Hemophilia Clotting Factors and Products

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Terms and Conditions

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Policy Summary

See Purpose

Overview

Hemophilia is a blood disease characterized by greatly prolonged coagulation time. The blood fails to clot and abnormal bleeding occurs. Hemophilia is usually inherited. 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.

In rare cases, hemophilia can develop after birth, which is called acquired hemophilia caused by the development of antibodies (immune system proteins) directed against the body's own VIII or IX blood clotting factors. Unlike inherited hemophilia, acquired hemophilia A is not a genetic disorder and affects both males and females. The development of acquired hemophilia A has been related to other medical conditions or health states, such as pregnancy, cancer, or the use of certain medications. However, in about half of the cases, no underlying cause can be found.

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 hemarthrosis 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).
- Congenital factor XI deficiency (Hemophilia C).
- 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).

Anti-Inhibitor Coagulant Complex (AICC)

Per CMS NCD 110.3, Anti-inhibitor coagulant complex, AICC, is a drug used to treat hemophilia in patients with factor VIII inhibitor antibodies. AICC has been shown to be safe and effective and has Medicare coverage when furnished to patients with hemophilia A and inhibitor antibodies to factor VIII who have major bleeding episodes and who fail to respond to other, less expensive therapies.

Applicable Codes

The following list(s) of procedure and/or diagnosis 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.

HCPCS Code	Description
J7170	Injection, emicizumab-kxwh, 0.5 mg
J7175	Injection, factor X, (human), 1 IU
J7179	Injection, von Willebrand factor (recombinant), (Vonvendi), 1 IU VWF:RCo
J7180	Injection, factor XIII (antihemophilic factor, human), 1 IU
J7181	Injection, factor XIII A-subunit, (recombinant), per IU
J7182	Injection, factor VIII, (antihemophilic factor, recombinant), (NovoEight), per IU
J7183	Injection, von Willebrand factor complex (human), Wilate, 1 IU vWF:RCo
J7185	Injection, factor VIII (antihemophilic factor, recombinant) (Xyntha®), per IU
J7186	Injection, antihemophilic factor VIII/von Willebrand factor complex (human), per factor VIII IU
J7187	Injection, von Willebrand factor complex (Humate-P®), per IU VWF:RCO
J7188	Injection, factor VIII (antihemophilic factor, recombinant), (Obizur), per IU
J7189	Factor VIIa (antihemophilic factor, recombinant), (NovoSeven RT), 1 mcg
J7190	Factor VIII (antihemophilic factor, human), per IU
J7191	Factor VIII (antihemophilic factor (porcine)), per IU
J7192	Factor VIII (antihemophilic factor, recombinant), per IU, not otherwise specified
J7193	Factor IX (antihemophilic factor, purified, non-recombinant), per IU
J7194	Factor IX, complex, per IU
J7195	Injection, factor IX (antihemophilic factor, recombinant) per IU, not otherwise specified
J7198	Anti-inhibitor, per IU
J7199	Hemophilia clotting factor, not otherwise classified
J7200	Injection, factor IX, (antihemophilic factor, recombinant), Rixubis®, per IU
J7201	Injection, factor IX, FC fusion protein (recombinant), Alprolix, 1 IU
J7202	Injection, factor IX, albumin fusion protein, (recombinant), Idelvion®, 1 IU
J7203	Injection factor IX, (antihemophilic factor, recombinant), glycopegylated, (Rebinyn), 1 IU
J7204	Injection, Factor VIII, antihemophilic factor (recombinant), (Esperoct), glycopegylated-exei, per IU
J7205	Injection, factor VIII Fc fusion protein (recombinant), per IU

HCPCS Code	Description
J7207	Injection, factor VIII, (antihemophilic factor, recombinant), PEGylated, 1 IU
J7208	Injection, factor VIII, (antihemophilic factor, recombinant), PEGylated-aucl, (Jivi), 1 IU
J7209	Injection, factor VIII, (antihemophilic factor, recombinant), (Nuwiq®), 1 IU
J7210	Injection, factor VIII, (antihemophilic factor, recombinant), (Afstyla®), 1 IU
J7211	Injection, factor VIII, (antihemophilic factor, recombinant), (Kovaltry®), 1 IU
J7212	Factor VIIa (antihemophilic factor, recombinant)-jncw (Sevenfact), 1 mcg
J7213	Injection, coagulation factor IX (recombinant), Ixinity, 1 IU (Effective 07/01/2023)
J7214	Injection, Factor VIII/von Willebrand factor complex, recombinant (Altuviiio), per Factor VIII IU (Effective 10/01/2023)

