

PHARMACY POLICY STATEMENT

Ohio Medicaid

DRUG NAME	Enjaymo (sutimlimab)
BENEFIT TYPE	Medical
STATUS	Prior Authorization Required

Enjaymo is a classical complement inhibitor indicated for the treatment of hemolysis in adults with cold agglutinin disease (CAD). It is an immunoglobulin G (IgG), subclass 4 (IgG4) monoclonal antibody (mAb) that inhibits the classical complement pathway (CP) and specifically binds to complement protein component 1, s subcomponent (C1s), a serine protease which cleaves C4. Inhibition of the classical complement pathway at the level of C1s prevents deposition of complement opsonins on the surface of RBCs, resulting in inhibition of hemolysis in patients with CAD.

CAD is a form of autoimmune hemolytic anemia (AIHA) in which cold agglutinins can cause clinical symptoms related to RBC agglutination in cooler parts of the body and hemolytic anemia. Cold agglutinins are IgM autoantibodies against red blood cell antigens that bind at cold temperatures. Primary CAD (also called idiopathic CAD) is used to refer to cold agglutinins that cause RBC agglutination and extravascular hemolysis in the absence of an underlying disorder.

Enjaymo (sutimlimab) will be considered for coverage when the following criteria are met:

Cold Agglutinin Disease (CAD)

For **initial** authorization:

1. Member is at least 18 years of age; AND
2. Medication must be prescribed by or in consultation with hematologist or CAD specialist; AND
3. Member has a diagnosis of primary CAD confirmed by **ALL** of the following:
 - a) Cold agglutinin titer of ≥ 64 ;
 - b) Positive polyspecific direct antiglobin test (DAT);
 - c) Positive monospecific DAT for C3d;
 - d) Immunoglobulin G (IgG) DAT $\leq 1+$;
 - e) Evidence of chronic hemolysis (such as elevated reticulocyte count or increased lactate dehydrogenase (LDH)); AND
4. Member has documentation of one or more of the following symptoms: symptomatic anemia, acrocyanosis, Raynaud's phenomenon, hemoglobinuria, disabling circulatory symptoms, or a major adverse vascular event; AND
5. Member has a documented hemoglobin level ≤ 10.0 g/dL; AND
6. Member has a documented bilirubin level above normal reference range; AND
7. Member does NOT have CAD secondary to infection, rheumatologic disease, systemic lupus erythematosus, or overt hematologic malignancy.
8. **Dosage allowed/Quantity limit:** Initiate Enjaymo intravenously weekly for the first two weeks, with administration every two weeks thereafter. Quantity Limit: 14 vials per 30 days.
For patients weighing 39 kg to less than 75 kg: 6,500 mg by intravenous infusion (6 vials).
For patients weighing 75 kg or more: 7,500 mg by intravenous infusion (7 vials).

If all the above requirements are met, the medication will be approved for 6 months.

For **reauthorization**:

1. Labs must show the member's hemoglobin has increased by at least 1.5 g/dL; OR
2. Chart notes must show improvement or stabilized signs and symptoms of disease (such as reduced fatigue, decrease in bilirubin, decrease in the number of blood transfusions, etc.).

If all the above requirements are met, the medication will be approved for an additional 12 months.

CareSource considers Enjaymo (sutimlimab) not medically necessary for the treatment of conditions that are not listed in this document. For any other indication, please refer to the Off-Label policy.

DATE	ACTION/DESCRIPTION
03/21/2022	New policy for Enjaymo created.
05/22/2023	Removed requirement of history of blood transfusion to align with FDA labeling; added criteria to confirm diagnosis per clinical trial; removed not using in combination with rituximab; adjusted hemoglobin increase goal in reauthorization criteria from 2 to 1.5 g/dL per updated clinical trial; added references; added a quantity limit; changed reauthorization example from QOL to reduced fatigue per clinical trial; included exclusion of CAD secondary to other causes.

References:

1. Enjaymo [package insert]. Waltham, MA; Bioverativ USA Inc. March 2023.
2. Tvedt THA, et al. Sutimlimab, an investigational C1s inhibitor, effectively prevents exacerbation of hemolytic anemia in a patient with cold agglutinin disease undergoing major surgery. *Am J Hematol.* 2022 Feb 1;97(2):E51-E54.
3. Röth A, et al. Sutimlimab in Cold Agglutinin Disease. *N Engl J Med.* 2021 Apr 8;384(14):1323-1334.
4. Gabbard AP, Booth GS. Cold Agglutinin Disease. *Clin Hematol Int.* 2020;2(3):95-100.
5. Röth A, Berentsen S, Barcellini W, et al. Sutimlimab in patients with cold agglutinin disease: results of the randomized placebo-controlled phase 3 CADENZA trial. *Blood.* 2022;140(9):980-991. doi:10.1182/blood.2021014955
6. Brugnara C, Berentsen S, Cold agglutinin disease. In: UpToDate, Post, TW (Ed), UpToDate, Waltham, MA, 2023.

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Revised date: 05/22/2023