This chapter provides answers to these questions:

- What is comprehensive care?
- Why is comprehensive care important?
- Why do you need to register your child at a hemophilia treatment centre?
- Who are the members of the comprehensive care team?
- What are the roles of the members of the core team?
- How do the extended members support the core team?
- How do specialized hospital services support the core team?
- Where are hemophilia treatment centres located?
- How can you get comprehensive care if you live outside a big city?
- What makes comprehensive care a success?

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Comprehensive Care for Hemophilia

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What is comprehensive care?

Hemophilia affects the body in many different ways. Beyond physical effects such as pain and joint damage, it can also have emotional and social impacts on the person with hemophilia and his family. Because of this, medical professionals who treat people with hemophilia take a comprehensive approach to care. The word “comprehensive” means “complete.” Comprehensive care is about providing a complete range of healthcare services to children and adults with hemophilia. In Canada, comprehensive care for people with hemophilia began in the 1970s.

The comprehensive care team is a group of health professionals that works together at a hemophilia treatment centre (HTC). It offers a continuum of healthcare services to people with hemophilia throughout their lives. It is also referred to as the care team or the HTC team.

Each member of the comprehensive care team is an expert in a different area. The main members of the team are the hematologist, hemophilia nurse coordinator, physiotherapist and social worker — together, they make up the core team. This team will work with your family to help you understand all the various aspects of hemophilia and how to prevent and treat bleeds. Together you will develop a plan of care for your child. Other health professionals are available to provide additional support to the core team if needed.

See “Who are the members of the comprehensive care team?” later in this chapter.

We would like to express our appreciation to the comprehensive care team at Ste-Justine Hospital, Montreal: Dr. Georges Rivard, Claudine Amesse, Nick Zourikian, Sylvie Lacroix, and to Seng-Kang Lin and his mother who allowed us to document his visit to the hemophilia treatment centre. Photos of his visit are used to illustrate this chapter and others throughout the Guide.
Why is comprehensive care important?

Comprehensive care means treating the whole person. It embraces all aspects of a person’s health — his body, his feelings and his psychosocial well-being (how well he fits in at school, work, home and with friends, etc.). The goal of comprehensive care is to help people with hemophilia lead healthy and active lives. The comprehensive care team will support you and your family step by step as your child grows. Everyone can feel secure knowing there is a care team just for him!

The HTC team will teach you, your child and other members of the family how to recognize and treat bleeds. If your child has severe hemophilia, you may need to learn how to give him clotting factor concentrate at home. If your child has moderate or mild hemophilia, you might not need to give him treatments at home. However, if a bleed occurs, you will need to take him to the hospital for treatment with factor concentrates.

See Chapter 7, Home Infusion, and Chapter 9, Mild and Moderate Hemophilia.

The care team will work with your family and your child’s pediatrician, dentist, daycare staff and schoolteachers to make sure he gets the best care possible. You should get to know the key members of your child’s care team. Do not be afraid to ask for their advice if you have a question. If you are not sure who to talk to about a problem, speak to the HTC nurse coordinator. She/he will tell you what you need to know, or arrange for you to speak to someone who can help you.
Why do you need to register your child at a hemophilia treatment centre?

It is very important that you register your child at a hemophilia treatment centre, even if he gets most of his routine health care close to where you live. Going to an HTC at least once a year will allow your family to:

- Get correct information about hemophilia from the comprehensive care team.
- Get blood tests that can only be done by a specialized coagulation lab.
- Have access to the newest treatments for hemophilia.
- Find out about the latest research on hemophilia.
- Make sure your child gets a comprehensive “hemophilia check-up” regularly.

Most children with hemophilia have a check-up at the HTC every 6 to 12 months. How often your child goes will depend on how severe his hemophilia is, and how often the care team wants to see him for an assessment. Having check-ups at the HTC is very important to your child’s care. As your child grows, his plan of care will change to meet his evolving needs.

