



PASSPORT to well-being

empowering people with bleeding disorders
to maximize their quality of life

HOME CARE THE ROAD TO INDEPENDENCE





Canadian Hemophilia Society
Help Stop the Bleeding

The Canadian Hemophilia Society (CHS) exists to improve the quality of life of persons with hemophilia and other inherited bleeding disorders and to find a cure.

The CHS consults qualified medical professionals before distributing any medical information. However, the CHS does not practice medicine and under no circumstances recommends particular treatments for specific individuals. In all cases, it is recommended that individuals consult a physician before pursuing any course of treatment.

The CHS would like to acknowledge those people who contributed to the development of *Home Care – The Road to Independence*.

Claudine Amesse, R.N.,
Hôpital Ste-Justine, Montreal, QC

Dorine Belliveau, R.N.,
South-East Health Care Corporation, Moncton, NB

Heather Carlson
Member, CHS Board of Directors, Toronto, ON

Clare Cecchini,
Program Coordinator, Canadian Hemophilia Society

Christine Keilbeck
Member, CHS Manitoba Chapter Board of Directors, Winnipeg, MB

David Page,
Blood Safety Coordinator, Canadian Hemophilia Society

Nora Schwetz, R.N.,
Health Sciences Centre, Winnipeg, MB

Peter Wilson
Member, CHS National Programme Committee, Halifax, NS

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For further information please contact:

Canadian Hemophilia Society
625 President Kennedy Avenue, Suite 505
Montreal, Quebec, H3A 1K2
Tel: (514) 848-0503
Toll Free: 1-800-668-2686
E-mail: chs@hemophilia.ca
Website: www.hemophilia.ca

*Note: Bleeding disorders affect both men and women.
The use of the masculine in this text refers to both.*

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Introduction

Watching a person with a bleeding disorder or his caregiver do a home treatment for the first time always brings the same reactions. **“Amazing!” “I could never do that.” “It must hurt!” “How long it must take to learn!”**

It is normal to feel stress at the idea of home care. After all, not many people take responsibility for their own medical care, nor that of their children. Few do injections at home. Even fewer do IV infusions!

This booklet, *Home Care – The Road to Independence*, describes how home care can empower you to take control over a bleeding disorder. Taking such a road requires clear directions and signposts along the way. It is a road that can be safely navigated... with the help of guides on your journey—the members of the comprehensive care team at the Hemophilia Treatment Centre (HTC). They are experienced in caring for people with bleeding disorders and are never more than a telephone call away.

The benefits and risks of home care

Benefits

Home care...

- allows the bleed to be treated at the earliest possible moment. This is the best way to stop bleeding, reduce damage to joints and speed recovery.
- avoids the delays in traveling to the hospital and waiting for treatment.
- makes it easier for the person to follow a prophylactic treatment program.
- reduces disruption to the family's daily activities.
- allows the person and his family to be more independent of the health care system.
- fosters a strong partnership between the family and the comprehensive care team.
- allows the person and his family to participate in normal social, school, and work activities.
- makes travel easier and safer.



- helps the family gain confidence in caring for a child with a bleeding disorder and a greater feeling of control over their lives.
- reduces the number of in-hospital stays and outpatient visits.



"Home infusion was our gift to everyone in the family. Good-bye clinic and good-bye emergency room! We gained management of our son's health and the ability to treat him right away. The clinic is still a central part of his team; we just don't miss the waiting, the frustration and the constant upheaval in all our lives."

Risks

Several problems may occur when a person is on a home care program.

- A bleed not being treated properly - This can be avoided through good communication with the HTC.
- Poor response to treatment - The patient may respond poorly to treatment and be slow to realize this. If a bleed has not shown improvement after one treatment, the comprehensive care team should be contacted.
- Mistaking another health problem for a bleed - This could lead to a delay in seeking treatment. When in doubt, consult the comprehensive care team.
- Improper storage of clotting factor concentrates and other medications.
- Infection or damage to veins or the venous access device.
- Injury to other family members - This can occur if needles are not properly disposed of in an approved "sharps" container.

