

## Medical Policy:

### Adzynma (ADAMTS13, recombinant-krhn) intravenous lyophilized powder for solution

POLICY NUMBER	LAST REVIEW	ORIGIN DATE
MG.MM.PH.404	February 8, 2025	February 8, 2024

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The treating physician or primary care provider must submit to EmblemHealth, or ConnectiCare, as applicable (hereinafter jointly referred to as “EmblemHealth”), the clinical evidence that the member meets the criteria for the treatment or surgical procedure. Without this documentation and information, EmblemHealth will not be able to properly review the request preauthorization or post-payment review. The clinical review criteria expressed below reflects how EmblemHealth determines whether certain services or supplies are medically necessary. This clinical policy is not intended to pre-empt the judgment of the reviewing medical director or dictate to health care providers how to practice medicine. Health care providers are expected to exercise their medical judgment in rendering appropriate care. Health care providers are expected to exercise their medical judgment in rendering appropriate care.

EmblemHealth established the clinical review criteria based upon a review of currently available clinical information (including clinical outcome studies in the peer reviewed published medical literature, regulatory status of the technology, evidence-based guidelines of public health and health research agencies, evidence-based guidelines and positions of leading national health professional organizations, views of physicians practicing in relevant clinical areas, and other relevant factors). EmblemHealth expressly reserves the right to revise these conclusions as clinical information changes and welcomes further relevant information. Each benefit program defines which services are covered. The conclusion that a particular service or supply is medically necessary does not constitute a representation or warranty that this service or supply is covered and/or paid for by EmblemHealth, as some programs exclude coverage for services or supplies that EmblemHealth considers medically necessary.

If there is a discrepancy between this guideline and a member's benefits program, the benefits program will govern. Identification of selected brand names of devices, tests and procedures in a medical coverage policy is for reference only and is not an endorsement of any one device, test or procedure over another. In addition, coverage may be mandated by applicable legal requirements of a state, the Federal Government or the Centers for Medicare & Medicaid Services (CMS) for Medicare and Medicaid members. All coding and web site links are accurate at time of publication.

EmblemHealth may also use tools developed by third parties, such as the MCG™ Care Guidelines, to assist us in administering health benefits. The MCG™ Care Guidelines are intended to be used in connection with the independent professional medical judgment of a qualified health care provider and do not constitute the practice of medicine or medical advice. EmblemHealth Services Company, LLC, has adopted this policy in providing management, administrative and other services to EmblemHealth Plan, Inc., EmblemHealth Insurance Company, EmblemHealth Services Company, LLC, and Health Insurance Plan of Greater New York (HIP) related to health benefit plans offered by these entities. ConnectiCare, an EmblemHealth company, has also adopted this policy. All of the aforementioned entities are affiliated companies under common control of EmblemHealth Inc.

## Definitions

ADZYNMA (ADAMTS13, recombinant-krhn) is a human recombinant “A disintegrin and metalloproteinase with thrombospondin motifs 13” (rADAMTS13) indicated for prophylactic or on demand enzyme replacement therapy (ERT) in adult and pediatric patients with congenital thrombotic thrombocytopenic purpura (cTTP). ADAMTS13, recombinant-krhn is a recombinant form of the endogenous ADAMTS13. ADAMTS13 is a plasma zinc metalloprotease that regulates the activity of von Willebrand factor (VWF) by cleaving large and ultra-large VWF multimers to smaller units and thereby reducing the platelet binding properties of VWF and its propensity to form microthrombi

## Length of Authorization

Initial: 6 months

Continuation: 12 months

## Dosing Limits [Medical Benefit]

- A. Routine prophylaxis: approve up to 40 IU/kg by intravenous infusion once weekly; AND/OR
- B. On demand therapy: approve as follows

- Treatment Day 1: 40 IU/kg
- Treatment Day 2: 20 IU/kg
- Treatment Day 3 and beyond: 15 IU/kg once daily until two days after the acute event is resolved.

## Guideline

### I. Initial

#### 1. **Congenital Thrombotic Thrombocytopenic Purpura.**

- A. Patient is at least 2 years of age; **AND**
- B. At baseline (prior to therapy) ADAMTS13 activity is < 10% (< 10 IU/dL); **AND**  
*Note: Baseline refers to before any treatment was received, such as Adzynma or plasma-based therapies.*
- C. Patient does not have anti-ADAMTS13 autoantibodies as determined by a diagnostic test; **AND**
- D. Patient has a pathogenic variant or a mutation in the ADAMTS13 gene; **AND**  
*Note: Pathogenic variants or gene mutations are usually homozygous or compound heterozygous.*
- E. Medication is prescribed by or in consultation with a hematologist.

### II. Renewal

1. Documentation of a positive clinical response (e.g. improvement in acute and subacute TTP events, platelet counts, microangiopathic hemolytic anemia episodes, strokes/transient ischemic attacks, clinical symptoms)

## Applicable Procedure Codes

Code	Description
J7171	Injection, adams13, recombinant-krhn, 10 iu

## Applicable NDCs

Code	Description
64764-0130-01	Adzynma 500 Units
64764-0135-01	Adzynma 1500 Units
64764-0140-05	Adzynma 500 Units
64764-0145-05	Adzynma 1500 Units

## ICD-10 Diagnoses

Code	Description
D69.42	Congenital and hereditary thrombocytopenia purpura

## Revision History

Company(ies)	DATE	REVISION
EmblemHealth & ConnectiCare	2/08/2025	Remove from on-demand therapy for Dosage: approve up to 135 IU/kg by intravenous infusion per week as needed for the treatment of acute event(s).Add

		in Treatment Day 1: 40 IU/kg, Treatment Day 2: 20 IU/kg, Treatment Day 3 and beyond: 15 IU/kg once daily until two days after the acute event is resolved.  Addition of: Patient is at least 2 years of age
EmblemHealth & ConnectiCare	02/08/2024	New Policy

## References

1. Product Information: ADZYNMA intravenous lyophilized powder for solution, ADAMTS13, recombinant-krhn intravenous lyophilized powder for solution. Takeda Pharmaceuticals U.S.A. Inc. (per FDA), Lexington, MA, 2023.