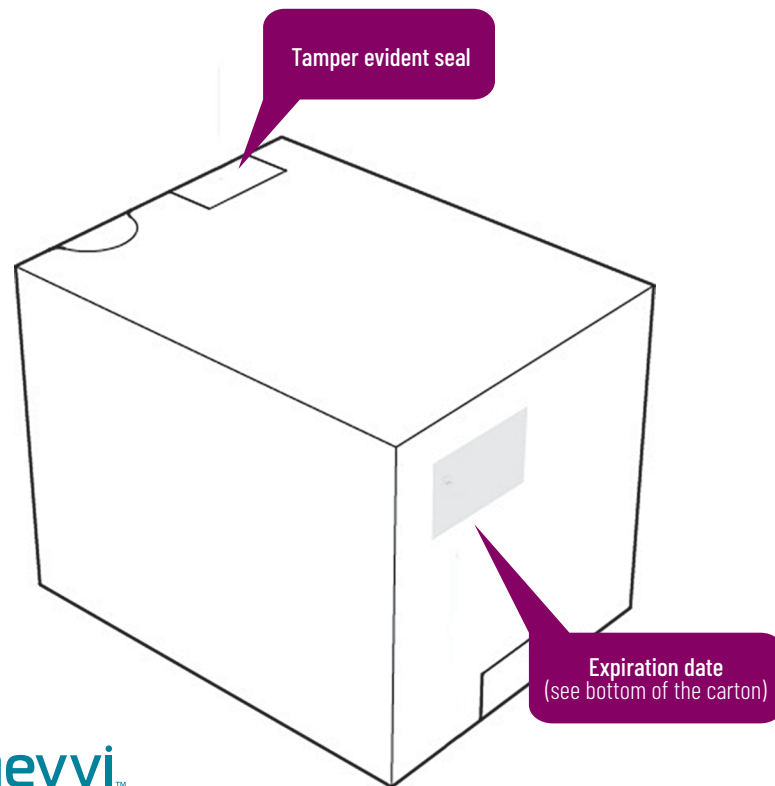


BEFORE YOU START

Supplies for preparing and taking or giving KYGEVVI.

KYGEVVI carton contents:

- ☐ KYGEVVI packets (x30)
- ☐ Instructions for Use (IFU)



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BEFORE YOU START

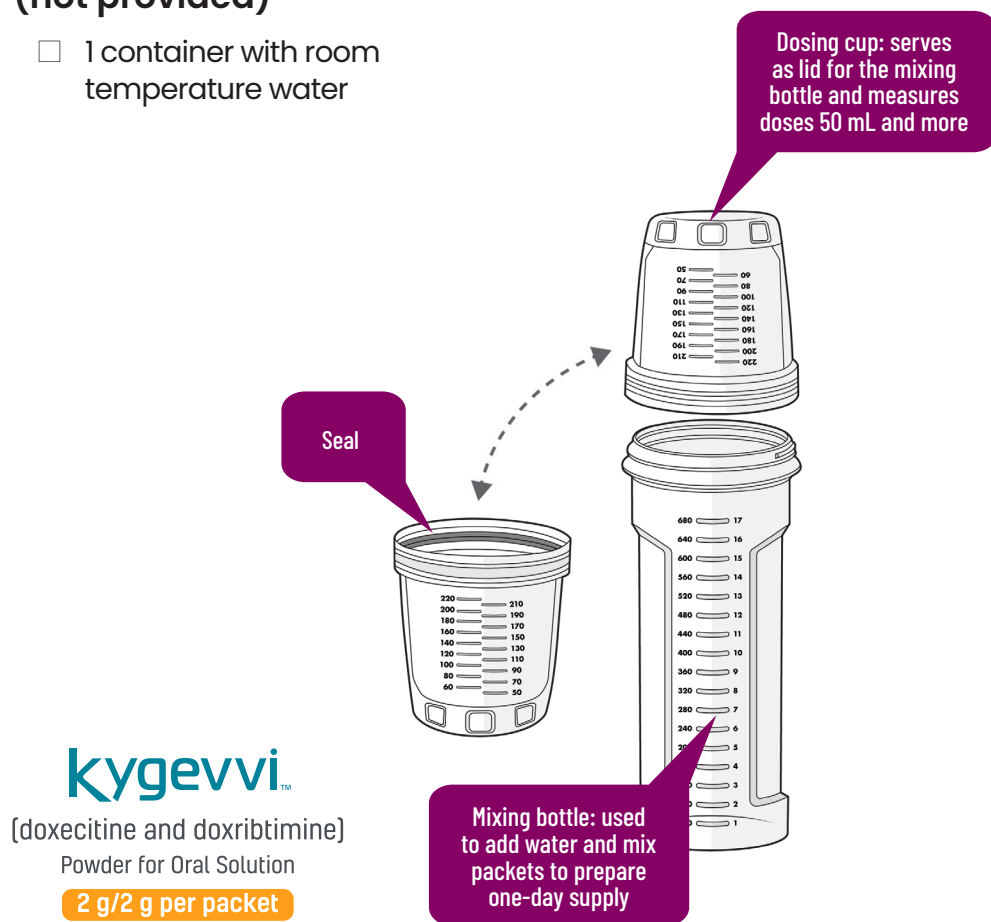
Supplies for preparing and taking or giving KYGEVVI.

