

Policy Name	Policy Number	Scope	
Agents for Hemophilia B	MP-RX-FP-03-23	🛛 МММ МА	🛛 MMM Multihealth
Service Category			
<ul> <li>Anesthesia</li> <li>Surgery</li> <li>Radiology Procedures</li> <li>Pathology and Laboratory Procedures</li> </ul>	Evaluatio	e Services and Proc on and Manageme osthetics or Supplie rugs	nt Services

#### Service Description

This document addresses the use of Factor IX Human, Purified [Alphanine], Factor IX Complex Human [Profilnine], Coagulation Factor IX Recombinant [Rixubis], Factor IX Fc Fusion Protein Recombinant [Alprolix], Factor IX Albumin Fusion Protein Recombinant [Idelvion], Coagulation Factor IX Recombinant, GlycoPEGylated [Rebinyn], drugs approved by the Food and Drug Administration (FDA) for the treatment of Hemophilia B.

#### **Background Information**

Factor replacement treatments can be created from blood products (human plasma-derived) and others that are manufactured (recombinant). Replacement therapy may be given on a routine, preventive basis which is also called prophylactic therapy. The infusion of factor replacements given to stop a bleeding episode is called on-demand or episodic therapy.

### Products in this document include:

- Coagulation Factor IX, Human plasma-derived: Alphanine SD
- Factor IX Complex, human plasma-derived: Profilnine SD
- Factor IX Recombinant: Rixubis, Benefix, Ixinity
- Coagulation Factor IX-Long-Acting
  - o Recombinant, Albumin Fusion Protein: Idelvion
  - o Recombinant coagulation factor IX, Fc Fusion Protein: Alprolix
  - o Recombinant coagulation factor IX, GlycoPEGylated: Rebinyn

Hereditary hemophilia B is the second most common type of hemophilia after hemophilia A (four times less common than hemophilia A). Although it is usually inherited, about one third of cases are caused by spontaneous mutations. Hemophilia A and B are clinically indistinguishable from one another, except by factor analysis. Hemophilia B is related to mutations in the gene coding for coagulation Factor IX (CDC 2014).

The U.S. National Hemophilia Foundation (NHF) and the World Federation of Hemophilia (Srivastava, 2020) both note there is a relationship of bleeding severity to the clotting factor level. Both entities list "severe" hemophilia as a clotting factor level < 1 IU/dl or < 1% of normal. A "mild" bleeding severity is identified as a clotting factor level of 5-40 IU/dl or 5 to < 40% of normal. A bleeding episode for individuals with mild risk includes severe bleeding with major trauma or surgery. Individuals with 1-5 IU/dl or 1-5% of normal are



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considered "moderate" risk for occasional spontaneous bleeding and prolonged bleeding with minor trauma or surgery (Srivastava, 2013).

#### Hemophilia severity:

- Severe hemophilia Severe hemophilia is defined as < 1 percent factor activity, which corresponds to < 1 IU/dL.
- Moderate hemophilia Moderate hemophilia is defined as a factor activity level ≥ 1 percent of normal and < 5 percent of normal, corresponding to ≥ 1 and < 5 IU/dL.
- Mild hemophilia Mild hemophilia is defined as a factor activity level ≥ 5 percent of normal and < 40 percent of normal (≥ 5 and < 40 IU/dL).

World Federation of Hemophilia 2020 Guidelines for treatment of hemophilia state that prophylaxis prevents bleeding and joint destruction, and that prophylaxis should enable those with hemophilia to lead healthy and active lives. Moreover, the updated 2020 guidelines proposes that the definition of prophylaxis be based on outcomes rather than doses or timing of initiation, and treatment 2 regimens that take into account the hemophilic phenotype of the individual in addition to factor levels. However, more studies are needed to determine if all individuals should remain on therapy as adults (that is, those with severe hemophilia vs. moderate or mild). The WFH 2020 guidelines have been endorsed by several societies worldwide, including the U.S. NHF. Short-term prophylaxis (of 4 to 8 weeks) may interrupt the bleeding cycle and benefit individuals with repeated bleeding into target joints. Prophylaxis does not reverse existing joint damage but reduces bleeding and may slow progression of joint damage. Prophylactic clotting factor administration is recommended prior to the individual engaging in activities with higher risk of injury. Randomized trials of prophylactic therapy of hemophilia have demonstrated a decreased incidence of arthropathy (Gringeri, 2011; Manco-Johnson, 2007).

