

## INSTRUCTIONS FOR ORAL DOSING

Shot Number	Visual	Caption	VO	Time Stamp / Approx. duration
1	<p>RAVICTI logo animation (5 seconds)</p> <ul style="list-style-type: none"> <li>• Fade up from black to white screen</li> <li>• Fade in type</li> <li>• Ribbon mark twists in</li> </ul> <p>Title fades in.</p> <p>RAVICTI logo transition (2 seconds)</p> <ul style="list-style-type: none"> <li>• Quick zoom in on logo</li> <li>• The top of the letter “I” in RAVICTI floods the screen with purple</li> </ul>	<p>Directions for taking RAVICTI® (glycerol phenylbutyrate) Oral Liquid by mouth</p>	<p>Welcome to the RAVICTI® (glycerol phenylbutyrate) Oral Liquid instructional dosing video.</p> <p>In this video you will see step-by-step instructions for the oral administration of RAVICTI for you or your child.</p>	00:00/ :12
2	<p>White text scrolls from bottom to top on purple background.</p>	<p><b>Use for RAVICTI® (glycerol phenylbutyrate) Oral Liquid</b></p> <p>RAVICTI is a prescription medicine used for long-term management of high blood levels of ammonia (hyperammonemia) caused by a condition called a urea cycle disorder (UCD). RAVICTI should be used if the UCD cannot be managed with a low-protein diet and dietary supplements alone. RAVICTI must be used along with a low-protein diet and in some cases, dietary supplements.</p> <p>RAVICTI is not used to treat extremely high levels of ammonia in the blood (hyperammonemic crisis) in people with UCDs.</p>	<p><b>Use for RAVICTI® (glycerol phenylbutyrate) Oral Liquid</b></p> <p>RAVICTI is a prescription medicine used for long-term management of high blood levels of ammonia (hyperammonemia) caused by a condition called a urea cycle disorder (UCD). RAVICTI should be used if the UCD cannot be managed with a low-protein diet and dietary supplements alone. RAVICTI must be used along with a low-protein diet and in some cases, dietary supplements.</p> <p>RAVICTI is not used to treat extremely high levels of ammonia in the blood (hyperammonemic crisis) in people with UCDs.</p>	:12/ 1:06

	<p>When “Before taking RAVICTI...” is spoken: Fade in knockout version of logo.</p> <p>RAVICTI logo transition (2 seconds)</p> <ul style="list-style-type: none"> <li>• Quick zoom in on logo</li> </ul> <p>The top of the letter “I” in RAVICTI floods the screen with white.</p>	<p>It is not known if RAVICTI is safe and effective for the treatment of <i>N</i>-acetylglutamate synthase (NAGS) deficiency.</p> <p>Please listen to the RAVICTI Important Safety Information at the end of this video. For additional important safety information, see the Medication Guide at RAVICTI.com and discuss with your doctor.</p>	<p>It is not known if RAVICTI is safe and effective for the treatment of <i>N</i>-acetylglutamate synthase (NAGS) deficiency.</p> <p>Please listen to the RAVICTI Important Safety Information at the end of this video. For additional important safety information, see the Medication Guide at RAVICTI.com and discuss with your doctor.</p>	
3	<p><b>WIDE</b>—View of the table, setting up the scene. Starts out of focus and slowly sharpens to introduce the patient and caregiver.</p>	<p>Always take RAVICTI® (glycerol phenylbutyrate) Oral Liquid exactly as directed by your doctor.</p> <p>Do not mix RAVICTI with formula or any other liquids.</p>	<p>This video is intended to complement the instructions you have already received from your doctor. Always take RAVICTI exactly as directed by your doctor.</p> <p>Do not mix RAVICTI with formula or any other liquids.</p> <p>Now, let’s get started.</p>	1:06/ :20
4	<p><b>MD</b>—Cut to a shot of the 3 items lined up on the table.</p> <p>Add labels for each item underneath that item as it is mentioned by the narrator.</p>	<p>Bottle of RAVICTI® (glycerol phenylbutyrate) Oral Liquid Vial shown is for demonstration use only.</p> <p>Blue AdaptaCap® Bottle Adapter</p> <p>Oral dosing syringe</p>	<p>Your RAVICTI shipment includes a 25-mL bottle of RAVICTI, a blue AdaptaCap® Bottle Adapter, and an oral dosing syringe.</p> <p>When dosing RAVICTI, be sure to use the oral dosing syringe provided with the product.</p>	1:20/ :32