ZX2000 administration kit

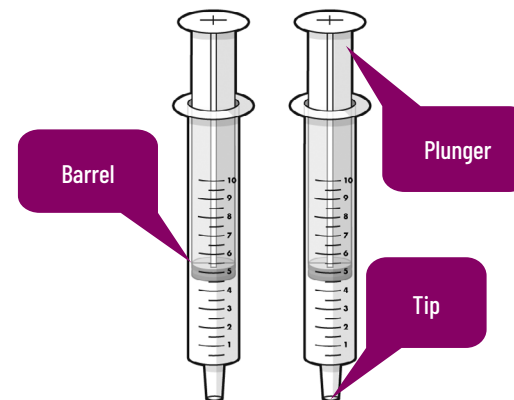
- ☐ 1 dosing cup
- ☐ 1 mixing bottle
- ☐ 2 oral syringes
- ☐ 2 spare seals

Other items needed (not provided)

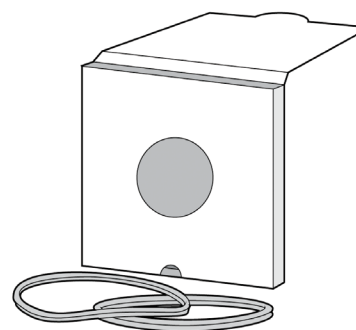
- ☐ 1 container with room temperature water



