CLOTTING FACTOR REPLACEMENT THERAPY

Non-Discrimination Statement and Multi-Language Interpreter Services information are located at the end of this document.

Coverage for services, procedures, medical devices and drugs are dependent upon benefit eligibility as outlined in the member’s specific benefit plan. This Medical Coverage Guideline must be read in its entirety to determine coverage eligibility, if any.

This Medical Coverage Guideline provides information related to coverage determinations only and does not imply that a service or treatment is clinically appropriate or inappropriate. The provider and the member are responsible for all decisions regarding the appropriateness of care. Providers should provide BCBSAZ complete medical rationale when requesting any exceptions to these guidelines.

The section identified as “Description” defines or describes a service, procedure, medical device or drug and is in no way intended as a statement of medical necessity and/or coverage.

The section identified as “Criteria” defines criteria to determine whether a service, procedure, medical device or drug is considered medically necessary or experimental or investigational.

State or federal mandates, e.g., FEP program, may dictate that any drug, device or biological product approved by the U.S. Food and Drug Administration (FDA) may not be considered experimental or investigational and thus the drug, device or biological product may be assessed only on the basis of medical necessity.

Medical Coverage Guidelines are subject to change as new information becomes available.

For purposes of this Medical Coverage Guideline, the terms "experimental" and "investigational" are considered to be interchangeable.

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Description:

Hemophilia and von Willebrand's disease (VWD) are the most common congenital bleeding disorders. The two main types of hemophilia are A and B. Hemophilia A (classic hemophilia) has low levels of clotting factor VIII, or antihemophilic factor (AHF). Hemophilia B (Christmas disease) has low levels of clotting factor IX.

AHF is an endogenous glycoprotein necessary for blood clotting and hemostasis. It is a cofactor that is necessary for factor IX to activate factor X in the intrinsic pathway.
CLOTTING FACTOR REPLACEMENT THERAPY (cont.)

Description: (cont.)

The main treatment for hemophilia is replacement of clotting factor VIII (for hemophilia A) or clotting factor IX (for hemophilia B). Administration of clotting factors is indicated for hemophilia when a bleeding episode arises (demand treatment) or when bleeding is anticipated or likely (prophylactic treatment).

Hemophilia A and B are classified as mild, moderate or severe, depending on the amount of clotting factor VIII or IX in the blood.

- Mild hemophilia: 5 – 40 percent of normal clotting factor
- Moderate hemophilia: 1 – 5 percent of normal clotting factor
- Severe hemophilia: Less than 1 percent of normal clotting factor

Definitions:

- Adult: Age 18 years and older

Criteria:

All Clotting factor replacement therapies will be reviewed by the medical director(s) and/or clinical advisor(s) and/or clinical pharmacist(s).

- If an individual has not been seen within 6 months but needs to continue therapy or begin initial therapy, a limited authorization will be given initially to allow for sufficient time for the individual to be evaluated by a provider affiliated with a Hemophilia Treatment Center (HTC). Individuals with ongoing therapy must have at least one yearly appointment with a HTC. Individuals in an active course of treatment will be allowed a 60-day transition of care period to permit ample time to be seen at a HTC. The Centers for Disease Control and Prevention (CDC) supports and funds the national network of HTCs. The link to the HTC directory is: [https://www2a.cdc.gov/ncbddd/htcweb/Dir_Report/Dir_Search.asp](https://www2a.cdc.gov/ncbddd/htcweb/Dir_Report/Dir_Search.asp).
CLOTTING FACTOR REPLACEMENT THERAPY (cont.)

Criteria: (cont.)

- Clotting factor replacement therapies are considered **medically necessary** with documentation of **ANY** of the following according to limitations shown: (cont.)

