INDIANA HEMOPHILIA & THROMBOSIS CENTER, INC.

Indiana's Only Federally Recognized Comprehensive Hemophilia Center
8402 Harcourt Road, Suite 500, Indianapolis, IN 46260 | T: 317-871-0000 1-877-CLOTTER (256-8837)

CONSENT FOR PROPHYLAXIS

You/your child has hemophilia (either factor VIII or IX deficiency). Programs of infusions given on a regular basis, prophylaxis, have been developed and have been shown to be effective in the prevention of the majority of hemophilia-related joint disease.

You/your child have been advised to have a central venous access device (CVAD) such as Hickman catheter, port-a-cath or port, placed in order to administer the infusions for this program. The placement of this device will be done under anesthesia and will require a short hospitalization (approximately three days), during which time you/your child will receive infusions to prevent bleeding after the surgery, as well as teaching related to the care and use of the CVAD. The surgeon who places the central venous access device will discuss with you the risks associated with the surgical procedure. After discharge from the hospital, you/your child will require further infusions to allow healing and prevent bleeding related to the surgery.

Purpose of the prophylaxis program:
The purpose of this program is to prevent patients with severe hemophilia (levels less than 1%) from developing joint disease related to joint bleeding. Recurrent bleeding into joints leads to damage, which may lead to disability and poor function of the affected joint and extremity. Severe joint destruction may require joint replacement as an adult in order to improve function.

Important issues related to prophylaxis:
Infusions on a regular basis will raise the factor level so that bleeding episodes are not as frequent. Not all bleeding episodes will be prevented on this program; bleeding episodes related to injury may still occur. However spontaneous bleeding episodes should not occur. Any spontaneous bleeding episode should be reported promptly to the hemophilia center. The dosage of factor concentrate or the frequency of infusions may need to be changed to meet your/your child's needs.

If you/your child has already developed a target joint prior to the start of this prophylaxis program, the progression of degenerative joint disease in that target joint **may not** be prevented. However, you/your child might expect to prevent the development of other target joints through this prophylaxis program.

Approximately 25% of patients with severe factor VIII deficiency, and less than 3% of patients with severe factor IX deficiency develop an inhibitor. An inhibitor is a antibody produced against the clotting factor you/your child are missing. The inhibitor prevents the ability of the factor concentrate from working, which may complicate the treatment of bleeding episodes and the prophylaxis program. The symptoms associated with an inhibitor are an increased frequency of bleeding episodes and/or poor response during these bleeding episodes to infusion of factor concentrate. It is not known why some patients develop inhibitors and others do not, but it appears to be related to individual variables. The use of a prophylaxis program **should not** increase the frequency of inhibitor development.

The presence of a CVAD requires the use of antibiotics before and possibly after dental work. This therapy is to prevent a possible infection of the CVAD related to the dental work. The staff of the hemophilia center or your dentist will provide you with information about the type and dosage of antibiotics required.

The CVAD may develop problems such as infection or a clot. The symptoms of an infection may include fever and chills, especially in relation to flushing or infusion through the port. The symptoms of a clot may include a feeling of resistance during an infusion or an inability to flush or infuse through the CVAD.

Under the best circumstances, central venous access devices may be expected to last about three to four years before requiring removal and/or replacement. About 20-25% of CVADs may develop the problems described above and may require interventions to save the device, or removal and/or replacement if the problem cannot be resolved.

A program to follow your/your child's progress has been developed by the hemophilia center, and you/your child are requested to participate in follow-up to assure appropriate care is provided. Your doctor and nurse will discuss this follow-up program with you. However, during prophylaxis therapy you/your child will have their factor levels monitored on a regular basis either at the IHTC or by a visiting nurse. You will be asked to return to the hemophilia center for ongoing assessments approximately every 6 months.

Benefits:
It is our hope to prevent joint disease or delay the development of target joints in you/your child.

Risks, discomforts and precautions:
Many of the risks associated with this program have been described above and include:

1. Bleeding associated with surgery
2. Need for antibiotic prophylaxis in association with dental work
3. Infection of the CVAD
4. Development of a clot in the CVAD

The side effects of factor concentrate are well known to you/your family. The most common problems and issues related to prophylaxis have been presented. Side effects unknown to and unexpected by the physicians may occur. All forms of toxicity are monitored to the greatest extent possible, and the program will be stopped if problems develop or you/your child wish to withdraw.

Alternatives:
The alternative to the prophylaxis program is the treatment of bleeding episodes when they occur which is referred to as episodic or on-demand therapy.

Consents:
I/my child have hemophilia that will be treated with a prophylaxis program. No compensation is available if an injury occurs due to this program. If emergency medical treatment is required as a result of treatment, I will be responsible for the cost of such treatment. If I have questions regarding this program or wish to terminate participation, I can reach my hematologist or nurse by calling the IHTC at 1-877-256-8837 24 hours per day, 7 days per week.

I acknowledge receipt of a copy of this informed consent statement.

_____ _____
Patient's name & signature Date

If patient is minor,

_____
Child's Name

_____ _____
Parent/Guardian's name & signature Date

_____
Witness name & signature