Hemophilia Infusion Therapy

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Related Medical Guideline
**Off-Label Use of FDA-Approved Drugs and Biologicals**

Dosage and Administration
Refer to drug-specific labeling

Applicable Products

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<td>Criteria II</td>
<td>Alphanate, Humate-P</td>
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### Definitions

<table>
<thead>
<tr>
<th>Condition</th>
<th>Description</th>
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<tr>
<td><strong>Hemophilia A</strong></td>
<td>Genetic disorder caused by missing or defective factor VIII, a clotting protein.</td>
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<tr>
<td>AKA factor VIII (FVIII) deficiency or classic hemophilia</td>
<td></td>
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<tr>
<td><strong>Hemophilia B</strong></td>
<td>Genetic disorder caused by missing or defective factor IX, a clotting protein.</td>
</tr>
<tr>
<td>AKA factor IX (FIX) deficiency or Christmas disease</td>
<td></td>
</tr>
<tr>
<td><strong>Congenital factor VII deficiency</strong></td>
<td>FXII deficiency is inherited in an autosomal recessive fashion, meaning both parents must carry the gene to pass it on to their children</td>
</tr>
<tr>
<td>AKA Hageman Factor</td>
<td></td>
</tr>
<tr>
<td><strong>Glanzmann Thrombasthenia</strong></td>
<td>Genetic disorder in which the platelets have qualitative or quantitative deficiencies of the fibrinogen receptor αIIbβ3</td>
</tr>
<tr>
<td><strong>Von Willebrand disease (VWD)</strong></td>
<td>Genetic disorder caused by missing or defective von Willebrand factor (VWF), a clotting protein. VWF binds factor VIII, a key clotting protein, and platelets in blood vessel walls, which help form a platelet plug during the clotting process.</td>
</tr>
</tbody>
</table>

### Guideline

Hemophilia medications are considered medically necessary when the following product-specific criteria are met:

**Criteria I — AlphaNine SD, Alprolix, Bebulin, BeneFIX, Profilnine SD, Mononine, Rixubis, and IXINITY**

1. **Hemophilia B**
   
   a. Congenital factor IX deficiency confirmed by blood coagulation testing; AND
   
   b. Used to treat at least 1:
      
      i. Control and prevention of acute bleeding episodes (hemorrhage)
      
      ii. Routine prophylaxis to prevent or reduce frequency of bleeding episodes
      
      iii. Perioperative management; AND
   
   c. Not used for induction of immune tolerance in members with Hemophilia (applicable to Alprolix, Rixubis and IXINITY only)

**Criteria II — Alphanate, Humate-P**

1. **Hemophilia A**
   
   a. Congenital factor VIII deficiency confirmed by blood coagulation testing; AND
   
   b. Used to treat at least 1:
      
      i. Control and prevention of acute bleeding episodes (hemorrhage)
      
      ii. Routine prophylaxis to prevent or reduce the frequency of bleeding episodes
      
      iii. Perioperative management

2. **Von Willebrand disease (VWD)**
a. Von Willebrand disease confirmed by blood coagulation and von Willebrand factor testing; AND
b. Used as surgical bleeding prophylaxis in members with VWD in whom desmopressin is either ineffective or contraindicated; OR to control episodes of acute bleeding (Humate-P only); AND
c. Alphanate is not indicated for members with severe (type 3) VWD undergoing major surgery

Criteria III — Advate, Eloctate, Helixate FS, Hemofil M, Koate-DVI, Kogenate FS, Monoclate-P, Novoeight, Recombinate, Xyntha and NUWIQ

1. Hemophilia A
   a. Congenital factor VIII deficiency confirmed by blood coagulation testing; AND
   b. Used to treat at least 1:
      i. Control and prevention of acute bleeding episodes (hemorrhage)
      ii. Routine prophylaxis to prevent or reduce frequency of episodic bleeding
      iii. Perioperative management

Criteria IV — Feiba NF

1. Hemophilia A
   a. Congenital factor VIII deficiency confirmed by blood coagulation testing; AND
   b. Confirmation that member has Factor VIII inhibitors; AND
d. Used to treat at least 1:
   i. Control and prevention of acute episodic bleeding (hemorrhage)
   ii. Routine prophylaxis to prevent or reduce the frequency of episodic bleeding
2. Hemophilia B
   a. Congenital factor IX deficiency confirmed by blood coagulation testing; AND
   b. Confirmation that member has Factor IX inhibitors; AND
e. Used to treat at least 1:
   i. Control and prevention of acute episodic bleeding (hemorrhage)
   ii. Routine prophylaxis to prevent or reduce the frequency of episodic bleeding
   iii. Perioperative management

