PreferredOne®

PRIOR AUTHORIZATION POLICY

POLICY: Hematology – Cablivi Prior Authorization Policy
- Cablivi® (caplacizumab-yhdp for injection, for intravenous or subcutaneous use - Genzyme)

REVIEW DATE: 02/03/2021

Overview

Cablivi, a von Willebrand factor (vWF)-directed antibody fragment, is indicated for the treatment of adult patients with **acquired thrombotic thrombocytopenic purpura** (aTTP), in combination with plasma exchange and immunosuppressive therapy.[1] Cablivi is given once a day during plasma exchange and continued for 30 days after the last plasma exchange session. If, after the initial treatment course, there are signs of persistent underlying disease such as suppressed ADAMTS13 (A Disintegrin And Metalloproteinase with ThromboSpondin-1 motif, member 13) levels, Cablivi therapy may be extended for a maximum of 28 days. Cablivi should be discontinued if the patient experiences more than two recurrences of aTTP while on Cablivi.

Disease Overview

Thrombotic thrombocytopenic purpura (TTP) is a rare but potentially fatal blood disorder.[2-5] TTP may be caused by an inherited severe deficiency of plasma ADAMTS13 activity resulting from mutations; this is referred to as hereditary or congenital TTP. More commonly, TTP is acquired and due to autoantibodies that inhibit plasma ADAMTS13 activity, referred to as immune-mediated TTP (iTTP). Reduced ADAMTS13 activity leads to accumulation of ultra-large vWF multimers in the blood, which bind to platelets and lead to excessive platelet clumping in the microvasculature, resulting in multi-organ failure and death. Cablivi is a nanobody that targets the ultra-large vWF and inhibits the interaction between vWF and platelets, thereby preventing platelet adhesion.[1-3,6]

Guidelines/Recommendations

The standard of care for treatment aTTP is plasma exchange and glucocorticoids.[7] Plasma exchange removes the ultra-large vWF and autoantibodies and replenishes ADAMTS13, and immunosuppressants inhibit autoantibody formation.[2,6,7] Rituximab can also be added to the aTTP treatment regimen.[3] Rituximab has been shown to reduce the incidence of aTTP relapse by diminishing the production of anti-ADAMTS13 antibodies and restoring ADAMTS13 activity.[3,4]

The International Society on Thrombosis and Haemostasis (ISTH) formed a multidisciplinary panel including hematologists and pathologists with clinical expertise in the diagnosis and management of TTP, clinicians from other relevant disciplines, and patient representatives to issue recommendations about treatment of TTP (2020).[8] For patients with aTTP or iTTP experiencing an acute event (first event or relapse), the panel suggests using Cablivi over not using Cablivi. The panel stressed that Cablivi should only be given under the guidance of an experienced clinician; ideally, a TTP expert (e.g., a hematologist or pathologist specialized in transfusion medicine with previous experience in treating the disease).

POLICY STATEMENT

Prior Authorization is recommended for prescription benefit coverage of Cablivi. All approvals are provided for the duration noted below. Because of the specialized skills required for evaluation and diagnosis of patients treated with Cablivi as well as the monitoring required for adverse events and efficacy, approval requires Cablivi to be prescribed by or in consultation with a physician who specializes in the condition

being treated. Note that one course of treatment consists of Cablivi to be administered in conjunction with plasma exchange and Cablivi to be administered for up to 60 days (one dose per day) following the last plasma exchange session.

Automation: None.

