Pharmacy Policy Bulletin: J-0913 Cablivi (caplacizumab-yhdp) – Commercial and Healthcare Reform	
Number: J-0913	**Category:** Prior Authorization
Line(s) of Business: ☒ Commercial ☒ Healthcare Reform ☐ Medicare	**Benefit(s):** **Commercial**: **Prior Authorization (1.):** 1. Miscellaneous Specialty Drugs Injectable = Yes w/Prior Authorization **Healthcare Reform**: Not applicable
Region(s): ☒ All ☐ Delaware ☐ New York ☐ Pennsylvania ☐ West Virginia	**Additional Restriction(s):** None
Version: J-0913-008	**Original Date:** 05/02/2019
Effective Date: 02/14/2025	**Review Date:** 01/29/2025

Drugs Product(s):	• Cablivi (caplacizumab-yhdp)
FDA-Approved Indication(s):	• Treatment of adult patients with acquired thrombotic thrombocytopenic purpura (aTTP), in combination with plasma exchange and immunosuppressive therapy.

Background:	• Cablivi is a von Willebrand factor (vWF)-directed antibody fragment. It targets the A1-domain of vWF and inhibits the interaction between vWF and platelets, thereby reducing both vWF-mediated platelet adhesion and platelet consumption.
	• The ADAM metallopeptidase with thrombospondin type 1 motif 13 (ADAMTS13) gene provides instructions for making an enzyme that is involved in blood clotting. Acquired thrombotic thrombocytopenic purpura is a thrombotic microangiopathy caused by severely reduced activity of the von Willebrand factor-cleaving ADAMTS13. This causes extensive clot formation in small blood vessels throughout the body leading to thrombocytopenia, microangiopathic hemolytic anemia, and ischemia.
	• Signs of persistent disease may include purple bruises (purpura) or small red or purple spots on the skin (petechiae), hematuria, fever, chest pain, headache, and seizures.
	• The International Society on Thrombosis and Haemostasis guidelines from thrombotic thrombocytopenic purpura recommend treatment with Cablivi for an acute event as a conditional recommendation with a moderate certainty in the evidence.
	• Prescribing Considerations:
	  ○ Cablivi is indicated in addition to the regimen of plasma exchange and immunosuppressive drugs currently used to treat aTTP episodes.
	  ○ When initiated, the first dose of Cablivi is administered intravenously at least 15 minutes prior to plasma exchange, followed by a dose given subcutaneously upon completion of plasma exchange.

	○ Subsequent doses are administered subcutaneously daily after plasma exchange. ○ Cablivi is then administered once daily for 30 days following the last plasma exchange. If after the initial treatment course, sign(s) of persistent underlying disease such as suppressed ADAMTS13 activity levels remain present, treatment may be extended for a maximum of 28 days (for a total treatment duration of 58 days). ○ Discontinue Cablivi if the patient experiences more than 2 recurrences of aTTP while on Cablivi. ○ Withhold Cablivi 7 days prior to elective surgery, dental procedures or other invasive interventions.

Approval Criteria

I. Initial Authorization

When a benefit, coverage of Cablivi may be approved when all of the following criteria are met **(A. through E.)**:

A. The member is 18 years of age or older.

B. Cablivi is prescribed by or in consultation with a hematologist.

C. The member has a diagnosis of acquired thrombotic thrombocytopenic purpura (aTTP) (specifically immune TTP) (ICD-10: D69.3)

D. Cablivi was started in an inpatient setting in conjunction with both of the following **(1. and 2.)**:

1. Plasma exchange

2. Immunosuppressive therapy (specifically systemic corticosteroids or rituximab)

E. The total treatment duration will be limited to 58 days beyond the last therapeutic plasma exchange.

II. Reauthorization

When a benefit, reauthorization of Cablivi may be approved when all of the following criteria are met **(A. and B.)**:

A. The request is for a new episode requiring the re-initiation of plasma exchange for the treatment of aTTP.

B. The member has not experienced more than 2 recurrences of aTTP while on Cablivi after the initial course of therapy.

III. An exception to some or all of the criteria above may be granted for select members and/or circumstances based on state and/or federal regulations.

Limitations of Coverage

I. Coverage of drugs addressed in this policy for disease states outside of the FDA-approved indications should be denied based on the lack of clinical data to support effectiveness and safety in other conditions unless otherwise noted in the approval criteria.

II. For Commercial or HCR members with a closed formulary, a non-formulary product will only be approved if the member meets the criteria for a formulary exception in addition to the criteria outlined within this policy.

Authorization Duration

Initial Authorization

- Commercial and HCR Plans: If approved, up to a 60-day authorization following the last plasma exchange that the patient has received may be granted.

Reauthorization

- Commercial and HCR Plans: If approved, up to a 60-day reauthorization may be granted. A maximum of three (3) courses per lifetime may be approved.

Automatic Approval Criteria

None

References:

1. Cablivi [package insert]. Cambridge, MA: Genzyme Corporation. October 2024.
2. Zheng, XL, et al. ISTH guidelines for treatment of thrombotic thrombocytopenic purpura. *J Thromb Haemost*. 2020; 18: 2496– 2502.