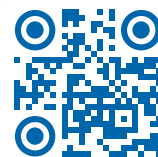


NULIBRY[®] (fosdenopterin) Dosing, Preparation, and Administration Overview



Detailed instructions for daily preparation and administration of NULIBRY can be viewed at nulibry.com/hcp/preparing-administering-nulibry



Scan here.

NULIBRY is indicated to reduce the risk of mortality in patients with molybdenum cofactor deficiency (MoCD) Type A.

Start NULIBRY if the patient has a diagnosis or presumptive diagnosis of MoCD Type A.

In patients with a presumptive diagnosis of MoCD Type A, confirm the diagnosis of MoCD Type A immediately after initiation of NULIBRY treatment. In such patients, discontinue NULIBRY if the MoCD Type A diagnosis is not confirmed by genetic testing.

Please see **additional Important Safety Information and full Prescribing Information including Instructions for Use** at <https://bit.ly/3tOMDHI>.

Indication and Important Safety Information



INDICATION

NULIBRY is indicated to reduce the risk of mortality in patients with molybdenum cofactor deficiency (MoCD) Type A.

IMPORTANT SAFETY INFORMATION

WARNINGS AND PRECAUTIONS

Potential for Photosensitivity

NULIBRY can make the patient oversensitive to sunlight. NULIBRY-treated patients or their caregivers are advised to avoid or minimize patient exposure to sunlight and artificial UV light and adopt precautionary measures when exposed to the sun, including wearing protective clothing and sunglasses, and use broad-spectrum sunscreen with high SPF in patients 6 months of age and older. If photosensitivity occurs, caregivers/patients are advised to seek medical attention immediately and consider a dermatological evaluation.

SIDE EFFECTS

The most common side effects in NULIBRY-treated patients were infusion catheter-related complications, pyrexia (fever), viral infection, pneumonia, otitis media (ear infection), vomiting, cough/sneezing, viral upper respiratory infection (common cold/flu-like infection), gastroenteritis (stomach flu-like symptoms), diarrhea, and bacteremia (bacteria in the blood). Side effects for rcPMP-treated patients were similar to the NULIBRY-treated patients.

PATIENT COUNSELING INFORMATION

Please read the FDA-approved NULIBRY Prescribing Information and Instructions for Use and follow the instructions on how to prepare and administer NULIBRY.

NULIBRY has a potential for photosensitivity; see Warnings and Precautions. Seek medical attention immediately if the patient develops a rash or if they notice symptoms of photosensitivity reactions (redness, burning sensation of the skin, blisters).

You are encouraged to report negative side effects of prescription drugs to the FDA. Visit www.fda.gov/medwatch, or call [1-800-FDA-1088](tel:1-800-FDA-1088).

TO LEARN MORE ABOUT NULIBRY, VISIT NULIBRY.COM/HCP

Please see additional Important Safety Information and full Prescribing Information including Instructions for Use at <https://bit.ly/3tOMDHI>.

Dosing for NULIBRY® (fosdenopterin) for injection



NULIBRY is administered as a daily IV infusion, after reconstitution. It must be reconstituted with Sterile Water for Injection USP. Dosing is individualized based on the patient's weight and may be administered at home by the child's caregiver if you deem it appropriate. NULIBRY comes as a lyophilized powder or cake in vials that must stay frozen until ready to be used (between -13°F and 14°F/-25°C and -10°C); then it needs to be brought to room temperature (between 59°F and 77°F/15°C and 25°C) when reconstituted. Reconstituted NULIBRY may be stored at room temperature 59°F to 77°F (15°C to 25°C) or refrigerated 36°F to 46°F (2°C to 8°C) for up to 4 hours including infusion time. Do not shake the NULIBRY vial, do not expose it to heat, and do not freeze after reconstitution.

NULIBRY is delivered by a specialty pharmacy. Because NULIBRY requires cold chain storage, a medical-grade freezer is required.

Start NULIBRY if the patient has a diagnosis or presumptive diagnosis of MoCD Type A. Discontinue NULIBRY if the MoCD Type A diagnosis is not confirmed by genetic testing.

See table below for weight-based dosing of patients <1 year of age.

