SENTARA COMMUNITY PLAN
(MEDICAID)

MEDICAL PRIOR AUTHORIZATION/STEP-EDIT REQUEST*

Directions: **The prescribing physician must sign and clearly print name (preprinted stamps not valid) on this request**. All other information may be filled in by office staff; **fax to 1-844-305-2331**. No additional phone calls will be necessary if all information (including phone and fax #s) on this form is correct. **If information provided is not complete, correct, or legible, authorization can be delayed**.

Drug Requested: Adzynma (ADAMTS13 recombinant-krhn) **(J7171) (Medical)**

MEMBER & PRESCRIBER INFORMATION: Authorization may be delayed if incomplete.

Member Name: _____

Member Sentara #: _____ **Date of Birth:** _____

Prescriber Name: _____

Prescriber Signature: _____ **Date:** _____

Office Contact Name: _____

Phone Number: _____ **Fax Number:** _____

DEA OR NPI #: _____

DRUG INFORMATION: Authorization may be delayed if incomplete.

Drug Name/Form/Strength: _____

Dosing Schedule: _____ **Length of Therapy**: _____

Diagnosis: _____ **ICD Code, if applicable:** _____

Weight: _____ **Date:** _____

☐ Standard Review. In checking this box, the timeframe does not jeopardize the life or health of the member or the member's ability to regain maximum function and would not subject the member to severe pain.

A. Quantity Limit (max daily dose) [NDC Unit]:

- Adzynma 500 IU single-dose vial for injection:
  - Prophylaxis: 2 vials every other week
  - On-Demand: 2 vials daily for 7 days
- Adzynma 1500 IU single-dose vial for injection: 64764-0135-xx
  - Prophylaxis: 3 vials every other week
  - On-Demand: 3 vials on day 1, then 2 vials on day 2 and 1 vial daily for 5 days more.

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B. Max Units (per dose and over time) [HCPCS Unit]:

Prophylaxis Therapy

- 4500 IU every other week (40 units/kg/dose once every other week; may increase frequency to once weekly)

On-Demand Therapy

- Treatment Day 1: 4500 IU (40 units/kg once)
- Treatment Day 2: 3000 IU (20 units/kg once)
- Treatment Day 3 and beyond: 1500 IU once daily x 5 days more (total of 7 days) (15 units/kg/dose once daily; continue for 2 days after the acute event has resolved)

CLINICAL CRITERIA: Check below all that apply. All criteria must be met for approval. To support each line checked, all documentation, including lab results, diagnostics, and/or chart notes, must be provided or request may be denied.

Initial Authorization:

- ❑ **For prophylaxis therapy -** coverage will be provided for 6 months and may be renewed annually
- ❑ **For on-demand therapy -** coverage is provided for 3 months per approval and is eligible for renewal

- ❑ Member is at least 2 years of age or older
- ❑ Prescribed by, or in consultation with, a hematologist/oncologist or a provider knowledgeable in genetic hematologic diseases
- ❑ Member has confirmed diagnosis of congenital thrombotic thrombocytopenic purpura (cTTP) due to inherited ADAMTS13 deficiency defined by **ALL** the following:
  - ❑ Submission of molecular genetic testing, documenting biallelic pathogenic variants in the ADAMTS13 gene located on chromosome 9q34
  - ❑ An ADAMTS13 activity of < 10 % as measured by the fluorescent resonance energy transfer- von Willebrand factor73 (FRETS-VWF73) assay (Note: Patients currently receiving prophylactic plasma infusion therapy that is clearly documented in current treatment regimen may exceed 10% ADAMTS13 activity at start of therapy)
  - ❑ Laboratory documentation confirms that anti-ADAMTS13 IgG inhibitory autoantibodies are **NOT** present
- ❑ Member does **NOT** have a diagnosis of other cTTP-like disorders (e.g., acquired TTP, immune TTP, other primary thrombotic microangiopathies, immune thrombocytopenia-ITP, Evans Syndrome)
- ❑ Laboratory measurement confirmation documents that the member does **NOT** a medical history or presence of a functional ADAMTS13 inhibitor
- ❑ Member does **NOT** have a known sensitivity to hamster protein

