SENTARA HEALTH PLANS

MEDICAL PRIOR AUTHORIZATION/STEP-EDIT REQUEST*

Directions: **The prescribing physician must sign and clearly print name (preprinted stamps not valid) on this request**. All other information may be filled in by office staff; **fax to 1-844-668-1550**. No additional phone calls will be necessary if all information (including phone and fax #s) on this form is correct. **If information provided is not complete, correct, or legible, authorization can be delayed**.

For Medicare Members: Medicare Coverage for outpatient (Part B) drugs is outlined in the Medicare Benefit Policy Manual (Pub. 100-2), Chapter 15, §50 Drugs and Biologicals. In addition, National Coverage Determination (NCD) and Local Coverage Determinations (LCDs) may exist and compliance with these policies is required where applicable. They can be found at: https://www.cms.gov/medicare-coverage-database/overview-and-quick-search.aspx. Additional indications may be covered at the discretion of the health plan.

Factor VIII Deficiency Therapy (Hemophilia A) (MEDICAL)

Drug Requested: (Select one drug below)

Human Plasma-derived Factor VIII Replacement Products		
❑ **J7183 Wilate® (+vWF)**	❑ **J7186 Alphanate® (+vWF)**	❑ **J7187 Humate-P® (+vWF)**
❑ **J7190 Hemofil M®, Koate®**		
Recombinant Factor VIII Replacement Products		
❑ **J7182 Novoeight®**	❑ **J7185 Xyntha®**	❑ **J7188 Obizur®**
❑ **J7192 Advate®, Recominate®, Kogenate®**	❑ **J7209 Nuwiq®**	❑ **J7210 Afstyla®**
❑ **J7210 Kovaltry®**		
Extended Half-life Factor VIII Replacement Products		
❑ **J7204 Esperoct®**	❑ **J7205 Eloctate®**	❑ **J7207 Adynovate®**
❑ **J7208 Jivi®**		
High Sustained Half-life Factor VIII Replacement Products		
❑ **J7214 ALTUVIIIO®**		

MEMBER & PRESCRIBER INFORMATION: Authorization may be delayed if incomplete.

Member Name: _____

Member Sentara #: _____ **Date of Birth:** _____

Prescriber Name: _____

Prescriber Signature: _____ **Date:** _____

Office Contact Name: _____

Phone Number: _____ **Fax Number:** _____

NPI #: _____

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DRUG INFORMATION: Authorization may be delayed if incomplete.

Drug Name/Form/Strength: _____

Dosing Schedule: _____ **Length of Therapy**: _____

Diagnosis: _____ **ICD Code, if applicable:** _____

Weight (if applicable): _____ **Date weight obtained:** _____

- ❑ Standard Review. In checking this box, the timeframe does not jeopardize the life or health of the member or the member's ability to regain maximum function and would not subject the member to severe pain.

Dosing Limits:

A. Quantity Limit (max daily dose) [NDC Unit]

- N/A

B. Max Units (per dose and over time) [HCPCS Unit]:

- Alphanate (von Willebrand complex): 55,200 billable units per 28 day supply
- Humate-P (von Willebrand complex): 55,200 billable units per 28 day supply
- Wilate (von Willebrand complex): 55,200 billable units per 28 day supply
- Advate: 64,400 billable units per 28-day supply
- Adynovate: 46,000 billable units per 28-day supply
- Afstyla: 69,000 billable units per 28-day supply
- Altuviiio: 23,000 billable units per 28-day supply
- Eloctate: 74,750 billable units per 30-day supply
- Esperoct: 40,250 billable units per 28-day supply
- Hemofil M: 55,200 billable units per 28-day supply
- Jivi: 41,400 billable units per 30-day supply
- Koate DVI: 55,200 billable units per 28-day supply
- Kogenate: 64,400 billable units per 28-day supply
- Kovaltry: 55,200 billable units per 28-day supply
- Novoeight: 69,000 billable units per 28-day supply
- Nuwiq: 64,400 billable units per 28-day supply
- Obizur: 115,000 billable units per 90-day supply
- Recombinate: 64,400 billable units per 28-day supply
- Xyntha/Xyntha Solofuse: 41,400 billable units per 28-day supply

CLINICAL CRITERIA: Check below all that apply. All criteria must be met for approval. To support each line checked, all documentation, including lab results, diagnostics, and/or chart notes, must be provided or request may be denied.

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Part I. Initial Authorization

