



9128 N. Lindbergh Dr., Peoria, IL, 61615 Phone: 309-692-5337 [www.ilbcdi.org](http://www.ilbcdi.org)

# Inhibitors Fact Sheet

## What is an inhibitor?

- Inhibitors are antibodies that are developed by the immune system (the body's natural defense system) because it sees the clotting factor that is being infused as a foreign substance. The immune system then thinks that this foreign substance needs to be destroyed. The antibodies that are formed neutralize the factor before the factor has time to work

## What are the symptoms of an inhibitor?

- A bleed does not quickly improve with a normal dose of factor
- A normal dose of factor treatment seems less and less effective
- Bleeding continues to be more and more difficult to control
- Breakthrough bleeding despite compliance with prophylactic infusions

## Who is at risk for developing an inhibitor?

- Inhibitors occur more often in individuals with severe Hemophilia than with moderate or mild, and more often in Hemophilia A than in Hemophilia B
- Approximately 25-30% of severe Hemophilia A patients develop inhibitors
- Only 2-5% of Hemophilia B patients develop inhibitors
- Most inhibitors develop during the first 50 exposures (or infusions) of factor

## How is an inhibitor diagnosed?

- A blood test called a **Bethesda Assay** is performed. The amount of inhibitor in a person's blood is measured in **Bethesda Units (BU)**.
  - High Titer = More than 5 BU. These inhibitors act more strongly and more quickly destroy infused factor. Must use a bypassing agent to stop bleeding
  - Low Titer = Less than 5 BU. These inhibitors act more weakly and more slowly destroy infused factor. May be able to use factor VIII (8) or factor IX (9) to overcome the inhibitor.

## **What are the treatment options for inhibitors?**

- Treating bleeds
  - Recombinant factor VIIa (NovoSeven™)
  - Activated prothrombin complex concentrates (aPCCs) (FEIBA™)
- Getting rid of the inhibitor
  - Immune tolerance induction therapy (ITI)
    - High dose factor VIII (8) or IX (9)
  - Immunosuppression (selected cases)

## **What is immune tolerance induction therapy (ITI)?**

- Daily high doses of factor over a period of weeks, months, or years
- Given with the goal of “teaching” the body to tolerate the factor and not see that factor product as foreign
- Often the patient is given bypassing agents as their prophylaxis regimen in addition to the high doses of factor product
  - Given to help prevent or control bleeding as the ITI will likely not give much factor coverage until the body learns to not recognize this as foreign anymore