### Applicable Codes

The following list(s) of procedure and/or diagnosis codes is provided for reference purposes only and may not be all inclusive. Inclusion or exclusion of a procedure, diagnosis or device code(s) does not constitute or imply member coverage or provider reimbursement policy. 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	Description
J7193	Factor IX (Anti-hemophilic factor, purified, non-recombinant) per IU [AlphaNine SD]
ICD-10	Description
D67	Hereditary factor IX deficiency [hemophilia B]
D68.311	Acquired hemophilia
Z29.8	Encounter for other specified prophylactic measure

### Coagulation Factor IX, Human plasma-derived (Alphanine SD)



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gents for Hemophilia	B MP-RX	(-FP-03-23	🛛 MMM MA	🛛 MMM Multihealth
Z79.899	Other long	term (current)	drug therapy [proph	vylactic]
actor IX Complex Hun	ıan (Profilnine SD)			
HCPCS		Desc	ription	
J7194	Fact	or IX complex,	per IU [Profilnine SD	]
ICD-10		Desc	ription	
D67	Heredit		eficiency [hemophili	a B]
Z29.8		-	ified prophylactic m	
Z79.899	Other long	term (current)	drug therapy [proph	iylactic]
actor IX Recombinant	(Benefi, Ixinity, Rixubis)			
HCPCS		Desc	ription	
J7200	Injection, factor IX, (A	Anti-hemophilio	factor, recombinan	t), Rixubis, per IU
J7195	Injection, factor IX (Anti	ection, factor IX (Anti-hemophilic factor, recombinant) per IU, not otherwise specified [Benefix, lxinity]		
J7213	Injection, coa	Injection, coagulation factor ix (recombinant) [Ixinity], 1 IU		
ICD-10		Desc	ription	
D67	Heredit	tary factor IX de	eficiency [hemophili	a B]
D68.311		Acquired	hemophilia	
Z29.8	Encounter	Encounter for other specified prophylactic measure		
Z79.899	Other long	term (current)	drug therapy [proph	iylactic]
•	-Long Acting Recombinant, Fc Fusion Protein (Alprolix)			•
ebinyn)		D		
ebinyn) HCPCS	Injection factors		ription	Alproliv 1 11
HCPCS J7201	-	IX, Fc fusion pro	otein (recombinant),	
HCPCS J7201 J7202	Injection, factor IX, a	IX, Fc fusion pro	otein (recombinant), protein, (recombina	nt), Idelvion, 1 IU
ebinyn) HCPCS J7201 J7202	-	IX, Fc fusion pro	otein (recombinant), protein, (recombina	nt), Idelvion, 1 IU
HCPCS         J7201         J7202         J7203	Injection, factor IX, a Injection factor ix, (antihem	IX, Fc fusion pro albumin fusion nophilic factor, Desc	otein (recombinant), protein, (recombinan recombinant), glyco ription	nt), Idelvion, 1 IU pegylated, Rebinyn, 1II
HCPCS         J7201         J7202         J7203         ICD-10         D67	Injection, factor IX, a Injection factor ix, (antihem	IX, Fc fusion pro Ibumin fusion pro nophilic factor, Desc tary factor IX de	otein (recombinant), protein, (recombinan recombinant), glyco ription eficiency [hemophili	nt), Idelvion, 1 IU pegylated, Rebinyn, 1II
HCPCS         J7201         J7202         J7203         ICD-10         D67         D68.311	Injection, factor IX, a Injection factor ix, (antihem Heredit	IX, Fc fusion pro Ibumin fusion Iophilic factor, Desc tary factor IX de Acquired	otein (recombinant), protein, (recombinant recombinant), glyco ription eficiency [hemophilia hemophilia	nt), Idelvion, 1 IU pegylated, Rebinyn, 1II a B]
HCPCS           J7201           J7202           J7203           ICD-10           D67	Injection, factor IX, a Injection factor ix, (antihem Heredit Encounter	IX, Fc fusion pro albumin fusion pro nophilic factor, Desc tary factor IX de Acquired r for other spec	otein (recombinant), protein, (recombinan recombinant), glyco ription eficiency [hemophili	nt), Idelvion, 1 IU pegylated, Rebinyn, 1IU a B] easure



### **Medical Necessity Guidelines**

When a drug is being reviewed for coverage under a member's medical benefit plan or is otherwise subject to clinical review (including prior authorization), the following criteria will be used to determine whether the drug meets any applicable medical necessity requirements for the intended/prescribed purpose.

Provider must submit documentation (such as office chart notes, lab results or other clinical information) supporting that member has met all approval criteria.

