HEMOPHILIA CLOTTING FACTORS

Guideline Number: MPG130.04  Approval Date: September 12, 2018

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POLICY SUMMARY

Overview
Hemophilia is a hereditary blood disease characterized by greatly prolonged coagulation time. The blood fails to clot and abnormal bleeding occurs. It is a sex-linked hereditary trait transmitted by normal heterozygous females who carry the recessive gene. It occurs almost exclusively in males. For purposes of Medicare coverage, hemophilia encompasses Factor VIII deficiency (classic hemophilia, hemophilia A), Factor IX deficiency (hemophilia B, Christmas disease, plasma thromboplastin component), and von Willebrand’s disease. Approximately 80% of those with hemophilia have type A, and both are associated with recurrent, spontaneous, and traumatic hemarthrosis.

The frequency and severity of hemorrhagic events induced by hemophilia are related to the amount of coagulation factor in the blood. Those with mild hemophilia (defined as having from 5% to 40% of normal coagulation factor activity) experience complications only after having undergone surgery or experiencing a major physical trauma. Those with moderate hemophilia (from 1% to 5% of coagulation factor activity) experience some spontaneous hemorrhage but normally exhibit bleeding provoked by trauma. Those with severe hemophilia (less than 1% of coagulation factor activity) exhibit spontaneous hemorrhosis and bleeding. Treatment for these patients is dependent on the severity of the disease and may include the administration of blood clotting factors such as Factor VIII, Factor IX, Factor VIIa and, Anti-inhibitors to control the bleeding.

Guidelines
Medicare provides coverage of self-administered blood clotting factors for hemophilia patients who are competent to use such factors to control bleeding without medical supervision. Medicare covers blood clotting factors for the following conditions:

- Factor VIII deficiency (classic hemophilia, hemophilia A)
- Factor IX deficiency (hemophilia B, Christmas disease, plasma thromboplastin component)
- Von Willebrand’s disease
- Acquired hemophilia (acquired Factor VIII autoantibodies most frequently) and other coagulation factor deficiencies, intrinsic circulating anticoagulants, antibodies or inhibitors.
- Congenital deficiencies of other clotting factors (such as congenital afibrinogenemia and others).

APPLICABLE CODES

The following list(s) of codes is provided for reference purposes only and may not be all inclusive. Listing of a code in this guideline does not imply that the service described by the code is a covered or non-covered health service. Benefit coverage for health services is determined by the member specific benefit plan document and applicable laws that may require coverage for a specific service. The inclusion of a code does not imply any right to reimbursement or guarantee claim payment. Other Policies and Guidelines may apply.

