TAKE CONTROL

Transitioning to Adult Care

"What should I know?"

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Take Control: Transitioning to Adult Care *"What should I know?"*

Name: _____

Date of Birth: _____

Diagnosis: _____

Level of Factor % (Factor VIII = 8, Factor IX = 9): _____

How would you explain your diagnosis to others?

_____

_____

_____

_____

Bleed Detection and Assessment:

List the major signs and symptoms you may experience with a:

1) Joint or Muscle Bleed: _____
2) Head Injury: _____
3) Neck Bleed: _____
4) Chest or Abdominal Bleed: _____

Always remember to call your hemophilia treatment centre (Phone number: _____) when you have a bleed for a follow-up plan.

There are some bleeds that are not as common in children/adolescents; however you may experience them as you age. If you should notice blood in your urine, stool, semen and bleeds associated with sexual activity, contact your hemophilia treatment centre for medical advice.

Treatment Protocol:

Do you have a factor first card? Yes ___ No ___

Do you wear a medical alert? Yes ___ No ___

Can you use DDAVP to treat bleeds? Yes ___ No ___

What is your brand of factor? _____

How do you document your infusions/treatments? (For example: paper diary, electronic diary)

_____

Are you on prophylaxis? Yes ___ No ___

If Yes:

Dose: _____ units Frequency (Infusion days): _____

If you had a major bleed, what product and dose would you use?

_____

If you had a moderate/minor bleed, what product and dose would you use?

_____

Always remember to call your hemophilia treatment centre (Phone number: _____) when you have a bleed for a follow-up plan.

Are you aware of your half-life (time in hours)? _____

Are you aware of your recovery (factor level 15-30 minutes post infusion of factor)? _____ %

Past History:

Have you ever had an inhibitor? Yes ___ No ___
Do you have an inhibitor currently? Yes ___ No ___

What are signs that you may have developed an inhibitor?

_____

_____

Have you ever experienced an intracranial hemorrhage (brain bleed)? Yes No ___

- If yes, when? _____
- How has this impacted your daily routine? (If applicable) _____

_____

_____

Do you have a target joint(s)? If so, where? _____

_____

Genetics of Hemophilia:

Do you have any relatives that also have hemophilia? (Family History)

_____

_____

_____

_____

For future knowledge, if you were to have children, what are the chances of having a baby boy with hemophilia? A baby girl who is a carrier?

_____

_____

General Wellness:

Routine Dental Care: Brush, floss and see a dentist routinely!

What is your routine treatment prior to a dental cleaning? (Check the appropriate boxes).

- ☐ None
- ☐ Tranexamic acid
- ☐ Factor
- ☐ DDAVP (Subcutaneous, Intranasal)

Surgeries/Dental Procedures: Surgeries and dental procedures can vary in the requirements needed. Please contact your hemophilia treatment centre well in advance in order for the appropriate pre-operative plan to be put in place. Do **NOT** undergo any procedure prior to communicating with your team.

Travel: Prior to planning a trip/vacation, go online to World Federation of Hemophilia website and find the local hemophilia treatment centre you will be closest to. Notify your team of your travel place and dates well in advance in order for a travel letter to be prepared and any prescriptions/factor to be ordered.

Physical Activity: Regular activity provides many benefits. It reduces fat composition, encourages weight loss and improves endurance. In addition to the cardiovascular benefits of exercise, musculoskeletal health is also enhanced. Joint health is particularly significant for patients with hemophilia. Reach out to your physiotherapist for recommendations on physical activity and sports choices for you.

What activities/sports do you currently participate in? _____

_____

Social Life: As a young adult, you have the opportunity to make individual decisions that can impact your overall health and wellbeing. Adolescence and young adulthood is a time of exploration, developing new boundaries and self discovery. There are some life choices that will put you at higher risk of bleeding in ways that you may not think of. For example, excessive consumption of alcohol can impair decision-making, and can lead to impaired gait resulting in falls or head injuries that may be mistaken for intoxication. It is important that those you socialize with are aware of your condition, as there may be times when you rely on their ability to advocate for care on your behalf.

Tattoos and piercing can put you at additional risk of bleeding. If these choices are ones in which you are considering, please contact your hemophilia treatment centre in order to discuss. Our goal of care is to work with you to minimize risks associated with your life choices.

Do you have any comments or questions that would be of benefit to mention to your hemophilia treatment team?

_____

_____

Do you know anyone with a similar bleeding disorder? Are you interested in receiving information on upcoming activities with others with similar conditions?

_____

_____

_____

Transitioning from pediatric to adult care can result in anxiety and be very overwhelming for you and your parents. This is a natural experience; however when you are knowledgeable about your condition and management this can assist in decreasing these feelings. Adult care focuses on you as the patient; however your family is always welcome as a support if you so choose.

