

Chapter 5

HEMOSTATIC AGENTS

Steven W. Pipe, Manuel Carcao, Kim Chew, Radoslaw Kaczmarek, Steve Kitchen, Johnny Mahlangu, Margareth C. Ozelo, Ekawat Suwantaroj, Jerzy Windyga, Glenn F. Pierce, Alok Srivastava

CLOTTING FACTOR CONCENTRATES

Hemostatic Agents for people with hemophilia **WITHOUT** Inhibitors

Clotting Factor Concentrates (CFCs) are the treatment of choice for PWH as they are very safe and effective for treating and preventing bleeds. There are 2 main types:

1 Virally inactivated **plasma-derived products** (donated plasma)

2 **Recombinant products** made from engineered cells and recombinant technology



- CFCs are used to prevent (prophylaxis) or treat (episodic) bleeding events for patients with Hemophilia A and B
- WFH does not indicate a preference of one type over the other
- Product selection should be based on product safety and quality, purity, viral inactivation, efficacy and availability in country
- Extended half-life (EHL) products have a longer half life and were developed to reduce the treatment burden of prophylaxis and to maintain higher factor trough levels to improve bleed prevention

NON-FACTOR REPLACEMENT THERAPIES

Hemostatic Agents for people with hemophilia **WITH AND WITHOUT** Inhibitors

New and emerging non-factor therapies are being developed with alternative modes of delivery.

Emicizumab is the first and only licensed non-factor replacement therapy at time of this publication.

Emicizumab is recommended as regular prophylaxis for patients with hemophilia A, with and without an inhibitor.

Key benefits of Emicizumab

- ✓ subcutaneous route of administration
- ✓ long half-life
- ✓ high efficacy in bleed prevention
- ✓ reduction of the frequency of bleed episodes in patients with Hemophilia A*

* Emicizumab is not used in patients with Hemophilia B.

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OTHER PLASMA PRODUCTS

Other Hemostatic Agents for people with hemophilia **WITHOUT** Inhibitors

Cryoprecipitate and **Fresh Frozen Plasma** can carry an increased risk of transmission of viral pathogens.

In situations where these are the only available or affordable treatment options, certain steps can be taken to minimize the risk, such as:

- Quarantining plasma until the donor has been tested for HIV, HCV and hepatitis B
- NAT testing to detect viruses

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CFCs should be used in preference to cryoprecipitate or fresh frozen plasma due to concerns about quality, safety, and efficacy. However, the WFH recognizes the reality that they are still widely used in countries around the world where they are the only available or affordable treatment options.

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BYPASSING AGENTS

Other Hemostatic Agents for people with hemophilia **WITH** Inhibitors

Bypassing agents are recommended for the treatment and prevention of bleeding complications in patients with hemophilia A or B who develop FVIII or FIX antibodies.

Activated prothrombin complex concentrate (aPCC)

Recombinant activated factor VIIa (rFVIIa)

OTHER HEMOSTATIC AGENTS

Other Hemostatic Agents used in specific circumstances

Desmopressin (DDAVP) is recommended for patients with mild or moderate hemophilia A and carriers of hemophilia A as an option for treatment.

Antifibrinolytics, such as **tranexamic acid**, are valuable in controlling mucocutaneous bleeding and for dental surgery or eruption of teeth.

Epsilon aminocaproic acid (EACA) is another antifibrinolytic but is less widely used.