Recommended initial dosage and titration schedule of NULIBRY for patients less than one year of age by gestational age		
Titration Schedule	Preterm neonates (gestational age <37 weeks)	Term neonates (gestational age ≥37 weeks)
Initial Dosage	0.4 mg/kg once daily	0.55 mg/kg once daily
Dosage at Month 1	0.7 mg/kg once daily	0.75 mg/kg once daily
Dosage at Month 3	0.9 mg/kg once daily	0.9 mg/kg once daily

For patients one year of age or older, the recommended dosage of NULIBRY is 0.9 mg/kg (based on actual body weight) administered as an intravenous infusion once daily.

Please see additional Important Safety Information and full Prescribing Information including Instructions for Use at <https://bit.ly/3tOMDHI>.

Handling of NULIBRY[®] and Supplies for Administration



Each NULIBRY delivery comes with:

- A quantity of NULIBRY vials based on the prescription you've written. NULIBRY comes in clear glass vials containing 9.5 mg of fosdenopterin
- The full Prescribing Information and the Instructions for Use for NULIBRY
- Every Welcome Kit includes a treatment log for the caregiver to fill out and bring to the appointments they will have with you

HOW IS NULIBRY FOR INJECTION STORED?

Frozen vials:

- Store NULIBRY in the freezer between -13°F to 14°F (-25°C to -10°C)
- Keep each NULIBRY vial in the original carton to protect from light until you are ready to use it

Vials of NULIBRY after mixing:

- Store vials of NULIBRY that have been mixed at room temperature 59°F to 77°F (15°C to 25°C) or refrigerated 36°F to 46°F (2°C to 8°C) for up to 4 hours, including infusion time

Vials of NULIBRY are for 1 time use only. Throw the vial away after use, even if there is medicine left in the vial. **Do not** save for later use.

Remind caregivers that if the freezer is not working properly to call [1-888-251-2800](tel:1-888-251-2800), Monday-Friday, 8 AM-8 PM ET. If it is after-hours, they can call and follow the prompt

Please see additional Important Safety Information and full Prescribing Information including Instructions for Use at <https://bit.ly/3tOMDHI>.

Handling of NULIBRY® and Supplies for Administration (continued)



Additional supplies needed to prepare and administer NULIBRY (please see Instructions for Use for NULIBRY for details):

- 1 vial of Sterile Water for Injection for each vial of NULIBRY needed for 1 dose
- 1 sterile 5 mL syringe for each vial of NULIBRY needed for 1 dose to mix (dissolve) NULIBRY with Sterile Water for Injection
- 1 sterile syringe large enough to hold the total amount of NULIBRY needed for one dose
- Sterile needles (18-gauge recommended)
- Alcohol wipes
- 1 sharps disposal container for throwing away used needles and syringes
- Gloves, if you require the caregiver to wear them
- 1 intravenous administration set with non-DEHP tubing and a 0.2 micron filter
- 1 infusion pump used to give the dose of NULIBRY per your instructions
- Other materials, if recommended by you for the caregiver to properly care for the child's intravenous access line or port before and after giving a dose of NULIBRY

Remind caregivers that if the freezer is not working properly to call [1-888-251-2800](tel:1-888-251-2800), Monday-Friday, 8 AM-8 PM ET. If it is after-hours, they can call and follow the prompt.

Please see additional Important Safety Information and full Prescribing Information including Instructions for Use at <https://bit.ly/3tOMDHI>.

Preparation and Administration

STEP 1 GATHER SUPPLIES

Use a clean, flat work surface.

Remove from the freezer the NULIBRY vials needed to prepare your child's prescribed dose. You may need more than 1 vial to prepare the total amount needed for 1 dose. Allow the NULIBRY vials to reach room temperature. This can be done by rolling each vial gently between your hands for 3 to 5 minutes as shown, or by allowing the vials to sit at room temperature for about 30 minutes (Figure A).