# Alphanine SD (Human plasma-derived, Coagulation Factor IX)

- **A.** Criteria for Initial Approval (Provider must submit documentation [such as office chart notes, lab results, pathology reports, imaging studies, and any other pertinent clinical information] supporting the patient's diagnosis for the drug and confirming that the patient has met **all** approval criteria.)
  - I. Individual has a diagnosis of hemophilia B (also called factor IX deficiency or Christmas disease); AND
  - II. Individual is using for the treatment of bleeding episodes;
  - OR
- III. Individual has a diagnosis of severe hemophilia B (defined as less than 1 IU/dL or 1% endogenous Factor IX) (NHF,Srivastava 2020); AND
- IN. Individual is using for routine prophylaxis to prevent or reduce the frequency of bleeding episodes;
   OR
- V. Individual has a diagnosis of mild to moderate hemophilia B (defined as endogenous Factor IX less than 40 IU/dL [less than 40%], but greater than or equal to 1 IU/dL) (NHF, Srivastava 2020); **AND**
- VI. Individual is using for routine prophylaxis to prevent or reduce the frequency of bleeding episodes; **AND**
- VII. Individual has one of the following:
  - a. One or more episodes of spontaneous bleeding into joint; **OR**
  - b. One or more episodes severe, life-threatening, or spontaneous bleeding as determined by prescriber; **OR**
  - c. Severe phenotype hemophilia determined by the individual's risk factors that increase the risk of a clinically significant bleed, including but not limited to, participation in activities likely to cause injury/trauma, procoagulant and anticoagulant protein levels, comorbid conditions affecting functional ability and physical coordination, or history of a clinically significant bleed.

### B. Criteria For Continuation of Therapy

- I. MMM considers continuation Alphanine SD therapy medically necessary in members requesting reauthorization for an indication listed in Section A above (Criteria for Initial Approval) when all of the following criteria is met:
  - a. Individual has had a positive therapeutic response to treatment (for example, reduction in frequency and/or severity of bleeding episodes).