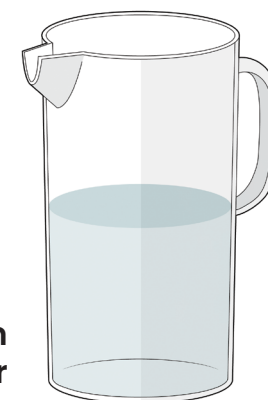
2 oral syringes



2 spare seals



Container with room temperature water



IMPORTANT INFORMATION

What you need to know before preparing and taking or giving KYGEVVI

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Powder for Oral Solution

2 g/2 g per packet

- You will prepare a **one-day supply** of KYGEVVI solution to be taken in **3 equal doses** throughout the day (about **6 hours** apart).
- KYGEVVI solution should be prepared on the same day as taking or giving your dose.
- KYGEVVI should be prepared and given by adults only.
- Take or give KYGEVVI exactly as prescribed by your healthcare provider.
- **Do not** change your dose without talking with your healthcare provider.
- **Do not** take or give more than the prescribed amount of KYGEVVI solution.
- **Take KYGEVVI solution with food.**
- **Do not** use KYGEVVI packets after the expiration date on the packet or carton.
- Only use the dosing cup, mixing bottle, and oral syringes provided with your ZX2000 administration kit. **Do not** use a household measuring cup or measuring spoon.
- Each ZX2000 administration kit comes with two oral syringes.
- Keep the second oral syringe as a spare.
- Rinse and dry the mixing bottle and dosing cup before first use. Do not use the dosing cup, mixing bottle, or oral syringe if it appears dirty or damaged.
- Each ZX2000 administration kit can be used for 6 months. Contact your healthcare provider when you need a replacement.
- Contact your healthcare provider or pharmacist for a replacement if your mixing bottle, dosing cup, or oral syringe is damaged or if the markings are missing or no longer readable.
- **Do not** use the packets if the tamper evident seal on the carton is broken.
- Mix KYGEVVI powder only with room temperature water. **Do not mix** KYGEVVI powder with any other liquids or foods.
- You may have KYGEVVI solution left over after taking or giving the 3 individual doses. Throw away (discard) any remaining KYGEVVI solution 16 hours after preparing or after taking or giving the 3 doses, whichever comes first.
- If powder spills out of a packet before use, **do not** use the packet. **Throw it away** and use a new KYGEVVI packet.
- If KYGEVVI solution comes in contact with your eyes or skin, rinse under cold water and contact your healthcare provider.
- KYGEVVI solution is compatible with most feeding tubes. Follow the steps in this Instructions for Use booklet to prepare your one-day supply of KYGEVVI. Then follow the feeding tube manufacturer instructions on how to give KYGEVVI using a feeding tube.
 - Draw up the prescribed individual dose using a syringe compatible with the feeding tube. Give the KYGEVVI solution right away through the feeding tube.
 - Flush any remaining KYGEVVI solution in the syringe or feeding tube until no solution is left. Ask your healthcare provider how much water to use.
 - Throw away (discard) any remaining KYGEVVI solution 16 hours after preparing or after taking or giving the 3 doses, whichever comes first.
- Store KYGEVVI packets at room temperature between 68°F to 77°F (20°C to 25°C).
- Store the prepared KYGEVVI solution at room temperature between 68°F to 77°F (20°C to 25°C) **or** in the refrigerator between 36°F to 46°F (2°C to 8°C).
- Make sure the dosing cup is screwed on tightly to the mixing bottle when storing your KYGEVVI solution to avoid spills or leaks.
- When storing or carrying the KYGEVVI solution, keep the mixing bottle upright.
- Use the KYGEVVI solution within 16 hours of preparing. **Throw away (discard) any remaining solution 16 hours after preparing or after taking or giving the 3 doses, whichever comes first.**
- **Keep KYGEVVI and all medicines out of the reach of children.**

PREPARING YOUR ONE-DAY SUPPLY OF KYGEVVI

GET SUPPLIES READY

STEP 1

- a) Wash your hands well with soap and water.
- b) Place the mixing bottle, dosing cup, and oral syringe (if you need one to measure your individual dose) on a clean, well-lit flat work surface. If the dosing cup is attached to the mixing bottle, unscrew it from the mixing bottle and set it down (see **Figure B**).
- c) Make sure “KYGEVVI” appears on the carton.
- d) Check the expiration date printed on the bottom of the KYGEVVI carton. When opening the KYGEVVI carton for the first time, break the tamper evident seal.
- e) Remove the prescribed number of KYGEVVI packets needed for your one-day supply of KYGEVVI out of the carton. Your one-day supply of KYGEVVI will be divided into 3 individual doses.
- f) **Do not** open the KYGEVVI packets until Step 2.

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Figure B



Note: The mixing bottle has markings on the front of the bottle in 40 mL increments. Each increment is equal to one packet of medicine.

The dosing cup has markings on the front and back of the cup in 10 mL increments. One side measures from 50 mL to 220 mL and the other side measures from 55 mL to 225 mL. Using both sides, you can measure KYGEVVI solution in 5 mL increments (from 50 mL to 225 mL).

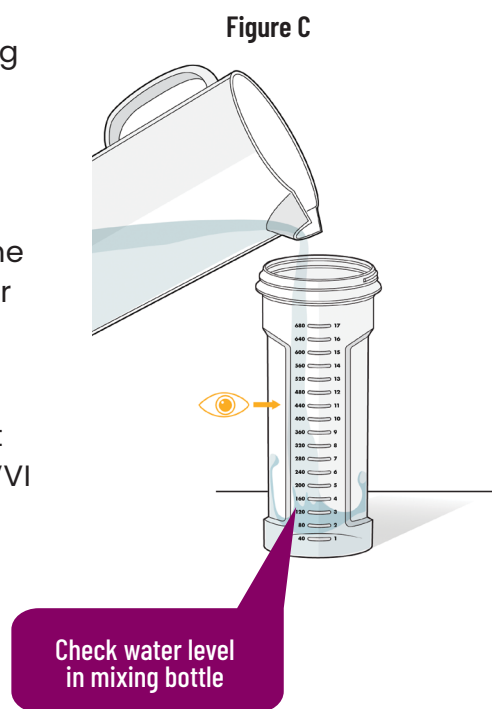
MEASURE WATER AND ADD POWDER FROM PACKETS