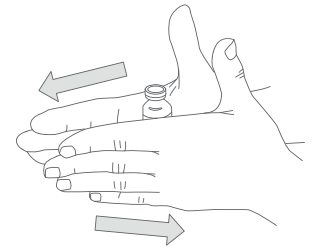
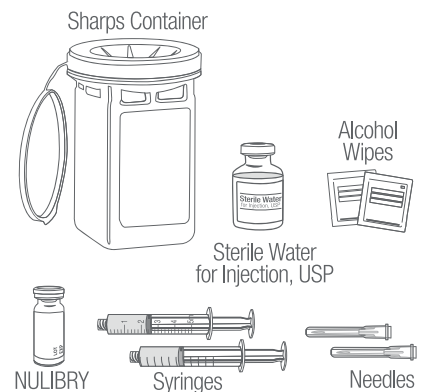


Figure A

Gather supplies needed to prepare and administer a dose of NULIBRY:

- 1 vial of Sterile Water for Injection for each vial of NULIBRY needed for 1 dose
 - Check the expiration date on the vial. **Do not** use the vial if the expiration date has passed
 - **Do not** use the vial if the flip-off seal on the vial is broken or missing
- 1 sterile 5 mL syringe for each vial of NULIBRY needed for 1 dose to mix (dissolve) NULIBRY with Sterile Water for Injection
- 1 sterile syringe large enough to hold the total amount of NULIBRY needed for one dose. Your healthcare provider should tell you what size and type of syringe to use
- sterile needles (18-gauge recommended)
- alcohol wipes
- 1 sharps disposal container for throwing away used needles and syringes. See **“How should I throw away (dispose of) used needles, and syringes?”**
- gloves, if your healthcare provider instructs you to wear gloves when preparing and giving NULIBRY
- 1 intravenous administration set with non-DEHP tubing and a 0.2 micron filter
- 1 infusion pump used to give the dose of NULIBRY as instructed by your healthcare provider
- other materials, if recommended by your healthcare provider to properly care for your child's intravenous access line or port before and after giving a dose of NULIBRY



Preparation and Administration (continued)

STEP 2 WASH YOUR HANDS

- Wash your hands well with soap and water. Use a clean towel to dry your hands or allow them to air dry
- If you have been told to wear gloves to prepare and give NULIBRY, put them on now

STEP 3 PREPARE THE VIALS

Remove the flip-off cap from each vial of Sterile Water for Injection needed to prepare 1 dose (Figure B).

Clean the rubber stopper of each vial with an alcohol wipe and allow to air dry. **Do not** blow on the stopper to dry it faster (Figure C).

Note: If you touch the vial stopper, you will need to clean it again with an alcohol wipe.

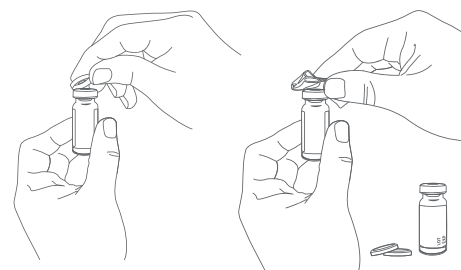


Figure B

Figure C

STEP 4 PREPARE THE SYRINGE USED FOR DRAWING UP STERILE WATER FOR INJECTION

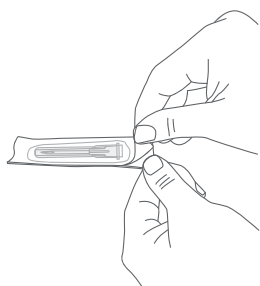


Figure D

Open the wrapper that contains 1 needle.

Do not remove the needle cover yet (Figure D).

Open the wrapper that contains a 5 mL syringe. Attach the needle to the syringe tip with a screw action in the direction of the arrow as shown. Your needle and syringe may look different than shown (Figure E).

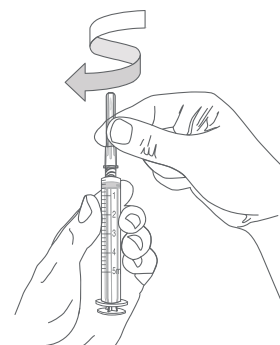


Figure E

Preparation and Administration (continued)

STEP 5 FILL SYRINGE WITH STERILE WATER FOR INJECTION

Remove the needle cover by pulling the cover straight off. **Do not** touch the needle or let the needle touch any surface (Figure F).

Hold the syringe with one hand. Use your other hand to pull back the plunger of the syringe until the top of the plunger reaches the 5 mL line on the syringe (Figure G).

