Adzynma® (ADAMTS13, recombinant-krhn)
(Intravenous)

Document Number: IC-0736

Last Review Date: 10/01/2026
Date of Origin: 12/07/2023
Dates Reviewed: 12/2023, 11/2024, 04/2025, 04/2026, 10/2026

I. Length of Authorization [1]

Prophylaxis Therapy

- Initial: Prior authorization validity will be provided initially for 6 months (180 days).
- Renewal: Prior authorization validity may be renewed every 12 months (365 days) thereafter.

On-Demand Therapy

- Initial: Prior authorization validity will be provided initially for 3 months (90 days).
- Renewal: Prior authorization validity may be renewed every 3 months (90 days) thereafter.

  Note: The cumulative amount of medication the member 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 member 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.

II. Dosing Limits

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

- 1800 billable units every 28 days

III. Initial Approval Criteria [1]

Prior authorization validity is provided in the following conditions:

- Member is at least 2 years of age; **AND**
- Member has not been diagnosed with other congenital thrombotic thrombocytopenic purpura (cTTP) like disorders (e.g., acquired TTP, immune TTP, other primary thrombotic microangiopathies, immune thrombocytopenia, Evans Syndrome, etc.); **AND**
- Member does not have a medical history or the presence of functional ADAMTS13 inhibitors prior to the start of therapy; **AND**

Universal Criteria [1-3]

- Member does NOT have any FDA-labeled contraindications to the requested agent (e.g., have manifested life-threatening hypersensitivity reactions to ADZYNMA or its components); **AND**

© 2026 Prime Therapeutics Management, LLC
Proprietary & Confidential.

- Member (including those who were previously exposed or naïve to plasma-based products) will have ADAMTS13 activity and ADAMTS13 neutralizing antibodies assessed prior to initiation and periodically during the course of treatment; **AND**

Congenital Thrombotic Thrombocytopenic Purpura (cTTP) † Φ [1-9]

- Member has a documented diagnosis of severe hereditary ADAMTS13 deficiency, defined as confirmed by molecular genetic testing, documenting biallelic pathogenic variants in *ADAMTS13*; **AND**
- Member has an ADAMTS13 activity of < 10 % as measured by the fluorescent resonance energy transfer- von Willebrand factor73 (FRETS-VWF73) assay *(Note: Members currently receiving prophylactic plasma infusion therapy may exceed 10% ADAMTS13 activity at start of therapy)*; **AND**
  - Treatment will be used as prophylactic therapy; **AND**
    - Member has a history of at least one TTP event or is currently receiving prophylactic plasma infusion therapy *(Note: Members who have been receiving prophylactic plasma-based therapies should discontinue routine use of those therapies after achieving a therapeutic response)*; **OR**
  - Treatment will be used as on-demand therapy and member is at risk of a disease exacerbation

† FDA Approved Indication(s); ‡ Compendium Recommended Indication(s); Φ Orphan Drug

IV. Renewal Criteria [1]

Prior authorization validity may be renewed based upon the following criteria:

- Member continues to meet the universal and other indication-specific relevant criteria identified in section III; **AND**
- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include: severe hypersensitivity or anaphylactic reactions, neutralizing antibodies, etc.; **AND**
- Member has responded to therapy compared to pre-treatment baseline with the following:
  - If symptomatic, improvement in the signs and symptoms of disease (e.g., neurological symptoms including confusion, dysphonia, dysarthria, focal or general motor symptoms, seizures; renal dysfunction, TTP-related pain, etc.); **AND**

Prophylaxis:

- Member has a reduction in or an absence of an acute TTP event, 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]); **OR**
- 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; **OR**

On-Demand*:

Proprietary Information. Restricted Access – Do not disseminate or copy without approval.