When he visits the HTC, the care team will review his history of bleeds and treatment plan. The team will want to know how many bleeds he has had and how much clotting factor concentrate he has infused since his last visit. You will need to keep record in a home infusion diary — this is very important because having accurate records of his bleeds and treatments will help the team decide if he is getting the right amount and frequency of clotting factor. The check-ups are also a good time to
let the care team know if your child is having frequent bleeds in one joint. You can then decide together whether to adjust his treatment plan to prevent the development of a target joint.

As your child grows and develops, his range of physical activities will expand and with this the frequency and dose of factor concentrate that he needs to treat or prevent bleeds could change. Regular visits to the HTC will allow you and the comprehensive care team to respond to his changing needs.
Who are the members of the comprehensive care team?

Many different health professionals and services are involved in providing comprehensive care for people with hemophilia. They fall into three general categories: the core team, extended members and specialized hospital services.

Core Team

The core team is made up of health professionals who have specific expertise in treating people with hemophilia. These members are based at an HTC and work together to provide comprehensive care — they are essential to any hemophilia program. Parents of children with hemophilia are also part of the team.

The members of the core team are:

- medical doctor (usually a hematologist)
- nurse coordinator
- physiotherapist
- social worker
- parent/caregiver

Your child is at the very centre of this core team. The core team will also work closely with your child’s pediatrician, dentist and teachers to help ensure that he gets the best possible care away from the HTC.

“It is essential to stay connected to your comprehensive care team, your family who will support and advise you, and to other parents of children with hemophilia.”
Extended Members

Other health professionals are also involved in comprehensive care but they are not permanent members of the care team. They are referred to as extended members of the team. All HTC programs must have these health professionals available to support the work of the core team. Extended members are usually based at the same hospital as the HTC — otherwise, the HTC will have agreements with other healthcare institutions on the referral of patients.

Extended members of the comprehensive care team include:

- dentist
- ear/nose/throat specialist
- experts in pain management
- child life specialist
- occupational therapist
- psychologist
- rheumatologist
- orthopedic surgeon
- geneticist/genetic counsellor
- gynecologist/obstetrician

The specific roles of these health professionals and specialized hospital services are described in the next section.

“Always have one really good question for your doctor. That’s how you learn.”
What are the roles of the members of the core team?

Medical Director

The medical director is the doctor who oversees patient health and the comprehensive care team. In many centres, the medical director of the comprehensive care team is a hematologist. A hematologist is a doctor who understands and treats blood problems. If the medical director is not a hematologist, she/he will call on a hematologist for advice.

The medical director’s job is to:

• Oversee the care team and the treatment of people with hemophilia.
• Help the family deal with any health or medical concerns and answer questions about hemophilia.
• Order blood tests and X-rays and review results and the treatment plan with the family.
• Keep up to date on the latest research and tell families about new developments in the management of hemophilia.

You should be aware that the medical director does not take the place of your family doctor or your child’s pediatrician. The family doctor or pediatrician can give you advice about general child health issues, such as the flu, chicken pox, ear infections and vaccinations. The medical director will communicate information and updates about your child’s health with his pediatrician and dentist.
Nurse Coordinator

The nurse coordinator is your key contact in the comprehensive care team. She/he coordinates when you and your child see other members of the team. In most HTCs, the nurse coordinator is the person who answers your telephone calls. If your child needs to go to the HTC to treat a bleed, the nurse coordinator is often the person who makes sure the infusion goes smoothly.

An important part of the nurse coordinator’s job is teaching people about hemophilia. She/he can go to your child’s daycare centre or school to educate teachers and staff. The nurse coordinator can also give you useful booklets and videos on hemophilia produced by the Canadian Hemophilia Society (CHS). There is a wide range of resources for different people involved in providing care for children with hemophilia such as family members, teachers, babysitters, and local nurses. The CHS also has colourful activity books and guides to help children understand hemophilia, how it is treated and what to do to stay healthy. Ask the HTC for copies of these educational materials, or contact the CHS.

When your child is old enough, the nurse coordinator will train you and your child to do home infusion, if this is needed. At a later date, when your child is older, the nurse coordinator will teach him how to self-infuse. See Chapter 7, Home Infusion.