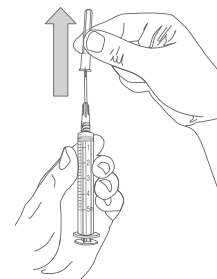


Figure F

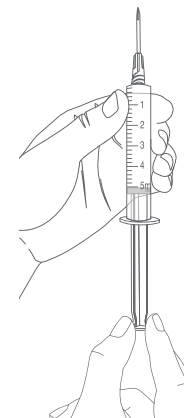


Figure G

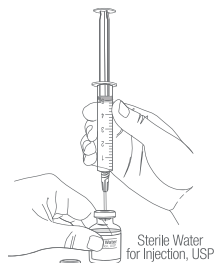


Figure H

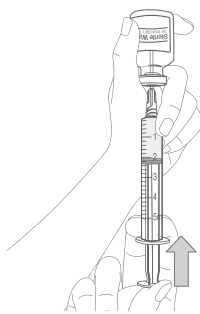


Figure I

Hold the vial of Sterile Water for Injection firmly on your work surface and insert the needle into the center of the vial stopper (Figure H).

Slowly turn the vial upside down. Check that the tip of the needle is not in the solution. Then, push up on the plunger to push all of the air from the syringe into the vial (Figure I).

Next, move the needle so that the tip is in the solution. Slowly pull down on the plunger of the syringe to fill the syringe with 5 mL of Sterile Water for Injection (Figure J).

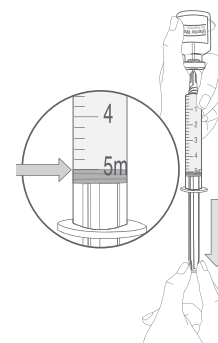


Figure J

Preparation and Administration (continued)

Nulibry[®]
(fosdenopterin)
for injection

STEP 5 FILL SYRINGE WITH STERILE WATER FOR INJECTION (CONTINUED)

Tap the syringe with your fingers until any air bubbles rise to the top of the syringe then gently push up on the plunger to push the air out of the syringe (Figures K & L).

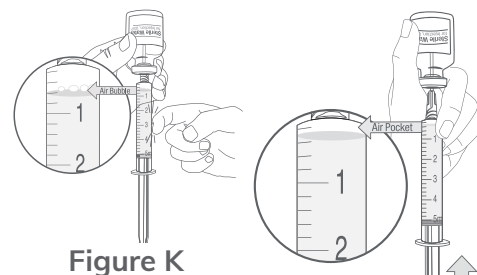


Figure K

Figure L

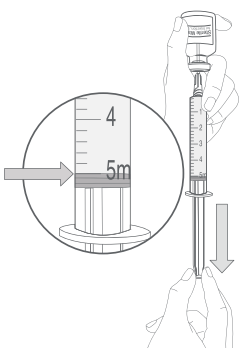


Figure M

After removing the air bubbles, check the syringe to be sure that 5 mL of solution is in the syringe before removing the needle from the vial (Figure M).

STEP 6 MIX (DISSOLVE) NULIBRY

Remove the flip-off cap from the vial of NULIBRY.

Wipe the rubber stopper on the vial of NULIBRY with a new alcohol wipe.

Hold the NULIBRY vial firmly on your work surface. Take the syringe with the Sterile Water for Injection and slowly insert the needle into the center of the vial stopper (Figure N).

Slowly push down on the plunger all the way to push the Sterile Water for Injection into the vial. Then carefully remove the needle from the vial. Throw away (dispose of) the used needle and syringe in your sharps disposal container right away.

Do not try to recap the needle. See the section “**How should I throw away (dispose of) used needles and syringes?**”

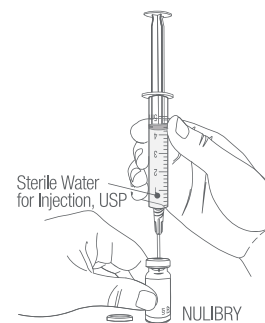


Figure N

Please see additional Important Safety Information and full Prescribing Information including Instructions for Use at <https://bit.ly/3tOMDHI>.

