Section A: Executive Summary

In 2013-14, the Canadian Hemophilia Society (CHS) conducted an assessment of the human and physical resources in the network of 25 Inherited Bleeding Disorder Comprehensive Care Centres (IBDCCCs) across Canada. The staff in these Centres care for the vast majority of Canadians with inherited bleeding disorders. Our goal was to evaluate their capacity to respect current national Standards of Care. Individual reports were provided to each Program's Centre Director. This national report summarizes the findings and presents key recommendations.

The CHS discovered that, despite dedicated and well-trained staff, the Programs are experiencing serious resource deficiencies. Twenty-three of the 25 Programs lack human resources in one or more of the following disciplines: hematology, physiotherapy, social work, clerical/data entry and nursing. In nine of the 25 Programs, no resources are allocated to certain core disciplines, notably physiotherapy and social work. The level of resources varies widely and inexplicably from Program to Program: from 0.2 to 1.5 Full Time Equivalents (FTEs) per 100 hemophilia A and B patients in nursing; from 0 to 0.4 FTEs per 100 patients in physiotherapy and social work, and 0 to 1.8 FTEs in clerical/data entry. Because of lack of clinic time or physical space, nine of the 25 Programs are unable to respect the schedule for regular health assessments as prescribed in the Standards of Care.

The cost to deliver care (including the cost of staffing, diagnostic and clinical support services, non-labour costs and office space, but excluding the cost of coagulation therapies) to the approximately 10,000 patients registered in IBDCCCs in Canada is estimated at approximately $20,000,000 (1). Many Programs are subject to increasing hospital, regional health authority or Ministry of Health pressures to limit or even reduce existing resources, despite a 75% increase in the number of patients registered in the 25 Programs in the period 2004-2014 (2) and increasing demands on services, in part because of an aging population. Only a few provinces have given provincial designation to their Programs for these rare inherited bleeding disorders (disease incidence of 1 in 10,000 to 1 in 1,000,000) requiring specialized and expert care. Budget restrictions at the Program level are viewed by the CHS as "penny-wise."

The cost of coagulation therapies (factors I, II, VII, VIII, IX, XI, XIII, VIII/VWF and factor VIII bypassing therapies) in 2013-14 was $204,000,000 for Canadian Blood Services (3) and $46,000,000 for Héma-Québec (4) for a total of $250,000,000, despite a significant decrease in the per unit cost of recombinant factor VIII—factor VIII represents close to 50% of the total coagulation factor budget—compared to previous years (5). Prescription of these biological products requires the expertise of hematologists experienced in the care of people with inherited bleeding disorders, expertise and experience that resides only in physicians working in IBDCCCs. Approximately ninety percent of these products are infused by patients and their caregivers via home infusion programs to prevent bleeding, a Standard of Care for many patients in Canada and around the world for the last several decades (6). Many Programs, however, describe a lack of capacity to adequately monitor the optimal utilization of these clotting factor concentrates used in home infusion protocols and their key clinical outcomes. Systems to report home infusions in accurate and timely ways are inadequate. Data entry resources cannot react to or solve any identified problems with the actual data entry process. Nurses are unable to respond quickly to critical care issues as identified by accurate and timely reporting of factor utilization. Under- and over-utilization occur. Sub-optimal health outcomes, including unnecessary bleeding and joint damage, are the results. Product wastage occurs in such an environment.

The cost of coagulation therapies, therefore, represents 90 to 95 percent of the total costs for the care of people with bleeding disorders. A silo approach to funding the care of people with inherited bleeding disorders is a barrier to rational resource allocation; hospitals are unaware of and perhaps unconcerned with the cost of factor concentrates reimbursed via the provincial blood budgets, and Ministries of Health have little control over hospitals' human resources. The CHS views the current under-funding of Programs, whose role is to facilitate the best health outcomes with such valuable coagulation therapies, as "pound-foolish."

Why is this a critical time to correct the problems?

A key indicator of the level of hemophilia care in a country is the number of International Units (IUs) of factor VIII consumed per capita. Canada currently uses 5.7 IUs per capita , 25-50% less factor VIII than developed countries such as United States, United Kingdom, Australia, Ireland and Sweden (7), in part due to historically conservative prophylactic protocols. In Canada over the last five years, utilization of factor VIII has been increasing at a rate of 6% per year (8). The recent 40-50% decrease in the per unit cost of factor VIII in Canada is very likely to be accompanied by an increase in utilization, as patients and physicians adopt prophylactic regimens that more effectively protect against bleeding and improve quality of life. At the same time, extended half-life factor VIII and IX concentrates, the first advance in the efficacy of factor therapies in four decades, are being introduced. The abilities to document utilization and evaluate health outcomes as clinical practices evolve are more critical than ever. This can only be done with adequate human resources in the Centres.

Moreover, to facilitate infusion reporting, data collection and health outcome research, the Association of Hemophilia Clinic Directors of Canada and McMaster University are launching the Canadian Bleeding Disorder Registry (CBDR) across Canada in 2015 to replace the outdated Canadian Hemophilia Assessment and Resource Management System (CHARMS). This information system has the potential to significantly improve home infusion reporting, data collection, health monitoring by Centre staff, patient decision-making, and health outcome research across the country and internationally. It will attain these goals, however, only if the CBDR is adequately funded in the medium to long term. The cost, including data entry resources in the Centres, is estimated at $600,000-$700,000 per year (9), or approximately one-quarter of one percent (0.25%) of the cost of coagulation therapies. Resources for accurate and timely data entry must be available in the 25 IBDCCCs; proper implementation of CBDR depends on it.

The centre assessments revealed that 52.2 FTEs are currently funded in the core disciplines of nursing, physiotherapy, social work and clerical/data entry across Canada. An additional 21.1 FTEs are considered necessary to help Program staff meet the current challenges of increasing numbers of patients and respect Standards of Care. At an average of $70,000 per FTE, this represents a national investment of less than $1,500,000, or approximately one-half of one percent (0.5%) of the current total budget of $270,000,000 for the care of people with inherited bleeding disorders.

Despite these shortcomings, it must be said patients and their families express very strong appreciation for the competence and dedication of the health care providers who work in the 25 IBDCCCs across Canada, and for the comprehensive care model that has evolved over the last four decades.

KEY RECOMMENDATIONS

1. That health officials in the provinces and territories consider the results of the individual Centre assessment reports and adjust human and physical resources in IBDCCCs so that Standards of Care can be respected and optimal care provided. Of paramount importance are the core team disciplines: physiotherapy, social work, administration/data entry, pediatric and adult physician, nursing.

3 & 4. That the provinces and territories support the medium and long-term operational costs of the Canadian Bleeding Disorder Registry so that utilization of coagulation therapies can be effectively monitored, individual patient care improved and overall health outcomes evaluated; That hospitals, regional health authorities and Ministries of Health (as appropriate) provide sufficient resources in data entry/management so that the Canadian Bleeding Disorder Registry can be used to its full potential.

11. That Inherited Bleeding Disorder Comprehensive Care Centres be recognized by Ministries of Health as having provincial mandates and that provincial or designated envelope funding be provided (where this is not already the case).

13. That IBDCCCs be given a provincial mandate for the prescription of clotting factor concentrates such that, except in emergency, they can only be prescribed to patients registered in a Program; and that outlying hospitals that distribute clotting factor concentrates be required to report distribution to the Program (where this is not already the case).

16. That IBDCCCs and the hospitals that house them support professional training and continuing education for core team members for these rare conditions through their national associations, namely: Canadian Association of Nurses in Hemophilia Care, Canadian Physiotherapists in Hemophilia Care and Canadian Social Workers in Hemophilia Care.

ALL 25 RECOMMENDATIONS CAN BE FOUND IN SECTION F.