Get to know your nurse coordinator. She/he is there to help you. Do not be afraid to ask questions, even if they seem simple or silly. You can be sure that parents of other children with hemophilia ask similar questions.

If you have a problem of any kind, it is important to solve it right away. Phoning your nurse coordinator does not always mean that you will have to take your child to the hospital. Often you will be able to solve the problem over the phone.

The nurse coordinator treats many children with hemophilia. This means she/he knows a lot about the bleeding disorder and is experienced in treating it.

“She always made me feel that my questions mattered. Our nurse has become our friend. She has guided us through some tough times. My son trusts her and has his own relationship with her.”
Training and support from the physiotherapist has helped us understand the importance of rest and ice when a joint bleed occurs. Physiotherapists know their stuff and are good at explaining the types of bleeds that my child gets, and what is needed to heal his bleeds.”

**Physiotherapist**

The physiotherapist is trained to keep muscles and joints healthy. The physiotherapist will give you advice on how to prevent or limit bleeding. She/he will also guide your child’s recovery from a joint or muscle bleed. The physiotherapist will help you:

- understand what a bleed is
- know whether a bleed is serious or not
- learn what to do to help your child get better after a muscle or joint bleed

The physiotherapist’s important role is to help your child recover quickly and safely from a bleed. Physiotherapists are skilled at determining what patients should do, in addition to factor replacement, to get better from a bleed. This might include:

- specific exercises that she/he will teach you and your child how to do
- possible use of assistive devices (braces, crutches, cane, wheelchair, etc.)
- the gradual return to activities (walking, running, sports, etc.)

The physiotherapist will also give you advice on ways for your child to stay active and healthy. Physical activity and exercise are important in a child’s development. Exercise keeps muscles strong. Strong muscles protect the joints and may help your child have fewer bleeds.
Social Worker

Most families of children with hemophilia meet the social worker when they first learn of the diagnosis. The social worker’s job is to help parents, siblings and the child himself deal with the psychological, social and potentially even financial impacts that hemophilia can have on their lives.

One of the early issues that you will face is deciding how much to protect your child. As your child gets older, other issues may come up such as how to train a babysitter and whether or not both parents should work outside the home. Discuss your concerns with the HTC social worker, who can help you find solutions.

Pain Management in Hemophilia

In various ways, all members of the core team are involved in the management of different aspects of pain. The person with hemophilia, and his parents in the case of a young child, are at the centre of the process.

- The nurse coordinator ensures that pain is assessed and treated by the appropriate team member.
- The hematologist develops a management plan for both acute and chronic pain, which may include medications.
- The physiotherapist makes recommendations for treating acute or chronic pain, with the overall goal being to prevent secondary complications such as tight muscles or reduced joint mobility.
- The social worker helps individuals and families living with hemophilia manage the complications that occur due to pain, and deal with related physical or psychosocial challenges to improve overall functioning and social well-being.
How do the extended members support the core team?

Dentist

It is very important to prevent dental problems in children with hemophilia. If your child does not take good care of his teeth by brushing/flossing regularly and seeing a dentist for check-ups, he could lose a tooth to decay or cavities.

He could also need dental surgery to fix a problem that has been allowed to worsen. For dental surgery, treatment will almost certainly be needed to prevent bleeding and can include the use of factor concentrates. Although most HTCs offer a dental service for children with hemophilia, many families see a local dentist. This is common when a family lives far away from the hemophilia centre.

It is also a good idea to discuss your child’s dental care with the HTC nurse coordinator or medical director. No matter where you live, you and your family dentist can get in touch with the dental service at the HTC with your questions or problems.

Ear/Nose/Throat Specialist (also called ENT)

The ear/nose/throat doctor treats all problems involving the ears, nose and throat. Should your child need to have tubes inserted in his ears because of ear infection, or have to have his tonsils or adenoids removed, the ENT doctor will perform the operation.
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Comprehensive Care for Hemophilia

Child Life Specialist
A child life specialist is often available to children and families living with hemophilia. The child life specialist usually works in a “play room” environment. She/he can help children who have a fear of needles and anxiety with factor infusions. Using play therapy, imagery and distraction techniques, the child life specialist helps children learn how to relax and learn coping skills for treatment and procedures.

