Medical Necessity Guidelines

Medical Benefit Drugs

Adzynma (ADAMTS13, recombinant-krhn)

Effective: January 14, 2025

Guideline Type	☒ Prior Authorization ☐ Non-Formulary ☐ Step-Therapy ☐ Administrative

Applies to:

Commercial Products

☒ Harvard Pilgrim Health Care Commercial products; Fax 617-673-0988

☒ Tufts Health Plan Commercial products; Fax 617-673-0988

CareLink[SM] – Refer to CareLink Procedures, Services and Items Requiring Prior Authorization

Public Plans Products

☒ Tufts Health Direct – A Massachusetts Qualified Health Plan (QHP) (a commercial product); Fax 617-673-0988

☐ Tufts Health Together – MassHealth MCO Plan and Accountable Care Partnership Plans; Fax 617-673-0939

☒ Tufts Health RITogether – A Rhode Island Medicaid Plan; Fax 617-673-0939

☐ Tufts Health One Care* – A Medicare-Medicaid Plan (a dual eligible product); Fax 617-673-0956

*The MNG applies to Tufts Health One Care members unless a less restrictive LCD or NCD exists.

Senior Products

☐ Harvard Pilgrim Health Care Stride Medicare Advantage; Fax 617-673-0956

☐ Tufts Health Plan Senior Care Options (SCO), (a dual-eligible product); Fax 617-673-0956

☐ Tufts Medicare Preferred HMO, (a Medicare Advantage product); Fax 617-673-0956

☐ Tufts Medicare Preferred PPO, (a Medicare Advantage product); Fax 617-673-0956

Note: While you may not be the provider responsible for obtaining prior authorization, as a condition of payment you will need to ensure that prior authorization has been obtained.

Overview

Food and Drug Administration - Approved Indications

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).

Clinical Guideline Coverage Criteria

The plan may authorize Adzynma for Members when the following criteria are met:

Initial Authorization Criteria

1. Documented diagnosis of congenital thrombotic thrombocytopenic purpura confirmed by both of the following:
   a. Molecular genetic testing
   b. ADAMTS13 activity less than 10%, as measured by the fluorescent resonance energy transfer-von Willebrand factor 73 assay

AND

2. Prescribed by or in consultation with a hematologist, oncologist, or specialist in rare genetic hematologic diseases

AND

3. If the request is for use as prophylactic therapy, documentation of **one (1)** of the following:
   a. The patient has a history of at least one documented thrombotic thrombocytopenic purpura event
   b. The patient is currently receiving prophylactic therapy with Adzynma to manage congenital thrombotic thrombocytopenic purpura

Reauthorization Criteria

1. Documented diagnosis of congenital thrombotic thrombocytopenic purpura confirmed by both of the following:
    a. Molecular genetic testing
    b. ADAMTS13 activity less than 10%, as measured by the fluorescent resonance energy transfer-von Willebrand factor 73 assay

AND

2. Prescribed by or in consultation with a hematologist, oncologist, or specialist in rare genetic hematologic diseases

AND

3. Documentation the patient has experienced a therapeutic response as defined by **one (1)** of the following:
    a. Reduction in or improvement in acute thrombotic thrombocytopenic purpura events defined as a drop in platelet count of greater than or equal to 50% of baseline, or platelet count less than 100,000 u/L AND an elevation of lactate dehydrogenase of greater than two times baseline or greater than two times the upper limit of normal)
    b. Reduction in or improvement in subacute thrombotic thrombocytopenic purpura event defined as 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
    c. Reduction in or improvement in thrombotic thrombocytopenic purpura manifestations defined as a drop in platelet count of greater than or equal to 25% of baseline, or platelet count of less than 150,000 u/L, or an elevation of lactate dehydrogenase greater than 1.5 x baseline or greater than 1.5 times the upper limit of normal

Limitations

- Initial Authorizations will be provided for 6 months. Reauthorizations will be provided for 12 months.
- Members new to the plan stable on Adzynma should be reviewed against Initial Authorization Criteria.