Section B: Introduction

Comprehensive care

In Canada, the minimum standard of care for patients with inherited bleeding disorders is known as ***comprehensive care*****.** Comprehensive care is principally delivered by a specialized interdisciplinary team to ensure accurate diagnosis; early and adequate factor replacement for bleeding episodes; "prophylaxis" or regular factor administration to prevent joint bleeding and preserve joint health; long-term management of joint and muscle damage and other sequelae; psychosocial support and education required to manage the bleeding disorder; and management of treatment associated complications. The latter can include the development of "inhibitors", antibodies that interfere with the function of the transfused coagulation factor, and/or chronic infections acquired through contaminated blood products. Core members of the specialist interdisciplinary team include:

- Physicians – pediatric and adult hematologists with additional training in the management of inherited bleeding disorders who provide medical oversight of care and provide expert advice to physicians province-wide.
- Clinical Nurse Specialist and Patient/Nurse Educator – advanced nursing care to individuals/families including teaching, case management and supporting patients in assessment of bleeds, navigating the health system with a bleeding disorder, home intravenous administration of factor and management of preventative doses of factor around major and minor procedures.
- Physiotherapist – with additional musculoskeletal training or experience has an active role in assessment of chronic joint health, bleeds and peri-operative recovery to promote optimal joint health, minimize disability and improve quality of life.
- Social Worker – to offer assessment, intervention and evaluation of psychosocial issues and support patients in managing the complexities of work, school, immigration or travel with a bleeding disorder. They play a major role with patients transitioning, securing financial resources to assist with travel, clinic attendance and other complexities such as compensation for HIV and hepatitis C infected patients.
- Administration/data entry – to provide clerical support to the comprehensive care team and record data related to the home use of clotting factor concentrates, including product utilization and key clinical outcomes.

Comprehensive care has been shown to reduce disease and treatment-associated morbidity and mortality. The minimum comprehensive care standard for adults is one clinic visit per annum as well as ongoing timely access to the care team as needed throughout the year. The standard for pediatric patient assessments is twice per annum.

Hemophilia and other inherited bleeding disorders

Bleeding disorders can be divided into four categories.

Hemophilia A and B

Hemophilia A and B are both lifelong, hereditary disorders caused by deficiencies in the specific blood proteins factor VIII or IX. In 1 out of 3 cases, there is no history in the family; the cause is a new genetic mutation. Thus hemophilia can affect any family. The two types of hemophilia, A and B, affect 3800 Canadians.

The blood of people with hemophilia doesn't clot normally; they don't bleed more profusely or more quickly than others … but for a longer period if not treated. External wounds are usually not serious. Far more important is internal bleeding into joints and muscles. Untreated, this is excruciatingly painful and leads to severe crippling. When bleeding occurs in a vital organ, especially the brain, it can be fatal. Where treatment is unavailable, life expectancy is less than 20 years. Today, with access to an optimal supply of clotting factor concentrates and excellent comprehensive care, young people with hemophilia can look forward to life expectancy very close to normal and full integration into society.

Hemophilia is an X-linked genetic disorder; therefore, the most severe forms of hemophilia affect almost only males. Women who are carriers, however, often have mild symptoms and can have bleeding problems that affect their quality of life.

In about 30% of people with severe hemophilia, the immune system rejects the clotting factors infused to stop or prevent bleeding. In one-third of these cases, this complication, called an inhibitor, is persistent, very serious and can be life-threatening.

The cost of clotting factor concentrates to treat hemophilia A and B in Canada in 2013-14 was close to 215 million dollars.

Von Willebrand disease (VWD) … the most common inherited bleeding disorder

One in 100 Canadians (300,000 people) carries the gene for von Willebrand disease. Symptoms affect an estimated 30,000 Canadians, both male and female. Many of these people have yet to be properly diagnosed; just over 4,000 are registered in IBDCCCs. Life-threatening hemorrhaging can occur after childbirth, surgery or trauma.

A woman's quality of life can be more seriously affected. Heavy menstrual bleeding can lead to hysterectomies. These can be avoided if the woman is properly diagnosed and treated.

Effective treatments, both plasma-derived clotting factor concentrates and chemical drugs, are available for von Willebrand disease.

The cost of von Willebrand factor concentrates in 2013-14 was approximately 28 million dollars.

Rare factor deficiencies

A small number of Canadians, fewer than 1500, suffer from rare factor deficiencies. These people have low levels of a specific blood protein, either factor I, II, V, VII, X, XI, or XIII. Like hemophilia and VWD, these are genetic conditions for which there are treatments in the form of clotting factor concentrates, but no cures. The cost of factor concentrates to treat rare factor deficiencies in 2013-14 was approximately seven million dollars.

Platelet function disorders

There are many different kinds of inherited platelet function disorders. In these disorders, blood platelets do not function normally, resulting in blood not clotting properly. In some

individuals, bleeding can be severe. Some people have no symptoms at all until they have a serious injury or surgery. As with von Willebrand disease, many cases go undiagnosed for decades.

More than 10,000 people with inherited bleeding disorders are registered in Canada's 25 inherited bleeding disorder comprehensive care centres (IBDCCCs).

Canadian Hemophilia Society

The Canadian Hemophilia Society (CHS) was founded in Montreal in 1953 by a small group of people with hemophilia, their families and physicians. Their dream at the time was to improve the quality of life and find a cure for hemophilia. The close collaboration among patients, health care providers and researchers was unique then and provides a model for the health care field today.

Through their efforts, the CHS quickly developed from a small, Montreal-based support group to a national volunteer patient organization.

Today, the Canadian Hemophilia Society is a national not-for-profit health charity, governed by a volunteer Board of Directors. Its national headquarters are in Montreal. The CHS has 10 autonomous provincial chapters, each with a volunteer Board of Directors. There are provincial chapter offices in three provinces: Quebec, Ontario and Manitoba. The CHS and its chapters have approximately 300 active volunteers and 20 staff across the country. The CHS is affiliated with the World Federation of Hemophilia, which is officially recognized by the World Health Organization.

The CHS provides programs and services to people with hemophilia A and B, von Willebrand disease, rare factor deficiencies and inherited platelet disorders, and to the health care providers who care for them.

The Canadian Hemophilia Society has five strategic priorities:

Care and Treatment – The CHS works in close collaboration with medical professionals—physicians, nurses, physiotherapists, social workers, and other related specialists—in the 25 specialized bleeding disorder treatment centres across the country. Our common goal is to ensure optimal inter-disciplinary care and treatment for all members of the family. We define this as *comprehensive care*.

Research – The CHS provides basic and clinical research grants and research fellowships to fund leading Canadian researchers working in the field of bleeding disorders in an effort to improve care and treatment, and ultimately find a cure.

Support and Education – The CHS is the primary source of educational materials designed for people with bleeding disorders, their families, health care professionals and the general public, and is world renowned for the quality of its programs and publications. Our website is recognized as the most comprehensive in the world.

Safe, Secure Supply of Therapies – In the 1970s and 1980s, 700 Canadians with hemophilia were infected with HIV from tainted blood. Two-thirds of these people have passed away. In addition, 1600 were infected with hepatitis C. The CHS played a key role in building a safer blood system for Canadians, leading to the creation of Canadian Blood Services and Héma-Québec. The CHS continues to be vigilant as the *watchdog* of the blood system on behalf of all Canadians. CHS experts serve on Canadian Blood Services and Hema-Quebec committees, provincial blood advisory committees, and monitor blood safety nationally and internationally.

The CHS receives program funding from all the companies in the Canadian clotting factor concentrate market. Our relationship with them is carefully regulated by our *Policy on Relationships with Companies in the Pharmaceutical Industry*, which is guided by best ethical practices. The CHS also receives significant donations and bequests from members of the general public and is one of 16 members of Healthpartners, a federal government workplace giving program. While the national organization receives no government funding, two of our chapters, Quebec and Ontario, receive provincial supporting grants.