Occupational Therapist
An occupational therapist develops individual and group programs to help people of all ages be as independent and functional as possible in different environments (home, daycare, school, workplace, etc.). She/he works closely with people affected by a medical condition or injury. She helps them maintain, restore or increase their ability to care for themselves and engage in work, school or leisure. This is done by assessing the individual’s environment, functional limitations, and specific needs and goals.

Psychologist
A psychologist in an expert in the human mind and how it functions. A psychologist can help someone who is having trouble dealing with his hemophilia or having difficulties at school, work or with peers.

Rheumatologist
A rheumatologist treats health problems and diseases that cause pain and swelling in and around joints and muscles.

“My son feels angry about receiving medical care; he doesn’t want to deal with hemophilia, doesn’t want to see hemophilia doctors; he’s in denial.”

“He got counseling with the family and by talking about the disease, he was reassured that he is ‘normal’.”
Orthopedic Surgeon

An orthopedic surgeon is a doctor who treats diseases that affect the bones and joints, including fractures. If your child needs to have an operation on a joint, it is the orthopedic surgeon who will perform it. For more information on joints, see Chapter 8, Complications of Hemophilia.

Geneticist/Genetic Counsellor

A geneticist studies genes and how people inherit diseases. A geneticist or genetic counsellor has specialized training in providing genetic counselling for families of children with hemophilia. For more information on inheritance, see Chapter 2, How a Child Gets Hemophilia.

Gynecologist/Obstetrician

A gynecologist is a doctor who understands and treats women’s gynecological issues. Some carriers can have heavy menstrual bleeding and a gynecologist can find ways to reduce the bleeding. An obstetrician is a doctor who specializes in conception, pregnancy and childbirth. Gynaecologists and/or obstetricians with expertise in bleeding disorders can work with women who are carriers of hemophilia. For more information on hemophilia carriers, see Chapter 2, How a Child Gets Hemophilia, and Chapter 14, Symptomatic Carriers of Hemophilia.
How do specialized hospital services support the core team?

The hospital where your HTC is based has several laboratories that perform many specialized laboratory tests and functions. Many of the blood tests that your child needs will be done by a specialized coagulation laboratory. This kind of lab also does specialized blood tests for inhibitors (a possible complication that can develop in some patients), factor VIII and factor IX levels (also called factor assays), and any other blood tests that have to do with blood clotting. The Hemophilia Mutation Laboratory in Kingston, Ontario, provides a national service for the genetic analysis of inherited bleeding disorders.

Transfusion Medicine Department

Every large hospital has a transfusion medicine department, also referred to as the blood bank. The blood bank is where blood products are stored in a hospital. When the HTC doctor or nurse coordinator “orders” factor concentrates, they usually come from the blood bank. At some centres, the nurse coordinator orders factor concentrates directly from the local branch of Canadian Blood Services or Héma-Québec.

In some parts of Canada, parents pick up the clotting factor concentrates from the blood bank in the hospital where their HTC is located. In other regions, clotting factor concentrates are sent to the local hospital nearest to the family’s home. No matter who orders your child’s clotting factor concentrates or where you pick them up, you need to keep up-to-date records of his use of factor concentrates in an electronic or paper home infusion diary. These home infusion records must be returned to your nurse coordinator as requested.
Diagnostic Imaging Department

This department takes pictures (x-rays, ultrasound, MRIs, CT scans) of your child’s joints and bones when needed. These tests and images help the members of the care team decide what kind of treatment is best for your child. They also show whether the factor replacement therapy is working well.

Emergency Care Department (24-hour ER)

The emergency department of a hospital is also called the emergency room (ER). This department is responsible for providing medical care to people arriving at the hospital in need of immediate care, such as in the case of a bleed that requires treatment with factor concentrate and/or other medical interventions.