Diagnosis Code	Description
For HCPCS Code J7170	
D66	Hereditary factor VIII deficiency
D68.318	Other hemorrhagic disorder due to intrinsic circulating anticoagulants, antibodies, or inhibitors
For HCPCS Codes J7179, J7183, J7186, and J7187	
D66	Hereditary factor VIII deficiency
D68.0	Von Willebrand's disease (Deleted 09/30/2022)
D68.01	Von Willebrand disease, type 1 (Effective 10/01/2022)
D68.020	Von Willebrand disease, type 2A (Effective 10/01/2022)
D68.021	Von Willebrand disease, type 2B (Effective 10/01/2022)
D68.022	Von Willebrand disease, type 2M (Effective 10/01/2022)
D68.023	Von Willebrand disease, type 2N (Effective 10/01/2022)
D68.03	Von Willebrand disease, type 3 (Effective 10/01/2022)
D68.04	Acquired von Willebrand disease (Effective 10/01/2022)
D68.09	Other von Willebrand disease (Effective 10/01/2022)
For HCPCS Codes J7175, J7180, and J7181	
D68.2	Hereditary deficiency of other clotting factors
For HCPCS Codes J7182, J7185, J7190, J7192, J7204, J7205, J7207, J7208, J7209, J7210, and J7211	
D66	Hereditary factor VIII deficiency
For HCPCS Codes J7188, J7189, and J7191	
D66	Hereditary factor VIII deficiency
D67	Hereditary factor IX deficiency
D68.2	Hereditary deficiency of other clotting factors
D68.311	Acquired hemophilia
D68.318	Other hemorrhagic disorder due to intrinsic circulating anticoagulants, antibodies, or inhibitors
D69.1	Qualitative platelet defects
For HCPCS Code J7198	
D66	Hereditary factor VIII deficiency
D67	Hereditary factor IX deficiency
D68.318	Other hemorrhagic disorder due to intrinsic circulating anticoagulants, antibodies, or inhibitors
For HCPCS Codes J7193, J7194, J7195, J7200, J7201, J7202, and J7203	
D67	Hereditary factor IX deficiency

Diagnosis Code	Description
For HCPCS Code J7212	
D66	Hereditary factor VIII deficiency
D67	Hereditary factor IX deficiency
D68.2	Hereditary deficiency of other clotting factors
For HCPCS Code J7212	
D68.318	Other hemorrhagic disorder due to intrinsic circulating anticoagulants, antibodies, or inhibitors

References

CMS National Coverage Determinations (NCDs)

NCD 110.3 Anti-Inhibitor Coagulant Complex (AICC)

CMS Local Coverage Determinations (LCDs) and Articles

LCD	Article	Contractor	Medicare Part A	Medicare Part B
N/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, WV
L33684 Hemophilia Clotting Factors **Retired 10/01/2022**	A56482 Billing and Coding: Hemophilia Factor Products	First Coast	FL, PR, VI	FL, PR, VI
L35111 Hemophilia Factor Products **Retired 10/01/2022**	A56433 Billing and Coding: Hemophilia Factor Products	Novitas	AR, CO, DC, DE, LA, MD, MS, NJ, NM, OK, PA, TX	AR, CO, DC, DE, LA, MD, MS, NJ, NM, OK, PA, TX
N/A	A56119 Billing and Coding: Billing Limitations for Pharmacies	Noridian		AS, CA, GU, HI, MP, NV
N/A	A56124 Billing and Coding: Billing Limitations for Pharmacies	Noridian		AK, AZ, ID, MT, ND, OR, SD, UT, WA, WY

CMS Benefit Policy Manual

Chapter 6; § 30 Drugs and Biologicals
Chapter 15; § 50.5.5 Hemophilia Clotting Factors

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

Other(s)

National Government Services, Factor VIII Billing
Noridian Healthcare Solutions Jurisdiction E Part B, Hemophilia Clotting Factor Billing
Noridian Healthcare Solutions Jurisdiction F Part B, Hemophilia Clotting Factor Billing
Palmetto GBA Jurisdiction J, Part B, Hemophilia Clotting Factors: Submitting the Number of Units
Palmetto GBA Jurisdiction M, Part B, Hemophilia Clotting Factors: Submitting the Number of Units

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.

Date	Summary of Changes
11/08/2023	**Applicable Codes** • Added HCPCS codes J7213 and J7214 **Supporting Information** • Updated *References* section to reflect the most current information • Archived previous policy version MPG130.13

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 NCDs, LCDs, LCAs, 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 above 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.

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.