Preparation and Administration (continued)

Nulibry[®]
(fosdenopterin)
for injection

STEP 6 MIX (DISSOLVE) NULIBRY (CONTINUED)

Gently swirl the vial continuously until the powder is completely dissolved. **Do not** shake the vial (Figure O).

Repeat steps 4 through 6 if more than 1 vial of NULIBRY is needed to prepare your child's prescribed NULIBRY dose. Use a new 5 mL syringe and new needle for each vial of NULIBRY.

Note: When mixed, the NULIBRY solution should be clear, and colorless to pale yellow.

Do not use the solution if it is discolored or cloudy, or if there are any particles in it. If the solution is discolored or cloudy or there are particles in it:

- **Do not** throw the vial away because the pharmacist may ask that you return it
- tell your pharmacist and ask for a replacement vial

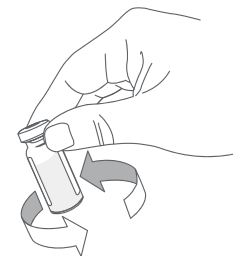


Figure O

STEP 7 PREPARE A SYRINGE WITH THE PRESCRIBED DOSE OF NULIBRY

If you stored the mixed NULIBRY solution in the refrigerator, remove the vials of mixed NULIBRY solution from the refrigerator and allow them to reach room temperature. This can be done by rolling each vial gently between your hands for 3 to 5 minutes as shown, or by allowing the vials to sit at room temperature for about 30 minutes (Figure P).

Open the wrapper that contains a new sterile needle. **Do not** remove the needle cover yet.

Open the wrapper that contains a sterile disposable syringe that is large enough to hold the total volume of NULIBRY needed for one dose. Attach the needle to the syringe tip with a screw action.

Do not remove the needle cover yet.

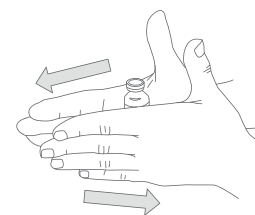


Figure P

Please see additional Important Safety Information and full Prescribing Information including Instructions for Use at <https://bit.ly/3tOMDHI>.

Preparation and Administration (continued)

STEP 7 PREPARE A SYRINGE WITH THE PRESCRIBED DOSE OF NULIBRY (CONTINUED)

Wipe the vial stopper of each mixed NULIBRY vial with a new alcohol wipe.

Remove the needle cover by pulling the cover straight off. **Do not** touch the needle or let the needle touch any surface.

Hold the syringe with one hand. Insert the needle into the center of the NULIBRY vial stopper then slowly turn the vial upside down (Figure Q).

Next, move the needle so that the tip is in the NULIBRY solution. Slowly pull back on the syringe plunger to fill the syringe with the amount of NULIBRY solution in mL for your child's prescribed dose.

Remove any air bubbles from the syringe. Tap on the syringe barrel so that any bubbles rise to the top of the vial. Push up on the plunger to push any air bubbles back into the vial. Check to make sure that the correct amount of NULIBRY solution has been drawn into the syringe. If needed, pull down slightly on the plunger until the prescribed amount of NULIBRY solution is in the syringe (Figure R).

Follow the steps below if more than 1 vial of NULIBRY is needed to make up the total amount of solution needed for 1 daily dose.

Remove the needle and syringe from the first vial of NULIBRY. Hold the syringe with one hand. Insert the needle into the center of the next NULIBRY vial stopper then slowly turn the vial upside down (Figure S).

Next, move the needle so that the tip is in the NULIBRY solution. Slowly pull back on the syringe plunger to fill the syringe with the amount of NULIBRY solution in mL for your child's prescribed dose.

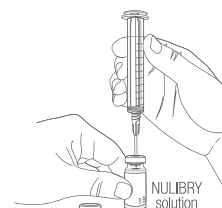


Figure Q

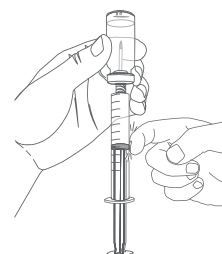


Figure R

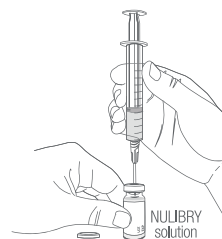


Figure S

Preparation and Administration (continued)

Nulibry[®]
(fosdenopterin)
for injection

STEP 7 PREPARE A SYRINGE WITH THE PRESCRIBED DOSE OF NULIBRY (CONTINUED)

Remove any air bubbles from the syringe. Tap on the syringe barrel so that any bubbles rise to the top of the vial. Push up on the plunger to push any air bubbles back into the vial. Then check to be sure that the prescribed amount of NULIBRY solution has been drawn into the syringe. If needed, pull down slightly on the plunger until the total prescribed amount of NULIBRY solution is in the syringe (Figure T).

