



Matty | All photos used with family permission

Managing intracerebroventricular (ICV) access device replacement for cerliponase alfa ▼

This brochure provides practical guidance and recommendations for planning, performing and managing ICV access device replacement in patients receiving cerliponase alfa. For additional information, refer to the Summary of Product Characteristics and your institution's policies and procedures.

Cerliponase alfa is indicated for the treatment of neuronal ceroid lipofuscinosis type 2 (CLN2) disease, also known as tripeptidyl peptidase 1 (TPP1) deficiency.¹

▼ This medicinal product is subject to additional monitoring. This will allow quick identification of new safety information. Healthcare professionals are asked to report any suspected adverse reactions.

Prescribing information and adverse event reporting information can be found [here](#) or at the end of this document.

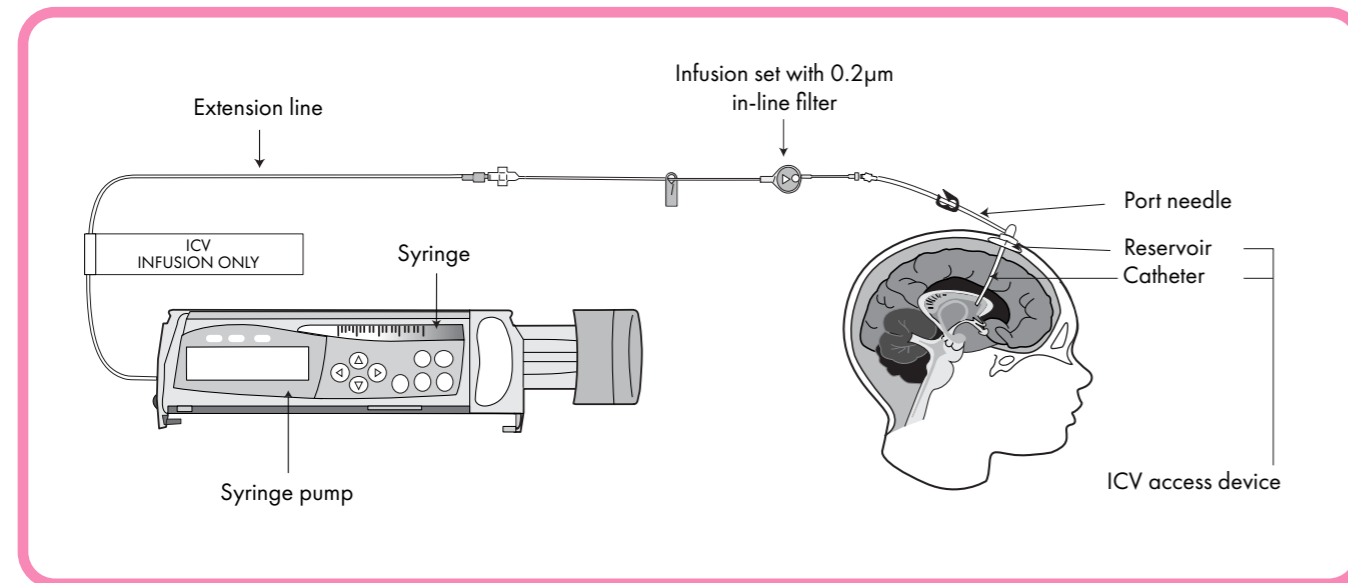
Developed and funded by BioMarin. For healthcare professionals only.

COM-ET-1523 April 2026

B:OMARIN[®]

Planning for ICV access device replacement

- ICV access for drug administration is an established and well-tolerated method for prolonged drug delivery to the brain²
- Cerliponase alfa is administered to the cerebrospinal fluid (CSF) by infusion via a surgically implanted reservoir and catheter (ICV access device)¹



Why device replacement is important¹

- Material degradation of the ICV access device reservoir has been reported after **approximately 4 years** of administration, which may impact the effective and safe use of the device¹
- During benchtop testing, such material degradation was recognised after **approximately 105 perforations** of the device¹
- In two clinical cases, the ICV access devices did not show signs of failure at the time of infusion; however, after removal, material degradation of the devices was apparent¹