Criteria V — Novoseven RT

1. Hemophilia A
   a. Congenital factor VIII deficiency confirmed by blood coagulation testing; AND
b. Confirmation that member has acquired Factor VIII inhibitors; AND

c. Used to treat at least 1:
   i. Control and prevention of acute episodic bleeding (hemorrhage); OR
   ii. Perioperative management

2. Acquired Hemophilia
   a. Diagnosis confirmed by blood coagulation testing; AND
   b. Used to treat at least 1:
      i. Control and prevention of acute episodic bleeding (hemorrhage); OR
      ii. Perioperative management

3. Hemophilia B
   a. Factor IX deficiency confirmed by blood coagulation testing; AND
   b. Confirmation that member has acquired Factor IX inhibitors; AND
   c. Used to treat at least 1:
      i. Control and prevention of acute episodic bleeding (hemorrhage); OR
      ii. Perioperative management

4. Congenital Factor VII Deficiency
   a. Factor VII deficiency confirmed by blood coagulation testing; AND
   b. Used to treat at least 1:
      i. Control and prevention of acute episodic bleeding (hemorrhage); OR
      ii. Perioperative management

5. Glanzmann’s Thrombasthenia
   a. Glanzmann’s Thrombasthenia has been confirmed by blood coagulation testing; AND
   b. Used to treat at least 1:
      i. Control and prevention of acute episodic bleeding (hemorrhage); OR
      ii. Perioperative management; AND
   c. The use of platelet transfusions is known or suspected to be ineffective or contraindicated

Criteria VI — Obizur

1. Acquired Hemophilia A
   a. Factor VIII deficiency confirmed by blood coagulation testing; AND
   b. Used to treat episodic bleeding; AND
   c. Not used for congenital Hemophilia A or von Willebrand disease
Criteria VII — Tretten

1. Congenital Factor XIII A-subunit deficiency
   a. Factor XIII A-subunit deficiency confirmed by blood coagulation testing; AND
   b. Used for routine prophylaxis of bleeding

Criteria VIII — Corifact

1. Congenital Factor XIII deficiency
   a. Factor XIII deficiency confirmed by blood coagulation testing; AND
   b. Used for routine prophylactic treatment; OR
   c. Used for perioperative management of surgical bleeding

Criteria IX — Wilate

1. Von Willebrand disease (VWD)
   a. VWD confirmed by blood coagulation and von Willebrand factor testing; AND
   b. Used as treatment of spontaneous and trauma-induced bleeding episodes in at least 1:
      i. Members with severe VWD; OR
      ii. Members with mild or moderate VWD in whom the use of desmopressin is known or suspected to be ineffective or contraindicated; AND
      iii. Is NOT being used for routine prophylactic treatment of spontaneous bleeding episodes OR perioperative management of surgical bleeding; AND
      iv. Is NOT being used for Hemophilia A
   c. Used for perioperative management of bleeding

Renewal Criteria

1. Member continues to meet clinical criteria above
2. Absence of unacceptable drug-toxicity (e.g.: symptoms of allergic-anaphylactic reactions [i.e., anaphylaxis, dyspnea, rash], thromboembolic events [thromboembolism, pulmonary embolism] and development of neutralizing antibodies [inhibitors]).

Limitations/Exclusions

Hemophilia medications are considered **investigational and not medically necessary** for any indications other than those listed above.
**Revision History**

6/16/2017 — added control of acute bleeding episodes as a covered indication for Humate-P.