STEP 2

- a) On a flat surface, pour the prescribed amount of room temperature water into the mixing bottle (see **Figure C**).
- **Do not** pour the water into the dosing cup.
 - **Important: Do not** add the powder from the packets to the mixing bottle before this step.

- b) Check to make sure the mixing bottle is filled with water up to the marking that matches the amount prescribed by your healthcare provider. The marking should also match the number of packets needed for your one-day supply (see **Figure C**).

- c) Check you have counted out the correct number of KYGEVVI packets for your one-day supply, as shown on your prescription.



- d) Tap the packet on a hard surface to settle the powder to the bottom of the packet away from the dotted line (see **Figure D**).

- e) Carefully fold and tear or cut along the dotted line (see **Figure E**). If you spill any powder, **do not** use it. Throw the packet away and use a new packet.

- f) Empty the entire packet contents into the mixing bottle. Be careful not to drop the packet into the mixing bottle (see **Figure F**).

- g) Pour only 1 packet into the mixing bottle at a time. Repeat **Steps 2d to 2f**, for each packet until you have poured the prescribed number of packets for your one-day supply.

Figure D

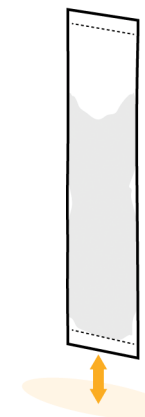


Figure E

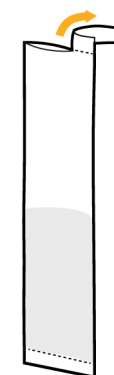
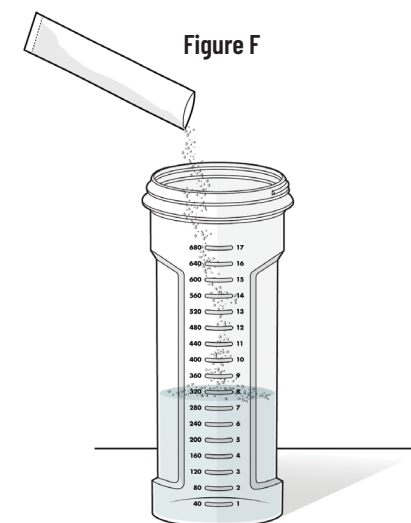