Context

In June 2007, after a lengthy process led by the multi-disciplinary Canadian Hemophilia Standards Group, a document entitled *Canadian Comprehensive Care Standards for Hemophilia and Other Inherited Bleeding Disorders* (*see* **Annex 4**), based on work done in Ontario, was adopted by the four health care provider associations whose members make up the core teams in hemophilia / inherited bleeding disorder programs: the Association of Hemophilia Clinic Directors of Canada (AHCDC), the Canadian Association of Nurses in Hemophilia Care (CANHC), the Canadian Physiotherapists in Hemophilia Care (CPHC) and the Canadian Social Workers in Hemophilia Care (CSWHC), and the national patient organization, the Canadian Hemophilia Society. The focus of these standards is on the structural and resource requirements necessary for a Hemophilia Treatment Centre (HTC) to effectively provide care, and on its functions and responsibilities.

In May 2010, the Canadian Hemophilia Standards Group published the results of a self-assessment survey conducted by 24 of the 25 IBDCCCs to validate the standards (10). The general level of acceptability of the standards was found to be high and it was concluded that they could be used for the purpose of external accreditation.

An external accreditation process, based on the audit processes developed by the United Kingdom Haemophilia Centres' Doctors Organization and the Irish Haemophilia Council, was developed by the Canadian Hemophilia Standards Group in 2011, but never implemented.

In 2013, considering the absence of a timetable to pursue a formal accreditation process, the Canadian Hemophilia Society developed its own assessment process. The overall goal is to identify any gaps in resources that may prevent IBDCCCs from delivering care and treatment according to the adopted Standards and that may lead to poor/sub-optimal patient outcomes. IBDCCC participation in the process was supported by the AHCDC. The primary objectives were to:

- Conduct a thorough assessment of the services and resources in Hemophilia Treatment Centres in each province;
- Prepare a detailed report and recommendations for hospital administrators and/or Ministry of Health officials in each province;
- Identify and meet key decision-makers in each province with the goal of maintaining and improving the care for people with inherited bleeding disorders;
- Follow up to push for implementation.

Methodology

The assessment is in two parts:

- interviews with core members of the comprehensive care team, either individually or in groups (*see* **Annex 1**), based on a questionnaire developed in 2011 by the Quebec Chapter of the Canadian Hemophilia Society and used to assess the four Quebec HTCs;
- a patient satisfaction survey, developed by the Canadian Hemophilia Standards Group for the accreditation process. A slightly modified version of the survey was used successfully in 2013 by the Irish Haemophilia Council in its audit of IBDCCCs in Ireland. A random selection of 3000 patients who had been seen in clinic in the last two years was made using the Canadian Hemophilia Registry (CHR) database. Questionnaires were sent by mail by the Centre staff. Answers were returned to the Canadian Hemophilia Society for compilation. Responses were anonymous. Twenty of the 25 Canadian centres distributed the patient satisfaction survey. Due to problems with CHR identifiers and varying degrees of participation by hemophilia / inherited bleeding disorder programs, it is not possible to state with any accuracy the number of questionnaires distributed; however, 347 were completed and received by the CHS. (*See complete national results in* **Annex 2**.)

The methodology is limited in that the assessors, for reasons of confidentiality, had no access to patient records and could not independently verify key indicators such as, for example, the capacity of the Programs to provide semi-annual or annual assessments as per the Standards. Concordance between the information provided by the Centre staff and the responses collected through the patient survey, however, often overcame this limitation.

PARTICIPATION

The participation of the centres was entirely voluntary. The four Quebec centres participated in the assessment in 2011 and it was not considered necessary to repeat the process in 2013-2014. Of the remaining 21 centres in the rest of Canada, 19 accepted to participate fully. One declined. In the remaining centre, only the adult hematologists could be interviewed. The patient satisfaction survey was, however, distributed. All 25 centres provided up-to-date information on the numbers of patients registered and on the staffing levels of core team members.

The visits to the centres occurred between October 2013 and July 2014. Draft reports were prepared and returned to the centre directors for review and validation. Final reports were distributed to centres between October 2014 and April 2015.

Assessors

The assessors were David Page, national executive director, Canadian Hemophilia Society; Sarah Crymble, Ontario hemophilia provincial coordinator; and Michel Long, national program manager, Canadian Hemophilia Society. Guidance and input on the assessment process was provided by Pam Wilton, RN, past-president of the Canadian Hemophilia Society and Board member of the World Federation of Hemophilia. (*See* **Annex 3** *for biographies.*)

Section C: Numbers of patients

Table 1 lists the numbers of patients by disease condition as reported by each of the 25 Programs between October 2013 and July 2014. These numbers are very similar to the data provided by the Canadian Hemophilia Registry in May 2014 to the World Federation of Hemophilia Global Survey (11).

Table 1: Numbers of bleeding disorder patients

Hemophilia A	Severe	850
	Moderate	288
	Mild	1,774
	Inhibitors	105
	Other*	74
	Total	**3,091**
Hemophilia B	Severe	175
	Moderate	259
	Mild	252
	Inhibitors	2
	Other*	21
	Total	**694**
von Willebrand disease	Type 1	3,410
	Type 2	499
	Type 3	998
	Undefined	181
	Total	**4,188**
Rare factor deficiencies		**1,200**
Inherited platelet disorders		**954**
Undiagnosed bleeding disorders*		**264**
Total		**10,391**

* Acquired, unclassified, carriers, referrals

Section D: Key observations by health care providers

1. The strengths of the Programs

The staff identified the following key strengths of the 25 Programs (the numbers in brackets indicate the number of Programs that made the same observation):

Human resources

- Committed, cohesive, stable and well-trained inter-disciplinary teams that have developed longitudinal experience and knowledge of patients, and that communicate well with each other; (16)
- A full complement of core team members; (6)
- A sufficient number of hematologists, providing flexibility and back-up; (4)
- Nurse practitioners as part of the team; (2)
- Flexibility in the nurses' work schedules to accommodate patient schedules, urgent care and support for surgical procedures; (2)
- Inclusion of an obstetrician-gynecologist and dentist as extended team members; (1)
- A designated physiotherapist; (1)
- Weekly meetings of the team; (1)
- Excellent support from managers; (1)
- Capacity to attend summer camp as part of job description. (1)

Services

- A sufficient number of clinics to assess patients every 3, 6, 12 or 24 months, according to the *Comprehensive Care Standards for Hemophilia and Other Inherited Bleeding Disorders*; (9)
- A strong home care program with re-certification that reduces in-patient and Emergency Department visits to a minimum; (8)
- Ability to participate in the Canadian Hemophilia Surveillance System (CHESS) and conduct high-level pre-clinical and clinical research; (8)
- An integrated pediatric-adult program and seamless transition for patients graduating from pediatric to adult care; (7)
- The capacity to hold travelling clinics in other centres; (6)
- An 18/7 or 24/7 nursing on-call system; (4)
- 24/7 coverage by the on-call hematology/oncology team; (4)
- A strong capacity to work with the recent immigrant population and the special needs they present to the Program; (4)
- Daily access to nursing care at the Centre; (2)
- Very short wait times for new patients referred to the Program; (2)
- A monthly Women and Bleeding Disorder clinic with OB-GYN expertise; (2)
- A patient-centered service; (1)
- Excellence in therapeutic services; (1)
- Good rapport with patients; (1)
- Very close follow-up with patients; (1)
- Having pediatric care and the women's program in the same building; all the sub-specialties are at hand; (1)

- A genetic counseling service; (1)
- A strong investment in newborns and very young children, which research has shown to be a key to preventing joint damage in later life; (1)
- A psychologist able to provide support for issues of stress and chronic pain, and to promote adherence to treatment protocols; (1)
- Copies of clinic letters sent to families/patients to reinforce treatment; (1)
- Saturday clinics for out-of-towners; (1)
- Genotyping of 100% of patients; (1)
- The capacity to treat infectious disease (HIV, HCV) complications of past clotting factor treatment; (1)
- Experience in immune tolerance induction therapies; (1)
- Success in working with patients to generate accurate home infusion records; (1)
- Capacity to conduct Saturday or after-school educational sessions with young patients. (1)

