Aetna Better Health® of New Jersey

Protocol for Cablivi® (caplacizumab-yhdp)
Updated July 2021
Approved October 2019

Addendum:
Addition of new diagnosis: thrombotic microangiopathy (TMA)

Background:
Cablivi is a von Willebrand factor (vWF)-directed antibody fragment indicated for the treatment of adult patients with acquired thrombotic thrombocytopenic purpura (aTTP), in combination with plasma exchange and immunosuppressive therapy. Acquired thrombotic thrombocytopenic purpura is also known as Moschcowitz disease and Microangiopathic hemolytic anemia.

Criteria for approval:

1. Patient is 18 years of age or older
2. Patient has a diagnosis of acquired thrombotic thrombocytopenic purpura (aTTP) confirmed by one of the following (a or b):
    a. Patient presents with thrombocytopenia AND Patient presents with microangiopathic hemolytic anemia (MAHA) *or thrombotic microangiopathy (TMA)* confirmed by red blood cell fragmentation (e.g., schistocytes) on peripheral blood smear; OR
    b. Patient's testing show ADAMTS13 activity levels of less than 10%
3. Medication is prescribed by or in consultation with a hematologist
4. Medication is prescribed to be given in combination with plasma exchange and immunosuppressive therapy (such as systemic glucorticosteroids, rituximab)
5. Medication will be discontinued if the patient experiences more than 2 recurrences of aTTP, while on treatment with Cablivi
6. If treatment needed beyond 30 days per episode: Additional therapy up to a maximum 28 additional days will be considered for approval if the prescriber submits documentation of remaining signs of persistent underlying disease (such as suppressed ADAMTS13 activity levels)
7. Total treatment duration per episode will be limited to 58 days beyond the last therapeutic plasma exchange
8. Medication is prescribed in accordance with Food and Drug Administration (FDA) established indication and dosing regimens or in accordance with medically appropriate off-label indication and dosing according to American Hospital Formulary Service, Micromedex, Clinical Pharmacology, or national guidelines.

Continuation of therapy:

1. Request is for a new (different) episode requiring the re-initiation of plasma exchange for the treatment of aTTP. (Documentation of date of prior episode and documentation date of new episode required)
2. Patient has not experienced more than 2 recurrences of aTTP, while on Cablivi
3. If treatment needed beyond 30 days per episode: Additional therapy up to a maximum 28 additional days will be considered for approval if the prescriber submits documentation of remaining signs of persistent underlying disease (such as suppressed ADAMTS13 activity levels)
4. Total treatment duration per episode will be limited to 58 days beyond the last therapeutic plasma exchange
5. Medication is prescribed in accordance with Food and Drug Administration (FDA) established indication and dosing regimens or in accordance with medically appropriate off-label indication and dosing according to American Hospital Formulary Service, Micromedex, Clinical Pharmacology, or national guidelines.
6. Medication is prescribed to be given in combination with plasma exchange and immunosuppressive therapy (such as systemic glucorticosteroids, rituximab)

References:

1. Cablivi [package insert]. Genzyme Corporation. Cambridge, MA 02142. February 2019
2. Clinical Pharmacology® Gold Standard Series [Internet database]. Tampa FL. Elsevier 2019. Updated periodically
3. George JN et al. Acquired TTP: Clinical manifestations and diagnosis. Post TW, ed. UpToDate. Waltham, MA: UpToDate Inc. https://www.uptodate.com (Accessed on May 17, 2021.)
4. National Heart, Lung, and Blood Institute. U.S. Department of Health & Human Services. Available at https://www.nhlbi.nih.gov/health-topics/thrombotic-thrombocytopenic-purpura. Accessed September 11, 2019
5. Scully M et al., Caplacizumab Treatment for Acquired Thrombotic Thrombocytopenic Purpura. NEJM. 2019;380:335- 346. Available at https://www.nejm.org/doi/10.1056/NEJMoa1806311. Accessed September 11, 2019.
6. Coppo P, Schwarzinger M, Buffet M, et al. Predictive features of severe acquired ADAMTS13 deficiency in idiopathic thrombotic microangiopathies: the French TMA Reference Center experience. PLoS One 2010;5(4):e10208-e10208.