Figure F

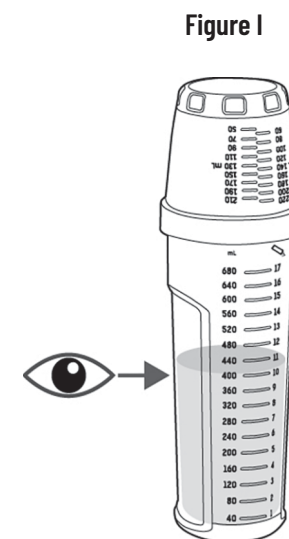
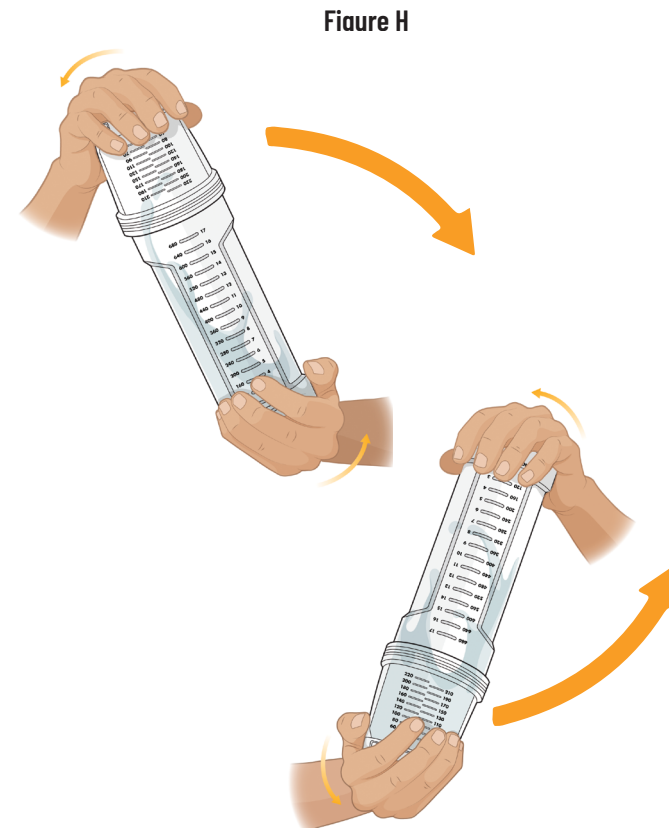
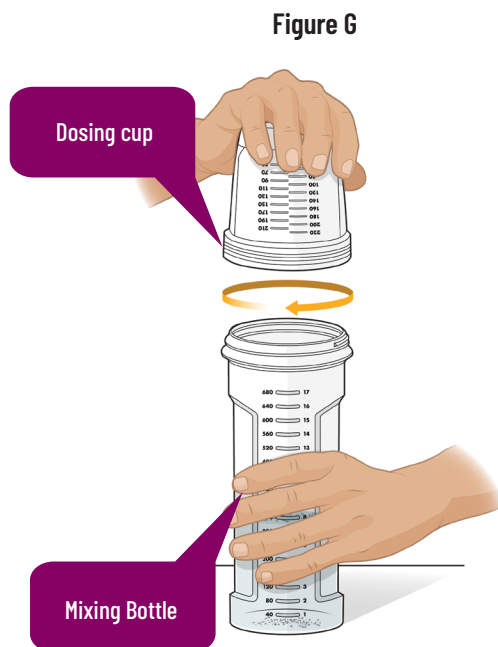


MIX AND INSPECT MEDICINE

STEP 3

- Screw the dosing cup tightly onto the mixing bottle (see **Figure G**).
- Place one hand at the end of the mixing bottle and the other hand at the end of the dosing cup. Gently turn the mixing bottle upside down and back at least **20 times**. (see **Figure H**).
- Check the solution. If you see any powder, keep turning the mixing bottle upside down and back until the powder dissolves (see **Figure I**).
 - The mixed solution may be cloudy and have some remaining powder at the bottom or top. This is normal.

You have now prepared your one-day supply of KYGEVVI solution for **3 individual doses**. Take KYGEVVI solution with food.



DOSING METHODS

HOW TO MEASURE YOUR INDIVIDUAL DOSE

There are 2 different methods to take or give KYGEVVI solution depending on your individual dose. Use the table below to identify which steps you should follow:

$\geq 50\text{ mL}$

Individual doses 50 mL or **more**



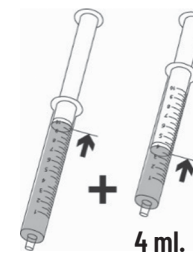
Example 100 mL

Use dosing cup

Follow **Step 4** on Pages 20–21

$< 50\text{ mL}$

Individual doses **less** than 50 mL
(dosing cup used for dose preparation only)



Example 14 mL

Use oral syringe

Follow **Step 5** on Pages 22–25



INDIVIDUAL DOSES 50 ML OR MORE

MEASURE AND TAKE OR GIVE YOUR INDIVIDUAL DOSE

STEP 4: DOSING CUP

- Check to make sure the dosing cup is screwed on tightly to the mixing bottle.
- Mix the already prepared solution by slowly turning the mixing bottle upside down and back at least 3 times.
- Unscrew the dosing cup from the mixing bottle and place it on a flat surface.