Ways of reducing the risks

There are many ways to reduce the risks of a home care program.

- Communicate immediately with the comprehensive care team when...
 - the person has an injury to a critical part of the body (head, neck, throat, chest, stomach, eye)
 - bleeding is heavy
 - a bleed does not respond to standard treatment
 - one joint has several bleeds in a short time
 - the person has an allergic reaction to the clotting factor concentrate
 - you are not sure what to do.
- Communicate frequently with the comprehensive care team to discuss general concerns around managing bleeds and home care.
- Be well informed. Frequently review educational material provided by the HTC.
- Keep accurate home care records of bleeds and treatments.
- Regularly review home treatment procedures with the comprehensive care team.

Types of home care

Home infusion with clotting factor concentrates

Today, many people with bleeding disorders receive home infusions with clotting factor concentrates. These are infused intravenously (IV). They include...

- most people with severe hemophilia A and B (factor VIII and IX deficiencies)
- some people with mild or moderate hemophilia A and B
- some people with other factor deficiencies (factors VII, X, XIII)
- some people with severe von Willebrand Disease (VWD).

THERE ARE TWO APPROACHES TO HOME INFUSION:

On-demand therapy - The infusion is given as soon as the patient or his caregiver is aware of a bleed. The goal is to promptly stop the bleed.

Prophylaxis therapy - The patient receives regular infusions of clotting factor concentrates, usually 1, 2 or 3 times a week. This is done in order to prevent bleeding episodes from happening. Most children in Canada, especially the

younger ones, receive prophylaxis therapy. Many adults do, too. This is because prophylaxis has been shown to be the best way to prevent joint damage.

THERE ARE THREE TYPES OF PROPHYLAXIS:

Long-term prophylaxis is the most common type of prophylaxis. Infusions of factor concentrates are given over a period of many years. The goal of this therapy is to prevent bleeding and avoid joint damage.

Short-term prophylaxis is often used to halt the development of a "target joint". This is a joint that suffers frequent bleeds. Joint damage can result. Thus, infusions are given over a period of 3 to 6 months, or until bleeding in the target joint stops happening.

Occasional prophylaxis - A person with a bleeding disorder sometimes takes part in activities that can cause bleeding. An infusion prior to the risky activities can prevent bleeding.

Home treatments with desmopressin

Some people with bleeding disorders are able to treat their conditions with a drug called desmopressin. These include people with...

- some types of VWD
- some types of platelet function disorders
- mild or moderate factor VIII deficiency (hemophilia A).

Desmopressin can be infused intravenously (in a hospital only), injected subcutaneously or inhaled by nasal spray.

"I'm on the running team at school. I don't want to miss practice because of bleeding. Now that I carry my own treatment, I am in control."



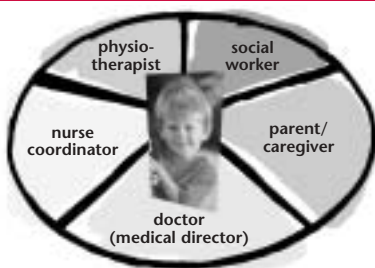
Other medications

Other medications to aid clotting, such as the antifibrinolytics Amicar® and Cyklokapron®, can also be administered at home. They are taken by mouth.



The comprehensive care team

Team members



In addition to the patient and/or caregiver, the comprehensive care team usually includes:

- a doctor, usually a hematologist, who recommends treatments and oversees the health of the patient.
- a nurse coordinator, who is the key contact in the comprehensive care team.
- a physiotherapist, who is trained to keep a person's muscles and joints healthy.
- a social worker and/or psychologist to help the patient and his family members deal with the impact of the bleeding disorder on their lives.