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C. A	uthorization Duration			
۱. ۱۱.	Initial Approval Durat Reauthorization Appr	-		
D. C	onditions Not Covered			
	ny other use is considere nay not be all inclusive):	d experimental, investigational,	or unproven, inclua	ling the following (this lis
۱. ۱۱.		l of coumarin-induced anticoagu coagulopathy associated with li		
III.	0	als with hemophilia A with inhil	•	
IV.		of other clotting factors which		
V.	When the above crite	ria are not met and for all other	r indications	
Profilniı	ne SD (Human plasr	na-derived, Factor IX Con	nplex)	
		<b>val</b> (Provider must submit doc , imaging studies, and any other		
		e drug and confirming that the p	atient has met <b>all</b> a	
<i>و</i> ا.		e drug and confirming that the p nosis of hemophilia B (also calle	atient has met <b>all</b> a	
I. 11.	Individual has a diagr <b>AND</b> Individual is using for		atient has met <b>all</b> a <sub>l</sub>	
I. 11.	Individual has a diagr AND Individual is using for R	nosis of hemophilia B (also calle the treatment of bleeding episo nosis of severe hemophilia B (de	atient has met <b>all</b> a <sub>l</sub> ed factor IX deficien odes,	cy or Christmas disease
۱. ۱۱. ۱۱۱. ۱۷.	Individual has a diagr AND Individual is using for R Individual has a diagn Factor IX) (NHF, Srivas Individual is using as r	nosis of hemophilia B (also calle the treatment of bleeding episo nosis of severe hemophilia B (de	atient has met <b>all</b> a ed factor IX deficien odes, fined as less than 1	cy or Christmas disease IU/dL or 1% endogenou
I. II. O III. IV.	Individual has a diagr AND Individual is using for R Individual has a diagn Factor IX) (NHF, Srivas Individual is using as r R Individual has a diagn	nosis of hemophilia B (also calle the treatment of bleeding episc nosis of severe hemophilia B (de stava 2020); <b>AND</b>	atient has met <b>all</b> a ed factor IX deficien odes, fined as less than 1 or reduce the freque ohilia B (defined as e	cy or Christmas disease IU/dL or 1% endogenou ncy of bleeding episodes endogenous Factor IX les
I. II. O III. IV. O	Individual has a diagr AND Individual is using for R Individual has a diagn Factor IX) (NHF, Srivas Individual is using as r R Individual has a diagn than 40 IU/dL [less tha	nosis of hemophilia B (also calle the treatment of bleeding episo nosis of severe hemophilia B (de stava 2020); <b>AND</b> routine prophylaxis to prevent o osis of mild to moderate hemop	atient has met <b>all</b> a ed factor IX deficien odes, fined as less than 1 or reduce the freque ohilia B (defined as e ual to 1 IU/dL) (NHF	cy or Christmas disease IU/dL or 1% endogenou ncy of bleeding episodes endogenous Factor IX les , Srivastava 2020); AND
I. II. III. IV. V.	Individual has a diagr AND Individual is using for R Individual has a diagn Factor IX) (NHF, Srivas Individual is using as r R Individual has a diagn than 40 IU/dL [less the Individual is using for AND Individual has one of t	nosis of hemophilia B (also calle the treatment of bleeding episo nosis of severe hemophilia B (de stava 2020); <b>AND</b> routine prophylaxis to prevent o osis of mild to moderate hemop an 40%], but greater than or eq routine prophylaxis to prevent o the following:	ed factor IX deficien odes, fined as less than 1 or reduce the freque ohilia B (defined as e ual to 1 IU/dL) (NHF or reduce the freque	cy or Christmas disease IU/dL or 1% endogenou ncy of bleeding episodes endogenous Factor IX les , Srivastava 2020); AND
I. II. III. IV. V. VI.	Individual has a diagr AND Individual is using for R Individual has a diagn Factor IX) (NHF, Srivas Individual is using as r R Individual has a diagn than 40 IU/dL [less that Individual is using for AND Individual has one of the a. One or more of b. One or more	nosis of hemophilia B (also calle the treatment of bleeding episo losis of severe hemophilia B (de stava 2020); <b>AND</b> routine prophylaxis to prevent o osis of mild to moderate hemop an 40%], but greater than or eq routine prophylaxis to prevent o the following: episodes of spontaneous bleedi episodes of severe, life-threate	ed factor IX deficien odes, fined as less than 1 or reduce the freque ohilia B (defined as e ual to 1 IU/dL) (NHF or reduce the freque	cy or Christmas disease IU/dL or 1% endogenou ncy of bleeding episodes endogenous Factor IX les , Srivastava 2020); <b>AND</b> ncy of bleeding episodes
I. II. III. IV. V. VI.	Individual has a diagr AND Individual is using for R Individual has a diagn Factor IX) (NHF, Srivas Individual is using as r R Individual has a diagn than 40 IU/dL [less that Individual is using for AND Individual has one of the a. One or more b. One or more by the prescri	nosis of hemophilia B (also calle the treatment of bleeding episo nosis of severe hemophilia B (de stava 2020); <b>AND</b> routine prophylaxis to prevent of osis of mild to moderate hemop an 40%], but greater than or eq routine prophylaxis to prevent of the following: episodes of spontaneous bleedi episodes of severe, life-threater iber; <b>OR</b>	ed factor IX deficien odes, fined as less than 1 or reduce the freque ohilia B (defined as e ual to 1 IU/dL) (NHF, or reduce the freque ng into joint; <b>OR</b> ning, or spontaneou	cy or Christmas disease IU/dL or 1% endogenou ncy of bleeding episodes endogenous Factor IX les , Srivastava 2020); AND ncy of bleeding episodes s bleeding as determine
I. II. III. IV. V. VI.	Individual has a diagr AND Individual is using for R Individual has a diagn Factor IX) (NHF, Srivas Individual is using as r R Individual has a diagn than 40 IU/dL [less the Individual is using for AND Individual has one of t a. One or more b. One or more by the prescri c. Severe pheno	nosis of hemophilia B (also calle the treatment of bleeding episo losis of severe hemophilia B (de stava 2020); <b>AND</b> routine prophylaxis to prevent o osis of mild to moderate hemop an 40%], but greater than or eq routine prophylaxis to prevent o the following: episodes of spontaneous bleedi episodes of severe, life-threate	atient has met <b>all</b> and ed factor IX deficien odes, fined as less than 1 or reduce the freque ohilia B (defined as e ual to 1 IU/dL) (NHF, or reduce the freque ng into joint; <b>OR</b> ning, or spontaneou	cy or Christmas disease IU/dL or 1% endogenou ncy of bleeding episodes endogenous Factor IX les , Srivastava 2020); <b>AND</b> ncy of bleeding episodes s bleeding as determine



