Medical Policy:

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

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

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

A. 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 up to 135 IU/kg by intravenous infusion per week as needed for the treatment of acute event(s).

Note: On demand therapy is given as a daily dose until 2 days after the acute event resolves; however, the total weekly dose should not exceed 135 IU/kg.

Guideline

I. Initial

1. **Congenital Thrombotic Thrombocytopenic Purpura.**
   A. 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.
   B. Patient does not have anti-ADAMTS13 autoantibodies as determined by a diagnostic test; **AND**
   C. 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.
   D. 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
J3590	Adzynma IV (ADAMTS13, recombinant-krhn), unclassified drugs
C9399	Adzynma IV (ADAMTS13, recombinant-krhn), unclassified drugs or biologics

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	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.