BILLING & CODING GUIDE*

This document is intended as a general guide for submitting information to payers for reimbursement. Use of this guide does not guarantee that the payer will provide coverage for CABLIVI and is not intended to be a substitute for, or an influence on, the independent medical judgment of the prescriber. Prescribers should follow payer-specific coding requirements and exercise clinical judgment when selecting codes and submitting claims to truthfully and accurately reflect the services and products furnished to a specific patient.

The coding information discussed in this guide is provided for informational purposes only, is subject to change, and should not be construed as legal advice. The codes listed herein may not apply to all patients or to all health plans. Conversely, additional codes not listed in this guide may apply to some patients.

*This information is subject to change, and providers should consult relevant references for the description of each code to determine its appropriateness.

INDICATIONS AND SELECTED IMPORTANT SAFETY INFORMATION

INDICATIONS:
CABLIVI (caplacizumab-yhdp) is indicated for the treatment of adult patients with acquired thrombotic thrombocytopenic purpura (aTTP), in combination with plasma exchange and immunosuppressive therapy.

CONTRAINDICATIONS:
CABLIVI is contraindicated in patients with a previous severe hypersensitivity reaction to caplacizumab-yhdp or to any of its excipients. Hypersensitivity reactions have included urticaria.

Please see full Prescribing Information.

Cablivi®
caplacizumab-yhdp
Injection 11 mg

Coverage

In some instances, Cablivi may be bundled into inpatient payment rates when administered in a hospital. The 3 MS-DRGs that represent the greatest number of potential patients who may be eligible for treatment involving CABLIVI are shown in the table below. This table may not be reflective of all MS-DRG codes that may be used for CABLIVI. The MS-DRG code is determined by the payer based on the primary diagnosis.

Potential MS-DRGs[1]

545	Connective Tissue Disorders with MCC
546	Connective Tissue Disorders with CC
547	Connective Tissue Disorders without CC/MCC

The New Technology Add-on Payment (NTAP) for CABLIVI expired permanently on October 1, 2022, which means Medicare will no longer provide NTAP add-on payments toward costs that exceed the standard DRG payment.

Institutions should be aware that CMS is recalculating net care costs associated with the administration of CABLIVI based on three years of average cost data, which may offset the cost impact on hospitals treating Medicare Fee-for-Service patients with this medication.

Other Reimbursement Considerations

The specifics of coverage may vary by payer. Please refer to the individual patient's plan to determine any applicable coverage requirements.

For patients covered by Medicare, drug costs for doses administered in the hospital are typically included in the MS-DRG payment and are covered under Medicare Part A. After inpatient discharge, most patients will self-administer CABLIVI at home; these drug costs are expected to be covered under the Medicare Part D (pharmacy) benefit.

CC, complication or comorbidity; DRG, diagnosis-related group; MCC, major complication or comorbidity; MS-DRG, Medicare Severity Diagnosis Related Group.

SELECTED IMPORTANT SAFETY INFORMATION (cont'd)

WARNINGS AND PRECAUTIONS:

Hemorrhage:

- CABLIVI increases the risk of bleeding. In clinical studies, severe bleeding adverse reactions of epistaxis, gingival bleeding, upper gastrointestinal hemorrhage, and metrorrhagia were each reported in 1% of subjects. Overall, bleeding events occurred in approximately 58% of patients on CABLIVI versus 43% of patients on placebo.
- In the postmarketing setting cases of life-threatening and fatal bleeding were reported in patients receiving CABLIVI.
- 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.
- Avoid concomitant use of CABLIVI with antiplatelet agents or anticoagulants. If clinically significant bleeding occurs, interrupt use of CABLIVI. Von Willebrand factor concentrate may be administered to rapidly correct hemostasis. If CABLIVI is restarted, monitor closely for signs of bleeding.
- Withhold CABLIVI for 7 days prior to elective surgery, dental procedures or other invasive interventions. If emergency surgery is needed, the use of von Willebrand factor concentrate may be considered to correct hemostasis. After the risk of surgical bleeding has resolved, and CABLIVI is resumed, monitor closely for signs of bleeding.

ADVERSE REACTIONS:

The most common adverse reactions (>15% of patients) were epistaxis (29%), headache (21%) and gingival bleeding (16%).

Diagnosis and Procedure Codes

Although there is no specific ICD-10 diagnosis code for aTTP, healthcare providers (HCPs) may use the code for thrombotic microangiopathy when submitting claims. However, there are three specific ICD-10 procedure codes available for the administration of caplacizumab. In addition, there are three codes that are not specific to caplacizumab that may be used by some hospitals. HCPs should always review payer-specific material and use their own clinical judgment when submitting claims for use of CABLIVI.