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	ecting functional ability and pl	hysical coordination	, or history of a clinically
significant blee	ed.		
medically necessary in above (Criteria for Initi a. Individual has	of Therapy nuation Profilnine SD (Human members requesting reautho ial Approval) when all of the fo had a positive therapeutic resp /or severity of bleeding episod	prization for an indi llowing criteria is m ponse to treatment	cation listed in Section A et:
C. Authorization Duration			
I. Initial Approval Duration II. Reauthorization Appro	-		
D. Conditions Not Covered			
	d experimental, investigational,	, or unproven, incluc	ling the following (this lis
may not be all inclusive): I. Individual has a diagno	osis of Factor VII deficiency; <b>OR</b>		
-	ia are not met and for all other		
Benefix, Ixinity, Rixubis (Rec	combinant Factor IX)		
	<b>val</b> (Provider must submit doc imaging studies, and any other drug and confirming that the p	r pertinent clinical in	formation] supporting the
I. Individual has a diagno AND	osis of hemophilia B, (also calle	ed factor IX deficier	icy or Christmas disease)
II. Individual is using for	one of the following:		
	of bleeding episodes; OR		
b. Peri-procedura <b>OR</b>	al management for surgical, inv	asive or interventio	nal radiology procedures
	osis of severe hemophilia B (de ava 2020); <b>AND</b>	efined as less than 1	IU/dL or 1% endogenou
	outine prophylaxis to prevent o	or reduce the freque	ncy of bleeding episodes
OR			
than 40 IU/dL [less tha	osis of mild to moderate hemop n 40%], but greater than or eq	ual to 1 IU/dL) (NHF	, Srivastava 2020); AND
VI. Individual is using for r	outine prophylaxis to prevent of	or reduce the freque	ency of bleeding episodes

VI. Individual is using for routine prophylaxis to prevent or reduce the frequency of bleeding episodes; AND



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MI ne	<ul> <li>b. One or more ep by the prescribe</li> <li>c. Severe phenotyp risk of a clinicall likely to cause i conditions affect significant bleed</li> <li>iteria for Continuation of</li> <li>MM considers continuation cessary in members requesting Initial Approval) when all</li> </ul>	isodes of spontaneous bleedin isodes of severe, life-threater r; <b>OR</b> pe hemophilia determined by ly significant bleed, including injury/trauma, procoagulant ting functional ability and ph l. <b>Therapy</b> ion Benefix, Ixinity, Rixubis esting reauthorization for an lof the following criteria is mo positive therapeutic response	the individual's risk but not limited to, and anticoagulant hysical coordination (Recombinant Fact indication listed in et:	a factors that increase the participation in activitie protein levels, comorbie , or history of a clinicall cor IX therapy medicall Section A above (Criteri
۱. ۱۱. <b>D. Co</b>	Initial Approval Duratior Reauthorization Approva nditions Not Covered	-		
	ay not be all inclusive): Treatment of other factor Treatment of individuals To reverse coumarin-ind Treatment of bleeding d Using for the induction of	experimental, investigational, or deficiencies (for example fa s with hemophilia A with inhib luced anticoagulation; <b>OR</b> ue to low levels of liver-depe of immune tolerance in indivio are not met and for all other	actors II, VII, VIII and bitors to factor VIII; ndent coagulation f duals with hemophi	I X); OR OR actors; OR
Alprolix (Recomb <b>A. Cri</b>	(Recombinant, Fc binant, glycoPEGylate iteria for Initial Approva	g-Acting, Albumin Fusio Fusion Protein Coa ed Coagulation Factor IX I (Provider must submit doc maging studies, and any other	gulation Facto <) <i>umentation [such c</i>	r IX), or Rebinyr