More and more HTC's are working closely with a gynecologist to treat women with bleeding disorders. The team also works closely with hospital labs, the blood bank and specialized personnel, as well as health care providers in rural areas.

"The people at the HTC don't only provide care and training. They also offer encouragement and support. Knowing they were there gave us the confidence we needed. That was very important to my husband and I."



Starting on a home care program

Age

There is no standard age for starting. The comprehensive care team and the family will make a joint decision about when it is best for the child to begin a home care program.

Both parents and child must be ready. Ideally, a home infusion program begins before the child starts school.

Normally, to be eligible, the person needing factor replacement must infuse an average of at least once per month. This is necessary to maintain skills.

Learning to do sub-cutaneous injections with desmopressin is easier. Using the nasal spray is easier still.

Parents are responsible for a child's home care therapy. However, even a very young child can become involved in his own treatment. Small tasks suitable for the child's age, such as taking the factor concentrate out of the refrigerator, disinfecting the skin with alcohol, and holding still, encourage the child to learn about his care. Participating gives a child a sense of control over a painful and stressful situation.

The child who has been involved with treatment at home learns the steps and becomes more responsible as he gets older, which, in turn, makes the management of the bleeding disorder easier for the family.

It is never too late to learn. A person who has chosen not to treat himself or his child may one day decide to do so.

"I learned to self-infuse as an adult. I had to have an operation and infusions every day after for a few weeks. It turned out to be easier than I thought it would be. Later, I just kept on. Now I travel everywhere with my product."



Skills the patient and/or caregiver need

Before starting a home care program, the patient or his caregiver must show that he can...

- recognize when a bleed is occurring and how serious it is
- use clean and safe techniques to prepare clotting factor concentrates or desmopressin
- safely infuse the clotting factor concentrates or inject the desmopressin subcutaneously
- seek assistance, when needed
- dispose of used equipment safely
- keep a diary of treatments
- store factor concentrates and desmopressin properly
- attend assessment clinics regularly.

Good communication with the comprehensive care team is essential.

"It's so important to always take your desmopressin with you. I went out of town to a conference and forgot my medication at home. I fell down stairs and had a bad bleed. You can't imagine the trouble I had getting desmopressin in a strange city on a weekend."



Assessing bleeds

When in doubt, treat. If for any reason you have doubts about how to assess a bleed, contact your HTC for advice or go to the nearest ER.

Be sure to work with the members of your comprehensive care team to learn how best to assess bleeds at home. You will need to learn...

- in which parts of the body bleeds are most serious
- what to look for and what to do
- when to go to the HTC or ER for help.

Locations that can be serious or life-threatening include the...

- head
- neck and throat
- chest
- abdomen
- eye.

Any bleed associated with a large blood loss is considered serious.

Any bleed that can result in permanent loss of movement such as in the forearm, the calf muscle, and iliopsoas muscle, is considered serious or limb-threatening.

Excellent resources to help assess bleeds are:

- *All About Hemophilia: A Guide for Families*, Chapter 5, Management of Bleeds. © Canadian Hemophilia Society, 2001.
- *All About von Willebrand Disease*. © Canadian Hemophilia Society, 2000.
- *A Home Treatment Guide and Notebook*. © Western Canadian Hemophilia Nurses Group, 2000.
- *Assessment and Treatment of Joint and Muscle Bleeds*. © Kathy Mulder, Children's Hospital, Winnipeg, 2000.

"We check our son over carefully, from head to toe, every evening at bath time. He doesn't even know we are checking him. But we look for bruises and swelling, and make sure he can move his joints all the way. It's a lot easier if we find something and treat it right then than to have to deal with it in the middle of the night."

"Every night when I get ready for bed I put on my pyjamas, brush my teeth and check my joints. If I need treatment, I tell my mom."

