PART III: CONSUMER INFORMATION

BeneFIX®
Coagulation Factor IX (Recombinant)

This leaflet is part III of a three-part "Product Monograph and is designed specifically for Consumers. This leaflet is a summary and will not tell you everything about BeneFIX. Contact your doctor or hemophilia treatment centre if you have any questions about the drug.

ABOUT THIS MEDICATION

What the medication is used for:

- The control and treatment of bleeding and the prevention of bleeding in people with hemophilia B.
- Routine prophylaxis.
- BeneFIX has been approved for use in hemophilia B for adults and children.
- Ask your doctor if you have any questions about why BeneFIX has been prescribed for you.

What it does:

- People with hemophilia B (Christmas disease) are deficient in coagulation factor IX.
- Factor IX is a protein produced naturally in the body. It helps the blood form clots to stop bleeding.
- When the body does not make enough factor IX, and you become injured, your blood will not form clots as it should, and you may bleed into and damage your muscles and joints.
- Injections of factor IX are used to treat hemophilia B.
- BeneFIX is created using recombinant technology that allows it to be made without human blood or plasma products, making it naturally free of blood borne pathogens.

When it should not be used:

- Do not use BeneFIX for the treatment of other coagulation factor deficiencies (e.g., factors II, VII and X), for the treatment of hemophilia A, in patients with inhibitors to factor VIII, for the reversal of coumarin-induced anticoagulation, nor for the treatment of bleeding due to low levels of liver-dependent coagulation factors.
- Do not use BeneFIX if you are allergic to hamster proteins or any of the nonmedicinal ingredients listed below.
- Do not use BeneFIX after the expiry date (printed on the bottle). If you take this medicine after the expiry date has passed, it may not work well.
- Do not use BeneFIX if the packaging is torn or shows signs of tampering.

If you are not sure whether you should use BeneFIX, talk to your doctor.

What the medicinal ingredient is:

- Recombinant coagulation Factor IX (Nonacog alfa)

What the important nonmedicinal ingredients are:

- Glycine
- Sucrose
- Histidine
- Polysorbate 80
- Sodium chloride solution

What dosage forms it comes in:

BeneFIX comes as a white powder in a glass vial, nominally containing 250, 500, 1000, 1500, 2000 and 3000 IU per vial. The actual amount of Factor IX is stated on the label of each bottle. BeneFIX must be reconstituted (dissolved) with the diluent syringe and the product contains approximately: 50, 100, 200, 300, 400 and 600 IU/mL, respectively.

WARNINGS AND PRECAUTIONS

Serious Warnings and Precautions

STOP taking BeneFIX and contact your doctor immediately if

- **You experience allergic reactions such as skin rash, itching, chest tightness, wheezing, dizziness, hives, faintness, rapid heartbeat, blurred vision, shortness of breath, and/or a swollen face. Severe allergic reactions to BeneFIX and other Factor IX products have been reported.**

Contact your doctor immediately if

- **Your bleeding does not stop as expected**

BEFORE you use BeneFIX talk to your doctor or hemophilia treatment centre if you:

- Are pregnant or planning to become pregnant
- Are breast feeding or planning to breast feed
- Are at risk of developing blood clots
- Have liver disease
- Have recently had surgery or are planning to have surgery, including dental surgery

INTERACTIONS WITH THIS MEDICATION

Drugs that may interact with BeneFIX include:

- There are no known interactions of BeneFIX with other medications.
- Tell your doctor or pharmacist if you are taking any other medicines, including any you buy without a prescription, including natural health products.

PROPER USE OF THIS MEDICATION

Usual dose:

- Your doctor will decide the dose of BeneFIX you will receive.
- BeneFIX is injected directly into the bloodstream.
- The dose, duration and frequency of infusion will depend on your individual needs for replacement factor IX and may be influenced by your age, weight, activity level and severity of bleed.
- Your doctor may periodically need to check laboratory blood test results following infusion to be sure that blood level of factor IX is high enough to allow satisfactory blood clotting.
- If you have been using plasma-derived factor IX, the dose of BeneFIX may differ from the dose of plasma-derived factor IX.
- Do not lower the dose of BeneFIX without checking with your doctor, unless you are having an allergic reaction.

Overdose:

- No symptoms of overdose are known.

For management of a suspected overdose, contact your health care practitioner or the nearest hospital emergency department or your nearest Poison Control Centre immediately, even though you may not feel sick.

Missed Dose:

- If you miss a dose of this medicine, check with your doctor as soon as possible for instructions.