Always remember your treatment plan changes as your lifestyle changes, and your hemophilia treatment centre will work with you through these life changes and adapt your plan accordingly. It is extremely important to remain connected and engaged with your hemophilia treatment team.

Important Information to Know About Your Condition

Bleed Detection and Assessment:

Signs & Symptoms of Bleeds

Joint/Muscle Bleeds:

- Joint and muscle bleeds may not have obvious physical signs when the bleed begins. You may be reluctant to use the limb, may be unable to bear weight due to pain and may have limited range of motion. If the bleed is in a joint or muscle, the area around the joint may begin to swell and the skin over the joint may be warm to the touch. Additionally, pain will continue to worsen as the bleeding continues.
- Joints that are most often affected are the knees, ankles, and elbows (these are called the index joints). However, muscle bleeds can occur anywhere in the body (more commonly in flexor muscles; for example, calf, hamstring).
- **Seek medical attention immediately** if you experience numbness, decreased circulation (limb is cool to touch) and decreased nerve innervation (decreased sensation to touch, tingling of limb). As these are warning signs that the bleed may have progressed into compartment syndrome (compression of nerves and main blood vessels of the area).

Head Injury:

- A head injury that can lead to an intracranial hemorrhage (bleed in the brain) is considered life-threatening and **you must seek medical attention immediately**. Symptoms to be concerned about are decreased level of consciousness or change in level of consciousness, lethargy, irritability or change in behaviours, nausea and/or vomiting, headache, and an unsteady gait. Occasionally a seizure may be a presenting symptom of a bleed inside the brain.

Neck Bleeds:

- A neck injury/bleed could be life-threatening and **you must seek medical attention immediately**, as the bleed can compromise your ability to breathe.
- Symptoms are pain and tenderness with touching the area and swallowing, as well as difficulty with swallowing and breathing. If the neck injury has resulted in bleeding into the spinal cord in the neck, you could experience numbness or pain in the arms or legs, or even weakness of these parts of the body.
- Sometimes with a neck injury, you could also stop passing urine (a neurologic symptom).

Chest/Abdominal Bleeds:

- A chest or abdominal injury can lead to bleeds and as a result are also life-threatening, so **medical attention should be sought immediately**. Usually the main symptom is pain at the site of trauma, and visible bruising may be present.

Treatment Management:

The main treatment for hemophilia is an infusion of the missing or deficient clotting factor (Factor (F)VIII or FIX). Factor concentrates are infused into a vein, usually in the arm. Hemophilia A is treated with clotting factor VIII concentrate; Hemophilia B is treated with clotting factor IX concentrate. The clotting factor is used to increase the level of the deficient factor to a level that initially stops bleeding and later prevents bleeding from recurring. When FVIII is infused 1U/kg, this increases the level by 2%; when FIX is infused 1U/kg, this increases the level by 1% (for example 50U/kg increases FVIII to 100%; 50U/kg increases FIX to 50%). This is known as a recovery.

PK stands for pharmacokinetics. Pharmacokinetics describes what the body does to the factor – how it absorbs, distributes, metabolizes and eliminates the factor infused. The half-life of clotting factor is an average time, when referring to the hemophilia population, because everyone's half-life varies depending on their own PK. Half-life refers to the time it takes for half of the infused factor circulating in the blood to disappear from your bloodstream. On average, the half-life for factor VIII is 8-12 hours; the half-life for factor IX is 18-24 hours.