<table>
<thead>
<tr>
<th>Brand Name of Medication</th>
<th>On demand treatment and control of bleeding episodes</th>
<th>Routine prophylaxis to reduce the frequency of bleeding episodes</th>
<th>Perioperative management of bleeding</th>
<th>Control and prevention of bleeding episodes</th>
<th>Reduce the risk of joint damage without pre-existing joint damage</th>
<th>Other Refer to specific indication</th>
</tr>
</thead>
<tbody>
<tr>
<td>Aminocaproic Acid</td>
<td>✓</td>
<td></td>
<td></td>
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<td></td>
<td></td>
</tr>
<tr>
<td>Desmopressin Acetate (DDAVP)</td>
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</tbody>
</table>

**Hemophilia A:**
Individuals with hemophilia A with factor VIII coagulant activity levels greater than 5%. Will often maintain hemostasis in individuals with hemophilia A during surgical procedures and postoperatively when administered 30 minutes prior to scheduled procedure. Desmopressin acetate will also stop bleeding in individuals with hemophilia A with episodes of spontaneous or trauma-induced injuries such as hemarthroses, intramuscular hematomas or mucosal bleeding. In certain clinical situations it may be justified to try desmopressin acetate in individuals with factor VIII levels between 2% to 5%, however, these individuals should be carefully monitored.

**Von Willebrand’s Disease (Type I):**
Individuals with mild to moderate von Willebrand’s disease (Type I) with factor VIII levels greater than 5%. Desmopressin acetate will often maintain hemostasis in individuals with mild to moderate von Willebrand’s disease during surgical procedures and postoperatively when administered 30 minutes prior to the scheduled procedure. Will usually stop bleeding in individuals with mild to moderate von Willebrand’s with episodes of spontaneous or trauma-induced injuries such as hemarthroses, intramuscular hematomas or mucosal bleeding.
CLOTTING FACTOR REPLACEMENT THERAPY (cont.)

Resources:

Literature reviewed 09/26/17. We do not include marketing materials, poster boards and non-published literature in our review.

CLOTTING FACTOR REPLACEMENT THERAPY (cont.)

Resources: (cont.)

CLOTTING FACTOR REPLACEMENT THERAPY (cont.)

Resources: (cont.)

CLOTTING FACTOR REPLACEMENT THERAPY (cont.)

Non-Discrimination Statement:

Blue Cross Blue Shield of Arizona (BCBSAZ) complies with applicable Federal civil rights laws and does not discriminate on the basis of race, color, national origin, age, disability or sex. BCBSAZ provides appropriate free aids and services, such as qualified interpreters and written information in other formats, to people with disabilities to communicate effectively with us. BCBSAZ also provides free language services to people whose primary language is not English, such as qualified interpreters and information written in other languages. If you need these services, call (602) 864-4884 for Spanish and (877) 475-4799 for all other languages and other aids and services.

If you believe that BCBSAZ has failed to provide these services or discriminated in another way on the basis of race, color, national origin, age, disability or sex, you can file a grievance with: BCBSAZ’s Civil Rights Coordinator, Attn: Civil Rights Coordinator, Blue Cross Blue Shield of Arizona, P.O. Box 13466, Phoenix, AZ 85002-3466, (602) 864-2288, TTY/TDD (602) 864-4823, crc@azblue.com. You can file a grievance in person or by mail or email. If you need help filing a grievance BCBSAZ’s Civil Rights Coordinator is available to help you. You can also file a civil rights complaint with the U.S. Department of Health and Human Services, Office for Civil Rights electronically through the Office for Civil Rights Complaint Portal, available at https://ocrportal.hhs.gov/ocr/portal/lobby.jsf, or by mail or phone at: U.S. Department of Health and Human Services, 200 Independence Avenue SW., Room 509F, HHH Building, Washington, DC 20201, 1–800–368–1019, 800–537–7697 (TDD). Complaint forms are available at http://www.hhs.gov/ocr/office/file/index.html

Multi-Language Interpreter Services:

Spanish: Si usted, o alguien a quien usted está ayudando, tiene preguntas acerca de Blue Cross Blue Shield of Arizona, tiene derecho a obtener ayuda e información en su idioma sin costo alguno. Para hablar con un intérprete, llame al 602-864-4884.