Recommended Authorization Criteria

Coverage of Cablivi is recommended in those who meet the following criteria:

FDA-Approved Indications

1. **Acquired Thrombotic Thrombocytopenic Purpura (aTTP).** Approve for one course of treatment (up to 60 days following the last plasma exchange session) if the patient meets ALL of the following criteria (A, B, C, D, and E):
   - **A)** Patient is ≥ 18 years of age; AND
   - **B)** Cablivi was initiated in the inpatient setting, in combination with plasma exchange therapy; AND
   - **C)** Patient is currently receiving at least one immunosuppressive therapy; AND
     Note: Examples include systemic corticosteroids, rituximab (or a rituximab product), cyclosporine, cyclophosphamide, mycophenolate mofetil, hydroxychloroquine, Velcade® [bortezomib for injection]).
   - **D)** If the patient has previously received Cablivi, he/she has not had more than two recurrences of aTTP while on Cablivi; AND
   - **E)** The medication is prescribed by or in consultation with a hematologist.

CONDITIONS NOT RECOMMENDED FOR APPROVAL

Coverage of Cablivi is not recommended in the following situations:

1. Coverage is not recommended for circumstances not listed in the Recommended Authorization Criteria. Criteria will be updated as new published data are available.

REFERENCES

1. Cablivi® for injection [prescribing information]. Cambridge, MA: Genzyme Corporation; September 2020.
2. Duggan S. Caplacizumab: first global approval. *Drugs*. 2018;78:1639-1642.
3. Coppo P, Cuker A, George JN. Thrombotic thrombocytopenic purpura: toward targeted therapy and precision medicine. *Res Pract Thromb Haemost*. 2019;3:26-37.
4. Joly BS, Coppo P, Veyradier A. Thrombotic thrombocytopenic purpura. *Blood*. 2017;129:2836-2846.
5. Zheng XL, Vesely SK, Cataland SR, et al. International Society on Thrombosis and Haemostasis (ISTH) guidelines for the diagnosis of thrombotic thrombocytopenic purpura. *J Thromb Haemost*. 2020;18:2486-2495.
6. Scully M, Cataland SR, Peyvandi F, et al. Caplacizumab treatment for acquired thrombotic thrombocytopenic purpura. *N Engl J Med*. 2019;380:335-346.
7. 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;158:323-335.
8. Zheng XL, Vesely SK, Cataland SR, et al. International Society on Thrombosis and Haemostasis (ISTH) guidelines for the treatment of thrombotic thrombocytopenic purpura. *J Thromb Haemost*. 2020;18:2496-2502.

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HISTORY

Type of Revision	Summary of Changes	Review Date
New Policy	--	02/20/2019
Selected Revision	Revised criterion "The patient is currently receiving daily plasma exchange and at least one immunosuppressant therapy (e.g., corticosteroids with or without a rituximab product" to "The patient is currently receiving at least one immunosuppressive therapy (e.g., systemic corticosteroids, rituximab [or a rituximab product], cyclosporine [Neoral®, Sandimmune®, generics], cyclophosphamide, mycophenolate mofetil, [CellCept®, Myfortic®, generics], hydroxychloroquine, Velcade® [bortezomib for injection]).	02/27/2019

Annual Revision	The following changes were made:
	• **Policy Statement:** This section was revised from "All approvals are provided for the duration noted below" to "All approvals are provided for one course of therapy. Note that one course of therapy consists of Cablivi to be administered in conjunction with plasma exchange and Cablivi to be administered for a maximum of 60 days (one dose per day) following the last plasma exchange".
	• **Acquired Thrombotic Thrombocytopenic Purpura (aTTP):**
	  • The approval duration was changed from 3 months to "one course of treatment (up to 60 days following the last plasma exchange session).
	  • Two criteria were added: Cablivi was initiated in the inpatient setting in combination with plasma exchange therapy; and If the patient has previously received Cablivi therapy, he/she has not had more than two recurrences of aTTP while on Cablivi therapy.
	  • Examples of immunosuppressant therapies (of which at least one is required to be used in conjunction with Cablivi) were removed from the criteria and changed to a note.
		01/29/2020

Annual Revision	No criteria changes.	02/03/2021

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Fax: 763.847.4010
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U.S. Department of Health and Human Services
200 Independence Avenue, SW
Room 509F, HHH Building
Washington, D.C. 20201
1-800-368-1019, 800-537-7697 (TDD)

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