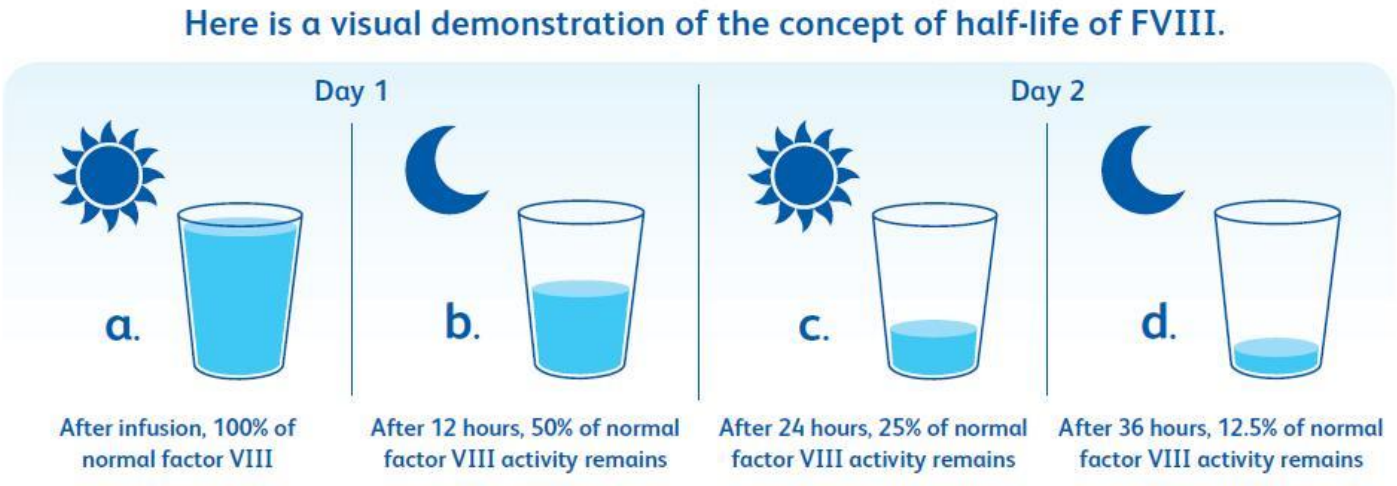


Image adapted and reproduced with permission from the Canadian Hemophilia Society (CHS) All About Hemophilia: A Guide for Families.

Prophylaxis treatment is regularly scheduled infusions of clotting factor concentrate (FVIII or FIX) to prevent bleeds. The number of prophylaxis infusions a week will vary between people depending on PK. The number of infusions will vary from one infusion a week to infusions given every day. Most people with severe hemophilia receiving currently available factor concentrates will, over time, receive infusions every other day. The reason for every other day infusions is to keep the factor level above 1%, thus decreasing the risk of bleeding.

Some people with mild or moderate hemophilia A are able to use Desmopressin (DDAVP) for the treatment of mild/moderate bleeds. In order for these people to use DDAVP, we first need to do what we call a DDAVP challenge to see if DDAVP increases their FVIII levels enough to prevent or treat bleeds.

DDAVP is not a blood product. It is a chemical that releases factor VIII from storage sites in the body temporarily (approximately 24 hours). It can be given intravenously, subcutaneously or intranasally.

Complications of Hemophilia:

Inhibitors:

- An inhibitor is an antibody.
- In people with hemophilia, the immune system reacts to the clotting factor concentrate that is infused into their bloodstream. The immune system recognizes the factor concentrate as foreign, so in some people antibodies against factor concentrate develop. These antibodies then eliminate the infused factor and therefore prevent it from working to stop/prevent a bleed.
- The development of an antibody (inhibitor) is more common in severe hemophilia A patients, and can more commonly occur within the first 50 factor infusions.
- If you develop an inhibitor, you will notice that you will not be responding as well to clotting factor therapy when you have a bleed; you may also be bleeding and bruising more frequently than usual.

Treatment for Inhibitors:

- The treatment that we will start is called immune tolerization.
- This treatment involves giving regularly scheduled high doses of factor VIII or IX (depending on whether you have hemophilia A or B with an inhibitor).
- High doses and regular exposure of the factor in the body wears out the antibody production and the immune system becomes used to the presence of the factor, and therefore no longer rejects it.
- It can take 1 to 2 years (or more) to achieve successful tolerance, such that the person with hemophilia and an inhibitor can be successfully treated with FVIII or FIX.

Target Joints:

