### Cablivi

#### Description

Cablivi (caplacizumab-yhdp)

#### Background

Cablivi (caplacizumab-yhdp) is a von Willebrand factor (vWF)-directed antibody fragment. Cablivi targets the A1-domain of vWF, and inhibits the interaction between vWF and platelets, thereby reducing both vWF-mediated platelet adhesion and platelet consumption (1).

#### Regulatory Status

FDA approved indication: Cablivi is indicated for the treatment of adult patients with acquired thrombotic thrombocytopenic purpura (aTTP), in combination with plasma exchange and immunosuppressive therapy (1).

The recommended dose of Cablivi is as follows: (1)

- First day of treatment: 11 mg bolus intravenous injection at least 15 minutes prior to plasma exchange followed by an 11 mg subcutaneous injection after completion of plasma exchange on day 1
- Subsequent treatment during daily plasma exchange: 11 mg subcutaneous injection once daily following plasma exchange
- Treatment after the plasma exchange period: 11 mg subcutaneous injection once daily for 30 days beyond the last plasma exchange
- If after 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

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<th>Section:</th>
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<td>July 1, 2019</td>
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<tr>
<td>Subsection:</td>
<td>Hematological Agents</td>
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<td>February 22, 2019</td>
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Last Review Date: June 20, 2019
Discontinue Cablivi if the patient experiences more than 2 recurrences of aTTP, while on Cablivi.

Cablivi increases the risk of bleeding. The risk of bleeding is increased in patients with underlying coagulopathies (e.g. hemophilia, other coagulation factor deficiencies). It is also increased with concomitant use of Cablivi with drugs affecting hemostasis and coagulation. Cablivi should be interrupted if clinically significant bleeding occurs. If needed, von Willebrand factor concentrate may be administered to rapidly correct hemostasis. If Cablivi is restarted, the patient should be monitored closely for signs of bleeding. Cablivi should be withheld for 7 days prior to elective surgery, dental procedures or other invasive interventions.

The safety and effectiveness of Cablivi in pediatric patients have not been established (1).

**Related policies**
IVIG, Nplate, Promacta, Rituxan, Tavalisse

**Policy**

*This policy statement applies to clinical review performed for pre-service (Prior Approval, Precertification, Advanced Benefit Determination, etc.) and/or post-service claims.*

Cablivi may be considered medically necessary in patients that are 18 years of age and older for the treatment of acquired thrombotic thrombocytopenic purpura (aTTP) and if the conditions indicated below are met.

Cablivi may be considered investigational in patients under 18 years of age and for all other indications.

**Prior-Approval Requirements**

**Age**
18 years of age or older

**Diagnosis**

Patient must have the following:

Acquired thrombotic thrombocytopenic purpura (aTTP)

AND ALL of the following:
1. Used in combination with plasma exchange and immunosuppressive therapy
2. Cablivi should be continued for 30 days following the last plasma exchange session
3. NO suspected thrombotic microangiopathies that were not associated with thrombotic thrombocytopenic purpura (TTP), such as hemolytic uremic syndrome
4. NO congenital TTP
5. Prescriber agrees to monitor for signs of bleeding
6. Prescriber agrees to discontinue therapy with Cablivi if the patient experiences more than 2 recurrences of aTTP, while on Cablivi

Prior–Approval Renewal Requirements
Same as above

Policy Guidelines

Pre–PA Allowance
None

Prior–Approval Limits

<table>
<thead>
<tr>
<th>Strength</th>
<th>Quantity</th>
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<tr>
<td>11 mg single-dose vials</td>
<td>60 vials</td>
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Duration 90 days

Prior–Approval Renewal Limits
Same as above

Rationale

Summary
Cablivi (caplacizumab-yhdp) is a von Willebrand factor (vWF)-directed antibody fragment. Cablivi targets the A1-domain of vWF, and inhibits the interaction between vWF and platelets, thereby reducing both vWF-mediated platelet adhesion and platelet consumption (1).

Prior approval is required to ensure the safe, clinically appropriate and cost effective use of Cablivi while maintaining optimal therapeutic outcomes.

References

<table>
<thead>
<tr>
<th>Date</th>
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<tbody>
<tr>
<td>February 2019</td>
<td>Addition to PA</td>
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<tr>
<td>June 2019</td>
<td>Annual review. Per SME, addition of: Cablivi should be continued for 30 days following the last plasma exchange session; no suspected thrombotic microangiopathies not associated with TTP; no congenital TTP</td>
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Keywords
This policy was approved by the FEP® Pharmacy and Medical Policy Committee on June 20, 2019 and is effective on July 1, 2019.