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- ❑ Provider is requesting Adzynma for **ONE** of the following treatment options:
  - ❑ Prophylactic Therapy: The provider has submitted documentation of past medical history recording the member having at least one TTP event or is currently receiving prophylactic plasma infusion therapy (Note: Patients who have been receiving prophylactic plasma-based therapies should discontinue routine use of those therapies after achieving a therapeutic response)
  - ❑ On-Demand Therapy: Patient is at risk of disease exacerbation

    NOTE: The cumulative amount of medication the patient has on-hand, indicated for the acute treatment of TTP events, will be considered for authorizations. The authorization will provide a sufficient quantity in order for the patient to have a cumulative amount of medication on-hand in order to treat up to 4 acute attacks per 4 weeks for the duration of the authorization.

Reauthorization: 12 months. Check below all that apply. All criteria must be met for approval. To support each line checked, all documentation, including lab results, diagnostics, and/or chart notes, must be provided or request may be denied.

❑ Prophylaxis Therapy

- ❑ Provider attests member has **NOT** reported any signs of unacceptable toxicity from treatment (e.g., severe hypersensitivity or anaphylactic reactions, severe abdominal pain, diarrhea, or headache)
- ❑ Member has **NOT** developed neutralizing antibodies to ADAMTS13
- ❑ Member has responded to therapy compared to pre-treatment baseline as documented by **ONE** of the following:
  - ❑ Member has a reduction in or an absence of an acute TTP event documented in the most recent progress notes since previous service authorization request [an acute TTP event will be defined by a drop in platelet count (≥ 50% of baseline or a platelet count <100,000/µL) and an elevation of lactate dehydrogenase (LDH) (> 2 × baseline or > 2 × upper limit normal (ULN)]
  - ❑ Member has a reduction in or an absence of a sub-acute TTP event, defined by a thrombocytopenia event or a microangiopathic hemolytic anemia event; and organ- specific signs and symptoms including but not limited to renal dysfunction events, neurological symptoms events, fever, fatigue/lethargy, and/or abdominal pain

Reauthorization: 3 months. Check below all that apply. All criteria must be met for approval. To support each line checked, all documentation, including lab results, diagnostics, and/or chart notes, must be provided or request may be denied.

❑ On-Demand Therapy

- ❑ Provider attests member has **NOT** reported any signs of unacceptable toxicity from treatment (e.g., severe hypersensitivity or anaphylactic reactions, severe abdominal pain, diarrhea, or headache)
- ❑ Member has **NOT** developed neutralizing antibodies to ADAMTS13

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- ❑ Member has responded to an acute TTP event with therapy as evidenced by improvement in thrombocytopenia (response will be defined as platelet count was ≥150,000/μL or platelet count now within 25% of baseline) or in microangiopathic hemolytic anemia (response will be defined as LDH ≤1.5 × baseline or ≤1.5 × ULN)

  NOTE: The cumulative amount of medication the patient has on-hand, indicated for the acute treatment of TTP events, will be considered for authorizations. The authorization will provide a sufficient quantity in order for the patient to have a cumulative amount of medication on-hand in order to treat up to 4 acute attacks per 4 weeks for the duration of the authorization (unless otherwise specified).

Medication being provided by: Please check applicable box below.

- ❑ **Location/site of drug administration:** _____

  NPI or DEA # of administering location: _____

  OR

- ❑ **Specialty Pharmacy – Proprium Rx**

For urgent reviews: Practitioner should call Sentara Health Plans Pre-Authorization Department if they believe a standard review would subject the member to adverse health consequences. Sentara Health Plan's definition of urgent is a lack of treatment that could seriously jeopardize the life or health of the member or the member's ability to regain maximum function.

Use of samples to initiate therapy does not meet step edit/ preauthorization criteria.

Previous therapies will be verified through pharmacy paid claims or submitted chart notes.*