disea II. Indivi III. Indivi a b C. OR IV. Indivi equal VI. Indivi a b C. B. Criteria fe	dual has a diagnosis se); AND dual has less than 1 l dual is using for one The treatment of Peri-procedural m OR Routine prophylax dual has a diagnosis dual has endogenou to 1 IU/dL (NHF,Sriv dual is using for one Individual is using radiology procedu Individual is using episodes for one o 1. Individual oR 2. Individual spont	bleeding episodes; <b>OR</b> hanagement for surgical, inv kis to prevent or reduce the of mild to moderate hemop is Factor IX level less than 4 vastava 2020); <b>AND</b> of the following: for the treatment of bleedi is for peri-procedural manage ures; <b>OR</b> is for routine prophylaxis to of the following: dual has had one or more of dual has had one or more	enous Factor IX (NHF vasive or interventior e frequency of bleedin ohilia B; <b>AND</b> 40 IU/dL (less than 4 ing episodes; <b>OR</b> gement for surgical, i prevent or reduce th episodes of spontan	, Srivastava 2020); <b>AND</b> nal radiology procedures; ng episodes; 40%) but greater than or nvasive or interventional he frequency of bleeding eous bleeding into joint;
disea II. Indivi III. Indivi a b C. OR IV. Indivi equal VI. Indivi a b C. B. Criteria fe	se); AND dual has less than 1 l dual is using for one The treatment of Peri-procedural m OR Routine prophylax dual has a diagnosis dual has endogenou to 1 IU/dL (NHF,Sriv dual is using for one Individual is using radiology procedu Individual is using episodes for one o 1. Individual Spont	IU/dL (less than 1%) endoge of the following: bleeding episodes; <b>OR</b> hanagement for surgical, inv kis to prevent or reduce the of mild to moderate hemop is Factor IX level less than 4 rastava 2020); <b>AND</b> of the following: for the treatment of bleedi for peri-procedural manage ares; <b>OR</b> g for routine prophylaxis to of the following: dual has had one or more of dual has had one or more	enous Factor IX (NHF vasive or interventior e frequency of bleedin ohilia B; <b>AND</b> 40 IU/dL (less than 4 ing episodes; <b>OR</b> gement for surgical, i prevent or reduce th episodes of spontan	, Srivastava 2020); <b>AND</b> nal radiology procedures; ng episodes; 40%) but greater than or nvasive or interventional he frequency of bleeding eous bleeding into joint;
II. Indivi III. Indivi a b C. OR IV. Indivi equal VI. Indivi a b C. B. Criteria fe	dual has less than 1 l dual is using for one The treatment of Peri-procedural m OR Routine prophylax dual has a diagnosis dual has a diagnosis dual has endogenou to 1 IU/dL (NHF,Sriv dual is using for one Individual is using radiology procedu Individual is using episodes for one o 1. Individual OR 2. Individual	of the following: bleeding episodes; <b>OR</b> hanagement for surgical, inv kis to prevent or reduce the of mild to moderate hemop is Factor IX level less than 4 vastava 2020); <b>AND</b> of the following: for the treatment of bleedi for peri-procedural manage ares; <b>OR</b> for routine prophylaxis to of the following: dual has had one or more of dual has had one or more	vasive or intervention frequency of bleedin ohilia B; <b>AND</b> 40 IU/dL (less than 4 ing episodes; <b>OR</b> gement for surgical, i prevent or reduce th episodes of spontan	nal radiology procedures; ng episodes; 40%) but greater than or nvasive or interventional he frequency of bleeding eous bleeding into joint;
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b C. OR IV. Indivi equal VI. Indivi a b C. B. Criteria fe	<ul> <li>Peri-procedural monostructure</li> <li>OR</li> <li>Routine prophylax</li> <li>dual has a diagnosis</li> <li>dual has endogenou</li> <li>to 1 IU/dL (NHF,Sriving</li> <li>dual is using for one</li> <li>Individual is using</li> <li>Individual is using</li> <li>Individual is using</li> <li>radiology procedu</li> <li>Individual is using</li> <li>episodes for one of</li> <li>1. Individual is</li> <li>OR</li> <li>2. Individual is</li> </ul>	hanagement for surgical, inv kis to prevent or reduce the of mild to moderate hemop is Factor IX level less than 4 rastava 2020); <b>AND</b> of the following: for the treatment of bleedi for peri-procedural manage ares; <b>OR</b> g for routine prophylaxis to of the following: dual has had one or more of dual has had one or more	e frequency of bleedin ohilia B; <b>AND</b> 40 IU/dL (less than 4 ing episodes; <b>OR</b> gement for surgical, i prevent or reduce th episodes of spontan	ng episodes; 40%) but greater than or nvasive or interventional he frequency of bleeding eous bleeding into joint;
C. OR IV. Indivi equal VI. Indivi a b C. B. Criteria fe	OR Routine prophylax dual has a diagnosis dual has endogenou to 1 IU/dL (NHF,Sriv dual is using for one Individual is using radiology procedu Individual is using episodes for one o 1. Individual OR 2. Individual spont	kis to prevent or reduce the of mild to moderate hemop is Factor IX level less than 4 vastava 2020); <b>AND</b> of the following: for the treatment of bleedi for peri-procedural manage ares; <b>OR</b> for routine prophylaxis to of the following: dual has had one or more of dual has had one or more	e frequency of bleedin ohilia B; <b>AND</b> 40 IU/dL (less than 4 ing episodes; <b>OR</b> gement for surgical, i prevent or reduce th episodes of spontan	ng episodes; 40%) but greater than or nvasive or interventional he frequency of bleeding eous bleeding into joint;
OR IV. Indivi equal VI. Indivi a b c. B. Criteria fe	dual has a diagnosis dual has endogenou to 1 IU/dL (NHF,Sriv dual is using for one Individual is using radiology procedu Individual is using episodes for one o 1. Individual OR 2. Individ spont	of mild to moderate hemop is Factor IX level less than 4 astava 2020); <b>AND</b> of the following: for the treatment of bleedi for peri-procedural manage ares; <b>OR</b> for routine prophylaxis to of the following: dual has had one or more of dual has had one or more	ohilia B; <b>AND</b> 40 IU/dL (less than 4 ing episodes; <b>OR</b> gement for surgical, i prevent or reduce th episodes of spontan	40%) but greater than or nvasive or interventional he frequency of bleeding eous bleeding into joint;
IV. Indivi V. Indivi equal VI. Indivi a b C.	dual has endogenou to 1 IU/dL (NHF,Sriv dual is using for one Individual is using radiology procedu Individual is using episodes for one o 1. Individual OR 2. Individual spont	as Factor IX level less than 4 vastava 2020); AND of the following: for the treatment of bleedi for peri-procedural manage ares; OR for routine prophylaxis to of the following: dual has had one or more of dual has had one or more	40 IU/dL (less than 4 ing episodes; <b>OR</b> gement for surgical, i prevent or reduce th episodes of spontan	nvasive or interventional he frequency of bleeding eous bleeding into joint;
V. Indivi equal VI. Indivi a b c. <b>B. Criteria f</b> e	dual has endogenou to 1 IU/dL (NHF,Sriv dual is using for one Individual is using radiology procedu Individual is using episodes for one o 1. Individual OR 2. Individual spont	as Factor IX level less than 4 vastava 2020); AND of the following: for the treatment of bleedi for peri-procedural manage ares; OR for routine prophylaxis to of the following: dual has had one or more of dual has had one or more	40 IU/dL (less than 4 ing episodes; <b>OR</b> gement for surgical, i prevent or reduce th episodes of spontan	nvasive or interventional he frequency of bleeding eous bleeding into joint;
equal VI. Indivi a b c. <b>B. Criteria f</b> e	to 1 IU/dL (NHF,Sriv dual is using for one Individual is using Individual is using radiology procedu Individual is using episodes for one o 1. Individ OR 2. Individ spont	vastava 2020); <b>AND</b> of the following: for the treatment of bleedi for peri-procedural manage ares; <b>OR</b> for routine prophylaxis to of the following: dual has had one or more of dual has had one or more	ing episodes; <b>OR</b> ement for surgical, i prevent or reduce tl episodes of spontan	nvasive or interventional he frequency of bleeding eous bleeding into joint;
VI. Indivi a b c. <b>B. Criteria f</b> e	dual is using for one Individual is using Individual is using radiology procedu Individual is using episodes for one o 1. Individ OR 2. Individ spont	of the following: for the treatment of bleedi for peri-procedural manage ares; <b>OR</b> for routine prophylaxis to of the following: dual has had one or more of dual has had one or more	ement for surgical, i prevent or reduce th episodes of spontan	he frequency of bleeding eous bleeding into joint;
a b C. <b>B. Criteria f</b> e	Individual is using Individual is using radiology procedu Individual is using episodes for one o 1. Individ OR 2. Individ spont	for the treatment of bleedi for peri-procedural managures; <b>OR</b> for routine prophylaxis to of the following: dual has had one or more of dual has had one or more	ement for surgical, i prevent or reduce th episodes of spontan	he frequency of bleeding eous bleeding into joint;
b c. <b>B. Criteria f</b> e	Individual is using radiology procedu Individual is using episodes for one o 1. Individ OR 2. Individ spont	; for peri-procedural manage ares; <b>OR</b> ; for routine prophylaxis to of the following: dual has had one or more o dual has had one or more	ement for surgical, i prevent or reduce th episodes of spontan	he frequency of bleeding eous bleeding into joint;
C. B. Criteria fe	radiology procedu Individual is using episodes for one o 1. Individ OR 2. Individ spont	ures; <b>OR</b> g for routine prophylaxis to of the following: dual has had one or more o dual has had one or more	prevent or reduce the pisodes of spontant	he frequency of bleeding eous bleeding into joint;
B. Criteria fe	Individual is using episodes for one o 1. Individ OR 2. Individ spont	for routine prophylaxis to of the following: dual has had one or more o dual has had one or more	episodes of spontan	eous bleeding into joint;
	episodes for one o 1. Indivio OR 2. Indivio spont	of the following: dual has had one or more o dual has had one or more	episodes of spontan	eous bleeding into joint;
	OR 2. Indivio spont	dual has had one or mo		
	2. Indivio spont		re episodes of sev	ere life-threatening or
	spont			
		aneous pieeding as determi	ined by the prescribe	_
	3. Sever	e phenotype hemophilia de		
		ase the risk of a clinically si	•	
		ipation in activities likely	-	-
	antico	oagulant protein levels, con	norbid conditions af	fecting functional ability
	and p	hysical coordination, or hist	tory of a clinically sig	nificant bleed.
MMM co	or Continuation of Tl	herapy		
	nsiders continuation	Idelvion (Recombinant Long	g-Acting, Albumin Fu	ision Protein Coagulation
Factor IX)	, Alprolix (Recombin	ant, Fc Fusion Protein Coag	gulation Factor IX), o	r Rebinyn (Recombinant,
glycoPEG	lated Coagulation	Factor IX) necessary in m	nembers requesting	reauthorization for an
indicatior	listed in Section A al	bove (Criteria for Initial Appı	roval) when all of the	e following criteria is met:
		sitive therapeutic response of bleeding episodes).	e to treatment (for	r example, reduction in
C. Authoriza	tion Duration			
I In:+:-				
I. Initial II. Reaut	Approval Duration:	1 year		