When to consider device replacement¹

- The ICV access device should be replaced **prior to 4 years** of single-puncture administrations, which equates to approximately 105 administrations of cerliponase alfa¹
- **Earlier replacement may be necessary** if there are any signs of device-related complications, such as leakage, infection or device failure¹




Consider tracking the number of ICV access device punctures, as this can help you gauge the usable life of the reservoir

Inspecting and managing the ICV access device¹

- Cerliponase alfa must be administered using aseptic technique, per your institution's standard of care, to reduce the risk of infection
- It should be administered by, or under the direction of, a healthcare professional experienced in ICV administration
- Prior to each infusion, inspect the scalp for signs of ICV access device leakage, failure or potential infection
- If you suspect infection or other issues, do not proceed with the infusion. Consult with a neurosurgeon to confirm the integrity or performance of the device



Device-related complications

-  Common signs of device leakage and device failure include swelling, erythema of the scalp, extravasation of fluid, or bulging of the scalp around or above the ICV access device. However, these signs may also occur in the context of device-related infections
-  The signs and symptoms of device-related infections may not be apparent; therefore, CSF samples should routinely be sent for testing to detect subclinical device infections
-  Cerliponase alfa treatment should be interrupted in cases of device failure and may require replacement of the access device prior to subsequent infusions

Accessing the ICV access device is a fundamental step for each cerliponase alfa infusion. Therefore, it is important to review procedures to support patient safety, reduce the potential risk of infection, and avoid unnecessary or premature ICV access device replacement

Abbreviated prescribing information (INTL)

cerliponase alfa®

Refer to Summary of Product Characteristics for full prescribing information.

▼ This medicinal product is subject to additional monitoring. This will allow quick identification of new safety information. Healthcare professionals are asked to report any suspected adverse reactions.

Name of Product: Cerliponase alfa (cerliponase alfa) 150 mg solution for infusion.
Presentation: Each vial of cerliponase alfa contains 150 mg cerliponase alfa in 5 ml of solution (30 mg/ml). Each presentation contains two vials of cerliponase alfa, and one vial of flushing solution. Cerliponase alfa is a recombinant form of human tripeptidyl peptidase 1 (rhTPP1). **Therapeutic indications:** Cerliponase alfa is indicated for the treatment of neuronal ceroid lipofuscinosis type 2 (CLN2) disease, also known as tripeptidyl peptidase 1 (TPP1) deficiency. **Dosage and administration:** Cerliponase alfa must only be administered by a trained healthcare professional knowledgeable in intracerebroventricular administration in a healthcare setting. The recommended dose is 300 mg cerliponase alfa administered once every other week by intracerebroventricular infusion. In patients less than 2 years of age, lower doses are recommended, see full Summary of Product Characteristics. Cerliponase alfa and the flushing solution must only be administered by the intracerebroventricular route. Each vial of cerliponase alfa and flushing solution are intended for single use only. Cerliponase alfa is administered to the cerebrospinal fluid (CSF) by infusion via a surgically implanted reservoir and catheter (intracerebroventricular access device). The intracerebroventricular access device must be implanted prior to the first infusion. The implanted intracerebroventricular access device should be appropriate for accessing the cerebral ventricles for therapeutic administration. **Contraindications:** Life-threatening anaphylactic reaction to the active substance or to any of the excipients, if re-challenge is unsuccessful. CLN2 patients with ventriculo-peritoneal shunts. Cerliponase alfa must not be administered if there are signs of acute intracerebroventricular access device leakage, device failure, or device-related infection. **Special Warnings and Precautions:** Cerliponase alfa must be administered using aseptic technique to reduce the risk of infection. Intracerebroventricular access device-related infections, including sub-clinical infections and meningitis, have been observed in patients treated with cerliponase alfa. Meningitis may present with the following symptoms: fever, headache, neck stiffness, light sensitivity, nausea, vomiting, and change in mental status. CSF samples should routinely be sent for testing to detect subclinical device infections. In clinical studies, antibiotics were administered, the intracerebroventricular access device was replaced, and cerliponase alfa treatment was continued. Healthcare professionals should inspect the scalp for skin integrity to ensure the intracerebroventricular access device is not compromised prior to each infusion. Common signs of device leakage and device failure include swelling, erythema of the scalp, extravasation of fluid, or bulging of the scalp around or above the intracerebroventricular access device. However, these signs may also occur in the context of device-related infections. Inspection of the infusion site and a patency check must be performed to detect intracerebroventricular access device leakage and/or failure prior to initiation of cerliponase alfa infusion. The signs and symptoms of device-related infections may not be apparent, therefore, CSF samples should routinely be sent for testing to detect subclinical device infections. Consultation with a neurosurgeon