Treating bleeds

Ten points to remember

1. Treat at the first sign of bleeding, even if the person is on prophylaxis! The earlier the treatment starts, the faster the bleed will get better.
2. If for any reason you feel unsure about how to proceed, contact your HTC for advice or go to the nearest ER.
3. Use rest, compression and elevation along with factor concentrate or desmopressin.
4. Use the right amount of clotting factor concentrate for the type of bleed. Giving too little factor may mean a second dose is needed.
5. Always use the complete vial of factor concentrate unless the HTC advises otherwise.
6. Never infuse/inject alone. Make sure someone else is there.
7. If the factor concentrate will not dissolve, if it is not clear, or you have doubts about it for any reason, do not use it. Call the HTC for advice. Do not throw it away!
8. Keep accurate home treatment records.
9. Keep your skills up to date by reviews at the HTC.
10. Remember!

There's much to gain and little to lose.

When in doubt, it's wise to infuse.

Calculating doses

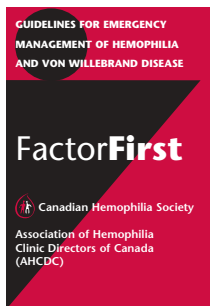
Deciding how much clotting factor to give is based on:

- body weight
- type of bleed
- how quickly the bleed is treated
- type of bleeding disorder



- the level of clotting factor recovery in a person's bloodstream after infusion. (Recovery is the amount of factor concentrate a person's body can actually use to stop bleeding compared to the amount infused.)

* To know the correct dose, fill out your FactorFirst card with the HTC personnel and keep it handy at all times.



For more information about calculating doses of FVIII and FIX, see:

- *A Home Treatment Guide and Notebook*. © Western Canadian Hemophilia Nurses Group, 2000.
- *All About Hemophilia: A Guide for Families*, Chapter 4. © Canadian Hemophilia Society, 2001.

Preparing factor concentrate and desmopressin

The staff at the HTC will show the patient and/or his caregiver how to prepare the particular treatment. Here are some general tips.

- Wash hands thoroughly with soap and dry them with a clean towel before beginning.
- Choose a working area which is quiet, flat, clean, comfortable, and well-lit.
- Follow the instructions provided by the staff at your HTC and included in the package insert.



"I learned to mix product at camp when I was seven. I started helping Mom and Dad with the treatment after that. I don't want to try to get the needle in the vein yet. Maybe when I am ten or older, I will be ready to do it myself."

Accessing veins

Venipuncture

The staff at the HTC will provide training to the patient and/or his caregiver in venipuncture (the placement of a butterfly needle into a vein to infuse clotting factor concentrate). What's more, from time to time, they will check a person's technique to be sure it is safe.

Items needed for venipuncture

- A tourniquet
- Alcohol wipes
- Butterfly needles
- Tape
- A syringe
- Cotton balls or gauze
- Clotting factor concentrate
- A "sharps" container



Tips for successful venipuncture

- Find three or four veins that are easy to access. Use these in rotation to keep veins healthy.
- Veins are larger and easier to see when a person is warm. Before infusing...
 - Do some exercise.
 - Take a warm shower or bathe the arm in warm water or a warm compress for 5 to 10 minutes.
 - Dress warmly.
 - Use a warm room in the house.
- Increase blood flow into the arm before applying the tourniquet.
- Hold the arm below the heart to fill the veins with blood.
- Squeeze a rubber ball.
- Apply the tourniquet for 2 or 3 minutes. After this, the vein will shrink. If this happens, remove the tourniquet and start again.
- Ask the staff at the HTC to suggest arm exercises which develop good veins.
- Remember. Practice makes perfect... or almost perfect. Everyone misses now and again.

"I was scared when I started treating my son at home. He was five and ready to let me try. I was surprised I would be allowed to do it. It was a bit nerve racking at first. In time, I gained confidence and feel good about it now. One time I even infused him on a hiking trip. →"

Even now, after five years of treating my son, I miss the vein the odd time. But he is used to getting needles now and knows that it isn't too painful."