Policy Name	Policy Number	Scope	
Agents for Hemophilia B	MP-RX-FP-03-23		🛛 MMM Multihealth
D. Conditions Not Covered			
Any other use is considered expe may not be all inclusive):	erimental, investigational,	or unproven, incluc	ling the following (this list
<ol> <li>Using for the induction of in</li> <li>When the above criteria are</li> </ol>		•	lia B; <b>OR</b>
Limits or Restrictions			
A. Therapeutic Alternatives			
The list below includes preferred be subject to prior authorization i. N/A		commended in the o	approval criteria and may
B. Quantity Limitations			
Approvals may be subject to compendia, and/or evidence recommendations as per the FD.	-based practice guidel	ines. The chart	
For the most up-to-date FD at <u>https://dailymed.nlm.nih.gov</u>			-
	r IX, Human esd.com/en/product-infor an plasma-derived: Profil		: Alphanine SD
Factor IX Recor	mbinant: Rixubis om), Ixinity ( <u>https://www</u>	(https://www.rixub	is.com), Benefix
<ul><li>Recombinant, A</li><li>Recombinant</li></ul>	Ibumin Fusion Protein: Id coagulation factor	elvion ( <u>https://wwv</u> IX, Fc Fusic	
<ul> <li>Recombinant (<u>https://www.re</u></li> </ul>	coagulation facto	or IX, Glyco	pPEGylated: Rebinyn
	coagulation facto	or IX, Glyco	oPEGylated: Reb