- ❑ Diagnosis of congenital factor VIII deficiency has been confirmed by blood coagulation testing
- ❑ Medication prescribed by a specialist familiar with treating patients with hemophilia (factor VIII deficiency)
- ❑ **For Jivi Requests:** Member must be at least 12 years of age
- ❑ **For Obizur Requests: ALL** the following must be met:
  - ❑ Member must be at least 18 years of age
  - ❑ Member has a diagnosis of acquired factor VIII deficiency has been confirmed by blood coagulation testing **[NOTE: Use for congenital Hemophilia A or von Willebrand's disease will NOT be approved]**
  - ❑ Provider is only requesting and dosing Obizur for on-demand treatment and control of bleeding episodes
  - ❑ Provider has submitted laboratory documentation that the member does not have baseline anti-porcine factor VIII inhibitor titer >20 Bethesda Units (BU)
- ❑ **For von Willebrand disease (vWD), complete the following:**
  - ❑ Diagnosis has been confirmed by blood coagulation and von Willebrand factor testing
  - ❑ For Alphanate and Humate P, use will be for treatment in **ONE** of the following:
    - ❑ On demand treatment for spontaneous and trauma-induced bleeding episodes
    - ❑ Surgical bleeding prophylaxis during major or minor procedures in patients with vWD in whom desmopressin is either ineffective or contraindicated **(Authorizations valid for 1 month)**
  - ❑ For Alphanate, will **NOT** be used in members with severe (type 3) vWD undergoing major surgery **OR** treatment of spontaneous/trauma-induced bleeding episodes
  - ❑ For Wilate, use will be for treatment in **ONE** of the following:
    - ❑ On demand treatment and control of bleeding episodes with severe vWD, or mild-moderate vWD in whom desmopressin is either ineffective or contraindicated
    - ❑ Perioperative management of bleeding **(Authorizations valid for 1 month)**
    - ❑ Routine prophylaxis to reduce frequency of bleeding episodes
  - ❑ Provider will **NOT** use any of the following for the treatment of von Willebrand's disease: Advate, Eloctate, Hemofil M, Koate/Koate DVI, Kogenate FS, Novoeight, Recombinate, Xyntha/Xyntha Solofuse, Nuwiq, Adynovate, Kovaltry, Afstyla, Jivi, Esperoct, Altuviiio
- ❑ Member was previously treated with valoctocogene roxaparvovec and factor VIII activity levels decreased and/or bleeding was **NOT** controlled
- ❑ Requested medication will be used as treatment in at least **ONE** of the following:
  - ❑ On-demand treatment and control of bleeding episodes

    Please Attach On-Demand Treatment Dosing Calculations [Dosage regimen to adhere to most current recommended FDA-label and/or compendia recommendations (see Part IV)]

(Continued on next page)

- ❑ Perioperative management **(Authorizations valid for 1 month)**

  Name Description of Procedure: _____

  Date of Procedure: _____

  Dosage regimen must adhere to most current recommended FDA-label and/or compendia recommendations (see Part IV):

  _____

- ❑ Routine prophylaxis

  Dosage regimen must adhere to most current recommended FDA-label and/or compendia recommendations (see Part IV):

  _____

- ❑ **FOR ROUTINE PROPHYLAXIS**: Requested medication will be used as treatment in at least **ONE** of the following:
  - ❑ Severe factor VIII deficiency (factor VIII level of <1%) **AND** member must meet **ONE** of the following:
    - ❑ Member has a history of life-threatening hemorrhage requiring on-demand use of Factor VIII therapy
    - ❑ Member has a history of repeated, serious spontaneous bleeding episodes requiring on-demand use of Factor VIII therapy

Part II. If Requesting Extended-Half Life (EHL) Products: J7205 Eloctate, J7207 Adynovate, J7208 Jivi, J7204 Esperoct, or J7214 Altuviiio

- ❑ Member must have had a trial and failure of a non-extended half-life clotting factor replacement product: Wilate, Humate, Alphanate, Hemofil, Novoeight, Xyntha, Advate, Recombinate, Kogenate, Nuwiq, Kovaltry, Afstyla **[NOTE: Submit past medical history, FVIII levels, trials of prior therapy, etc. to convey that the member is NOT a candidate for a non-extended half-life product]**
- ❑ Provider must submit a half-life study to determine the appropriate dose and dosing interval of the EHL product when initiated
- ❑ **FOR ROUTINE PROPHYLAXIS**, **ALL** the following must be met:
  - ❑ Requested medication will **NOT** be used in combination with other FVIII products, Hemlibra® (emicizumab-kxwh) in those with hemophilia A as prophylactic therapy, or Hympavzi® (marstacimab-hncq), Alhemo® (concizumab-mtci), and/or Qfitlia® (fitusiran) in those with hemophilia A as prophylactic therapy
  - ❑ If the request exceeds any of the following dosing limits, documentation must be submitted specifying why the member is **NOT** a suitable candidate prophylaxis therapy with Hympavzi® (marstacimab-hncq), Alhemo® (concizumab-mtci), or Qfitlia® (fitusiran):
    - ❑ Eloctate: 50 IU/kg every 4 days (total weekly dose of 87.5 IU/kg)
    - ❑ Adynovate: 40 IU/kg twice weekly (total weekly dose of 80 IU/kg)
    - ❑ Jivi: 60 IU/kg every 5 days (total weekly dose of 84 IU/kg)
    - ❑ Esperoct: 50 IU/kg every 4 days (total weekly dose of 87.5 IU/kg)
    - ❑ Altuviiio: 50 IU/kg every 4 days (total weekly dose of 87.5 IU/kg)

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Part III. Renewal Clinical Authorization

- ❑ Requested medication will be used as treatment in at least **ONE** of the following:
  - ❑ On-demand treatment and control of bleeding episodes **(Authorization will be approved for 6 months)**