Preparation and Administration:

RECONSTITUTION

Always wash your hands before performing the following procedures. Aseptic technique (meaning clean and germ-free) should be used during the reconstitution procedure. All components used in the reconstitution and administration of this product should be used as soon as possible after opening their sterile containers to minimize unnecessary exposure to the atmosphere.

BeneFIX is administered by intravenous (IV) infusion after reconstitution with the supplied diluent (0.234% sodium chloride diluent).

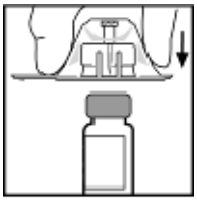
1. If refrigerated allow the vial of lyophilized BeneFIX® and the pre-filled diluent syringe to reach room temperature.

2. Remove the plastic flip-top cap from the BeneFIX® vial to expose the central portions of the rubber stopper.

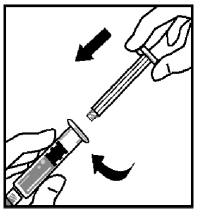
3. Wipe the top of the vial with the alcohol swab provided, or use another antiseptic solution, and allow to dry. After cleaning, do not touch the rubber stopper with your hand or allow it to touch any surface.

4. Peel back the cover from the clear plastic vial adapter package. **Do not remove the adapter from the package**.

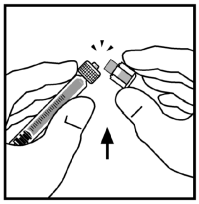
5. Place the vial on a flat surface. While holding the adapter in the package, place the vial adapter over the vial. Press down firmly on the package until the adapter snaps into place on top of the vial, with the adapter spike penetrating the vial stopper. Leave the adapter package in place.



6. Grasp the plunger rod as shown in the diagram. Avoid contact with the shaft of the plunger rod. Attach the threaded end of the plunger rod to the diluent syringe plunger by pushing and turning firmly.

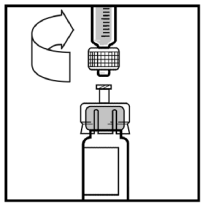


7. Remove the tamper-resistant, plastic-tip cap from the diluent syringe by bending the cap up and down to break the perforation. Do not touch the inside of the cap or the syringe tip. The cap may need to be replaced, so place the cap on its side on a clean surface in a spot where it would be least likely to become environmentally contaminated.

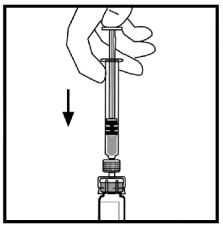


8. Lift the package away from the adapter and discard the package.

9. With the vial on a flat surface, connect the diluent syringe to the vial adapter by inserting the tip of the syringe into the adapter opening while firmly pushing and turning the syringe clockwise until the connection is secured.



10. Slowly depress the plunger rod to inject all the diluent into the BeneFIX® vial.

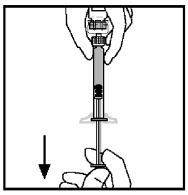


11. With the syringe still connected to the adapter, **gently** swirl the contents of the vial until the powder is dissolved.

12. Inspect the final solution for specks before administration. The solution should appear clear and colorless.
Note: If you use more than one vial of BeneFIX® per infusion, reconstitute each vial by following the previous instructions.

13. Ensuring that the syringe plunger rod is still fully depressed, invert the vial. Slowly draw the solution into the syringe.

Note: If you prepared more than one vial of BeneFIX®, remove the diluent syringe from the vial adapter, leaving the vial adapter attached to the vial. Quickly attach a separate large luer lock syringe and draw back the reconstituted contents as instructed above. Repeat this procedure with each vial in turn. Do not detach the diluent syringes or the large luer lock syringe until you are ready to attach the large luer lock syringe to the next vial adapter.



14. Detach the syringe from the vial adapter by gently pulling and turning the syringe counterclockwise. Discard the vial with the adapter attached.

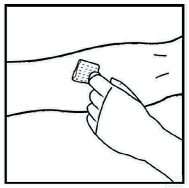
Note: If the solution is not to be used immediately, the syringe cap should be carefully replaced. Do not touch the syringe tip or the inside of the cap.

BeneFIX should be administered within 3 hours after reconstitution. The reconstituted solution may be stored at room temperature prior to administration.

ADMINISTRATION (Intravenous Injection)

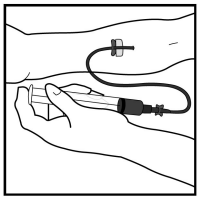
1. Attach the syringe to the luer end of the infusion set tubing provided.

2. Apply a tourniquet and prepare the injection site by wiping the skin well with an alcohol swab provided in the kit.



Once you learn how to self infuse you can follow the instructions in this insert.