**Applicable Procedure Codes**

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<tr>
<th>Code</th>
<th>Description</th>
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<tbody>
<tr>
<td>J7175</td>
<td>Injection, factor X (human), 1 i.u. (New Code: 01/01/2017)</td>
</tr>
<tr>
<td>J7179</td>
<td>Injection, von willebrand factor (recombinant), (vonvendi), 1 i.u. vwf:rco (New Code: 01/01/2017)</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>
</tr>
<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>
</tr>
<tr>
<td>J7190</td>
<td>Factor VIII (antihemophilic factor, human) per IU</td>
</tr>
<tr>
<td>J7192</td>
<td>Factor VIII (antihemophilic factor, recombinant) per IU, not otherwise specified</td>
</tr>
<tr>
<td>J7193</td>
<td>Factor IX (antihemophilic factor, purified, nonrecombinant) per IU</td>
</tr>
<tr>
<td>J7194</td>
<td>Factor IX complex, per IU</td>
</tr>
<tr>
<td>J7195</td>
<td>Injection, factor IX (antihemophilic factor, recombinant) per IU, not otherwise specified</td>
</tr>
<tr>
<td>J7198</td>
<td>Antiinhibitor, per IU</td>
</tr>
<tr>
<td>J7199</td>
<td>Hemophilia clotting factor, not otherwise classified</td>
</tr>
<tr>
<td>J7200</td>
<td>Injection, factor IX, (antihemophilic factor, recombinant), Rixubis, per IU</td>
</tr>
<tr>
<td>J7201</td>
<td>Injection, factor ix, fc fusion protein, (recombinant), alprolix, 1 i.u. (Revised Code: 01/01/2017)</td>
</tr>
<tr>
<td>J7202</td>
<td>Injection, factor ix, albumin fusion protein, (recombinant), idelvion, 1 i.u. (New Code: 01/01/2017)</td>
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<tr>
<td>J7205</td>
<td>Injection, factor VIII Fc fusion protein (recombinant), per IU</td>
</tr>
<tr>
<td>J7207</td>
<td>injection, factor viii, (antihemophilic factor, recombinant), pegylated, 1 i.u. (New Code: 01/01/2017)</td>
</tr>
<tr>
<td>J7209</td>
<td>Injection, factor viii, (antihemophilic factor, recombinant), (nuwiq), 1 i.u. (New Code: 01/01/2017)</td>
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<tr>
<td>96365</td>
<td>Intravenous infusion, for therapy, prophylaxis, or diagnosis (specify substance or drug); initial, up to 1 hour</td>
</tr>
<tr>
<td>96366</td>
<td>Intravenous infusion, for therapy, prophylaxis, or diagnosis (specify substance or drug); each additional hour (List separately in addition to code for primary procedure)</td>
</tr>
<tr>
<td>96367</td>
<td>Intravenous infusion, for therapy, prophylaxis, or diagnosis (specify substance or drug); additional sequential infusion of a new drug/substance, up to 1 hour (List separately in addition to code for primary procedure)</td>
</tr>
<tr>
<td>96368</td>
<td>Intravenous infusion, for therapy, prophylaxis, or diagnosis (specify substance or drug); concurrent infusion (List separately in addition to code for primary procedure)</td>
</tr>
<tr>
<td>96369</td>
<td>Subcutaneous infusion for therapy or prophylaxis (specify substance or drug); initial, up to 1 hour, including pump set-up and establishment of subcutaneous infusion site(s)</td>
</tr>
<tr>
<td>96370</td>
<td>Subcutaneous infusion for therapy or prophylaxis (specify substance or drug); each additional hour (List separately in addition to code for primary procedure)</td>
</tr>
<tr>
<td>96371</td>
<td>Subcutaneous infusion for therapy or prophylaxis (specify substance or drug); additional pump set-up with establishment of new subcutaneous infusion site(s) (List separately in addition to code for primary procedure)</td>
</tr>
<tr>
<td>96372</td>
<td>Therapeutic, prophylactic, or diagnostic injection (specify substance or drug); subcutaneous or</td>
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</tbody>
</table>
Intramuscular

- Therapeutic, prophylactic, or diagnostic injection (specify substance or drug); intra-arterial

- Therapeutic, prophylactic, or diagnostic injection (specify substance or drug); intravenous push, single or initial substance/drug

- Therapeutic, prophylactic, or diagnostic injection (specify substance or drug); each additional sequential intravenous push of a new substance/drug (List separately in addition to code for primary procedure)

- Therapeutic, prophylactic, or diagnostic injection (specify substance or drug); each additional sequential intravenous push of the same substance/drug provided in a facility (List separately in addition to code for primary procedure)

- Application of on-body injector (includes cannula insertion) for timed subcutaneous injection

- Unlisted therapeutic, prophylactic, or diagnostic intravenous or intra-arterial injection or infusion

- Home infusion/specialty drug administration, per visit (up to 2 hours);

- Home infusion/specialty drug administration, per visit (up to 2 hours); each additional hour (List separately in addition to code for primary procedure)

Applicable ICD-10 Diagnosis Codes

- D66 Hereditary factor VIII deficiency
- D67 Hereditary factor IX deficiency
- D68.0 Von Willebrand’s disease
- D68.1 Hereditary factor XI deficiency
- D68.2 Hereditary deficiency of other clotting factors
- D68.311 Acquired hemophilia
- D68.9 Coagulation defect, unspecified
- D69.1 Qualitative platelet defects
- R58 Hemorrhage, not elsewhere classified

References


Specialty-matched clinical peer review.