    Please Attach On-Demand Treatment Dosing Calculations [Dosage regimen to adhere to most current recommended FDA-label and/or compendia recommendations (see Part IV)]
  - ❑ Perioperative management **(NO RENEWAL AUTHORIZATIONS – PLEASE COMPLETE PART I)**
  - ❑ Routine prophylaxis **(Authorization will be approved for a 12-month period)**

    Dosage regimen must adhere to most current recommended FDA-label and/or compendia recommendations (see Part IV):

    _____

    NOTE: Provider must submit clinical rationale (i.e., past medical records, weight gain, half-life study results, increase in breakthrough bleeding when patient is fully adherent to therapy) for an increase in dose
- ❑ Provider must confirm **ALL** the following:
  - ❑ Member has experienced an absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include: anaphylaxis and hypersensitivity reactions (e.g., angioedema, chest tightness, hypotension, urticaria, wheezing, dyspnea, etc.), thromboembolic events (pulmonary embolism, venous thrombosis, and arterial thrombosis), development of neutralizing antibodies (inhibitors), nephrotic syndrome, etc.
  - ❑ Member continues to meet criteria in Part I and other indication-specific relevant criteria such as concomitant therapy requirements (not including prerequisite therapy), performance status, etc.
  - ❑ Member has demonstrated a beneficial response to therapy (i.e., the frequency of bleeding episodes has decreased from pre-treatment baseline)

Part IV. Dosage/Administration Information

NOTE:

Hemophilia Dosing For BMI
• For members with a BMI ≥ 30, a half-life study should be performed to determine the appropriate dose and dosing interval. • For minimally treated patients (< 50 exposure days to factor products) previously receiving a different factor product, inhibitor testing is required at baseline, then at every comprehensive care visit (yearly for the mild and moderate patients, semi-annually for the severe patients)

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Indication	Dose
Advate	
On-demand treatment and control of bleeding episodes Congenital Hemophilia A	Dose (IU/kg) = desired factor VIII rise (IU/dL or % of normal) x 0.5 (IU/kg per IU/dL) **Minor** Circulating Factor VIII required (% of normal) (20-40%) = 10-20 IU/ kg - Repeat every 12-24 hours as needed (every 8 to 24 hours for patients underage of 6). Continue until the bleeding episode is resolved (approximately 1 to 3 days). **Moderate** Circulating Factor VIII required (% of normal) (30-60%) = 15-30 IU/ kg - Repeat every 12-24 hours as needed (every 8 to 24 hours for patients underage of 6). Continue until the bleeding episode is resolved (approximately 3 days or more). **Major** Circulating Factor VIII required (% of normal) (60-100%) = 30-50 IU/ kg - Repeat every 8-24 hours as needed (every 6 to 12 hours for patients underage of 6). Continue until the bleeding episode is resolved.
Routine prophylaxis Congenital Hemophilia A	For prophylaxis regimen to prevent or reduce frequency of bleeding episodes, dose between 20 to 40 IU per kg every other day (3 to 4 times weekly). Alternatively, an every third day dosing regimen targeted to maintain FVIII trough levels ≥ 1% may be employed. Adjust dose based on the patient's clinical response.
Perioperative management Congenital Hemophilia A	Dose (IU/kg) = desired factor VIII rise (IU/dL or % of normal) x 0.5 (IU/kg per IU/dL) **Minor** Circulating Factor VIII required (% of normal) (60-100%) = 30-50 IU/ kg – Single dose within one hour of the operation. Repeat after 12- 24 hours for optional additional dosing as needed to control bleeding. **Major** Circulating Factor VIII required (% of normal) (80-120%) = Preoperative: 40-60 IU/ kg to achieve 100% activity. Followed by a repeat dose every 8-24 hours (every 6 to 24 hours for patients underage of 6) postoperatively until healing is complete.
Adynovate	
On-demand treatment and control of bleeding episodes Congenital Hemophilia A	Dose (IU) = Body Weight (kg) x Desired factor VIII rise (IU/dL or % of normal) x 0.5 (IU/kg per IU/dL) **Minor** Target Factor VIII level (IU/dL or % of normal) (20-40%) = 10-20 IU/kg - Repeat every 12-24 hours until the bleeding episode is resolved **Moderate** Target Factor VIII level (IU/dL or % of normal) (30-60%) = 15-30 IU/kg - Repeat every 12-24 hours until the bleeding episode is resolved **Major** Target Factor VIII level (IU/dL or % of normal) (60-100%) = 30-50 IU/kg - Repeat every 8-24 hours until the bleeding episode is resolved.

