Reference number(s)
2871-A

SPECIALTY GUIDELINE MANAGEMENT

CABLIVI (caplacizumab-yhdp)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indications
Cablivi is indicated for the treatment of adult patients with acquired thrombotic thrombocytopenic purpura (aTTP), in combination with plasma exchange and immunosuppressive therapy.

All other indications are considered experimental/investigational and not medically necessary.

II. DOCUMENTATION

Submission of the following information is necessary to initiate the prior authorization review:
For continuation of therapy: medical record documentation of signs of persistent underlying aTTP

III. CRITERIA FOR INITIAL APPROVAL

Acquired thrombotic thrombocytopenic purpura (aTTP)
Authorization of 30 days may be granted for treatment of acquired thrombotic thrombocytopenic purpura (aTTP), after the plasma exchange period in the inpatient setting, when all of the following criteria are met:
A. The member received the requested medication with plasma exchange.
B. The requested medication will be given in combination with immunosuppressive therapy.
C. The member will not receive the requested medication beyond 30 days from the cessation of plasma exchange unless the member has documented persistent aTTP.
D. The member has not experienced more than 2 recurrences of aTTP while on the requested medication. (A recurrence is when the member needs to reinitiate plasma exchange. A 28-day extension of therapy does not count as a recurrence.)

IV. CONTINUATION OF THERAPY

Authorization of 28 days may be granted for continuation of therapy for aTTP when all of the following criteria are met:
A. The request for continuation of therapy is for extension of therapy after the initial course of the requested medication (initial course: treatment with the requested medication during and 30 days after plasma exchange).
B. The member has either of the following documented signs of persistent underlying aTTP:
   1. ADAMTS13 activity level less than 10% or

2. All of the following:
    a. Microangiopathic hemolytic anemia (MAHA) documented by the presence of schistocytes on peripheral smear
    b. Thrombocytopenia (platelet count below normal per laboratory reference range), and
    c. Elevated lactate dehydrogenase (LDH) level (LDH level above normal per laboratory reference range)

C. The requested medication will be given in combination with immunosuppressive therapy.
D. The member has not received a prior 28-day extension of therapy after the initial course of the requested medication for this course of treatment.
E. The member has not experienced more than 2 recurrences of aTTP while on the requested medication. (A recurrence is when the member needs to reinitiate plasma exchange. A 28-day extension of therapy does not count as a recurrence.)

V. REFERENCES

1. Cablivi [package insert]. Cambridge, MA: Genzyme Corporation; April 2023.
2. Scully M, Cataland SR, Peyvandi F; et al. Caplacizumab treatment for acquired thrombotic thrombocytopenic purpura. *N Engl J Med*. 2019;380(4):335-346.
3. Sadler JE. Pathophysiology of thrombotic thrombocytopenic purpura. *Blood.* 2017;130(10):1181-1188.
4. Scully M, Cataland S, Coppo P, et al. Consensus on the standardization of terminology in thrombotic thrombocytopenic purpura and related thrombotic microantiopathies. *J Thromb Haemost*. 2017; 15(2):312-322.
5. Scully M, Hunt BJ, Benjamin S, et al. Guidelines on the diagnosis and management of thrombotic thrombocytopenic purpura and other thrombotic microangiopathies. *Br J Haematol*. 2012;158(3)323-335.
6. Westwood JP, Thomas M, Alwan F, et al. Rituximab prophylaxis to prevent thrombotic thrombocytopenic purpura relapse: outcome and evaluation of dosing regimens. *Blood Adv.* 2017; 1(15):1159-1166.