5	<b>MD—Woman from the waist up.</b> View caregiver’s hands as she picks up the RAVICTI bottle and twists off the cap.	Remove the cap.	First, remove the childproof cap on the RAVICTI bottle. To do that, push down on the cap while twisting it to the left.	1:33/ :40
6	<b>CU—Hands, bottle, AdaptaCap.</b> Caregiver’s hands as she twists on the AdaptaCap	Attach blue AdaptaCap® Bottle Adapter.	Then, attach the blue AdaptaCap® Bottle Adapter onto the bottle. Put it on top of the bottle and twist it to the right.  Make sure it is secured tightly on the bottle.	1:40/ :48
7	<b>CU—Hands, bottle, syringe.</b> Caregiver’s hands as she inserts the syringe into the AdaptaCap	Place tip of the oral dosing syringe into AdaptaCap® Bottle Adapter.	While holding the RAVICTI bottle securely, place the tip of the oral dosing syringe into the AdaptaCap® Bottle Adapter.	1:48/ :54
8	<b>MD to CU—Hands, bottle, syringe.</b> Caregiver’s hands as she turns bottle upside down	Turn bottle upside down.	Turn the bottle upside down with the oral dosing syringe still inserted.  Now you’re ready to fill the oral dosing syringe with RAVICTI.	1:55/ :01
9	<b>CU to MD—Hands, bottle, syringe.</b> Camera pulls back to show upside down bottle, syringe, and hands as plunger is pulled back to fill syringe with liquid.  Then, the caregiver turns the bottle upright.	Pull plunger back slowly.  Note: Individual dosage amounts may vary.  Bubble  1 2 3 4  Turn bottle upright.	Get the medicine into the oral dosing syringe by pulling the plunger back slowly. Pulling slowly will help avoid large air bubbles from forming in the medicine.  Pull the plunger back until the first line of the black rubber stopper is even with the dosing mark. The dosing marks are on the barrel of the oral dosing syringe. Match the dosing mark with the amount of medicine prescribed by your doctor.  Then, turn the bottle upright again.	2:02 :27

10	<p><b>ECU—Syringe.</b> Syringe showing air bubbles</p>	<p>Check for large air bubbles.</p> <p>If you take too much RAVICTI, call your doctor or go to the nearest hospital emergency room right away.</p> <p>You should consult your doctor if you have missed a dose of RAVICTI.</p> <p>Do not mix RAVICTI with formula or any other liquids.</p>	<p>If there are large air bubbles, push the medicine back into the bottle. Do this slowly.</p>	2:27/ :33
11	<p><b>MD to CU—Hands, bottle, syringe.</b> Caregiver pushes medicine back into the bottle.</p> <p>Turns bottle over</p> <p>Draws medicine into the syringe without bubbles</p>		Then draw up the medicine again.	2:33/ :35
12	<p><b>MD to CU— Woman’s hands, patient’s head, syringe.</b> Shot of patient opening his mouth and caregiver pressing on the syringe plunger to administer liquid. Child closes his mouth and swallows with a pleasant expression on his face.</p>	Squirt medicine into mouth.	<p>Once you have your oral dosing syringe ready, place it into the mouth. Push the plunger to squirt the medicine directly into the mouth.</p> <p>Make sure all of the medicine is pushed out of the oral dosing syringe and swallowed.</p> <p>Put the cap on the oral dosing syringe.</p>	2:35/ :49

13	<p><b>MD to WIDE— Patient upper torso, tabletop.</b> Patient having a large sip of water with a basic salad in view that has tomato, lettuce, carrot, and cucumber with no dressing.</p>	<p>Drink liquid or eat food.</p>	<p>Drink liquid or eat some food right after taking RAVICTI.</p> <p>This helps ensure that all of the medicine is swallowed.</p>	<p>2:49/ :55</p>
14	<p><b>CU—Hand, bottle with AdaptaCap.</b> Caregiver’s hands as she closes the blue tab on the AdaptaCap</p>	<p>Close tab on AdaptaCap® Bottle Adapter.</p>	<p>Close the tab on the AdaptaCap® Bottle Adapter.</p> <p>Make sure it’s closed tightly.</p>	<p>2:56/ :00</p>
15	<p><b>MD—shot of the 3 items lined up on the table.</b></p> <p>Camera pans out to shot of caregiver and a patient talking.</p> <p>Fades to purple background with on screen text.</p>	<p>If you take too much RAVICTI, call your doctor or go to the nearest hospital emergency room right away.</p> <p>You should consult your physician if you have missed a dose of RAVICTI.</p> <p>Do not mix RAVICTI with formula or any other liquids.</p>	<p>You can use 1 oral dosing syringe and 1 blue AdaptaCap® Bottle Adapter for all doses each day. You can also choose to use a new oral dosing syringe with each dose. Do not rinse the AdaptaCap® Bottle Adapter or oral dosing syringe between doses.</p> <p>After the last dose of the day, dispose of the oral dosing syringe.</p> <p>If you take too much RAVICTI, call your doctor or go to the nearest hospital emergency room right away.</p> <p>You should consult your physician if you have missed a dose of RAVICTI.</p> <p>Do not mix RAVICTI with formula or any other liquids.</p>	<p>3:00/ :32</p>