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Indication	Dose
Adynovate	
Perioperative management Congenital Hemophilia A	Dose (IU) = Body Weight (kg) × Desired factor VIII rise (IU/dL or % of normal) × 0.5 (IU/kg per IU/dL) **Minor** Target Factor VIII required (% of normal) (60-100%) = 30-50 IU/ kg – Single dose within one hour before the operation. Repeat after 24 hours, if necessary, single dose or repeat as needed until bleeding is resolved. **Major** Target Factor VIII required (% of normal) (80-120%) (pre- and post- operative) = 40-60 IU/ kg within 1 hour before the operation to achieve 100% activity. Repeat dose every 8- 24 hours (every 6 to 24 hours for patients under age of 12) to maintain FVIII activity within the target range and continue until adequate wound healing.
Routine prophylaxis Congenital Hemophilia A	Administer 40-50 IU per kg body weight 2 times per week in children and adults (12 years and older). Administer 55 IU per kg body weight 2 times per week in children (<12 years) with a maximum of 70 IU per kg. Adjust the dose based on the patient's clinical response.
Afstyla	
On-demand treatment and control of bleeding episodes Congenital Hemophilia A	Dose (IU) = Body Weight (kg) x Desired factor VIII rise (IU/dL or % of normal) x 0.5 (IU/kg per IU/dL) **Minor** Target Factor VIII level (IU/dL or % of normal) 20-40% - Repeat every 12-24 hours until the bleeding episode is resolved. **Moderate** Target Factor VIII level (IU/dL or % of normal) 30-60% - Repeat every 12-24 hours until the bleeding episode is resolved. **Major** Target Factor VIII level (IU/dL or % of normal) 60-100% - Repeat every 8-24 hours until the bleeding episode is resolved.
Perioperative management Congenital Hemophilia A	**Minor** Target Factor VIII level (IU/dL or % of normal) 30-60%- Repeat every 24 hours, for at least one day, until healing is achieved. **Major** Target Factor VIII level (IU/dL or % of normal) 80-100%- Repeat every 8-24 hours until adequate wound healing, then continue for at least another 7 days to maintain a Factor VIII activity of 30-60% (IU/dL).
Routine prophylaxis Congenital Hemophilia A	Adults and adolescents (≥12yrs old): Administer 20-50 IU per kg body weight 2 to 3 times per week. Adjust the dose based on the patient's clinical response. Children (<12 yrs old): Administer 30-50 IU per kg body weight 2 to 3 times per week. Adjust the dose based on the patient's clinical response.

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Indication	Dose
Altuviiio	
On-demand treatment and control of bleeding episodes Congenital Hemophilia A	**Minor/Moderate** Single dose of 50 IU/kg. For minor and moderate bleeding episodes occurring within 2 to 3 days after a prophylactic dose, a lower dose of 30 IU/kg dose may be used. Additional doses of 30 or 50 IU/kg every 2 to 3 days may be considered. **Major** Single dose of 50 IU/kg. Additional doses of 30 or 50 IU/kg every 2 to 3 days can be considered. Note: For resumption of prophylaxis (if applicable) after treatment of a bleed, it is recommended to allow an interval of at least 72 hours between the last 50 IU/kg dose for treatment of a bleed and resuming prophylaxis dosing. Thereafter, prophylaxis can be continued as usual on the patient's regular schedule.
Perioperative management Congenital Hemophilia A	**Minor** Single dose of 50 IU/kg. An additional dose of 30 or 50 IU/kg after 2 to 3 days may be considered. **Major** Single dose of 50 IU/kg. Additional doses of 30 or 50 IU/kg every 2 to 3 days may be administered as clinically needed for perioperative management.
Routine prophylaxis Congenital Hemophilia A	The recommended dosing for routine prophylaxis for adults and children is 50 IU/kg of Altuviiio administered once weekly.
• For the dose of 50 IU/kg, the expected in vivo peak increase in Factor VIII level expressed as IU/dL (or % of normal) is estimated using the following formula: • Estimated Increment of Factor VIII (IU/dL or % of normal) = 50 IU/kg x 2 (IU/dL per IU/kg) • To achieve a specific target Factor VIII activity level, use the following formula: Dosage (IU) = Body Weight (kg) x Desired Factor VIII Increase (IU/dL or % normal) x 0.5 (IU/kg per IU/dL).	
Eloctate	
On-demand treatment and control of bleeding episodes Congenital Hemophilia A	Dose (IU/kg) = Desired factor VIII rise (IU/dL or % of normal) x 0.5 (IU/kg per IU/dL) **Minor and Moderate** Circulating Factor VIII required (% of normal) (40-60%) = 20-30 IU/ kg - Repeat every 24-48 hours as needed (every 12 to 24 hours for patients under age of 6). Continue until the bleeding episode is resolved. **Major** Circulating Factor VIII required (% of normal) (80-100%) = 40-50 IU/ kg - Repeat every 12-24 hours as needed (every 8 to 24 hours for patients under age of 6). Continue until the bleeding episode is resolved (approximately 7-10 days).

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<table>
<tr><th>Indication</th><th>Dose</th></tr>
<tr><td colspan="2">Eloctate</td></tr>
<tr><td>Routine prophylaxis Congenital Hemophilia A</td><td>Adults and adolescents ≥ 6: The recommended starting regimen is 50 IU/kg administered every 4 days. The regimen may be adjusted based on patient response with dosing in the range of 25-65 IU/kg at 3–5-day intervals.