Tips for keeping veins healthy

- Stop infusing if swelling appears near the site of the infusion. The factor is going into the tissues, not the vein.
- If a vein has not healed from a previous infusion, and there is a bruise, use a different site.
- Do not use veins which are inflamed, surrounded by red skin or are sore to the touch.
- Never put pressure over the puncture site before or during withdrawal of the needle. Wait until after the needle has been removed.
- Apply pressure with a cotton swab for at least 5 minutes after withdrawal to prevent bleeding and bruising. Check after 5 minutes and, if there is any oozing, continue with pressure. If the puncture site was in the elbow, keep the arm straight during this time. Then apply a clean dressing to the site and keep it for one hour.
- Apply a lanoline cream daily over infusion sites to keep the skin smooth and soft.



Implanted devices

If access to the veins is difficult, as it sometimes is with small children and even some adults, a *central venous access device*, for example, a *Port-a-Cath*, can be surgically implanted. Parents are taught how to care for and use this device to infuse the factor concentrate. The use of such a device means that the parent does not have to insert a needle into a vein for each treatment. This can make it easier for the family to move from the hospital to the home setting.

For more information on healthy veins, see...

- *Looking after the Veins*. Peter Jones, MD. © World Federation of Hemophilia, Treatment of Hemophilia #32, December 2003. (www.wfh.org)

"I have Type 3 von Willebrand Disease and my veins are terrible. I couldn't self-infuse so I got a Port-a-Cath. Now I'm free from all those trips to the ER."

Record-keeping

All HTC's have a form for recording home treatment records. This form may be paper or electronic, and is sent to the HTC regularly. Information reported includes:

- patient's name
- date of the treatment
- site of the bleed /or prophylaxis
- name of the particular product infused / injected / inhaled
- dose
- lot number (for clotting factor concentrates)
- adverse reactions, if any.



Keeping accurate records of each home treatment has many benefits for the patient. (See *Charting the Course*, Canadian Hemophilia Society, 2004.)

Allergic reactions

Clotting factor concentrates may cause *allergic reactions*. These are very rare. Desmopressin may also cause adverse reactions such as facial flushing, headache, nausea and light-headedness.

It is very important to recognize allergic reactions, and to notify your HTC and seek medical attention when they occur.

Review with your HTC the guidelines for managing allergic reactions including...

- recognizing signs of an allergic reaction
- stopping infusion at the first sign
- managing allergic reactions at home
- recognizing signs of a serious allergic reaction requiring immediate action (medications and/or hospitalization).

Storage

Medications to treat bleeding disorders, especially clotting factor concentrates, are very valuable. Handle them with care by following these instructions.

- Read the product insert. Every medication has an insert which explains how your specific product should be stored and used.

- Store factor concentrates at the proper temperature. (If frozen, bottles could be damaged, compromising sterility. If exposed to heat, the clotting activity could be lost.)
- Store supplies needed for treatments (syringes, needles, etc.) in a clean, safe place where children will not be able to get into them.
- Pay attention to expiry dates. It's the same principle as milk: use the product which will expire first before using the others.
- Discard used needles and supplies that have blood on them into an approved "sharps" container.
- Talk to your HTC staff if you have any questions. If you think the product has not been stored properly, get advice. Do not discard any products on your own.



"It is good practice to always keep your clotting factor concentrates in the fridge."

Transport

- When traveling with desmopressin or clotting factor concentrates which require refrigeration, keep them in a cooler with ice.
- Ask your doctor for a letter which authorizes the patient to carry these medications. The letter should specify the name of the product. When crossing a border or taking an airplane, the letter will answer many questions.

"I traveled around Europe for a month last summer. Airport security examined my bag containing my product. But I had a letter from my doctor and there was no problem."



Conclusion

This booklet describes how home care can empower a person with a bleeding disorder and help him along the road to independence. But it is important to remember you are not alone! The members of the comprehensive care team at the Hemophilia Treatment Centre will work with you to ensure that the road is smooth, and that home care is a success.