olicy I	Name	Policy Number	Scope
gents	for Hemophilia B	MP-RX-FP-03-23	🛛 MMM MA 🛛 MMM Multihea
fere	nce Information		
2. 3. 4. 5. 6. 7.	http://www.clinicalpharmac DailyMed. Package inserts. http://dailymed.nlm.nih.gov DrugPoints® System [electr periodically. Lexi-Comp ONLINE™ with Al National Hemophilia Foun September 29, 2022. National Hemophilia Found Treatment of Hemophilia https://www.hemophilia.or Council-MASAC/MASAC-Red Licensedfor-the-Treatment- 2022.	latabase online]. Tampa, cology.com. Updated periodica U.S. National Library of Med //dailymed/about.cfm. Access onic version]. Truven Health HFS™, Hudson, Ohio: Lexi-Com dation (NHF). Available at: dation (NHF). Recommendati a and Other Bleeding Dis g/Researchers-Healthcare-Pro commendations/MASAC-Recon of-Hemophilia-and-Other-Bleed	FL: Gold Standard, Inc.: 2022. U ally. 6 icine, National Institutes of Health webs ed: September 29, 2022. Analytics, Greenwood Village, CO. Upda pp, Inc.; 2022; Updated periodically. http://www.hemophilia.org/. Accessed ons Concerning Products Licensed for sorders. September 2020. Available viders/Medicaland-Scientific-Advisory- mmendations-Concerning-Products- eding-Disorders. Accessed: September
8. 9.	management of hemore https://onlinelibrary.wiley.cc CMS IOM Publication 100-0 Hemophilia Clotting Factor A56482, Billing and Codin	ohilia. Haemophilia. 3rd om/doi/epdf/10.1111/hae.14 4, Medicare Claims Processing s, Section 80.4.1 Clotting Fac g: Hemophilia Factor Produc	ederation of Hemophilia. Guidelines for edition. August 2020. Available 046. Accessed: September 29, 2022. Manual, Chapter 17, Section 80.4 Billing etor Furnishing Fee. Local Coverage Art cts (Revision Effective Date: 10/01/20 r Products (A56482) (cms.gov)
		ements, contract language, ar er the application of this clinic	nd Plan utilization management program al criteria.
	any means, electronic, mech	•	rieval system or transmitted, in any form wise, without permission from the health
più			



Revision Type	Summary of Changes	P&T Approval Date	MPCC Approval Date
Annual Review 11/15/2024	Added Duration of Therapy and Quantity Limits section; Wording, and formatting changes; Coding Reviewed: No changes.	2/18/2025	3/6/2025
Policy Inception 11/18/2022	Elevance Health's Medical Policy adoption.	N/A	11/30/2023