Physical and information resources

- Excellent space for treatment rooms in the Centre or an excellent dedicated clinic space; (5)
- Good physical space for offices in proximity to clinic space; (4)
- A fully deployed Electronic Patient Record; (4)
- Access to a 24/7 hematology out-patient clinic that provides emergency care out-of-hours, and helps to avoid recourse to the Emergency Department; (3)
- Ability to use Telemedicine to diagnose and treat acute bleeds in patients in remote locations; (3)
- On-line information on both the pediatric and adult programs, including location; (3)
- Easy access for patients to the clinic (drop-off at door); (2)
- Up-to-date manuals to promote quality assurance. (2)

Training, Continuing Education

- Ability to attend regional and national meetings of professional associations to share best practices (medical, nursing, physiotherapy, social work); (13)
- Involvement with national professional associations (medical, nursing, physiotherapy, social work). (9)

Relationships, outreach

- Capacity to liaise with and do outreach to other health care providers and Emergency Departments at other hospitals; (12)
- Good relationships with and a well-functioning hemostasis lab; (8)
- Good working relationship with the CHS Chapter; (7)
- Good access to and support from other health care services and hospitals; (6)
- In-service training by bleeding disorder nurses for in-patient care and in the Emergency Department; (6)
- Capacity to do outreach to schools; (3)
- Close working relationship/communications with the adult Program; (1)
- Treatment instructions for all patients coming to Emergency. (1)

Funding

- Designated provincial funding; (2)
- Funding for patients to attend clinics; (2)
- Access to a fund (through industry and the local chapter of the CHS) for certain patient supplies. (1)

2. The weaknesses of the Programs

The staff identified the following areas that need to be addressed to best meet the Standards:

Human resources

- Serious lack of adequate human resources, vulnerability to hospital-level decisions; (23)
- Insufficient or no dedicated time for the social work position; (19)
- Physiotherapy position not adequately funded; (17)
- Insufficient or no dedicated time for administration / data entry; (15)
- FTE in nursing is insufficient; (13)
- Core team is incomplete; (9)
- Difficulty with staff retention; (1)
- No dentistry; (1)
- No psychologist; (1)
- No access to pain specialist; (1)
- Sub-specialty support is deficient: the Program has no dedicated rheumatologist, orthopedic surgeon or dentist. (1)

Services

- Insufficient clinic time (usually adults); (9)
- Lack of resources to follow up on home infusion diaries; (6)
- No capacity to conduct outreach clinics; (4)
- Restrictions on testing in coagulation lab on weekends; (3)
- Lack of involvement in national research to assess health outcomes; (3)
- No specialized clinic for women with bleeding disorders with an assigned obstetrician-gynecologist; (2)
- Clinic inaccessible from 9 to 5; (1)
- Lack of 24/7 access to Centre personnel; (1)
- Lack of capacity to follow schedule of annual, semi-annual and quarterly who cannot come to centre or attend travelling clinics; (1)
- Lack of support for patients to travel; (1)
- A long waiting list for assessment and surgery; (1)
- Need for improvement in transitioning teens; (1)
- Inability to conduct home visits to do education about home infusion; (1)
- No oral surgeon serving the Program; (1)
- An inability to follow mild patients treated in the community and who are not registered in the treatment centre. (1)

Physical and information resources

- Insufficient, inadequate clinic space (usually adults); (9)
- Information technology; (8)
- Lack of robust quality assurance programs, written policies and procedures for assessing patients are lacking; (7)
- Need for more efficient data entry of patient care information; (4)
- Need for more on-line information on Program and to direct patients to the Centre; (3)
- Office space not optimal; (2)
- Lack of tools (such as GRASP) to help track workload; (1)
- Program can no longer supply infusion equipment. (1)

Training, Continuing Education

- Core team members not supported in pursuing continuing education, largely through meetings of their national associations. (4)

Relationships, Outreach

- Lack of visibility within the hospital and with health authorities, no provincial recognition; (4)
- Transition program difficult to coordinate through two different health authorities; (1)
- Decreasing support from chapter, especially in terms of effective advocacy; (1)
- Not all VWD patients are referred to the Program. (1)

Funding

- Need for increased support from the chapter to help patients (e.g. financial support to attend clinic). (1)

3. Human resources

Current levels

Following consultation with experts in the IBDCCCs, the number of hemophilia A and B patients in a centre was chosen as the best criterion for comparing staffing levels. All people with hemophilia A and B, whether severe, moderate or mild, are very likely to be diagnosed and followed in an IBDCCC. It was decided to give the same weight in this calculation to each patient, independently of the burden of severe disease, the presence of inhibitors, infectious disease complications or pediatric status. Policies on registration of patients with von Willebrand disease, on the other hand, are quite variable from Program to Program and made their total numbers a less reliable benchmark. Other factor deficiencies are exceedingly rare and even more variable from centre to centre.

Table 2 lists the full-time equivalent (FTE) positions for the four core program disciplines of nursing, physiotherapy, social work and administration/data entry in the 25 IBDCCCs per 100 hemophilia A and B patients. Centres 1 to 5 are pediatric Programs; Centres 6 to 10 provide services only to adults; Centres 11 to 25 are Programs that serve pediatric and adult populations with the same core team.

Table 2: Full-time equivalents (FTEs) per 100 hemophilia A and B patients in all centres

Centre	Nursing	Physiotherapy	Social work	Admin/data	Comments
1	1.5	0.4	0.3	0	These are pediatric centres
2	1.0	0.2	0.3	0.6	
3	1.0	0.3	0.2	0.14	
4	0.9	0.1	0.2	1.1	
5	1.6	0.4	0.4	1.0	
6	0.6	0.2	0.2	0.2	These are adult centres
7	0.6	0.1	0.1	0.6	
8	0.4	0.1	0.01	0.2	
9	0.4	0	0	0.2	
10	0.6	0	0.1	0.1	
11	1.2	0.14	0.2	0.5	These are combined pediatric / adult centres
12	0.5	0.2	0.05	0.6	
13	0.8	0.2	0.3	0.8	
14	0.5	0.1	0.1	0.6	
15	1.5	0.25	0.1	0.4	
16	0.6	0.1	0.05	0.01	
17	1.0	0.1	0	0.6	
18	1.3	0	0	0.6	
19	1.4	0.4	0.4	1.8	
20	1.0	0.05	0	0	
21	0.6	0.1	0	1.0	
22	1.0	0.25	0.02	0.25	
23	1.1	0.07	0.07	0	
24	0.2	0.01	0.01	0.1	
25	0.4	0.01	0	0.4	
Total	21.7	3.88	3.11	12.36	
Mean	0.87	0.15	0.13	0.47	

To compare centres of similar size and with similar populations, the 25 IBDCCCs were divided into five categories:

- the five pediatric centres
- the five adult centres
- the five largest combined pediatric/adult centres
- the five medium-sized combined pediatric/adult centres
- the five smallest combined pediatric/adult centres.

Their ratios of FTEs to 100 hemophilia A and B patents are presented in Table 3.

Table 3: Comparison of full-time equivalents (FTEs) per 100 hemophilia A and B patients by category of centre

	Nursing	Physiotherapy	Social work	Admin/data
Pediatric centres	1.20	0.30	0.30	0.57
Adult centres	0.50	0.08	0.08	0.26
5 largest combined centres	0.82	0.15	0.06	0.47
5 medium-sized centres	1.1	0.17	0.13	0.68
5 smallest combined centres	0.74	0.05	0.06	0.38
Mean	0.87	0.15	0.13	0.47

One can observe from Table 3 that…

- Human resources tend to be highest in pediatric centres;
- Nursing resources tend to be lowest in adult centres;
- Physiotherapy and psychosocial resources tend to be lowest in adult centres and in the smallest combined pediatric/adult centres;
- Beyond the above three observations, there is wide variation in human resource/patient ratios in all core disciplines from centre to centre with no consistent or clear explanation or justification.