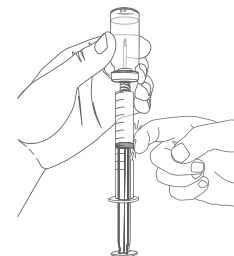


Figure T

Repeat this step if additional vials of NULIBRY are needed to make up your child's dose.

When the needle is removed from the last vial of NULIBRY, all of the mixed dose of NULIBRY is in 1 syringe.

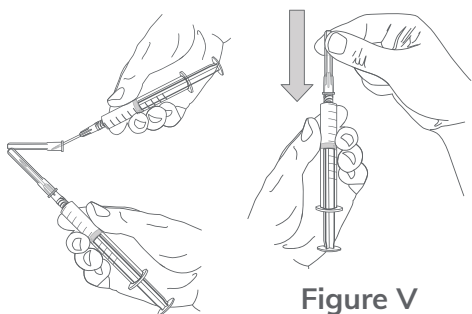


Figure U

Figure V

Replace the needle cover before removing the needle from the syringe by placing the cover on a flat surface and sliding the needle into the cover as shown. With one hand, hold the syringe and use the needle to "scoop up" the cover. Once the cover is on the needle, use the other hand to secure the cover on the needle hub (Figures U & V).

Remove the covered needle from the syringe tip with a screw action in the direction of the arrow as shown (Figure W).

Do not touch the tip of the syringe after removing the needle.

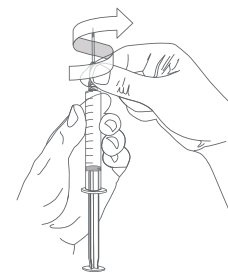


Figure W

Throw away (dispose of) the needle in the sharps disposal container right away (Figure X). See **"How should I throw away (dispose of) used needles and syringes?"**

Throw away the used NULIBRY vial(s) after use, even if there is medicine left in the vial.

The dose of NULIBRY is now ready to be given to your child.



Figure X

Please see additional Important Safety Information and full Prescribing Information including Instructions for Use at <https://bit.ly/3tOMDHI>.

Preparation and Administration (continued)

STEP 8 GIVING A DOSE OF NULIBRY

NULIBRY is given into your child's vein (intravenously) through a special access catheter or port placed by your healthcare provider.

When NULIBRY is given through an infusion pump, infuse NULIBRY at a rate of 1.5 mL per minute.

If the amount (volume) in mL for your child's prescribed dose of NULIBRY is less than 2 mL, your healthcare provider may tell you to give NULIBRY by a slow intravenous push using a syringe.

Follow your healthcare provider's instructions for how to give your child's dose of NULIBRY by slow intravenous push.

Follow your healthcare provider's instructions for proper care of your child's intravenous access catheter or port before and after giving a dose of NULIBRY.

STEP 9 RECORD THE INFUSION

After giving each dose of NULIBRY, record information about the dose in the treatment log sheet. See the section of this Instructions for Use called **“Important information you need to know before preparing and giving NULIBRY.”**

STEP 10 HOW SHOULD I THROW AWAY (DISPOSE OF) USED NEEDLES AND SYRINGES?

Put your used needles and syringes in an FDA-cleared sharps disposal container right away after use (Figure Y).