ICD-10-CM Code[2]	
M31.1	Thrombotic Microangiopathy*
ICD-10-PCS Codes[3-8]	
XW013W5 -or- 3E013GC	Introduction of Caplacizumab into Subcutaneous Tissue, Percutaneous Approach, New Technology Group 5 Introduction of Other Therapeutic Substance into Subcutaneous Tissue, Percutaneous Approach
XW033W5 -or- 3E033GC	Introduction of Caplacizumab into Peripheral Vein, Percutaneous Approach, New Technology Group 5 Introduction of Other Therapeutic Substance into Peripheral Vein, Percutaneous Approach
XW043W5 -or- 3E043GC	Introduction of Caplacizumab into Central Vein, Percutaneous Approach, New Technology Group 5 Introduction of Other Therapeutic Substance into Central Vein, Percutaneous Approach

*Thrombotic Microangiopathy has the inclusion note of thrombotic thrombocytopenic purpura, which would include aTTP.

Product Information[9]

How supplied	Lyophilized powder in single-use vials containing 11 mg per vial
Quantity and units per case	1 vial per kit
NDC	NDC 58468-0225-1

ICD-10-CM, international classification of diseases, 10th revision, clinical modification; ICD-10-PCS, international classification of diseases, 10th revision, procedural coding system; NDC, national drug code.

SELECTED IMPORTANT SAFETY INFORMATION (cont'd)

CONCOMITANT USE OF ANTICOAGULANTS AND ANTIPLATELET AGENTS:
Concomitant use of CABLIVI with any anticoagulant or antiplatelet agent may increase the risk of bleeding. Avoid concomitant use when possible. Assess and monitor closely for bleeding with concomitant use.

PREGNANCY:
There are no available data on CABLIVI use in pregnant women to inform a drug associated risk of major birth defects and miscarriage.

- **Fetal/neonatal adverse reactions:** CABLIVI may increase the risk of bleeding in the fetus and neonate. Monitor neonates for bleeding.
- **Maternal adverse reactions:** All patients receiving CABLIVI, including pregnant women, are at risk for bleeding. Pregnant women receiving CABLIVI should be carefully monitored for evidence of excessive bleeding.

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References: 1. Centers for Medicare & Medicaid Services. Draft ICD-10-CM/PCS MS-DRGv28 Definitions Manual: MDC 8 Diseases & Disorders of the Musculoskeletal System & Connective Tissue Disorders. https://www.cms.gov/icd10m/version37-fullcode-cms/fullcode_cms/P0011.html. Accessed March 7, 2021. **2.** Centers for Disease Control and Prevention. ICD-10-CM tabular list of diseases and injuries. Accessed October 20, 2022. https://ftp.cdc.gov/pub/Health_Statistics/NCHS/Publications/ICD10CM/2022/icd10cm-tabular-2022-April-1.pdf#:~:text=ICD-10-CM%20TABULAR%20LIST%20of%20DISEASES%20and%20INJURIES%20Table,%28D50-D89%29%204%20Endocrine%2C%20nutritional%20and%20metabolic%-20diseases%20%28E00-E89%29 **3.** ICD10data. 2023 ICD-10-PCS Procedure Code XW013W5. Accessed October 20, 2022. https://www.icd10data.com/ICD10PCS/Codes/X/W/0/1/XW013W5 **4.** ICD10data. 2023 ICD-10-PCS Procedure Code 3E013GC. Accessed October 20, 2022. https://www.icd10data.com/ICD10PCS/Codes/3/E/0/1/3E013GC **5.** ICD10data. 2023 ICD-10-PCS Procedure Code XW033W5. Accessed October 20,2022. https://www.icd10data.com/ICD10PCS/Codes/X/W/0/3/XW033W5 **6.** ICD10data. 2023 ICD-10-PCS Procedure Code 3E033GC. Accessed October 20, 2022. https://www.icd10data.com/ICD10PCS/Codes/3/E/0/3/3E033GC **7.** ICD10data. 2023 ICD-10-PCS Procedure Code XW043W5. Accessed October 20, 2022. https://www.icd10data.com/ICD10PCS/Codes/X/W/0/4/XW043W5 **8.** ICD10data. 2023 ICD-10-PCS Procedure Code 3E043GC. Accessed October 20, 2022. **9.** CABLIVI [package insert]. Cambridge, MA: Genzyme corporation.

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