See tables 5 to 9 in **Annex 5** *for a breakdown by Centre.*

Optimal levels

The assessors gathered views from all Program staff on the recommended staffing levels to optimally respect Standards of Care and also considered comments made through the patient satisfaction survey. The numbers in brackets represent the additional staffing in FTEs that is recommended.

Table 4: IBDCCC current FTEs and recommended additional staffing

Centre		Hemophilia A and B patients	Nursing	Physiotherapy	Social work	Admin/data
1	Pediatric	103	1.6 NP	0.4	0.3	0 (1.0)
2	Pediatric	63	0.6 (1.0)	0.15 (0.3-0.4)	0.2	0.35
3	Pediatric	167	1.6 (2.0)	0.5 (1.0)	0.33 (1.0)	0.25 (1.0)
4	Pediatric	56	0.5 (0.7)	0.05 (0.2)	0.1 (0.2)	0.2 (0.4)
5	Pediatric	57	0.9 (1.1)	0.2 (0.4)	0.2 (0.3)	0.2 (0.5)
6	Adult	215	1.3 (2.0)	0.4 (0.8)	0.4 (0.8)	0.5 (1.5)
7	Adult	175	1.0* (1.6)	0.2 (0.4)	0.2 (0.4)	1.0 (1.5)
8	Adult	404	1.5 (2.0)	0.4 (0.6)	0.1 (0.2)	0.8 (1.0)
9	Adult	184	1.0	0 (0.2)	0 (0.2)	0.4
10	Adult	118	0.7 (1.0)	0 (0.2)	0.1 (0.2)	0.1 (0.2)
11	Ped./adult	196	1.5** (2.0)	0.3 (0.4)	0.4 (0.8)	1.0 (1.5)
12	Ped./adult	245	1.2	0.6 (1.0)	0.1	1.5
13	Ped./adult	60	0.5 (1.0)	0.1 (0.2)	0.2 (0.4)	0.5 (1.0)
14	Ped./adult	174	0.8	0.2	0.2	1.0
15	Ped./adult	158	2.3	0.4	0.2 (0.3)	0.6 (1.0)

Centre		Hemophilia A and B patients	Nursing	Physiotherapy	Social work	Admin/data
16	Ped./adult	179	1.0	0.2	0.1	0.02 (0.5)
17	Ped./adult	104	1.0	0.1	On call (0.2)	0.6
18	Ped./adult	40	0.5 (1.0)	0 (0.2)	0 (0.2)	0.25
19	Ped./adult	139	2.0	0.5	0.6	2.5
20	Ped./adult	62	0.6	0.025 d/y (0.2)	0(0.1)	0 (0.2)
21	Ped./adult	211	1.4	0.2	0 (0.2)	1.0 (1.5)
22	Ped./adult	396	4.0	1.0	0.1 (0.5)	1.0
23	Ped./adult	134	1.5 (2.0)	0.1 (0.2)	0.1 (0.2)	0 (1.0)
24	Ped./adult	89	0.2 (0.5)	0.008 (0.1)	0.008 (0.1)	0.1
25	Ped./adult	56	0.2	0.008 (0.2)	0 (0.1)	0.2
Total		3,785	28.4 (5.6)	5.9 (3.7)	3.9 (4.1)	14.0 (7.7)

NP – Nurse Practitioner*

1.6 for all blood/immune disorders**

2.5 for all blood/immune disorders

4. Staff recommendations to sustain/improve optimal care to meet the Standards

The principal recommendations are:

Human resources

- Increase the FTEs in nursing, physiotherapy and/or psychosocial support where currently needed and evaluate as the Programs grow; (23)
- Increase funding for administrative support and data entry so as to have the capacity to enter data into CHARMS in a timely way in support of patient care and health outcome research; (10)
- Allocate dedicated physiotherapy and social work time to the Program; (9)
- Add a psychologist to the core team; (2)
- Increase time for the hematologist; (2)
- Identify and assign to the Program an oral surgeon with interest in treating people with bleeding disorders; (2)
- Increase access to pain management specialist; (2)
- Identify and assign to the Program an OB-GYN with interest in treating girls and women with bleeding disorders. (1)

Services

- Increase number and/or time of clinics; (9)
- Increase involvement in clinical research; (3)
- Increase capacity to do PK testing and use data; (2)
- Improve the formal transition program from pediatric to adult care; (2)
- Expand outreach clinics; (1)
- Create an effective on-call system; (1)
- Improve clotting factor concentrate distribution and utilization; (1)
- Hold regular clinics for girls and women with bleeding disorders; (1)

- Increase use of Telemedicine; (1)
- Increase education of patients on bleed recognition. (1)

Physical and information resources

- Create an improved electronic patient reporting system (e.g. a SmartPhone app) and a renewed commitment by patients/families to accurately report utilization; (8)
- Build more capacity in the coagulation lab; (5)
- Develop formal written policies and procedures to promote quality care; (4)
- Have dedicated clinic space; (2)
- Buy Smartphones for the Centre; (2)
- Obtain appropriate physiotherapy equipment for the exam rooms; (1)
- Crete a web-based educational tool for teens to improve adherence to prophylaxis schedules and infusion reporting. (1)

Training, Continuing Education

- Support Continuing Education for all core team members; (9)
- Create a mentorship program in physiotherapy to support training and continued education. (1)

Relationships, Outreach

- Make the Program more visible within the hospital and with health authorities; (5)
- Continue and, where warranted, expand travelling clinics. Where patients live in remote communities, the Ministry of Health should provide travel assistance to allow them to attend annual or semi-annual clinic assessments; (2)
- Create more awareness by administrators of the health care and financial benefits of home infusion programs by administration; (1)
- Do more training in Emergency Departments; (1)
- Do more education by the CHS and its chapters on infusion. (1)

Organizational

- Require by government mandate (following the lead of Quebec and Manitoba) that all patients who use clotting factor concentrates be registered in a IBDCCC. Accompany this expanded mandate with sufficient resources to carry it out; (6)
- Mandate a bar code system for clotting factor infusion reporting, which allows scanning both by patients in the home and by health care providers, linked to a national registry. (2)

Funding

- Provide Ministry recognition of Programs with envelope funding; (4)
- Provide more financial assistance from chapters to families in need to attend clinic (gas, parking, food) and summer camp, and for specialized equipment such as helmets and mobility aids. (4)

Section E: Key observations by patients

A random selection of 3000 patients who had been seen in clinic in the last two years was made using the Canadian Hemophilia Registry (CHR) database. Patient satisfaction surveys were sent by mail by 20 of the 25 Canadian centres in the first half of 2014. Due to problems with CHR identifiers and varying degrees of participation by hemophilia / inherited bleeding disorder programs, it is not possible to state with any accuracy the number of questionnaires actually distributed; however, 347 were completed and returned to the Canadian Hemophilia Society for compilation. Responses were anonymous. Not all questions applied to each respondent.

This summary presents the quantitative results, some comments from the assessors and patient quotes to illustrate particular issues. The full results of the survey are available in **Annex 2**.

1. 96% (237/248) are satisfied with the availability of the centre team when needing to be seen for an urgent problem.

   Sixteen people indicated that they live between 1 to 9 hours away from the Centre, which prevents them from going to the clinic without an appointment for an urgent need or a new problem. Some indicated that they therefore tend to rely more on phone and email communications or they visit their local Emergency Department. Among these, many indicated being satisfied with the phone support. In one case, they were directed to a local hospital with which the HTC was in touch and in another the nurse coordinator arranged the necessary treatment in preparation for when the patient was to arrive at the HTC.

2. 96% (230/240) are satisfied with the care they receive when needing to be seen for an urgent problem.