Figure J

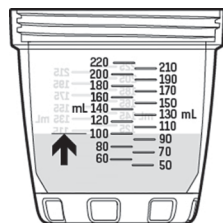


Figure K

- Pour KYGEVVI oral solution from the mixing bottle into the dosing cup until it reaches the marking on the dosing cup for your prescribed individual dose (see **Figure J**).
- ✦ **Note:** Your dose may be different than the dose shown in **Figure J**.
- Drink or give the entire solution from the dosing cup (see **Figure K**).
 - When it is time for the **second or third individual dose**, repeat **Steps 4a to 4e** for each individual dose.
 - After the **first or second individual dose**, go to **Step 6** (p.26) for instructions on how to clean your supplies and store KYGEVVI solution. After the **third individual dose**, go to **Step 7** (p.28) for instructions on how to clean your supplies and dispose of unused KYGEVVI solution.



INDIVIDUAL DOSES LESS THAN 50 ML

MEASURE AND TAKE OR GIVE
YOUR INDIVIDUAL DOSE

STEP 5: ORAL SYRINGE

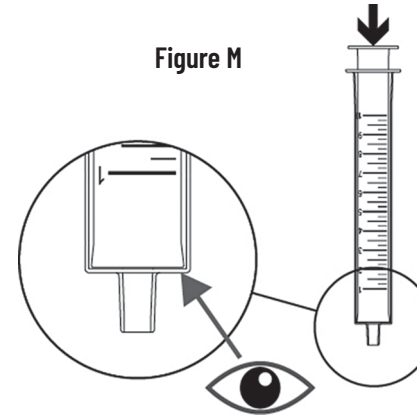
If your individual dose is more than 10 mL, you will need to use the same oral syringe more than 1 time.

- a) Check to make sure the dosing cup is screwed on tightly to the mixing bottle.
- b) **Mix the already prepared solution by slowly turning the mixing bottle upside down and back at least 3 times.**
- c) Unscrew the dosing cup from the mixing bottle and place on a flat surface.

Figure L



Figure M



- d) Pour slightly more than the amount of KYGEVVI solution needed for your prescribed individual dose into the dosing cup (see **Figure L**).
- e) Push the plunger of the oral syringe all the way down to make sure there is no air in the oral syringe when measuring the dose (see **Figure M**).

f) Place the tip of the oral syringe into the dosing cup with the oral solution. Fill the oral syringe by pulling the plunger back until it reaches the marking on the oral syringe that matches your prescribed individual dose (see **Figure N**). **Note:** If your individual dose is more than 10 mL, you will need to use the same oral syringe more than 1 time. Follow your healthcare provider's instructions on how to measure your dose.



g) Place the tip of the oral syringe into the mouth and point the tip towards the inside of either cheek (see **Figure O**).

- **If you are giving KYGEVVI to a young child, the child must be seated and held in place to prevent the oral solution from going down the wrong way or choking.**

h) Slowly push the plunger all the way down until the oral syringe is empty (see **Figure O**).

- i) If your prescribed individual dose is more than 10 mL, repeat **Steps 5d to 5h** until you take or give the full individual dose.
- j) Pour back any remaining solution from the dosing cup into the mixing bottle.
- k) When it is time for the **second or third individual dose**, repeat **Steps 5a to 5j** for each individual dose.
- l) After the **first or second individual dose**, go to **Step 6** (p.26) for instructions on how to clean your supplies and store KYGEVVI solution. After the **third individual dose**, go to **Step 7** (p.28) for instructions on how to clean your supplies and dispose of unused KYGEVVI solution.

Figure O



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[doxycitine and doxribtimine]

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BETWEEN INDIVIDUAL DOSES

CLEAN UP AFTER FIRST AND SECOND INDIVIDUAL DOSE

STEP 6

After you complete the first or second individual dose:

- a) Rinse the dosing cup with cold water after each use (see **Figure P**).
- b) Dry the dosing cup with a clean, dry towel.
- c) After the dosing cup is dry, screw the dosing cup tightly onto the mixing bottle (see **Figure Q**) and store it at room temperature or in the refrigerator until it is time for the next individual dose.