Additional Healthcare Services

The comprehensive care team will also coordinate access to additional healthcare services as needed. These include:

- ongoing education and training in the management of hemophilia
- home infusion program with education and training
- other medical and allied health expertise

The care team can also provide educational services to other healthcare services and outside agencies, and outreach and education to daycare staff and teachers.
Where are hemophilia treatment centres located?

Most hemophilia treatment centres are based in large hospitals in big cities. Some hospitals have separate clinics for children and adults, while others treat both adults and children at the same HTC.

If hemophilia is diagnosed or suspected, your family doctor or a specialist will refer your family to an HTC close to where you live. If you happen to live in or near a city that has a HTC, then this is where your child will receive his ongoing care and assessments.

To find a hemophilia treatment centre close to where you live, see Chapter 17, For More Information.

How can you get comprehensive care if you live outside a big city?

If your family lives in a small town, a rural area or remote part of Canada, there are three ways to get comprehensive care:

- Regular check-ups – It is very important for you and your child to visit a HTC once or twice a year for a full check-up. During these visits, your child will see the doctor, nurse coordinator and physiotherapist, and have blood tests done. In some cases, the social worker may meet with you. The care team will work with you to develop a plan for your child’s day-to-day care.
• **Day-to-day care** – Your local health professionals can help you with day-to-day concerns related to hemophilia. The comprehensive care team will gladly work with your family doctor and local hospital staff to help them understand hemophilia and the correct treatment for your child. You may want to leave a copy of your child’s care plan at the hospital in your area. In many cases, treatments such as the infusion of clotting factor concentrates or another treatment product called *desmopressin* will be given in the ER of your local hospital. The HTC doctor or nurse coordinator can contact the ER staff to make sure they know the correct treatment and dose for your child’s bleeding episodes.

Your care team may be able to set up physiotherapy treatment close to your home so that you won’t have to make as many long trips to the city.

• **Special travelling clinics** – Your child may also be able to attend a travelling hemophilia clinic. These clinics are usually staffed by a doctor, nurse coordinator and physiotherapist. Travelling clinics are often set up to go to more remote places that do not have an HTC but where there are several people with hemophilia. The purpose is to reduce the travel time for people with hemophilia and their family members. Sometimes the comprehensive care team holds these travelling clinics at local hospitals, where they can also teach the ER staff and doctors how to treat hemophilia.
What makes comprehensive care a success?

Success depends on many things and many people. It is very important that you, your child and the comprehensive care team have strong, open lines of communication. Parents should feel comfortable enough with the comprehensive care team to be able to pick up the phone and call any time with a question or concern. As your child gets older, he should feel comfortable calling the nurse coordinator or physiotherapist when he has a question. Growing together as a team will increase everyone’s confidence and comfort level. Knowing that when you call the HTC team, you won’t have to start at “the very beginning” makes it easier to solve problems as they come up.

There may be times when waiting a long time at the ER will make you upset with the system. Call your comprehensive care team to talk about what you went through and how you feel. By working together, you may be able to set up new ways to get your child the treatment that is needed.

As you and your child work with the members of the comprehensive care team, you will gain a lot from their knowledge and experience. If you wish, your care team can put you in touch with other families who also have a child with hemophilia. Families can be a great help to each other by sharing what they have learned and experienced.

“We need CHS to be a strong advocate, working with centres regarding standards of care, that is, with medical staffing and programs. We need to work together to continue to advocate for the safety of the blood system.”
Success also depends on respect and being responsible. To get the most from comprehensive care, a family must try to follow the care team’s recommendations. This means following your child’s treatment plan, and possibly making changes in lifestyle or the sports he plays. It’s very important to keep good records of bleeds and clotting factor infusions. These records show whether or not your child’s treatment plan is working. Adjustments can be made if needed.

The comprehensive care team is large but works closely together. The same goal is shared by everyone — a child that despite hemophilia is healthy and active. This is the best way to measure success!

“As you learn more you feel more empowered. We joined the Society and met other families at social events. We needed them so much, to learn how to manage different bleeds. Our nurse gave us so much perspective. She explained what muscle or joint was having the problem, how common it was, how long it would take to resolve and how we could help our son at home.”