   Nineteen people commented being very satisfied with the care and attention they receive at the Centre.

   I phoned (really out of the blue) because I had a previously diagnosed blood disorder, needed dental surgery (which was not going to be done until I went to the blood clinic). I was seen immediately AND compassionately.

3. 88% (302/342) are supplied with a *FactorFirst* wallet card to facilitate care at the Emergency Department.

   A wallet card with bleeding disorder info for a triage/emergency nurse/doctor would help especially for 1) unsuccessful home infusion 2) urgent/ new medical problem.

4. 62% (167/271) reported the medical information on the card is updated annually.

   Nine people indicated the *FactorFirst* gets updated every two years and, for many, during their appointments set every other year. Eight people indicated it is updated as needed if their conditions change.

5. 96% (152/158) are satisfied with the arrangements that are in place for home treatment (for example, telephone access to nurse coordinator, education in home infusion techniques, supply of factor concentrates, supply of infusion equipment).

   Nine people commented being very satisfied and have found the system efficient; however, some patient concerns were raised.

We are required annually to do a one-on-one clinic with our nurse to ensure we are doing techniques correctly and sterilizing correctly.

Our nurse coordinator is impossible to reach, I always get her voicemail and can go days without a call back only to find out she's on holiday, sick or just days off. Our clinic really needs a full-time nurse coordinator.

We have to purchase the infusion equipment and we have no medical coverage. It's very expensive because our son has a port. 12 needles are $200.

6. 59% (141/240) on *home infusion* regularly fill out an electronic or paper diary about home treatment/bleeding events; 5% (7/148) do not fill out a diary even though they are asked; 20% (38/240) report they are not asked to fill out a diary about home treatment/bleeding events.

   I always fill out the diary (maybe not always in a timely manner;) way easier with the online services now.

   I've never been asked for it, so found it to be a needless waste of my time, so I stopped.

   There is no follow through and it is very old school (paper). Very inefficient, would prefer electronic process online.

   Would be nice if our clinic used EZ log or any digital log.

   Usually phone in to data clerk at HTC for record of bleed.

   I don't think the comprehensive care centre is reviewing these records. There is no contact from clinic during periods of high usage to cope with serious bleeds or trauma.

7. 18% (60/339) report they visited their centre for a scheduled check-up every 6 months; 49% (167/339) report they visited their centre for a scheduled check-up every 12 months; 14% (49/339) report they visited their centre for a scheduled check-up every 24 months; 5% (16/339) report they never visited their centre for a scheduled check-up; 14% (47/339) report they visited their centre on a different schedule.

   Should be once a year but it is always very difficult to get an appointment. I think I have waited up to 3 years.

   We are invited to attend the Wednesday clinics but opt not to because of our distance.

   Tough to get to from our town to Vancouver, especially when clinic days are Tuesdays.

   The team has been to my hometown hospital a few times for clinics and I have seen them when they were here. I live 5 hrs away from the clinic and I have only attended a clinic a few times except when I was hospitalized in St John's.

   Usually done by Telemedicine from my community to HTC.

8. 95% (318/334) report they meet their physicians at the time of their regular check-up; 96% (321/334) report they meet their nurses at the time of their regular check-up;

52% (173/334) report they meet their physiotherapists at the time of their regular check-up; 33% (112/334) report they meet their social workers at the time of their regular check-up.

Other specialties mentioned in comments: dentist (17); rheumatologist (9); orthopedic surgeon (5); genetics counselor (5); surgeon (5); medical student/resident (5); administrative assistant-secretary-receptionist (5); research nurse (4); ob-gyn (3); lab personnel related to blood work (3); pediatrician (2); psychiatrist (1) nutritionist (1); gastroenterologist (1).

We spend more time waiting in between members than actually with them so feel this too is a waste of time.

The social worker presently at the clinic has indicated a lack of understanding of hemophilia. Several years ago, she pointed to judo and other martial arts as a good form of exercise!

My physiotherapist is awesome.

Very often other team members (i.e. social work, physiotherapist) are not available.

9. 92% (301/326) are satisfied with the arrangements in place for regular check-ups.

   Fifteen people indicated wait times are too long.

   We are also happy that they are now willing to offer an outreach clinic in our community once a year.

   Infrequent appointment times: "We have an opening 6 months from now or 9 months from now."

10. 98% (317/325) report all their questions are answered to their satisfaction at regular check-ups.

11. 93% (171/183) report they are satisfied with the physiotherapy and orthopedic services that are available for the management of joint problems.

    Five people, surprisingly, indicated not being aware this service existed.

    Physio services have been available for about a year. My physiotherapist is great, very knowledgeable and professional.

    Having a physio available has made a great difference in preventing joint bleeds.

    The physio does not appear to receive much training regarding joint health and bleeding disorders.

    A friend and four other people had their surgery canceled twice because of lack of nurses to administer product after surgery, which I found disheartening.

12. 21% (73/343) report that dental services are offered through their clinic; 28% (95/343) report that dental services are not offered through their clinic; 51% (175/343) report they do not know if dental services are offered through their clinic.

    Eleven people indicated not being sure if this service was offered.

    My son has had dental services arranged in the past for problems with his teeth/gums. This service was provided in conjunction with the nurse coordinator and it was excellent.

It would be a great benefit since dental procedures are a big issue, despite regular cleaning and check-ups several times a year.

Most times we see a dentist at a clinic visit but we do have our own family dentist and always consult with the bleeding disorders clinic before any procedures are performed.

13. 78% (46/60) report they are satisfied with the dental services offered through the Program.

    We had to make the appointment and coordinate with clinic for treatment; there were no issues.

14. 50% (56/112) report that the cost of dental services is covered.

15. 55% (149/272) report they have access to a social worker if needed; 5% (13/272) report they do not have access to a social worker if needed; 40% (110/272) report they do not know if they have access to a social worker if needed.

    The children's clinic has not been able to maintain a social worker, but the social worker at the adult clinic was available and more than happy to help and VERY knowledgeable.

    Excellent. Our child has been bullied due to his bleeding disorder and she helps him cope.

    Would be great to have regular phone 'visits' with new parents to see how they are doing. Don't wait for the parent to call the SW; make a weekly (?) call simply part of the program to keep lines of communication open. Can lower number of calls as parents become more comfortable with their new BD child. Great way to lower stress and allow gradual supported learning.

16. 27% (73/267) report they have access to a psychologist if needed; 7% (21/267) report they do not have access to a psychologist if needed; 65% (173/267) report they do not know if they have access to a psychologist if needed.

    I think a psychologist would be an asset as my son has had resentment and feelings I was unsure how to handle with regards to frequent infusions.

    The HTC appears unable to retain psychologists; they are like trying to see God or Jesus Christ in the flesh.

17. 85% (68/80) report they are satisfied with the psychosocial services offered through the Program.

    We chat briefly with a social worker but that's not long term counseling, which would be beneficial if offered.

18. 91% (39/43) report that they were satisfied with how the clinic recently helped with transition from pediatric to adult care.

    I started with the clinic at 16 and I am now 19. I didn't notice any changes if there were any.

    My oldest son graduated to adult care and he found the adult clinic to be not nearly as accessible.

19. 78% (238/307) report the clinic team provides information and advice to other health care providers (for example, pediatricians, family physicians, dentists, etc.) who care for the them or their children; 5% (16/307) report the clinic team does not provide this information and advice; 17% (53/307) report they do not know if the clinic team provides this information and advice.

    They always communicate with our local doctor and send him updates after our yearly appointments.

    They have been amazing at this and have made a real difference to my care.

    Team sent note to my family doctor and cardiologist (needed his input re DDAVP). They are available if dentist or other ER, etc. need advice. Have prescription and instruction for upcoming dental work.

    Clinic provides my physician with all information/results of annual check-up.

    This is something that needs to be done better. The GPs don't understand and aren't communicated with.

