Medical Drug Clinical Criteria

Subject:	Adzynma (ADAMTS13, recombinant-krhn)		
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Overview

This document addresses the use of Adzynma (ADAMTS13, recombinant-krhn), a recombinant ADAMTS13 protein. It replaces the missing or deficient ADAMTS13 enzyme in patients diagnosed with thrombotic thrombocytopenic purpura (TTP). Adzynma was approved by the U.S. Food and Drug Administration (FDA) November 9, 2023.

Adzynma (ADAMTS13, recombinant-krhn) was granted orphan drug designation by the FDA for the treatment and prevention of congenital (cTTP), acquired idiopathic and secondary forms of TTP. Adzynma has also been granted Fast Track Designation for the treatment and prevention of acute episodes of TTP in patients with congenital ADAMTS13 deficiency. The phase 3, pivotal study (NCT03393975) included subjects with cTTP only and excluded any other TTP-like disorders (microangiopathic hemolytic anemia), including acquired TTP. Individuals had a documented diagnosis of severe hereditary ADAMTS13 deficiency, defined as: confirmed molecular genetic testing and ADAMTS13 activity less than 10% as measured by the fluorescent resonance energy transfer von Willebrand factor 73 (FRETS-VWF73) assay in their medical history or at screening.

Clinical Criteria

When a drug is being reviewed for coverage under a member's medical benefit plan or is otherwise subject to clinical review (including prior authorization), the following criteria will be used to determine whether the drug meets any applicable medical necessity requirements for the intended/prescribed purpose.

Adzynma (ADAMTS13, recombinant-krhn)

Initial requests for Adzynma (ADAMTS13, recombinant-krhn) may be approved if the following criteria are met:

I. Individual has a diagnosis of severe congenital thrombotic thrombocytopenic purpura (cTTP); **AND**
II. Individual is using for on-demand treatment; **AND**
III. Documentation is provided that individual has the following (A and B) (NCT03393975, NCT04683003):
    A. Molecular genetic testing showing mutation in the ADAMTS13 gene; **AND**
    B. ADAMTS13 activity testing showing less than 10% of normal ADAMTS13 activity; **AND**
IV. Documentation is provided that individual is experiencing a 50% or greater drop in platelet count or platelet count is less than 100,000/microliter; **AND**
V. Lactate dehydrogenase elevation (LDH) is more than 2 times baseline or more than 2 times upper limit of normal (ULN) as defined by laboratory values;

OR

VI. Individual has a diagnosis of severe congenital thrombotic thrombocytopenic purpura (cTTP); **AND**
VII. Individual is using for prophylactic treatment; **AND**
VIII. Documentation is provided that individual has the following (A and B) (NCT03393975, NCT04683003):
    A. Molecular genetic testing showing mutation in the ADAMTS13 gene; **AND**
    B. ADAMTS13 activity testing showing less than 10% of normal ADAMTS13 activity; **AND**
IX. Documentation is provided that individual presents with platelet count greater than 100,000/ microliter (NCT03393975, NCT04683003); **AND**

X. Individual presents with lactate dehydrogenase (LDH) less than 2 times the upper limit of normal (ULN) as defined by laboratory values (NCT03393975, NCT04683003).

Continuation requests for Adzynma (ADAMTS13, recombinant-krhn) use may be approved if the following criteria are met:

I. Documentation is provided that individual is using on-demand treatment and platelet counts increase to at least 150,000/microliter or increases to 25% from baseline platelet counts; **OR**
II. Documentation is provided that individual is using for prophylactic treatment and individual has decreased number of TTP events.

Requests for Adzynma (ADAMTS13, recombinant-krhn) may not be approved for the following:

I. Individual is diagnosed with acquired idiopathic or secondary forms of thrombotic thrombocytopenic purpura; **OR**
II. When the above criteria are not met and for all other indications.

Coding

The following codes for treatments and procedures applicable to this document are included below for informational purposes. Inclusion or exclusion of a procedure, diagnosis or device code(s) does not constitute or imply member coverage or provider reimbursement policy. Please refer to the member's contract benefits in effect at the time of service to determine coverage or non-coverage of these services as it applies to an individual member.

HCPCS

J7171	Injection, adamts13, recombinant-krhn, 10 iu

ICD-10 Diagnosis

D69.42	Congenital and hereditary thrombocytopenia purpura

Document History

Reviewed: 05/17/2024
Document History:

- 05/17/2024 – Annual Review: No Change. Coding Reviewed: Effective 7/1/2024 Remove HCPCS J3590, J9399, C9167. Remove All diagnoses pend. Effective 7/1/2024 Add HCPCS J7171. Add ICD-10-CM D69.42.
- 11/17/2023 – New: Create clinical criteria for Adzynma. Coding Reviewed: Added HCPCS J3590, C9399. All diagnoses pend. Effective 4/1/2024 Added HCPCS C9167. Revised code description.

References

1. Clinical Pharmacology [database online]. Tampa, FL: Gold Standard, Inc.: 2024. URL: http://www.clinicalpharmacology.com. Updated periodically.
2. DailyMed. Package inserts. U.S. National Library of Medicine, National Institutes of Health website. http://dailymed.nlm.nih.gov/dailymed/about.cfm. Updated periodically.
3. DrugPoints® System [electronic version]. Truven Health Analytics, Greenwood Village, CO. Updated periodically.
4. 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]). ClinicalTrials.gov identifier: NCT03393975. Updated February 28, 2023. Accessed October 12, 2023. https://clinicaltrials.gov/study/NCT03393975
5. 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). ClinicalTrials.gov identifier:

NCT04683003. Updated May 22, 2023. Accessed: October 12, 2023. https://clinicaltrials.gov/study/NCT04683003
6. Sukumar, Senthil et al. “Thrombotic Thrombocytopenic Purpura: Pathophysiology, Diagnosis, and Management.” Journal of clinical medicine vol. 10,3 536. 2 Feb. 2021, doi:10.3390/jcm10030536
7. Zheng, X Long et al. “ISTH guidelines for treatment of thrombotic thrombocytopenic purpura.” Journal of thrombosis and haemostasis: JTH vol. 18,10 (2020): 2496-2502. doi:10.1111/jth.15010

Federal and state laws or requirements, contract language, and Plan utilization management programs or polices may take precedence over the application of this clinical criteria.

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