Codes

The following code(s) require prior authorization:

Table 1: HCPCS Codes

HPCPS Codes	Description
J7171	INJECTION ADAMTS13 RECOMBINANT-KRHN 10 IU

References

1. Alwan F, et al. Characterization and treatment of congenital thrombotic thrombocytopenic purpura. Blood. 2019;133(15):1644-1651.
2. Asmis, LM, et al. Recombinant ADAMTS13 for hereditary thrombotic thrombocytopenic purpura. N Engl J Med. 2022;387(25):2356-2361
3. Scully M, et al. A British society of haematology guideline: diagnosis and management of thrombotic thrombocytopenic purpura and thrombotic microangiopathies. British Journal of Haematology. 2023;203(4):546-563.
4. Scully M, et al. S305: Phase 2 randomized, placebo-controlled, double-blind, multicenter study of recombinant ADAMTS13 in patients with immune-mediated thrombotic thrombocytopenic purpura. Hemasphere. 2023;7(Suppl):e8651306. Published August 8, 2023.
5. Scully M, et al. Recombinant ADAMTS-13: first-in-human pharmacokinetics and safety in congenital thrombotic thrombocytopenic purpura. Blood. 2017;130(19):2055-2063.
6. Sukumar S, et al. Thrombotic thrombocytopenic purpura: pathophysiology, diagnosis, and management. J Clin Med. 2021;10(3):536.
7. Zheng XL, et al. Good practice statements (GPS) for the clinical care of patients with thrombotic thrombocytopenia purpura. J Thromb Haemost. 2020;18:2503–2512.
8. Zheng XL, et al. ISTH guidelines for the diagnosis of thrombotic thrombocytopenic purpura. J Thromb Haemost. 2020;18:2486–2495.
9. Adzynma (ADAMTS13, recombinant-krhn) [prescribing Information]. Lexington, MA: Takeda Pharmaceuticals U.S.A., Inc.; June 2024.

Approval And Revision History

March 12, 2024: Reviewed by Pharmacy and Therapeutics Committee (P&T)

Subsequent endorsement date(s) and changes made:

- July 1, 2024: Administrative update: Removed expired C Code C9167 and added J Code J7171 as part of Quarterly HCPC code updates (eff 7/1/24).
- January 14, 2025: No changes (eff 1/14/25)

Background, Product and Disclaimer Information

Medical Necessity Guidelines are developed to determine coverage for benefits and are published to provide a better understanding of the basis upon which coverage decisions are made. We make coverage decisions using these guidelines, along with the Member's benefit document, and in coordination with the Member's physician(s) on a case-by-case basis considering the individual Member's health care needs.

Medical Necessity Guidelines are developed for selected therapeutic or diagnostic services found to be safe and proven effective in a limited, defined population of patients or clinical circumstances. They include concise clinical coverage criteria based on current literature review, consultation with practicing physicians in our service area who are medical experts in the particular field, FDA and other government agency policies, and standards adopted by national accreditation organizations. We revise and update Medical Necessity Guidelines annually, or more frequently if new evidence becomes available that suggests needed revisions.

For self-insured plans, coverage may vary depending on the terms of the benefit document. If a discrepancy exists between a Medical Necessity Guideline and a self-insured Member's benefit document, the provisions of the benefit document will govern. For Tufts Health Together (Medicaid), coverage may be available beyond these guidelines for pediatric members under age 21 under the Early and Periodic Screening, Diagnostic and Treatment (EPSDT) benefits of the plan in accordance with 130 CMR 450.140 and 130 CMR 447.000, and with prior authorization.

Treating providers are solely responsible for the medical advice and treatment of Members. The use of this guideline is not a guarantee of payment or a final prediction of how specific claim(s) will be adjudicated. Claims payment is subject to eligibility and benefits on the date of service, coordination of benefits, referral/authorization, utilization management guidelines when applicable, and adherence to plan policies, plan procedures, and claims editing logic.