3. Perform venipuncture. Insert the needle on the infusion set tubing into the vein, and remove the tourniquet. The reconstituted BeneFIX product should be injected intravenously over several minutes. The rate of administration should be determined by the patient's comfort level.



Reconstituted BeneFIX **should not be administered in the same tubing or container with other** medicinal products.

Following completion of BeneFIX treatment, remove the infusion set and discard. Dispose of all unused solution, empty vial(s), and used needles and syringes in an appropriate container for throwing away waste that might hurt others if not handled properly.

Agglutination of red blood cells in the tubing/syringe has been reported with the administration of BeneFIX®. No adverse events have been reported in association with this observation. To minimize the possibility of agglutination, it is important to limit the amount of blood entering the tubing. Blood should not enter the syringe.

Note: If red blood cell agglutination is observed in the tubing or syringe, discard all material (tubing, syringe and BeneFIX® solution) and continue administration with a new package.

SIDE EFFECTS AND WHAT TO DO ABOUT THEM

- During your treatment with BeneFIX, your blood will be checked for inhibitors to factor IX activity. Inhibitors are antibodies against Factor IX, which are made by your immune system. The inhibitors stop the factor IX from working as well as it used to.

Tell your doctor immediately if you are using increasing amounts of BeneFIX in order to control a bleed.

- Injection of any medicine intravenously may have side effects. Often they are not serious but sometimes they can be. You may need medical treatment if you experience some of the side effects in the table below.

SERIOUS SIDE EFFECTS, HOW OFTEN THEY HAPPEN AND WHAT TO DO ABOUT THEM

Symptom / effect	**STOP taking BeneFIX and call your doctor immediately**
The following side effects could mean you are having an allergic reaction. **These side effects are rare.**	
• A skin rash	✓
• Itching	✓
• Chest tightness	✓
• Wheezing	✓
• Dizziness	✓
• Hives	✓
• Faintness	✓
• Rapid heartbeat	✓
• Shortness of breath	✓
• A swollen face	✓
• Blurred vision	✓

Tell your doctor if you notice any of the following side effects and they worry you:

- *Headache*
- *Runny or blocked nose*
- *Light-headedness*
- *Fever*
- *Chills*
- *Flushing*
- *Nausea*
- *Vomiting*
- *Diarrhea*
- *Feeling tired, drowsy or a lack of energy*
- *Discomfort or swelling at the injection site*
- *Altered taste*
- *Coughing*
- *Burning sensation in the jaw or skull*
- *Changes in your vision*

These are all mild side effects of BeneFIX injection and will usually disappear on their own. Tell your doctor if you are concerned or if they continue.

This is not a complete list of side effects. For any unexpected effects while taking BeneFIX, contact your *doctor or hemophilia treatment centre.*

HOW TO STORE IT

Before preparation (BeneFIX powder):

DO NOT freeze.

BeneFIX can be stored at room temperature (below 30°C) or under refrigeration. Store the diluent syringe between 2°C to 30°C. Throw away any unused BeneFIX and diluent after the expiration date.

Keep BeneFIX (and needles) where young children cannot reach it.

BeneFIX must be used by the expiry date on the label. Do not use BeneFIX beyond the date (month and year) on the label, even if it has been stored properly.

After preparation (BeneFIX solution):

To avoid bacterial contamination of the solution, use the reconstituted BeneFIX as soon as possible or within 3 hours.

REPORTING SUSPECTED SIDE EFFECTS

You can report any suspected adverse reactions associated with the use of health products to the Canada Vigilance Program by one of the following 3 ways:

- **Report online at www.healthcanada.gc.ca/medeffect**
- **Call toll-free at 1-866-234-2345**
- **Complete a Canada Vigilance Reporting Form and:**
  - **Fax toll-free to 1-866-678-6789, or**
  - **Mail to:**
    Canada Vigilance Program
    Health Canada
    Postal Locator 0701E
    Ottawa, Ontario
    K1A 0K9

Postage paid labels, Canada Vigilance Reporting Form and the adverse reaction reporting guidelines are available on the MedEffect™ Canada Web site at www.healthcanada.gc.ca/medeffect.

NOTE: Should you require information related to the management of side effects, contact your health professional. The Canada Vigilance Program does not provide medical advice.

MORE INFORMATION

This document plus the full product monograph, prepared for health professionals, can be found at: www.pfizer.ca or can be obtained by contacting the sponsor, Pfizer Canada Inc., at 1-800-463-6001 (Medical Information).

This leaflet was prepared by Pfizer Canada Inc.

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