    This should be done in all situations. Not all other care providers are necessarily and sufficiently informed about hemophilia issues.

    Reports sent to family doctor. Excellent when travelling, providing factor and notifying me and the area that I am visiting in case of emergency.

    The doctors actually did a seminar for the dentists in our area to educate them on bleeding disorders.

20. 58% (131/226) report the clinic team provides information and advice to people in their community (for example, teachers, daycare workers, employers) if requested; 6% (14/226) report the clinic team does not provide this information and advice; 36% (81/226) report they do not know if the clinic team provides this information and advice if requested.

    The nurse came to our daycare and talked to the supervisor.

    They regularly offer to provide information, advice or visits to our sons' teachers.

    I asked my son's nurse to do a presentation as he starts kindergarten in September but she is not able to.

    They have given school presentations, calls, etc. Fantastic support.

21. 82% (256/311) are satisfied with access to the centre, for example: parking, after-hours entrance, signage, availability of wheelchairs in the building; 18% (55/311) are not satisfied.

    Twenty-five people indicated parking as very costly. Nineteen commented that access to the Centre itself is problematic and directions non-existent.

    The clinic is quite difficult to find and information services were not helpful and did not know that the clinic existed. Parking is too expensive. This could really impact a family that needed to visit frequently.

    Parking increases the stress of the situation; can the CHS fund a parking program?

    Shuttle service should be provided from parking lot to hospital.

22. 91% (203/222) report it is easy to book an appointment if they or their children need to go to the centre for help between regularly scheduled appointments for a non-urgent medical reason.

    The clinic team always makes accommodations to get us in for an appointment.

    Not always. Particular staff seem to be away different days of week. It may help if everyone had a regular schedule.

    If I call the clinic, they are always available to talk to me or will call me back in a timely fashion.

    I had difficulty booking appointment, not clear who to call.

23. 23% (40/171) report they are seen immediately; 40% (68/171) report they are seen the same day; 10% (17/171) report they are seen the next day; 27% (46/171) report they are seen 2 or 3 days later.

    Every time I have requested a visit it has always been on the day I requested.

    4 to 6 weeks. I have had a hard time convincing the clinic nurse that I need to be seen.

    Very accommodating to me! Nurse is amazing and tireless!

    However, I have a concern about the after-hours on-call phone service being eliminated. This is valuable. In the past, I have had ER treatment given using the wrong protocol (i.e. intravenous instead of subcutaneous). This resulted in a shock response driving home. The on-call nurse had not been consulted by ER. If the on call service were eliminated, the potential for these incidents would only increase.

    It depends on the issue and who I'm willing to see about it. My regular nurse only works part-time and I'd rather wait to see her than explain my whole history to someone for a simple question.

24. 86% (273/316) report they know whom to contact for help if the comprehensive care clinic is closed or after hours.

25. 70% (166/236) report they are satisfied with the care they have received at the Emergency Department.

    Thirty-nine people indicated that the main issue is that Emergency personnel, especially in those hospitals that do not house an IBDCCC, lack knowledge about bleeding disorders.

    We don't visit the ER anymore now that we do home infusions, but it was very frustrating and time consuming at our local hospital. Always had to be admitted and send to PEDS instead of treated in ER.

    We get very good (although maybe not as knowledgeable) service in emerg. There, we are people and not an inconvenience. The docs are great (know hemophilia) but the nurses don't always. As a parent, I have to make sure I know the treatment plan because the clinic doesn't communicate quickly with emerg.

    Not always, they often do not believe you have a bleeding disorder even if you show your card.

    Had a joint bleed and waited 4 hours before getting factor (now we self-infuse).

Emergency departments do not seem to be helpful to people with bleeding disorders. They do not seem to believe the patient or know what to do.

They don't understand and have tried to kill me a few times doing things they shouldn't.

I consider myself to be very well educated on the subject and find ER visits to be a very frustrating experience. Last time we saw SIX doctors before he was given his factor and two of them told me to give him ibuprofen.

Local ER doctors and medical staff have very limited hemophilia knowledge. I know more than they, and have them contact my clinical team for advice.

We have had some good and some horrible experiences. However, the nurses at our clinic are very efficient with contacting ER to check on our visits and complaints.

I feel that rural hospitals do not have enough information regarding bleeding disorders or the implications of a severe joint injury ... and because of that I will transport my child to Saskatoon when the need arises.

The emergency department at the local hospital does not provide proper care.

Not always: most doctors have little knowledge of hemophilia and when to give the factor needed.

26. What do you think is very good about your comprehensive care clinic?

The vast majority, 89 of the respondents who commented, indicated that it is the staff team that makes their Centre very good. Most indicated that they find them to be very caring, kind, knowledgeable, personable, understanding, sensitive, empathetic, reassuring, friendly, courteous, helpful and committed. Continuity and minimal turnover of staff were also mentioned as positive elements.

The quality of the care and treatment was also rated very highly by a vast majority underlining efficiency, competency, compassion, dedication and professionalism. Services were said to be well organized and meticulous. Many feel they have established trusting, strong, long-term relationships with their Program team members particularly with the nurse coordinators. They feel they are treated with respect.

Many respondents find the Program staff very accessible, accommodating, attentive, thorough, informative and centered on their needs both as a patient but also as a human being. Many said the Centres have a family feeling.

Having a multi-disciplinary team that has wide and varied expertise was also deemed to be very valuable. This gives better access to specialized physicians, and reduces wait time for appointments. They work together at troubleshooting problems and finding solutions.

Regarding communications, respondents indicated they appreciate the open communications, being listened to and the ease with which they can communicate with the members of their Program team. Being very accessible by phone and email when needed and providing answers to questions quickly are also much appreciated, as well as follow-ups and the occasional unsolicited phone check-up to find out how they are doing. Knowing they can reach someone 24/7 is also much appreciated.

Regarding the Centre itself, many stated that having a comprehensive care centre for bleeding disorders with the specialized programs and services offered all in one

location, including good coagulation labs, is a major asset. The coordination by medical personnel to meet all specialties in one appointment instead of having to return several times for separate visits is valued. Having a Centre in the city where they live or which is close by is also appreciated. Travelling clinics that provide service at a distance from the main Centre are also highly appreciated. People feel they can be seen quickly and efficiently. An IBDCCC provides access to most medical specialties. The information Program staff provide to other health care service professionals and the coordination of care beyond the Centre itself are seen as very valuable. The patient and their family members are well guided regarding the treatment and care of bleeding disorders.

The fact that factor concentrates are available 24/7 or can be sent quickly and good ordering / pick-up services are all highly appreciated. Up-to-date medical information and educational services are valued by many as well as the research activities that are conducted. Being able to be access services in English and French was also identified as important.

See **Annex 2** *for all comments.*

27. What do you think is most in need of improvement at your comprehensive care clinic?

    Fifty-eight (58) people indicated nothing needed to be improved or that they were very satisfied with everything.

    Seventeen (17) people indicated more personnel, including nurses, physiotherapists, physicians, social workers and psychologists, are needed. Of note, a few mentioned the need for greater access to nurse practitioners.

    Sixteen (16) people identified long wait times as an issue and suggested that scheduling of appointments needs to be improved to make better use of time both for the clinic staff and for the patients. People request less wait time between meetings with clinic members. More flexibility with appointment dates and times was also suggested. It was requested that health care providers be punctual and stick to schedules.

    Fifteen (15) people raised issues about the physical resources such as the need for more beds/rooms, a more permanent location for the clinic, and larger and more adequate work spaces, including better waiting rooms. More and better up-to-date equipment and waiting rooms with toys and other things to keep the children busy and amused were mentioned. One person mentioned that the signage needs to be improved as the Centre is hard to find.

    Thirteen (13) people indicated parking as an area needing improvement. Some suggested instituting a special rate for patients and also having the CHS chapters help out patients with a special parking subsidy program.