Children < 6 years of age: The recommended starting regimen is 50 IU/kg administered twice weekly. The regimen may be adjusted based on patient response with dosing in the range of 25-65 IU/kg at 3–5-day intervals. More frequent or higher doses up to 80 IU/kg may be required.</td></tr>
<tr><td>Perioperative management Congenital Hemophilia A</td><td>Dose (IU/kg) = Desired factor VIII rise (IU/dL or % of normal) x 0.5 (IU/kg per IU/dL)
<u>Minor</u>
Circulating Factor VIII required (% of normal) (50-80%) = 25-40 IU/ kg - Repeat every 24 hours as needed (every 12 to 24 hours for patients underage of 6). Continue at least 1 day until healing is achieved.
<u>Major</u>
Circulating Factor VIII required (% of normal) (80-120%) = Preoperative: 40-60 IU/ kg – Followed by a repeat dose of 40-50 IU/kg after 8-24 hours (6 to 24 hours for patients under age of 6). Continue every 24 hours until adequate wound healing; then continue therapy for at least 7 days to maintain FVII activity within the target range.</td></tr>
<tr><td colspan="2">Esperoct</td></tr>
<tr><td>On-demand treatment and control of bleeding episodes Congenital Hemophilia A</td><td>One IU of Factor VIII activity corresponds to the quantity of Factor VIII in one milliliter of normal human plasma. The calculation of the required dosage of Factor VIII is based on the empirical finding that one IU of Factor VIII per kg body weight raises the plasma Factor VIII activity by two IU/dL.

To achieve a specific target Factor VIII activity level, use the following formula: Dosage (IU) = Body Weight (kg) × Desired Factor VIII Increase (IU/dL or % normal) × 0.5 ; OR
<table>
<tr><th>Type of bleeding</th><th>Adolescents /Adults ≥12 years Dose (IU/kg)</th><th>Children <12 years Dose (IU/kg)</th><th>Additional doses</th></tr>
<tr><td>Minor
Early hemarthrosis, mild muscle bleeding, or oral bleeding</td><td>40</td><td>65</td><td>One dose should be sufficient</td></tr>
<tr><td>Moderate
More extensive hemarthrosis, muscle bleeding, or hematoma</td><td>40</td><td>65</td><td>An additional dose may be administered after 24 hours</td></tr>
<tr><td>Major
Life- or limb-threatening hemorrhages, gastro- intestinal bleeding, intracranial, intra-abdominal or intrathoracic bleeding, fractures</td><td>50</td><td>65</td><td>Additional dose(s) may be administered approximately every 24 hours</td></tr>
</table></td></tr>
</table>

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<table>
<tr><th>Indication</th><th>Dose</th></tr>
<tr><td colspan="2">Esperoct</td></tr>
<tr><td>Routine prophylaxis
Congenital Hemophilia A</td><td>• Adults and adolescents (≥ 12 years): The recommended starting dose is 50 IU per kg body weight every 4 days. This regimen may be individually adjusted to less or more frequent dosing based on bleeding episodes.
• Children (< 12 years): A dose of 65 IU per kg body weight twice weekly. This regimen may be individually adjusted to less or more frequent dosing based on bleeding episodes.</td></tr>
<tr><td rowspan="3">Perioperative management
Congenital Hemophilia A</td><td>To achieve a specific target Factor VIII activity level, use the following formula: Dosage (IU) = Body Weight (kg) × Desired Factor VIII Increase (IU/dL or % normal) × 0.5 ; OR</td></tr>
<tr><td>
<table>
<tr><th>Type of surgery</th><th>Adolescents/Adults ≥12 years Dose (IU/kg)</th><th>Children <12 years Dose (IU/kg)</th><th>Additional doses</th></tr>
<tr><td>Minor
Including tooth extraction</td><td>50</td><td>65</td><td>Additional dose(s) can be given after 24 hours if necessary</td></tr>
<tr><td>Major
Intracranial, intra-abdominal, intrathoracic, or joint replacement surgery</td><td>50</td><td>65</td><td>Additional doses can be given every 24 hours for the first week and then approximately every 48 hours until wound healing has occurred</td></tr>
</table>
</td></tr>
<tr><td colspan="2">Hemofil M</td></tr>
<tr><td>On-demand treatment and control of bleeding episodes
Congenital Hemophilia A</td><td>Dose (IU/kg) = Desired factor VIII rise (IU/dL or % of normal) x 0.5 (IU/kg per IU/dL)
<u>Early hemarthrosis or muscle bleed or oral bleed</u>
Circulating Factor VIII required (% of normal) (20-40%) = Begin infusion every 12 to 24 hours for one-three days until the bleeding episode as indicated by pain is resolved or healing is achieved.
<u>More extensive hemarthrosis, muscle bleed, or hematoma</u>
Circulating Factor VIII required (% of normal) (30-60%) = Repeat every 12-24 hours for usually three days or more until pain and disability are resolved.
<u>Life threatening bleeds such as head injury, throat bleed, severe abdominal pain</u>
Circulating Factor VIII Required (% of normal) (60-100%) = Repeat every 8-24 hours until the bleeding threat is resolved.</td></tr>
<tr><td>Perioperative management
Congenital Hemophilia A</td><td><u>Minor</u>
Circulating Factor VIII required (% of normal) (60-80%) A single infusion plus oral antifibrinolytic therapy within one hour is sufficient in approximately 70% of cases.
<u>Major</u>
Circulating Factor VIII required (% of normal) (80-100% pre- and post-operative): Repeat dose every 8-24 hours depending on state of healing.</td></tr>
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Indication	Dose
Jivi	
On-demand treatment and control of bleeding episodes Congenital Hemophilia A	Dose (IU/kg) = Desired factor VIII rise (IU/dL or % of normal) x reciprocal of expected recovery (or observed recovery, if available) (e.g., 0.5 for a recovery of 2 IU/dL per IU/kg) **Minor** Circulating Factor VIII required (% of normal) (20-40%) – 10-20IU/kg repeat dose every 24-48 hours until bleed resolves **Moderate** Circulating Factor VIII required (% of normal) (30-60%) – 15-30IU/kg repeat dose every 24-48 hours until bleed resolves **Major** Circulating Factor VIII Required (% of normal) (60-100%) – 30-50IU/kg repeat dose every 8-24 hours until bleed resolves
Perioperative management Congenital Hemophilia A	**Minor** Circulating Factor VIII required (% of normal) (30-60%) – 15-30IU/kg repeat dose every 24 hours for at least 1 day until healing is achieved **Major** Circulating Factor VIII required (% of normal) (80-100%) – 40-50IU/kg repeat dose every 12-24 hours until adequate wound healing is complete, then continue therapy for at least another 7 days to maintain Factor VIII activity of 30–60% (IU/dL)
Routine prophylaxis Congenital Hemophilia A	The recommended initial regimen is 30–40 IU/kg twice weekly. Based on the bleeding episodes, the regimen may be adjusted to 45–60 IU/kg every 5 days or may be further individually adjusted to less or more frequent dosing.
Koate/Koate DVI	
On-demand treatment and control of bleeding episodes Congenital Hemophilia A	Dose (IU/kg) = Desired factor VIII rise (IU/dL or % of normal) x 0.5 (IU/kg per IU/dL) **Minor** Circulating Factor VIII required (% of normal) (30%) = 15 IU/kg repeat dose every 12 hours until hemorrhage stops and healing has been achieved. **Moderate** Circulating Factor VIII required (% of normal) (50%) = 25 IU/kg repeat dose every 12 hours until healing has been achieved. **Major** Circulating Factor VIII Required (% of normal) (80-100%) = Initial: 40-50 IU/kg. Maintenance dose 25 IU/kg. Repeat every 12 hours for at least 3 – 5 days until healing has been achieved for up to 10 days.

