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ALL ABOUT INHIBITORS

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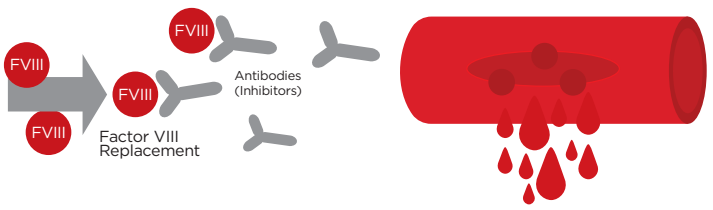
WHAT IS **AN INHIBITOR?**[1,2]

An inhibitor is an immune system response to infused clotting factor concentrates, which renders standard replacement therapy ineffective. The Inhibitor:

- Identifies factor concentrates as a foreign substance
- Attacks factor concentrates
- Neutralizes factor activity
- Makes factor ineffective in stopping bleeding.



This can develop in about 20–30% of patients who have severe Hemophilia A and 2–5% of Hemophilia B

WHEN DO **INHIBITORS DEVELOP?**[1]

Inhibitor can develop only after a Persons with Hemophilia (PwH) has been treated with replacement clotting factor. Most inhibitors occur early in life, usually within the first 50 days of treatment with replacement clotting factor.

WHAT IS THE **IMPACT OF AN INHIBITOR?** [3,4]

Patients with inhibitors do not bleed more frequently than other PwH, but their bleeds are more difficult to manage because they do not always respond to standard treatment. As a result, inhibitor patients have an increased risk of developing complications such a joint damage. Initially, a joint bleed or hemarthrosis occurs. Joint bleeding usually occurs in the knees, elbows and ankles and it can be very painful. Repeated bleeding into a joint may cause permanent joint damage, termed arthropathy, that results in a loss of normal joint function and, often, visible deformity.

WHO IS AT RISK OF **DEVELOPING INHIBITORS?**[3]

Anyone with Hemophilia can develop an inhibitor. However, there are some factors that may increase your risk. These include:

Genetic Risk Factors:

- Disease severity (increased in people with severe disease)[3]
- Certain types of genetic mutation[3,4]
- Family history of inhibitors[4]
- Race and ethnicity (increased in people of African or Hispanic origin)[3,5]

Treatment Related Risk Factors

- Exposure to exogenous FVIII
- Periods of intensive treatment

WHEN SHOULD I GET **TESTED FOR INHIBITORS?**[4]

A person may not always have clear symptoms associated with the presence of an inhibitor. In cases where individuals are symptomatic, an inhibitor is usually suspected when bleeding does not stop even after treatment doses.

HOW IS **INHIBITOR CONFIRMED?**[4]

Once an inhibitor is suspected, the only way to confirm a case is through laboratory testing during a regular physician visit. Confirmation is made using a blood test called the “Bethesda (inhibitor) Assay.” This test measures the presence and amount of antibodies directed against a coagulation factor in a person's blood after being exposed to factor.

WHAT ARE THE POSSIBLE **BETHESDA TEST RESULTS?**[4]

If the results of the Bethesda Assay are positive, it means that there is a detectable level of antibodies working against the deficient coagulation factor being produced as a result of being treated with factor. The degree to which this affects a person is measured in "units." Inhibitors are classified into two categories based upon unit levels achieved. Those with a ‘5 or higher’ Bethesda units test result are classified as having a "high-responding" inhibitor level; those who measure ‘below 5 Bethesda units’ despite repeated exposure to factor concentrate, are classified as having a "low-responding" inhibitor level.

People with ‘high-responding’ inhibitor levels often have quick and strong immune system responses directed against Factor (VIII or IX), meaning that the inhibitor level can increase to very high levels of antibodies.

In cases where a person has a ‘low-responding’ inhibitor level, the body's immune response to factor is slow—and it produces a persistently low level of antibodies despite the person's continual exposure to factor concentrate.

WOULD A PERSON WITH INHIBITOR POSITIVE, **ALWAYS REMAIN INHIBITOR POSITIVE?** [4]

A positive test result does not mean a person will always have an inhibitor. Treatments may work to keep the inhibitor in check. In certain cases, inhibitor levels may drop over a period of months to years, to an undetectable range. Some inhibitors may reappear with further exposure, often months later. These cases are often a special type of ‘low-responding’ inhibitor cases, classified as ‘transient’ inhibitors.

Inhibitors pose a very challenging medical complication because of the uncertainty of how they can spontaneously disappear, respond to various treatments or in other cases require ongoing management for years.

TREATMENT OPTIONS FOR **PATIENTS WITH INHIBITORS.** [3]

Management of bleeding in patients with inhibitors must be in consultation with a center experienced in the management of such patients and all serious bleeds should be managed in these centers. Depending on the inhibitor levels in the body they are treated with either increased dose of factors or bypassing agents or both together. Bypassing agents for Hemophilia inhibitor patients include recombinant Factor VII and prothrombin complex concentrates, including the activated ones such as APCC (Activated Prothrombin Coagulation Complex).

DO'S AND DON'TS[4,5]

DO'S	DON'TS
Do regular exercise to stay fit. Discuss with your doctor to know about which exercises to undertake.	Avoid high-contact sports such as football, boxing, volley ball, kabaddi, cricket and wrestling, etc.
Be prepared with protective gear that is appropriate for a sport or activity for e.g. helmet, knee cap, shoes while cycling.	Don't miss your doses of your medication (factors or other appropriate products) prescribed by your doctor.
Promptly report to your treating doctor any headache, tingling in the joint, change in voice, breathing difficulty, blackish stools, blood mixed stools, etc.	Don't ignore a possible bleed in joint, muscle, or for that matter anywhere in the body.
Keep a regular health check up visit with your doctor to monitors your treatment and to check on joints and muscles status.	Avoid Aspirin. This will increase bleeding tendency.
While taking vaccinations, get injected under the skin (subcutaneous), not into the muscles (intra-muscular).	Don't tie your bandage too tight.
Discuss with your doctor about the healthy body weight suitable for you. Being overweight puts undue stress on joints and would increase the need for factor in case of bleeds.	Avoid furniture with sharp edge at home.
If you are travelling, carry your first aid kit with badge, ice pack, injection syringes, factors (as per doctors advice) etc.	
If you are going out of your home for vacation, carry your prescriptions and make a note of hemophilia treatment centre at the place you are visiting.	
If you happen to meet any doctor other than the on who is treating you, disclose to him/her that you have hemophilia and show your prescriptions. Discuss with them that no intramuscular or intra-articular (in to the joint) injections to be given.	
Follow the guidance of your treating physician	

If your doctor has suspected inhibitors and recommended you inhibitor testing, do not delay. Purna Samparq team can support you for inhibitor testing,

Contact The Team now.

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18001034260 purna.samparq@indegene.com

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CONTACT US: Takeda Biopharmaceuticals India Pvt. Ltd.
(formerly known as Baxalta Bioscience India Pvt. Ltd.)
6th Floor, Tower C, Building No. 8, DLF Cyber City, DLF Phase II, Gurgaon, Haryana- 122001, India.
Tel: +91-124-455 9100 | www.takeda.com

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