Figure P

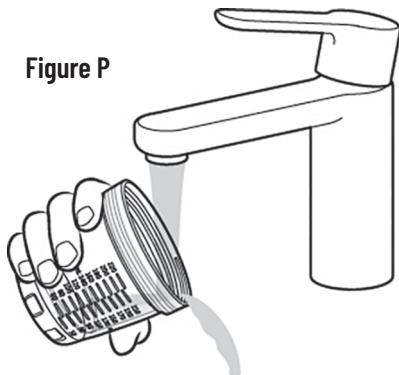
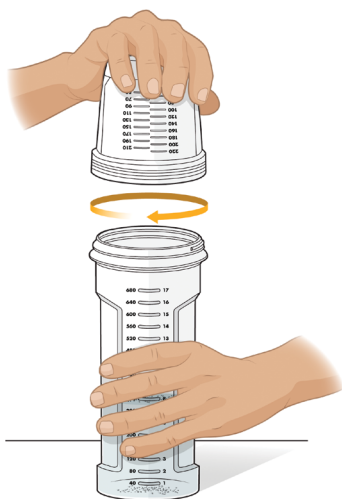


Figure Q

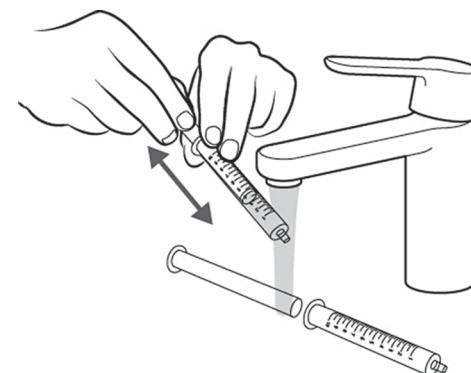


d) If you used the oral syringe, clean it with cold water:

- i. Rinse the oral syringe with cold water by filling the oral syringe with water and pushing it back out (see **Figure R**). Then remove the plunger from the barrel and rinse the plunger and barrel under running tap water until it is clean (see **Figure R**).
- ii. Let the oral syringe barrel and plunger dry in the open air. After the oral syringe barrel and plunger are dry, put the plunger back into the barrel.

Do not wash the dosing cup or oral syringe in the dishwasher.

Figure R



END-OF-DAY CLEANUP

POUR OUT AND CLEAN UP AFTER THIRD INDIVIDUAL DOSE

STEP 7

After you take or give the third individual dose, throw away any remaining KYGEVVI solution in the sink.

Do not save KYGEVVI solution for another day.

- Remove the seal from the dosing cup to thoroughly clean it (see **Figure S**).
- Clean the mixing bottle, dosing cup and seal by hand with soap and warm water. Use a brush to remove any residue left in the mixing bottle or dosing cup (see **Figure T**).
- Dry the mixing bottle, dosing cup and seal with a clean towel. Put the dry seal back into the dosing cup, with **the thin side of the seal** facing the groove.

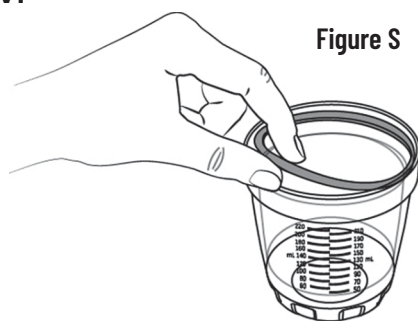
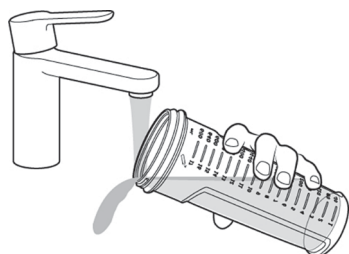


Figure S

Figure T



kygevvi

[doxecitine and doxribtimine]

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d) If you used the oral syringe, clean it with cold water:

- Rinse the oral syringe with cold water by filling the oral syringe with water and pushing it back out (see **Figure U**). Then remove the plunger from the barrel and rinse the plunger and barrel under running tap water until it is clean (see **Figure U**).
- Let the oral syringe barrel and plunger dry in the open air. After the oral syringe barrel and plunger are dry, put the plunger back into the barrel.