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Indication	Dose
Koate/Koate DVI	
On-demand treatment and control of bleeding episodes Congenital Hemophilia A	Dose (IU/kg) = Desired factor VIII rise (IU/dL or % of normal) x reciprocal of expected recovery (or observed recovery, if available) (e.g., 0.5 for a recovery of 2 IU/dL per IU/kg) **Minor** Circulating Factor VIII required (% of normal) (20-40%) – 10-20IU/kg repeat dose every 24-48 hours until bleed resolves **Moderate** Circulating Factor VIII required (% of normal) (30-60%) – 15-30IU/kg repeat dose every 24-48 hours until bleed resolves **Major** Circulating Factor VIII Required (% of normal) (60-100%) – 30-50IU/kg repeat dose every 8-24 hours until bleed resolves
Routine prophylaxis Hemophilia A	25-40 IU/kg three times weekly or 15-30 IU/kg three times weekly. Adjust dosing regimen based on individual response.
Perioperative management Congenital Hemophilia A	**Prior to surgery** Circulating Factor VIII Required (% of normal) (80-100%) = 40-50 IU/kg for one dose prior to surgery. **After surgery** Circulating Factor VIII Required (% of normal) (60-100%) = 30-50 IU/kg repeat dose every 12 hours for the next 7 – 10 days or until healing has been achieved.
Kogenate FS	
On-demand treatment and control of bleeding episodes Congenital Hemophilia A	Dose (IU/kg) = Desired factor VIII rise (IU/dL or % of normal) x 0.5 (IU/kg per IU/dL) **Minor** Circulating Factor VIII required (% of normal) (20-40%) = 10-20 IU/ kg - Repeat dose if there is evidence of further bleeding and continue until the bleeding episode is resolved. **Moderate** Circulating Factor VIII required (% of normal) (30-60%) = 15-30 IU/ kg - Repeat every 12-24 hours as needed. Continue until the bleeding episode is resolved. **Major** Circulating Factor VIII Required (% of normal) (80-100%) = Initial: 40-50 IU/ kg; Repeat 20-25 IU/kg every 8-12 hours until the bleeding episode is resolved.
Routine prophylaxis Congenital Hemophilia A	**Routine Prophylaxis in Adults** 25 units per kg of body weight three times per week. Routine Prophylaxis in **Children** 25 IU/kg of body weight every other day.

