

# Request to Facilitate Emergency Treatment for Patient with Plasminogen Deficiency Type 1 (PLGD-1)

Patient name: \_\_\_\_\_

DOB: \_\_\_\_\_

## PLGD-1 Healthcare Management Contact Information:

- Primary provider: \_\_\_\_\_
- Treatment facility: \_\_\_\_\_
- Contact information/emergency on call #: \_\_\_\_\_

The patient named above has been diagnosed with Plasminogen Deficiency Type 1 (PLGD-1). PLGD-1 is a lifelong, multi-system disorder in which the protein plasminogen is either missing or present at inadequate levels. As a result, fibrin-rich lesions can develop in organs throughout the body. Without treatment, such lesions can result in loss of vision, lung obstruction, or other severe and debilitating medical conditions.

- This patient is receiving RYPLAZIM (plasminogen, human-tvmh) to prevent the recurrence of existing lesions or the development of new lesions, as follows: dosage: 6.6 mg/kg of body weight; frequency: every \_\_\_\_\_ days.
- This patient is not currently receiving Ryplazim (plasminogen, human-tvmh). Please contact their primary provider (above) to determine the best treatment in an emergency situation.

In emergency situations, it is imperative that this patient have ready access to RYPLAZIM:

- Prior to and during a required surgical procedure
- Should life-threatening lesions develop (such as pulmonary lesions)

In all emergency situations for patients with PLGD-1, RYPLAZIM (plasminogen, human-tvmh) should be administered. Surgical removal of lesions usually results in regrowth which can be rapid without appropriate medical therapy, and can contribute to long-term scarring and organ damage. The goal of treatment is to adequately increase the blood level of plasminogen. Consult the primary specialist named above for recommended dose and frequency of administration both during the emergency and after its resolution. Clotting factor concentrate should never be wasted, as it is very expensive. Each infusion should come as close to the recommended dose as possible, ideally  $\pm 10\%$  of the above recommended dose, while using the entire vial.

**Because RYPLAZIM is not readily available at most facilities, patients should be allowed to bring their prescribed supply of RYPLAZIM from home for use during emergencies.**

If the patient does not have a supply of RYPLAZIM, please contact their primary MD (listed above) to facilitate ordering an emergency supply. If the primary MD cannot be reached, the ER / hospital team can email [Medicalinfo@kedrion.com](mailto:Medicalinfo@kedrion.com). An assessment of the case will be performed by an independent expert. If the case is considered an emergency, a limited supply can be purchased directly from Kedrion to overcome the emergency situation.

Basic guidelines for RYPLAZIM administration (Please see full prescribing information for complete administration guidelines):

- Filtration is necessary during the infusion. A syringe is required for transferring the sterile water into the plasminogen lyophilizate, and a syringe disc filter must be attached to the butterfly needle before infusing RYPLAZIM.
- Once the plasminogen has been reconstituted, it must be infused within 3 hours.
- Dosing frequency may change depending on emergency situation. Contact HCP listed for guidance.

Please contact the primary provider's office with questions regarding administration or refer to the package insert.

Sincerely,

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