©2026 Prime Therapeutics Management, LLC

- Member has responded to an acute TTP event with therapy as evidenced by improvement in thrombocytopenia *(defined as a drop in platelets ≥25% of baseline or a platelet count less than <150,000/mcg/L)* or in microangiopathic hemolytic anemia *(defined as an elevation of LDH > 1.5 times of baseline or >1.5 times ULN)*

  **Note: The cumulative amount of medication the member 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 member 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).*

V. Dosage/Administration [1]

Indication	Dose
Congenital Thrombotic Thrombocytopenic Purpura (cTTP)	**Prophylactic Therapy** The recommended prophylactic dosage regimen is as follows: – Administer 40 IU/kg body weight once every other week. *Note: The prophylactic dosing frequency may be adjusted to 40 IU/kg body weight once weekly based on prior prophylactic dosing regimen or clinical response.* **On-Demand Therapy** A guide for dosing Adzynma for on-demand treatment of an acute event is 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.
– Each vial of Adzynma is labeled with the actual rADAMTS13 activity, measured in terms of its potency in International Units (IU). – Calculate administration dose and volume based on the member's body weight using the actual potency (and not the nominal potency) as printed on Adzynma vial. – For Intravenous (IV) Infusion at a rate of 2 to 4 mL per minute.	

VI. Billing Code/Availability Information

HCPCS code:

- J7171 – Injection, adamts13, recombinant-krhn, 10 iu; 1 billable unit = 10 units

NDC:

- Adzynma 500 IU single-dose vial for injection: 64764-0130-xx
- Adzynma 1500 IU single-dose vial for injection: 64764-0135-xx

VII. References

1. Adzynma [package insert]. Cambridge, MA; Takeda Pharmaceuticals U.S.A., Inc.; August 2026. Accessed September 2026.

Proprietary Information. Restricted Access – Do not disseminate or copy without approval.

©2026 Prime Therapeutics Management, LLC

2. Asmis, L. M., Serra, A., Krafft, A., Licht, A., Leisinger, E., Henschkowski-Serra, J., et al. 2022. Recombinant ADAMTS13 for Hereditary Thrombotic Thrombocytopenic Purpura. N Engl J Med, 387, 2356-2361.
3. Alwan, F., C. Vendramin, R. Liesner, A. Clark, W. Lester, T. Dutt, W. Thomas, R. Gooding, T. Biss, H. G. Watson, N. Cooper, R. Rayment, T. Cranfield, J. J. van Veen, Q. A. Hill, S. Davis, J. Motwani, N. Bhatnagar, N. Priddee, M. David, M. P. Crowley, J. Alamelu, H. Lyall, J. P. Westwood, M. Thomas and M. Scully (2019). "Characterization and treatment of congenital thrombotic thrombocytopenic purpura." Blood 133(15): 1644-1651.
4. Sukumar, S., B. Lämmle and S. R. Cataland (2021). "Thrombotic Thrombocytopenic Purpura: Pathophysiology, Diagnosis, and Management." J Clin Med 10(3): 536.
5. Zheng XL, Al-Housni Z, Cataland SR, et al. 2025 focused update of the 2020 ISTH guidelines for management of thrombotic thrombocytopenic purpura. J Thromb Haemost. 2025;23(11):3711-3732. https://doi.org/10.1016/j.jtha.2025.06.002
6. ClinicalTrials.gov. *A Phase 3, Prospective, Randomized, Controlled, Open-label, Multicenter, 2 Period Crossover Study With a Single Arm Continuation Evaluating the Safety And Efficacy of BAX 930 (rADAMTS13) in the Prophylactic And On-demand Treatment of Subjects With Severe Congenital Thrombotic Thrombocytopenic Purpura (cTTP, Upshaw-Schulman Syndrome [USS], Hereditary Thrombotic Thrombocytopenic Purpura [hTTP])*. https://clinicaltrials.gov/study/NCT03393975?intr=tak-755&rank=4.
7. ClinicalTrials.gov. *A Phase 3b, Prospective, Open-label, Multicenter, Single Treatment Arm, Continuation Study of the Safety and Efficacy of TAK-755 (rADAMTS13, Also Known as BAX 930/SHP655) in the Prophylactic and On-demand Treatment of Subjects With Severe Congenital Thrombotic Thrombocytopenic Purpura (cTTP; Upshaw-Schulman Syndrome, or Hereditary Thrombotic Thrombocytopenic Purpura)*. https://clinicaltrials.gov/study/NCT04683003?intr=tak-755&rank=1.
8. Cuker A, Chaturvedi S (Jan 2026). Congenital thrombotic thrombocytopenic purpura (cTTP). Crowther M, Tirnauer JS (Eds.). *UpToDate*. Accessed February 24, 2026. Available from: https://www.uptodate.com/contents/congenital-thrombotic-thrombocytopenic-purpura-cttp?search=hereditary%20thrombotic%20thrombocytopenic%20purpura&source=search_result&selectedTitle=1~40&usage_type=default&display_rank=1
9. Scully M, Antun A, Cataland SR, et al. 2024 "Recombinant ADAMTS13 in Congenital Thrombotic Thrombocytopenic Purpura" N Engl J Med. 2024 May 2;390(17):1584-1596. doi: 10.1056/NEJMoa2314793.

