

Chapter 8

INHIBITORS TO CLOTTING FACTOR

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Inhibitors: WHAT YOU NEED TO KNOW

What are inhibitors?

Inhibitors are antibodies to factor treatment that some people with hemophilia develop. Inhibitors are a serious complication of treatment for hemophilia. They can prevent your usual clotting factor replacement therapy from working to stop your bleeds.

How are inhibitors treated?

If you develop an inhibitor, you will be treated by the medical team at your Hemophilia Treatment Center with medications to stop your body from making inhibitors. Communication with your medical team is very important.



When should I be tested for inhibitors?

Inhibitor testing is done through a special blood test, called a Nijmegen-modified Bethesda assay. You should be tested for inhibitors at specific times during your treatment.

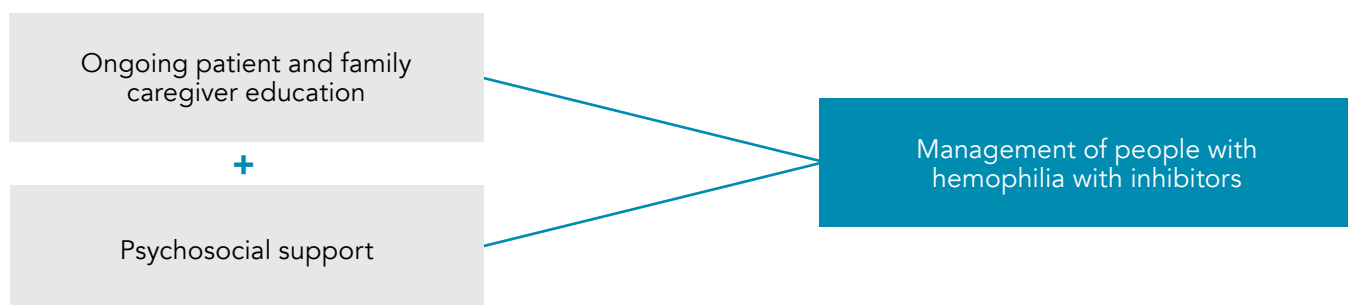
When should I get tested for inhibitors ?

- ✓ After you start treatment with CFCs and then every year
- ✓ After intensive CFC exposure, e.g., daily exposure for more than 5 days
- ✓ Before surgery
- ✓ If your CFCs no longer stop you from bleeding

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Good communication with your Hemophilia Treatment Center, and a well-coordinated plan of care is essential for the best care of people with hemophilia who develop inhibitors

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People with Hemophilia A and B can both develop inhibitors, but there are differences in inhibitor incidence, management, and response to treatment between the two.

Hemophilia A and inhibitors

approximately

30% incidence among previously untreated **severe Hemophilia A**

Inhibitors are more frequent in persons with severe hemophilia A than in those with moderate or mild hemophilia

Hemophilia B and inhibitors

approximately

5% incidence among **severe Hemophilia B**

Inhibitors are uncommon in patients with hemophilia B

Treatment is based on managing your bleeds while you have an inhibitor, and treating the inhibitor until it disappears

MANAGEMENT OF BLEEDS IN PWH WITH INHIBITORS

If you do get a bleed while you have an inhibitor, your physician will decide the best treatment to stop your bleed. This depends on the inhibitor titer, your clinical response to the treatment product, previous infusion reactions, the site and nature of the bleed, and the products available in your country. Treatment may include one of the following therapies:

Management of bleeds in PWH with inhibitors

- ✓ Clotting factor concentrate replacement therapy
- ✓ Bypassing agent (recombinant factor VIIa or activated prothrombin complex concentrate)
- ✓ Emicizumab (for Hemophilia A only)

TREATMENT TO ERADICATE THE INHIBITOR

Immune tolerance induction therapy, known as ITI, is often used to eradicate inhibitors. ITI is a regimen of clotting factor replacement therapy given over a longer period of time. The exact dose and frequency is individualized by the physician. Inhibitor eradication by ITI is successful in 70%-80% of patients with severe hemophilia A, but has a lower success rate in patients with hemophilia B.