may be needed to confirm the integrity of the device. Cerliponase alfa treatment should be interrupted in cases of device failure and may require replacement of the access device prior to subsequent infusions. Material degradation of the intracerebroventricular access device reservoir occurs after long periods of use according to preliminary results of benchtop testing and as observed in clinical trials with approximately 4 years of use. In two clinical cases, the ICV access devices did not show signs of failure at the time of infusion; however, after removal, material degradation of the devices were apparent and consistent with data from benchtop testing of ICV access devices. The access devices were replaced and patients resumed treatment with cerliponase alfa. Access device replacement should be considered prior to 4 years of regular administration of cerliponase alfa, however, it must always be ensured that the intracerebroventricular access device is used in accordance with the provisions of the respective medical device manufacturer. In case of intracerebroventricular access device-related complications, refer to the manufacturer's labelling for further instruction. Caution should be taken in patients prone to complications from intracerebroventricular medicinal product administration, including patients with obstructive hydrocephalus. Anaphylactic reactions have been reported with cerliponase alfa. As a precautionary measure, appropriate medical support should be readily available when cerliponase alfa is administered. If anaphylactic reactions occur, immediately discontinue the infusion and initiate appropriate treatment. Observe patients closely during and after the infusion. If anaphylaxis occurs, caution should be exercised upon re-administration. **Undesirable Effects:** Very common adverse reactions: Device-related infection, hypersensitivity, irritability, convulsion events, headache, CSF pleocytosis, vomiting, pyrexia, CSF protein increased, ECG abnormalities, CSF protein decreased. Very common product issues: Device leakage, needle issue, device malfunction. Common adverse reactions include: bradycardia, anaphylactic reaction, gastrointestinal disorder, rash, urticaria, feeling jittery, medical device site irritation. Common product issues: Device occlusion and device breakage. Meningitis and device dislocation were also reported at unknown frequency. **List of Excipients:** Sodium phosphate dibasic heptahydrate, sodium dihydrogen phosphate monohydrate, sodium chloride, potassium chloride, magnesium chloride hexahydrate, calcium chloride dihydrate, water for injections. **Incompatibilities:** This medicinal product must not be mixed with other medicinal products. **Storage and Use:** Store upright in a freezer (-25°C to -15°C). Thawed cerliponase alfa and flushing solution should be used immediately. Product should only be withdrawn from the unopened vials immediately prior to use to protect from light. If immediate use is not possible, unopened vials of cerliponase alfa or flushing solution should be stored at 2–8°C and used within 24 hours. **Preparation of Cerliponase alfa Infusion:** See full Summary of Product Characteristics. **Legal Category:** Prescription only medicine **Marketing Authorisation Holder:** BioMarin International Limited, Shanbally, Ringaskiddy, County Cork, Ireland. **Marketing Authorisation Number(s):** EU/1/17/1192/001 **Date of First Authorisation:** 30 May 2017 **Date of API revision:** December 2023.

Healthcare professionals should report adverse events in accordance with their local requirements.

Adverse events should also be reported to BioMarin on +1 415 506 6179 or drugsafety@bmrn.com

References:

1. Cohen-Pfeffer JL, et al. *Pediatr Neuro*. 2017;67:23–35.
2. Cerliponase alfa Summary of Product Characteristics.