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Indication	Dose
Kogenate FS	
Perioperative management Congenital Hemophilia A	Dose (IU/kg) = Desired factor VIII rise (IU/dL or % of normal) x 0.5 (IU/kg per IU/dL) **Minor** Circulating Factor VIII required (% of normal) (30-60%) = 15-30 IU/ kg – Repeat every 12- 24 hours until bleeding is resolved. **Major** Circulating Factor VIII required (% of normal) (100%) = Preoperative: 50 IU/ kg to achieve 100% activity. Followed by a repeat dose every 6-12 hours to keep FVIII activity in desired range. Continue until healing is complete.
Kovaltry	
On-demand treatment and control of bleeding episodes Congenital Hemophilia A	• Required dose (IU) = body weight (kg) x desired Factor VIII rise (% of normal or IU/dL) x reciprocal of expected/observed recovery (e.g., 0.5 for a recovery of 2 IU/dL per IU/kg) • Estimated Increment of Factor VIII (IU/dL or % of normal) = [Total Dose (IU)/body weight (kg)] x 2 (IU/dL per IU/kg) **Minor** (Early hemarthrosis, minor muscle, oral bleeds) Factor VIII level required (IU/dL or % of normal): 20-40 – repeat every 12-24 hours for at least 1 day, until bleeding episode as indicated by pain is resolved or healing is achieved. **Moderate** (More extensive hemarthrosis, muscle bleeding, or hematoma) Factor VIII level required (IU/dL or % of normal): 30-60 – repeat every 12-24 hours for 3 to 4 days or more until pain and acute disability are resolved. **Major** (Intracranial, intra-abdominal or intrathoracic hemorrhages, gastrointestinal bleeding, central nervous system bleeding, bleeding in the retropharyngeal or retroperitoneal spaces, or iliopsoas sheath, life or limb threatening hemorrhage) Factor VIII level required (IU/dL or % of normal): 60-100 – repeat every 8-24 hours until bleeding is resolved.
Routine prophylaxis Congenital Hemophilia A	Individualize the patient's dose based on clinical response: • Adults and adolescents: 20 to 40 IU of KOVALTRY per kg of body weight two or three times per week. • Children ≤12 years old: 25 to 50 IU of KOVALTRY per kg body weight twice weekly, three times weekly, or every other day according to individual requirements

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Indication	Dose
Kovaltry	
Perioperative management Congenital Hemophilia A	• Required dose (IU) = body weight (kg) x desired Factor VIII rise (% of normal or IU/dL) x reciprocal of expected/observed recovery (e.g., 0.5 for a recovery of 2 IU/dL per IU/kg) • Estimated Increment of Factor VIII (IU/dL or % of normal) = [Total Dose (IU)/body weight (kg)] x 2 (IU/dL per IU/kg) **Minor** (Such as tooth extraction) Factor VIII level required (IU/dL or % of normal): 30-60 (pre- and post-operative) – repeat every 24 hours for at least 1 day until healing is achieved. **Major** (Such as intracranial, intraabdominal, intrathoracic, or joint replacement surgery) Factor VIII level required (IU/dL or % of normal): 80-100 (pre- and post-operative) – repeat every 8-24 hours until adequate wound healing is complete, then continue therapy for at least another 7 days to maintain Factor VIII activity of 30-60% (IU/dL).
Novoeight	
On-demand treatment and control of bleeding episodes Congenital Hemophilia A	Dose (IU/kg) = Desired factor VIII rise (IU/dL or % of normal) x 0.5 (IU/kg per IU/dL) **Minor** Circulating Factor VIII required (% of normal) (20-40%), every 12 – 24 hours for at least 1 day until the bleeding episode is resolved **Moderate** Circulating Factor VIII required (% of normal) (30-60%), every 12 – 24 hours until pain and acute disability are resolved, approximately 3-4 days **Major** Circulating Factor VIII Required (% of normal) (60-100%), every 8 – 24 hours until resolution of bleed, approximately 7-10 days.
Perioperative management Hemophilia A	Dose (IU/kg) = Desired factor VIII rise (IU/dL or % of normal) x 0.5 (IU/kg per IU/dL) **Minor** Circulating Factor VIII required (% of normal) (30-60%) every 24 hours for at least 1 day until healing is achieved. **Major** Circulating Factor VIII required (% of normal) (80-100%) every 8 – 24 hours until adequate wound healing, then continue therapy for at least 7 days to maintain a factor VIII activity of 30 – 60% (IU/dL)
Routine prophylaxis Hemophilia A	**Adults and adolescents (≥12 yrs):** 20-50 IU/kg three times weekly **OR** 20-40 IU/kg every other day **Children (<12 yrs):** 25-60 IU/kg three times weekly **OR** 25-50 IU/kg every other day

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Indication	Dose
Nuwiq	
On-demand treatment and control of bleeding episodes Congenital Hemophilia A	**Dose** Required IU = body weight (kg) x desired Factor VIII rise (%) (IU/dL) x 0.5 (IU/kg per IU/dL) Expected Factor VIII rise (% of normal) = 2 x administered IU/body weight (kg) **Minor** Required peak post-infusion Factor VIII activity (% of normal or IU/dL): 20-40 every 12 – 24 hours for at least 1 day, until the bleeding episode is resolved **Moderate to Major** Required peak post-infusion Factor VIII activity (% of normal or IU/dL): 30-60 every 12 – 24 hours for 3-4 days or more until the bleeding episode is resolved **Life-threatening** Required peak post-infusion Factor VIII activity (% of normal or IU/dL): 60-100 every 8 – 24 hours bleeding risk is resolved
Routine prophylaxis Congenital Hemophilia A	**Dose** Required IU = body weight (kg) x desired Factor VIII rise (%) (IU/dL) x 0.5 (IU/kg per IU/dL) Expected Factor VIII rise (% of normal) = 2 x administered IU/body weight (kg) **Adolescents (12-17 years) and adults** 30 – 40 IU/kg every other day **Children (2-11 years)** 30 – 50 IU/kg every other day or three times per week
Perioperative management Congenital Hemophilia A	**Dose** Required IU = body weight (kg) x desired Factor VIII rise (%) (IU/dL) x 0.5 (IU/kg per IU/dL) Expected Factor VIII rise (% of normal) = 2 x administered IU/body weight (kg) **Minor** Required peak post-infusion Factor VIII activity (% of normal or IU/dL): 30-60 (pre- and post-operative) every 24 hours for at least 1 day until healing is achieved **Major** Required peak post-infusion Factor VIII activity (% of normal or IU/dL): 80-100 (pre- and post-operative) every 8 - 24 hours until adequate wound healing, then continue therapy for at least another 7 days to maintain Factor VIII activity of 30% to 60% (IU/dL)