<table>
<thead>
<tr>
<th>HCPCS Code</th>
<th>Description</th>
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<tbody>
<tr>
<td>C9140</td>
<td>Injection, factor VIII (antihemophilic factor, recombinant) (Afstyla), 1 IU (Deleted 12/31/2017)</td>
</tr>
<tr>
<td>HCPCS Code</td>
<td>Description</td>
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<td>------------</td>
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</tr>
<tr>
<td>J7175</td>
<td>Injection, factor X, (human), 1 IU</td>
</tr>
<tr>
<td>J7179</td>
<td>Injection, von Willebrand factor (recombinant), (Vonvendi), 1 IU VWF:RCo</td>
</tr>
<tr>
<td>J7180</td>
<td>Injection, factor XIII (antihemophilic factor, human), 1 IU</td>
</tr>
<tr>
<td>J7181</td>
<td>Injection, factor XIII A-subunit, (recombinant), per IU</td>
</tr>
<tr>
<td>J7182</td>
<td>Injection, factor VIII, (antihemophilic factor, recombinant), (NovoEight), per IU</td>
</tr>
<tr>
<td>J7183</td>
<td>Injection, von Willebrand factor complex (human), Wilate, 1 IU VWF:RCo</td>
</tr>
<tr>
<td>J7185</td>
<td>Injection, factor VIII (antihemophilic factor, recombinant) (Xyntha®), per IU</td>
</tr>
<tr>
<td>J7186</td>
<td>Injection, antihemophilic factor VIII/von Willebrand factor complex (human), per factor VIII i.u.</td>
</tr>
<tr>
<td>J7187</td>
<td>Injection, von Willebrand factor complex (Humate-P®), per IU VWF:RCO</td>
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<tr>
<td>J7188</td>
<td>Injection, factor VIII (antihemophilic factor, recombinant), per IU</td>
</tr>
<tr>
<td>J7189</td>
<td>Factor VIII (antihemophilic factor, recombinant), per 1 mcg</td>
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<tr>
<td>J7190</td>
<td>Factor VIII (antihemophilic factor, human), per IU</td>
</tr>
<tr>
<td>J7191</td>
<td>Factor VIII (antihemophilic factor (porcine), per IU</td>
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<tr>
<td>J7192</td>
<td>Factor VIII (antihemophilic factor, recombinant), per IU, not otherwise specified</td>
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<tr>
<td>J7193</td>
<td>Factor IX (antihemophilic factor, purified, nonrecombinant), per IU</td>
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<tr>
<td>J7194</td>
<td>Factor IX complex, per IU</td>
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<tr>
<td>J7195</td>
<td>Factor IX (antihemophilic factor, recombinant), per IU</td>
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<tr>
<td>J7198</td>
<td>Antiinhibitor, per IU [See the Medicare Advantage Policy Guideline titled Anti-Inhibitor Coagulant Complex (AICC) (NCD 110.3)]</td>
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<td>J7199</td>
<td>Hemophilia clotting factor, not otherwise classified</td>
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<tr>
<td>J7200</td>
<td>Injection, factor IX, (Antihemophilic Factor, Recombinant), Rixubis®, Per IU</td>
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<tr>
<td>J7201</td>
<td>Injection, factor IX, FC Fusion Protein (Recombinant), per IU</td>
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<td>J7202</td>
<td>Injection, factor IX, albumin fusion protein, (recombinant), Idelvion®, 1 IU</td>
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<td>J7205</td>
<td>Injection, factor VIII Fc fusion protein (recombinant), per IU</td>
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<td>J7207</td>
<td>Injection, factor VIII, (antihemophilic factor, recombinant), PEGylated, 1 IU</td>
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<td>J7209</td>
<td>Injection, factor VIII, (antihemophilic factor, recombinant), (Nuwiq®), 1 IU</td>
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<td>Injection, Factor VIII, (antihemophilic factor, recombinant), (Afstyla®), 1 IU (Effective 01/01/2018)</td>
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<td>J7211</td>
<td>Injection, Factor VIII, (antihemophilic factor, recombinant), (Kovaltry®), 1 IU (Effective 01/01/2018)</td>
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<table>
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<tr>
<th>ICD-10 Diagnosis Code</th>
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<tr>
<td><strong>For HCPCS Code J7179</strong></td>
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</tr>
<tr>
<td>D66</td>
<td>Hereditary factor VIII deficiency</td>
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<tr>
<td>D68.0</td>
<td>Von Willebrand's disease</td>
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<td><strong>For HCPCS Codes J7175, J7180, and J7181</strong></td>
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<td>Von Willebrand's disease</td>
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<td>D68.1</td>
<td>Hereditary factor XI deficiency</td>
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<td>D68.2</td>
<td>Hereditary deficiency of other clotting factors</td>
</tr>
<tr>
<td>D68.311</td>
<td>Acquired hemophilia</td>
</tr>
<tr>
<td>D68.312</td>
<td>Antiphosholipid antibody with hemorrhagic disorder</td>
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<tr>
<td>D68.318</td>
<td>Other hemorrhagic disorder due to intrinsic circulating anticoagulants, antibodies, or inhibitors</td>
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<tr>
<td>D68.4</td>
<td>Acquired coagulation factor deficiency</td>
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Hemophilia Clotting Factors
UnitedHealthcare Medicare Advantage Policy Guideline

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<td>Hereditary factor VIII deficiency</td>
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<td>D69.1</td>
<td>Qualitative platelet defects</td>
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<td><strong>HCPCS Codes J7201 and J7202</strong></td>
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<td>D67</td>
<td>Hereditary factor IX deficiency</td>
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**PURPOSE**

The Medicare Advantage Policy Guideline documents are generally used to support UnitedHealthcare Medicare Advantage claims processing activities and facilitate providers’ submission of accurate claims for the specified services. The document can be used as a guide to help determine applicable:

- Medicare coding or billing requirements, and/or
- Medical necessity coverage guidelines; including documentation requirements.

UnitedHealthcare follows Medicare guidelines such as LCDs, NCDs, and other Medicare manuals for the purposes of determining coverage. It is expected providers retain or have access to appropriate documentation when requested to support coverage. Please utilize the links in the References section below to view the Medicare source materials used to develop this resource document. This document is not a replacement for the Medicare source materials that outline Medicare coverage requirements. Where there is a conflict between this document and Medicare source materials, the Medicare source materials will apply.