<p>17</p>	<p>Important Safety Information scrolls from bottom to top.</p> <p>Black type on white background.</p>	<p><b>IMPORTANT SAFETY INFORMATION</b></p> <p><b><u>What is the most important safety information I should know about RAVICTI?</u></b></p> <p><b>RAVICTI may cause serious side effects, including:</b>  <b>Nervous system side effects (Neurotoxicity).</b> The breakdown of RAVICTI produces the byproduct phenylacetate (PAA), which may cause nervous system side effects.  <b>Call your doctor or get medical help right away if you have any of these symptoms while taking RAVICTI:</b></p> <ul style="list-style-type: none"> <li>• sleepiness</li> <li>• lightheadedness</li> <li>• change in taste</li> <li>• problems with hearing</li> <li>• confusion</li> <li>• problems with memory</li> <li>• worsening of numbness, tingling, or burning in your hands or feet</li> <li>• headache</li> <li>• feeling very tired (fatigue)</li> <li>• nausea</li> <li>• vomiting</li> </ul> <p><b><u>Who should not take RAVICTI?</u></b></p> <p><b>Do not take RAVICTI if you are allergic to phenylbutyrate.</b> Call your doctor or go to the nearest hospital emergency room if you experience any of the following symptoms of an allergic reaction while taking RAVICTI:</p> <ul style="list-style-type: none"> <li>• wheezing</li> <li>• shortness of breath</li> <li>• cough</li> <li>• low blood pressure</li> <li>• flushing</li> <li>• nausea</li> <li>• skin rash</li> </ul> <p><b><u>What should I tell my doctor before taking RAVICTI?</u></b></p>	<p><b>IMPORTANT SAFETY INFORMATION</b></p> <p><b><u>What is the most important safety information I should know about RAVICTI?</u></b></p> <p><b>RAVICTI may cause serious side effects, including:</b>  <b>Nervous system side effects (Neurotoxicity).</b> The breakdown of RAVICTI produces the byproduct phenylacetate (PAA), which may cause nervous system side effects.  <b>Call your doctor or get medical help right away if you have any of these symptoms while taking RAVICTI:</b></p> <ul style="list-style-type: none"> <li>• sleepiness</li> <li>• lightheadedness</li> <li>• change in taste</li> <li>• problems with hearing</li> <li>• confusion</li> <li>• problems with memory</li> <li>• worsening of numbness, tingling, or burning in your hands or feet</li> <li>• headache</li> <li>• feeling very tired (fatigue)</li> <li>• nausea</li> <li>• vomiting</li> </ul> <p><b><u>Who should not take RAVICTI?</u></b></p> <p><b>Do not take RAVICTI if you are allergic to phenylbutyrate.</b> Call your doctor or go to the nearest hospital emergency room if you experience any of the following symptoms of an allergic reaction while taking RAVICTI:</p> <ul style="list-style-type: none"> <li>• wheezing</li> <li>• shortness of breath</li> <li>• cough</li> <li>• low blood pressure</li> <li>• flushing</li> <li>• nausea</li> <li>• skin rash</li> </ul> <p><b><u>What should I tell my doctor before taking RAVICTI?</u></b></p>	<p>3:33/ 8:20</p>
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18	RAVICTI logo animation <ul style="list-style-type: none"> <li>• Fade in type</li> <li>• Ribbon mark</li> </ul> Website URL fades in.	RAVICTI and the HORIZON logo are trademarks owned by or licensed to Horizon. All other referenced trademarks are the property of their respective owners. © 2020 Horizon Therapeutics plc P-RVT-00473 05/20		8:21/ :26
19	Fade to white. Fade in Horizon logo. Fade to black.			8:26/ :31