Division: Pharmacy Policy	Subject: Prior Authorization Criteria
Original Development Date: Original Effective Date: Revision Date:	January 26, 2024 March 11, 2024

Adzynma™ (ADAMTS 13, recombinant-krhn)

LENGTH OF AUTHORIZATION:
Initial Therapy: On Demand enzyme replacement therapy (ERT) - Up to 2 months
Prophylactic enzyme replacement therapy (ERT) - 6 months
Continuation of Therapy: Prophylactic enzyme replacement therapy (ERT) - 6 months

REVIEW CRITERIA:

- Patient must be ≥ 2 years of age; **AND**
- Patient must have a documented diagnosis of congenital thrombotic thrombocytopenic purpura (cTTP) confirmed by the following:
  - Molecular genetic testing showing mutation in the ADAMTS13 gene; **AND**
  - ADAMTS 13 activity testing showing less than 10% of normal activity.
- Drug must be prescribed for Prophylactic **OR** On Demand ERT, and the following requirements (per diagnosis) must apply *(official labs required)*:
  - **For Prophylactic ERT**:
    - Platelet count > 100,000/µL; **AND**
    - Lactate dehydrogenase (LDH) < 2 times the upper limit of normal (ULN) as defined by laboratory values.
  - **For On Demand ERT**, the patient must present with the following:
    - ≥ 50% drop in platelet count or platelet count < 100,000/µL; **AND**
    - LDH > 2 times the ULN as defined by laboratory values.

CONTINUATION OF THERAPY:

- Patient met initial review criteria; **AND**
- Documentation of improved clinical response (e.g., decrease in incidence of acute/subacute TTP events and TTP manifestations, a decreased incidence of supplemental therapy administration); **AND**
- Patient has not experienced any treatment-restricting adverse effects; **AND**
- Dosing is appropriate as per labeling or is supported by compendia.

DOSING AND ADMINISTRATION:

- Refer to product labeling at https://www.accessdata.fda.gov/scripts/cder/daf/
- Available as a lyophilized powder in single-dose vials containing nominally 500 or 1500 international units.