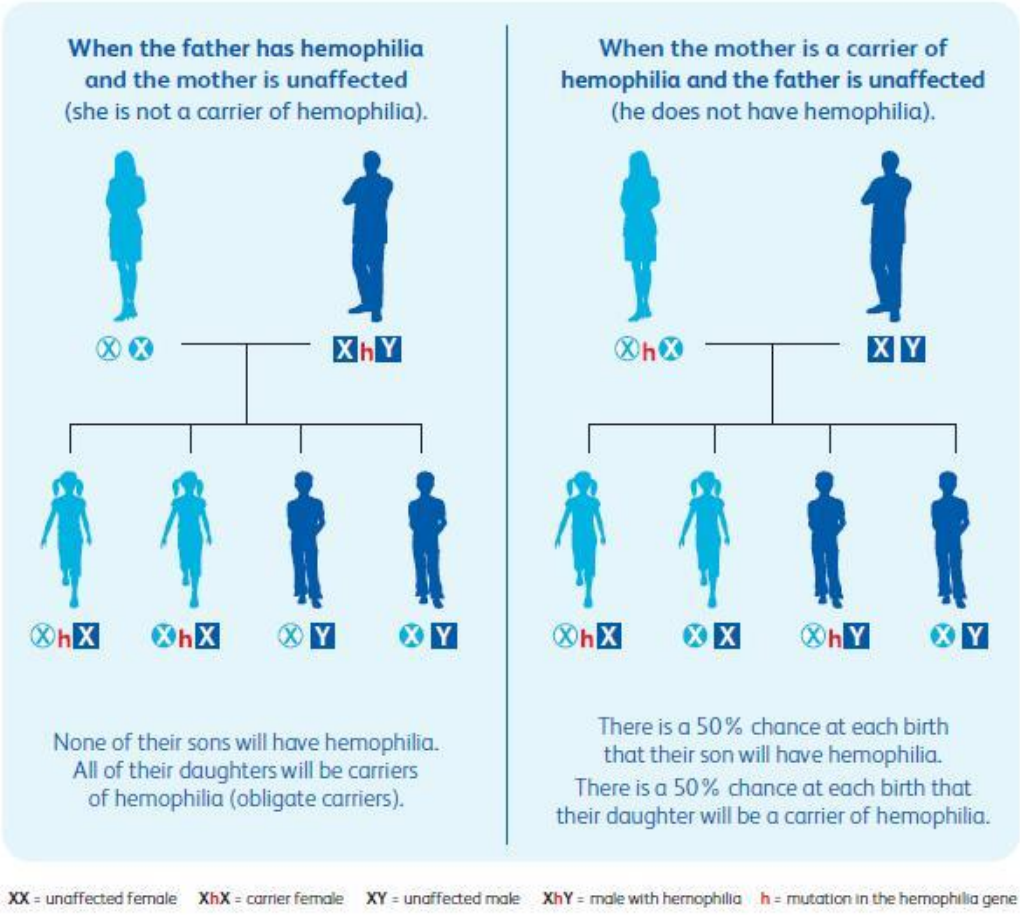
- Joints that are most often affected are the knees, ankles and elbows. If you typically experience bleeds in one joint (it is more prone to subsequent bleeds), this is called a target joint.

Genetics of Hemophilia:

Hemophilia is an inherited bleeding condition that is passed down to sons by the mother. This is known as an x-linked condition [the mother passes her affected X chromosome to the son (you)]. The X chromosome passed to the son (you) from the mother has DNA which contains multiple genes.

Hemophilia is caused by a "mutant gene" – this is called the mutation. A mutation is simply a change in the gene sequence. If the mutation occurs on the factor VIII gene, the person will have hemophilia A; if the mutation occurs on the factor IX gene, the person will have hemophilia B. There are multiple types of mutations that can occur. The type of mutation will result in the level of severity of hemophilia.

As the diagram below demonstrates, hemophilia can be passed down by a mother who is a carrier via an affected X chromosome. A mother who is a carrier has a 50% chance of having a boy with hemophilia and a 50% chance of having a girl who is a carrier of hemophilia. If the father has hemophilia, there is 0% chance he will have a son with hemophilia. This is because he passes on his Y chromosome, which is not affected by hemophilia, to the son. However, if he has a daughter, she will certainly be a carrier of hemophilia (this is known as an obligate carrier), since she receives his affected X chromosome.



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Physical Activity and Sports:

Risks and benefits must be weighed before making a decision as to what options are best for you. It is best to discuss with your hemophilia team when thinking about enrolling into a sport or activity – questions to consider are:

- What is your physical fitness level? Will this activity or sport be at a level that is too intense at this time? Would you benefit from strengthening or stretching exercises in preparation for the activity?
- What is your worst joint(s)? Will this sport/activity be overexerting that joint(s)?
- Will this sport/activity assist in the stability and strength of that joint(s)?
- Is this sport/activity one that will be difficult or increase in risk as you get older?
- Does it become more competitive and will it likely increase the level of physical contact over time (for example, ice hockey)?

It is important to note that regardless of your physical fitness level some sports are not recommended for someone with hemophilia. These are a few sports that are not recommended: boxing, American football, contact ice hockey, lacrosse, rugby and wrestling. Downhill skiing/snowboarding, waterskiing and skateboarding are higher risk activities, however proper protective equipment can assist in minimizing some of the risk.

Prophylaxis will assist with decreasing the frequency and the severity of a joint and muscle bleed, but it will not prevent all bleeds. Even though prophylaxis will increase factor levels (VIII or IX), this does not change the fact that you will have increased risk of joint and muscle bleeds in comparison to the person who does not have hemophilia. Remember, joint damage is cumulative and most likely irreversible.

This is a summary of information to review and reflect upon as you transition to being more independent with your hemophilia care. Remember family, peers and your hemophilia team are available for any questions or support you may require.

More extensive reference material can be found at:
Canadian Hemophilia Society www.hemophilia.ca

World Federation of Hemophilia www.wfh.org

Transition is a journey, not a destination. Enjoy the ride!

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