**REFERENCES**

**CMS National Coverage Determinations (NCDs)**

NCD 110.3 Anti-Inhibitor Coagulant Complex (AICC)

**CMS Local Coverage Determinations (LCDs)**

<table>
<thead>
<tr>
<th>LCD</th>
<th>Medicare Part A</th>
<th>Medicare Part B</th>
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<tr>
<td>L33684 (Hemophilia Clotting Factors) First Coast</td>
<td>FL, PR, VI</td>
<td>FL, PR, VI</td>
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<td>L35111 (Hemophilia Factor Products) Novitas</td>
<td>AR, CO, DC, DE, LA, MD, MS, NJ, NM, OK, PA, TX</td>
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**CMS Articles**

<table>
<thead>
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<th>Article</th>
<th>Medicare Part A</th>
<th>Medicare Part B</th>
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<tr>
<td>A55016 (Hemophilia clotting factors revision to the Part A and Part B LCD) First Coast</td>
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<tr>
<td>A55153 (Hemophilia clotting factors revision to LCD) First Coast</td>
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## Article

### Medicare Part A

A56065 (Billing and Coding Guidance for Anti-Inhibitor Coagulant Complex (AICC) National Coverage Determination (NCD) 110.3) Palmetto

AL, GA, NC, SC, TN, VA, W.VA

### Medicare Part B

AL, GA, NC, SC, TN, VA, W.VA

## CMS Benefit Policy Manual

Chapter 15; § 50 Drugs and Biologics

## CMS Claims Processing Manual

Chapter 17; § 40 Discarded Drugs and Biologicals, § 80.4-80.4.1 Billing for Hemophilia Clotting Factors/Clotting Factor Furnishing Fee, § 90.2 Drugs, Biologicals, and Radiopharmaceuticals

## MLN Matters

Article MM9759, Annual Clotting Factor Furnishing Fee Update 2017

Article MM9930, January 2017 Update of the Hospital Outpatient Prospective Payment System

Article MM10254, Annual Clotting Factor Furnishing Fee Update 2018

Article MM10918, Annual Clotting Factor Furnishing Fee Update 2019

## UnitedHealthcare Commercial Policies

Clotting Factors and Coagulant Blood Products

## Others

CMS ASP Drug Pricing Files

Medicare Annual Blood Clotting Factor Furnishing Fee Schedules

Hemophilia Clotting Factors: Submitting the Number of Units Palmetto GBA Jurisdiction 11 Part B, Palmetto Website

Hemophilia, U.S. Department of Health and Human Services: National Institutes of Health, NIH Website

Medicare Prescription Drug Benefit Manual, Chapter 6 Appendix C: Part B Drugs and Part D Coverage Chart

## GUIDELINE HISTORY/REVISION INFORMATION

Revisions to this summary document do not in any way modify the requirement that services be provided and documented in accordance with the Medicare guidelines in effect on the date of service in question.

<table>
<thead>
<tr>
<th>Date</th>
<th>Action/Description</th>
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<tbody>
<tr>
<td>04/15/2019</td>
<td>• Reorganized policy template; relocated Terms and Conditions and Purpose section</td>
</tr>
<tr>
<td></td>
<td>• Reformatted list of applicable ICD-10 diagnosis codes</td>
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<tr>
<td>09/12/2018</td>
<td>• Annual review for MAPG Committee presentation and approval</td>
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</table>

## TERMS AND CONDITIONS

The Medicare Advantage Policy Guidelines are applicable to UnitedHealthcare Medicare Advantage Plans offered by UnitedHealthcare and its affiliates.

These Policy Guidelines are provided for informational purposes, and do not constitute medical advice. Treating physicians and healthcare providers are solely responsible for determining what care to provide to their patients. Members should always consult their physician before making any decisions about medical care.

Benefit coverage for health services is determined by the member specific benefit plan document* and applicable laws that may require coverage for a specific service. The member specific benefit plan document identifies which services are covered, which are excluded, and which are subject to limitations. In the event of a conflict, the member specific benefit plan document supersedes the Medicare Advantage Policy Guidelines.

Medicare Advantage Policy Guidelines are developed as needed, are regularly reviewed and updated, and are subject to change. They represent a portion of the resources used to support UnitedHealthcare coverage decision making. UnitedHealthcare may modify these Policy Guidelines at any time by publishing a new version of the policy on this website. Medicare source materials used to develop these guidelines include, but are not limited to, CMS National Coverage Determinations (NCDs), Local Coverage Determinations (LCDs), Medicare Benefit Policy Manual, Medicare Claims Processing Manual, Medicare Program Integrity Manual, Medicare Managed Care Manual, etc. The information presented in the Medicare Advantage Policy Guidelines is believed to be accurate and current as of the date of publication, and is provided on an "AS IS" basis. Where there is a conflict between this document and Medicare source materials, the Medicare source materials will apply.
You are responsible for submission of accurate claims. Medicare Advantage Policy Guidelines are intended to ensure that coverage decisions are made accurately based on the code or codes that correctly describe the health care services provided. UnitedHealthcare Medicare Advantage Policy Guidelines use Current Procedural Terminology (CPT®), Centers for Medicare and Medicaid Services (CMS), or other coding guidelines. References to CPT® or other sources are for definitional purposes only and do not imply any right to reimbursement or guarantee claims payment.

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*For more information on a specific member's benefit coverage, please call the customer service number on the back of the member ID card or refer to the Administrative Guide.