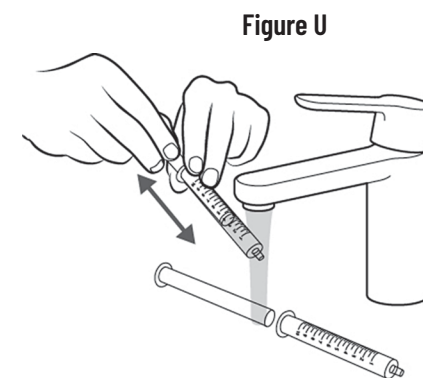


Figure U

Do not wash the mixing bottle, dosing cup, seal, or oral syringe in the dishwasher.

e) Store all supplies in a clean, dry area out of the reach of children for the next day's use.

DOSING CUP MAINTENANCE

REPLACING THE SEAL IF MISPLACED OR DAMAGED

Changing the dosing cup seal

If you misplace the dosing cup seal or you notice leakage when the dosing cup is screwed on tightly to the mixing bottle, change the seal using one of the two spare seals provided in the ZX2000 administration kit. Follow these steps to replace the seal:

- a) Remove the seal in the dosing cup (see **Figure V**). Skip this step if you misplaced the seal.
- b) Wash the dosing cup groove with warm water (see **Figure W**).
- c) Get a new seal from the spare seal box (see **Figure X**).
- d) Insert the seal into the groove of the dosing cup with the **thin side of the seal** facing the groove (see **Figure Y**).

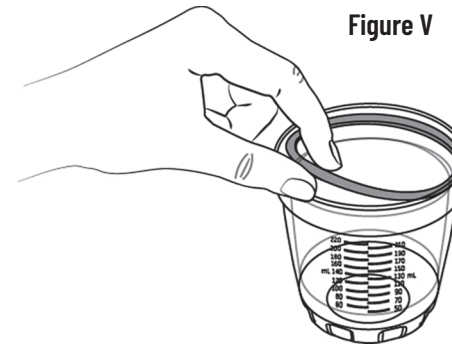


Figure V

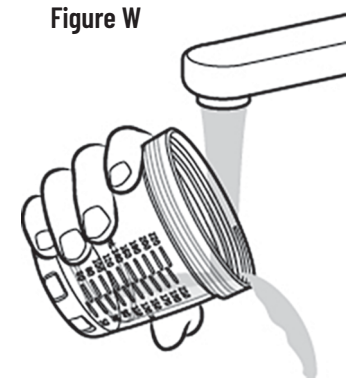


Figure W

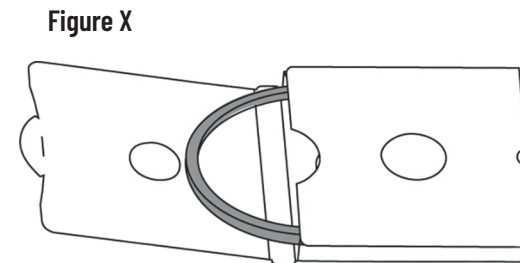


Figure X

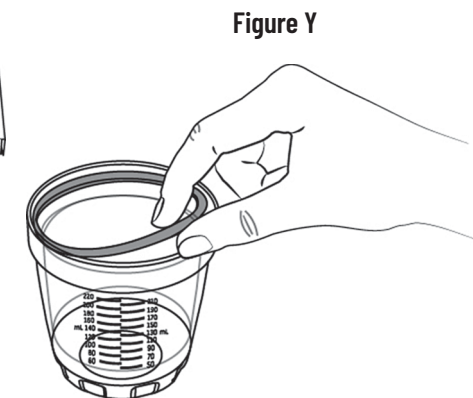


Figure Y

CONTACT INFORMATION

Contact your healthcare provider or pharmacist if you have any questions about this Instructions for Use.

You may also contact UCBCares® at 1-844-599-CARE (2273) for assistance.

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[illegible][illegible]

JOIN ONWARD®

There are three ways to get started.

ASK your doctor to sign you up.

OR

VISIT **ucbONWARD.com/KYGEVVI**

to download an ONWARD start form and take it to your doctor.

OR

CALL 1-844-ONWARD-1

(844-669-2731)

Monday through Friday,

8 AM – 8 PM ET.



Scan the code
for more KYGEVVI
resources and
treatment support.

kygevvi™

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