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Indication	Dose
Obizur	
On-demand treatment and control of bleeding episodes Acquired Hemophilia A	**Minor and Moderate** Loading dose: 200IU/kg; Maintenance dose: Titrate to maintain recommended FVIII trough levels at 50-100 IU/dL every 4 to 12 hours **Major** Loading dose: 200 IU/kg; Maintenance dose: Titrate to maintain recommended FVIII trough levels at 100-200 (to treat an acute bleed), then 50-100 IU/dL (after acute bleed is controlled) every 4 to 12 hours
Recombinate	
On-demand treatment and control of bleeding episodes Congenital Hemophilia A	Dose (IU/kg) = Desired factor VIII rise (IU/dL or % of normal) x 0.5 (IU/kg per IU/dL) **Early hemarthrosis or muscle bleed or oral bleed** Circulating Factor VIII required (% of normal) (20-40%) - Begin infusion every 12 to 24 hours for one-three days until the bleeding episode as indicated by pain is resolved or healing is achieved. **More extensive hemarthrosis, muscle bleed, or hematoma** Circulating Factor VIII required (% of normal) (30-60%) - Repeat every 12-24 hours for usually three days or more until pain and disability are resolved. **Life threatening bleeds such as head injury, throat bleed, severe abdominal pain** Circulating Factor VIII Required (% of normal) (60-100%) - Repeat every 8-24 hours until the bleeding threat is resolved.
Routine prophylaxis Hemophilia A	25-40 IU/kg three times weekly or 15-30 IU/kg three times weekly. Adjust dosing regimen based on individual response.
Perioperative management Congenital Hemophilia A	**Minor** Circulating Factor VIII required (% of normal) (60-80%) - A single infusion plus oral antifibrinolytic therapy within one hour is sufficient in approximately 70% of cases. **Major** Circulating Factor VIII required (% of normal) (80-100% pre- and post-operative) - Repeat dose every 8-24 hours depending on state of healing.
Xyntha/Xyntha Solofuse	
On-demand treatment and control of bleeding episodes Congenital Hemophilia A	Dose (IU/kg) = Desired factor VIII rise (IU/dL or % of normal) x 0.5 (IU/kg per IU/dL) **Minor** Circulating Factor VIII required (% of normal) (20-40%) - Repeat dose every 12- 24 hours for least 1 day, depending upon the severity of the bleeding episode. **Moderate** Circulating Factor VIII required (% of normal) (30-60%) - Repeat every 12-24 hours as needed. Continue for 3-4 days or until adequate local hemostasis is achieved. **Major** Circulating Factor VIII Required (% of normal) (60-100%) - Repeat every 8-24 hours until bleeding is resolved.

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Indication	Dose
Xyntha/Xyntha Solofuse	
Perioperative management Congenital Hemophilia A	**Minor** Circulating Factor VIII required (% of normal) (30-60%) - Repeat every 12- 24 hours. Continue for 3-4 days or until adequate local hemostasis is achieved. For tooth extraction, a single infusion plus oral antifibrinolytic therapy within 1 hour may be sufficient. **Major** Circulating Factor VIII required (% of normal) (60-100%) - Repeat every 8-24 hours. Continue until threat is resolved, or in the case of surgery, until adequate local hemostasis and wound healing are achieved.
Routine prophylaxis Hemophilia A	**Adults and adolescents (≥12 years):** The recommended starting regimen is 30 IU/kg of Xyntha administered 3 times weekly. **Children (<12 years):** The recommended starting regimen is 25 IU/kg of Xyntha administered every other day. More frequent or higher doses may be required in children <12 years of age to account for the higher clearance in this age group. Note: Adjust the dosing regimen (dose or frequency) based on the patient's clinical response.

Medication being provided by: Please check applicable box below.

- ❑ **Location/site of drug administration:** _____
  NPI or DEA # of administering location: _____

  OR

- ❑ **Specialty Pharmacy – Proprium Rx**

For urgent reviews: Practitioner should call Sentara Health Plans Pre-Authorization Department if they believe a standard review would subject the member to adverse health consequences. Sentara Health Plan's definition of urgent is a lack of treatment that could seriously jeopardize the life or health of the member or the member's ability to regain maximum function.

Use of samples to initiate therapy does not meet step edit/ preauthorization criteria.

Previous therapies will be verified through pharmacy paid claims or submitted chart notes.

***Approved by Pharmacy and Therapeutics Committee**: 5/22/2025
REVISED/UPDATED/REFORMATTED: ~~9/24/2025;~~ **10/24/2025**