Navajo: Díí kweʼé atah nilinígíí Blue Cross Blue Shield of Arizona haadá yiitʼéégo biñaʼdidíkidgo éí doodago Háída býá aniyeedíííí tʼáado leʼéé yínaʼdidíkidgo beehazʼanii hóól dií tʼáá hazaadíʼehí háká aʼdoowolgo bee haza doo baąh ilínígóó. Ataʼ halneʼíígíí kojí bíchíʼjí hodíílínii 877-475-4799.

Chinese: 如果您，或是您正在協助的對象，有關於插入項目的名稱 Blue Cross Blue Shield of Arizona 項目的問題，您有權利免費以您的母語得到幫助和訊息。洽詢一位翻譯員，請撥電話 在此插入數字 877-475-4799。

Vietnamese: Nếu quý vị, hay người mà quý vị đang giúp đỡ, có câu hỏi về Blue Cross Blue Shield of Arizona quý vị sẽ có quyền được giúp và có thể thông tin bằng ngôn ngữ của mình miễn phí. Đèn nói chuyện với một thông dịch viên, xin gọi 877-475-4799.

Arabic: إن كان لديك أي سؤال تساعد أسلحة ضمن بصمك الالكتروني للحصول على المساعدة والمعلومات Blue Cross Blue Shield of Arizona، للإجابة على كل سؤال، حجزة مترجم تم ترجمة من دون أي كلفة للتحدث مع مترجم أتصل ب 877-475-4799.
CLOTTING FACTOR REPLACEMENT THERAPY (cont.)

Multi-Language Interpreter Services: (cont.)

Tagalog: Kung ikaw, o ang iyong tinutulungan, ay may mga katanungan tungkol sa Blue Cross Blue Shield of Arizona, may karapatan ka na makakuha ng tulong at impormasyon sa iyong wika ng walang gastos. Uapang makauaspang isang tagasalin, tumawag sa 877-475-4799.

Korean: 만약 귀하 또는 귀하가 돕고 있는 어떤 사람이 Blue Cross Blue Shield of Arizona에 관해서 질문이 있다면 귀하는 그러한 도움과 정보를 귀하의 언어로 비용 부담 없이 얻을 수 있는 권리가 있습니다. 그렇게 통역사와 얘기하기 위해서는 877-475-4799로 전화하십시오.

French: Si vous, ou quelqu’un que vous êtes en train d’aider, a des questions à propos de Blue Cross Blue Shield of Arizona, vous avez le droit d’obtenir de l’aide et l’information dans votre langue à aucun coût. Pour parler à un interprète, appelez 877-475-4799.

German: Falls Sie oder jemand, dem Sie helfen, Fragen zum Blue Cross Blue Shield of Arizona haben, haben Sie das Recht, kostenlose Hilfe und Informationen in Ihrer Sprache zu erhalten. Um mit einem Dolmetscher zu sprechen, rufen Sie bitte die Nummer 877-475-4799 an.

Russian: Если у вас или лица, которому вы помогаете, имеются вопросы по поводу Blue Cross Blue Shield of Arizona, то вы имеете право на бесплатное получение помощи и информации на вашем языке. Для разговора с переводчиком позвоните по телефону 877-475-4799.

Japanese: ご本人様、またはお客様の身の回りの方でも、Blue Cross Blue Shield of Arizonaについてご質問がございましたら、ご希望の言語でサポートを受けたり、情報を入手したりすることができます。料金はかかりません。通訳をお話される場合、877-475-4799 までお電話ください。

Farsi:
آگر شما، یا کسی که شما به ایکم میکنید، سوال در مورد اطلاعات به وابسته به موضوع رایگان دریافت نمایید 877-475-4799.

Assyrian:


Thai: หากคุณ หรือคุณทำงานหรือผู้ที่คุณช่วยเหลือ Blue Cross Blue Shield of Arizona คุณมีสิทธิ์ที่จะได้รับความช่วยเหลือและข้อมูลในภาษาของคุณโดยไม่คิดค่าใช้จ่าย โทรที่ 877-475-4799.