Appendix A – Non-Quantitative Treatment Limitations (NQTL) Factor Checklist

Non-quantitative treatment limitations (NQTLs) refer to the methods, guidelines, standards of evidence, or other conditions that can restrict how long or to what extent benefits are provided under a health plan. These may include things like utilization review or prior authorization. The utilization management NQTL applies comparably, and not more stringently, to mental health/substance use disorder (MH/SUD) Medical Benefit Prescription Drugs and medical/surgical (M/S) Medical Benefit Prescription Drugs. The table below lists the factors that were considered in designing and applying prior

Proprietary Information. Restricted Access – Do not disseminate or copy without approval.

©2026 Prime Therapeutics Management, LLC

authorization to this drug/drug group, and a summary of the conclusions that Prime's assessment led to for each.

Factor	Conclusion
Indication	Yes: Consider for PA
Safety and efficacy	Yes: Consider for PA
Potential for misuse/abuse	No: PA not a priority
Cost of drug	Yes: Consider for PA

Appendix 1 – Covered Diagnosis Codes

ICD-10	ICD-10 Description
D69.42	Congenital and hereditary thrombocytopenia purpura

Appendix 2 – Centers for Medicare and Medicaid Services (CMS)

The preceding information is intended for non-Medicare coverage determinations. Medicare coverage for outpatient (Part B) drugs is outlined in the Medicare Benefit Policy Manual (Pub. 100-2), Chapter 15, §50 Drugs and Biologicals. In addition, National Coverage Determinations (NCDs) and/or Local Coverage Determinations (LCDs) may exist and compliance with these policies is required where applicable. Local Coverage Articles (LCAs) may also exist for claims payment purposes or to clarify benefit eligibility under Part B for drugs which may be self-administered. The following link may be used to search for NCD, LCD, or LCA documents: https://www.cms.gov/medicare-coverage-database/search.aspx. Additional indications, including any preceding information, may be applied at the discretion of the health plan.

Medicare Part B Covered Diagnosis Codes (applicable to existing NCD/LCA/LCD): N/A

Medicare Part B Administrative Contractor (MAC) Jurisdictions		
Jurisdiction	**Applicable State/US Territory**	**Contractor**
E (1)	CA, HI, NV, AS, GU, CNMI	Noridian Healthcare Solutions, LLC
F (2 & 3)	AK, WA, OR, ID, ND, SD, MT, WY, UT, AZ	Noridian Healthcare Solutions, LLC
5	KS, NE, IA, MO	Wisconsin Physicians Service Insurance Corp (WPS)
6	MN, WI, IL	Wellpoint Federal
H (4 & 7)	LA, AR, MS, TX, OK, CO, NM	Novitas Solutions, Inc.
8	MI, IN	Wisconsin Physicians Service Insurance Corp (WPS)
N (9)	FL, PR, VI	First Coast Service Options, Inc.
J (10)	TN, GA, AL	Palmetto GBA
M (11)	NC, SC, WV, VA (excluding below)	Palmetto GBA
L (12)	DE, MD, PA, NJ, DC (includes Arlington & Fairfax counties and the city of Alexandria in VA)	Novitas Solutions, Inc.
K (13 & 14)	NY, CT, MA, RI, VT, ME, NH	Wellpoint Federal

Proprietary Information. Restricted Access – Do not disseminate or copy without approval.

©2026 Prime Therapeutics Management, LLC

Medicare Part B Administrative Contractor (MAC) Jurisdictions		
Jurisdiction	**Applicable State/US Territory**	**Contractor**
15	KY, OH	CGS Administrators, LLC

Proprietary Information. Restricted Access – Do not disseminate or copy without approval.

©2026 Prime Therapeutics Management, LLC