If you do not have an FDA-cleared sharps disposal container, you may use a household container that is:

- made of heavy-duty plastic,
- can be closed with a tight-fitting, puncture-resistant lid, without sharps being able to come out,
- upright and stable during use,
- leak-resistant, and
- properly labeled to warn of hazardous waste inside the container

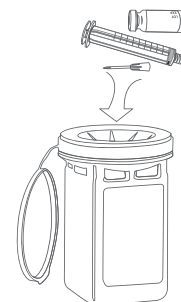
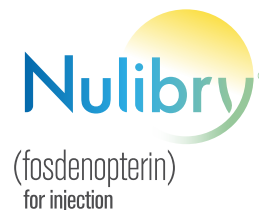


Figure Y

Please see additional Important Safety Information and full Prescribing Information including Instructions for Use at <https://bit.ly/3tOMDHI>.

Preparation and Administration (continued)



STEP 10 HOW SHOULD I THROW AWAY (DISPOSE OF) USED NEEDLES AND SYRINGES? (CONTINUED)

When your sharps disposal container is almost full, you will need to follow your community guidelines for the right way to dispose of your sharps disposal container. There may be state or local laws about how you should throw away used needles and syringes. For more information about safe sharps disposal, and for specific information about sharps disposal in the state that you live in, go to the FDA's website at: <http://www.fda.gov/safesharpsdisposal>.

Do not dispose of your used sharps disposal container in your household trash unless your community guidelines permit this. Do not recycle your used sharps disposal container.

Keep the sharps disposal container out of the reach of children.

STEP 11 WHAT SHOULD I AVOID DURING TREATMENT WITH NULIBRY?

NULIBRY may cause skin reactions.

Limit or avoid your child's exposure or time in sunlight and artificial ultraviolet (UV) light, such as UVA or UVB phototherapy.

Your child should wear clothing, a hat, and sunglasses that protect against sun exposure.

If your child is 6 months of age or older apply a broad-spectrum sunscreen with high sun protection factor.

Get medical help right away if your child develops a rash, or if you notice symptoms of a skin reaction, including: skin redness, blisters, or if your child feels like their skin is burning.

What are the ingredients in NULIBRY?

Active ingredient: fosdenopterin

Inactive ingredients: ascorbic acid, mannitol, sucrose. Sodium hydroxide and hydrochloric acid are used to adjust the pH.

For more information, call 1-888-507-5206 or go to www.NULIBRY.com.

Please see additional Important Safety Information and full Prescribing Information including Instructions for Use at <https://bit.ly/3tOMDHI>.

Indication and Important Safety Information



INDICATION

NULIBRY is indicated to reduce the risk of mortality in patients with molybdenum cofactor deficiency (MoCD) Type A.

IMPORTANT SAFETY INFORMATION

WARNINGS AND PRECAUTIONS

Potential for Photosensitivity

NULIBRY can make the patient oversensitive to sunlight. NULIBRY-treated patients or their caregivers are advised to avoid or minimize patient exposure to sunlight and artificial UV light and adopt precautionary measures when exposed to the sun, including wearing protective clothing and sunglasses, and use broad-spectrum sunscreen with high SPF in patients 6 months of age and older. If photosensitivity occurs, caregivers/patients are advised to seek medical attention immediately and consider a dermatological evaluation.

SIDE EFFECTS

The most common side effects in NULIBRY-treated patients were infusion catheter-related complications, pyrexia (fever), viral infection, pneumonia, otitis media (ear infection), vomiting, cough/sneezing, viral upper respiratory infection (common cold/flu-like infection), gastroenteritis (stomach flu-like symptoms), diarrhea, and bacteremia (bacteria in the blood). Side effects for rcPMP-treated patients were similar to the NULIBRY-treated patients.

PATIENT COUNSELING INFORMATION

Please read the FDA-approved NULIBRY Prescribing Information and Instructions for Use and follow the instructions on how to prepare and administer NULIBRY.

NULIBRY has a potential for photosensitivity; see Warnings and Precautions. Seek medical attention immediately if the patient develops a rash or if they notice symptoms of photosensitivity reactions (redness, burning sensation of the skin, blisters).

You are encouraged to report negative side effects of prescription drugs to the FDA. Visit www.fda.gov/medwatch, or call 1-800-FDA-1088.

TO LEARN MORE ABOUT NULIBRY, VISIT NULIBRY.COM/HCP

Reference: 1. NULIBRY [prescribing information]. <https://dailymed.nlm.nih.gov/dailymed/drugInfo.cfm?setid=4f67cc4e-84ed-4f4e-a5d9-6ffbfb84eddd>.

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