    Ten (10) people indicated issues related to the fact that they live far from the clinic which involves long travel times and expenses. Some would like to see clinics closer to home or more outreach clinic activities/services and better training for their local Emergency Departments.

    Four (4) people indicated that weekend and after hours coverage should be in place.

    See **Annex 2** *for all comments.*

28. 4% (13/341) report having ever made a complaint.

29. 27% (3/11) report the complaint process was made clear to them.

30. 62% (5/8) report the complaints were handled to their satisfaction.

Section F: Canadian Hemophilia Society observations and recommendations

This section presents a summary of the observations and recommendations included in the individual centre assessments.

Human resources

Despite dedicated and well-trained staff, 23 of the 25 Programs lack human resources in one or more of the following core disciplines: hematology, nursing, physiotherapy, social work and clerical/data entry (key indicator 1-2). In nine of the 25 Programs, no resources are allocated to certain core disciplines, notably physiotherapy and social work. The level of resources varies widely and inexplicably from Program to Program: from 0.2 to 1.5 Full Time Equivalents (FTEs) per 100 hemophilia A and B patients in nursing; from 0 to 0.4 FTEs per 100 patients in physiotherapy and social work, and 0 to 1.8 FTEs in clerical/data entry. The centre assessment revealed that 52.2 FTEs are currently funded in the core disciplines of nursing, physiotherapy, social work and clerical/data entry across Canada. An additional 21.1 FTEs are considered necessary to help Program staff meet the current challenges of increasing numbers and respect Standards of Care. At an average of $70,000 per FTE, this represents a national investment of less than $1,500,000, or approximately one-half of one percent (0.5%) of the current total budget of $270,000,000 for the care of people with inherited bleeding disorders.

Many Programs are subject to increasing hospital, regional health authority or Ministry of Health pressure to limit or even reduce existing resources, despite a 75% increase in the number of patients registered in the 25 Programs in the period 2004-2014 and increasing demands on services. Only a few provinces have given provincial designation to their Programs for these rare inherited bleeding disorders (disease incidence of 1 in 10,000 to 1 in 1,000,000) requiring specialized and expert care. Budget restrictions at the Program level are viewed by the CHS as “penny-wise.”

Recommendation 1: That health officials in the provinces and territories consider the results of the individual Centre assessment reports and adjust human and physical resources in IBDCCCs so that Standards of Care can be respected and optimal care provided. Of paramount importance are the core team disciplines: physiotherapy, social work, administration/data entry, pediatric and adult physician, nursing.

A number of Programs indicated a lack of availability of extended team members, as prescribed in the Standards of Care (key indicator 1-7a). Specific disciplines identified include dentistry, obstetrics-gynaecology, rheumatology, orthopedics and pain management.

Recommendation 2: That Centre Directors strive to establish effective relationships with extended team members (where they do not already exist) so that care can be truly comprehensive. Where Centres are small or where expertise is unavailable, a referral network should be established with experts in neighbouring Programs.

Data collection

Hemophilia care is changing quickly with the advent of a new generation of factor concentrates that will require long-term research to measure their impact on health outcomes. Provincial and territorial governments are requiring increased monitoring of the utilization of blood products and their recombinant alternatives. Sufficient human resources for data collection and entry are critical. Research and involvement in national databases are

identified as two of the 11 responsibilities of a HTC in the Standards document (key indicators 1-8a, 1-8b, 1-8c, 2-2b). Twenty-one of the Programs reported issues with the collection and analysis of patient data as a key barrier to optimal health care delivery.

To facilitate infusion reporting, data collection and health outcome research, the Association of Hemophilia Clinic Directors of Canada and McMaster University are developing the Canadian Bleeding Disorder Registry (CBDR) to replace the outdated Canadian Hemophilia Assessment and Resource Management System (CHARMS). This new integrated information system will be implemented at Canadian IBDCCCs starting in 2015, which will result in improved data collection for individual patient care, clinical research and utilization monitoring. This information system has the potential to significantly improve home infusion reporting, data collection and health monitoring by Centre staff, and health outcome research across the country and even internationally. It will attain these goals, however, only if the CBDR is adequately funded in the medium to long term. The cost is estimated at $300,000-$400,000 per year to house, maintain and develop the registry, and an additional $300,000 per year for badly needed data entry resources in the Centres. This represents approximately one-quarter of one percent of the cost of coagulation therapies.

Recommendation 3: That the provinces and territories support the medium and long-term operational costs of the Canadian Bleeding Disorder Registry so that utilization of coagulation therapies can be effectively monitored, individual patient care improved and overall health outcomes evaluated.

Recommendation 4: That hospitals, regional health authorities and Ministries of Health (as appropriate) provide sufficient resources in data entry/management so that the Canadian Bleeding Disorder registry can be used to its full potential.

Clinics and outpatient services

Because of lack of clinic time or space, nine of the 25 Programs are unable to respect the schedule for regular health assessments as prescribed in the Standards of Care. In at least six Centres, clinic space is inadequate to allow patients to be seen by the multi-disciplinary teams that make up comprehensive care centres. It is critical that patients are able to access the full comprehensive care team both during regularly scheduled appointments and during walk-in visits for acute bleeding problems (key indicators 3-2a, 3-3). Several centres are unable to set aside time and space for dedicated clinics for girls and women with inherited bleeding disorders (key indicator 3-2a). Both health care providers and patients expressed a strong appreciation for travelling clinics.

Recommendation 5: That hospitals ensure that there is adequate time and space for interdisciplinary clinics so that patients can be assessed as per the schedules defined in the Standards of Care.

Recommendation 6: That travelling clinics be continued and, where numbers warrant, expanded.

Recommendation 7: That where patients live in remote communities, the provincial governments provide travel assistance to allow them to attend annual or semi-annual clinic assessments.

Physical resources

New techniques in point of care ultrasound imaging to evaluate specific aspects of acute and chronic joint health are being introduced in some Canadian IBDCCCs. As this bedside equipment becomes incorporated into joint health assessment, personnel need to be trained and skills maintained to facilitate its use (key indicator 3-1). Used appropriately, this technology has the potential to identify bleeds that require treatment and avoid treatment when no bleed is occurring, as well as monitor long-term joint health and facilitate educational discussions between the team and patient at the bedside. Eleven of the Programs expressed a keen interest in adopting this technology.

Recommendation 8: That ultrasound diagnostic imaging be introduced at the bedside in Canadian IBDCCCs as this equipment becomes accepted as standard of care, and personnel be trained in its use.

Several Programs have started to use SmartPhones to take advantage of increasingly popular ways of communicating, including texting. This can decrease appointment no-shows, and increase adherence to treatment protocols and factor concentrate utilization reporting.

Recommendation 9: That Programs be provided with Smartphones to take advantage of modern ways of communicating, and that privacy regulations be developed in hospitals/health authorities to facilitate this style of communication.

Policy

The very success of the Programs—a home care model of treatment for even the most severely affected patients—results in a low number of in-person visits to the Centres and to the Emergency Departments, and hence lower visibility. The role of the teams in providing support to patients and remote health care providers needs to be recognized and supported (Scope of Care Standard 3, key indicator 1-3c). In many cases, directions to the Centres need to be improved, both on site and on the hospital websites.

Recommendation 10: That Programs make efforts to be more visible to hospital administrations, Ministries of Health, outside agencies and patients.

Inherited Bleeding Disorder Comprehensive Care Programs serve patients across large geographies and multiple health regions. Some individual Programs serve entire provinces. Dedicated provincial funding from Ministries of Health, similar in design to that of the Saskatchewan Bleeding Disorders Program, should be provided to Programs.

Recommendation 11: That Inherited Bleeding Disorder Comprehensive Care Programs be recognized by Ministries of Health as having provincial mandates and that provincial or designated envelope funding be provided (where this is not already the case).

Recommendation 12: That Provincial Inherited Bleeding Disorder Councils, made up of health officials, representatives from the province's IBDCCCs and the patient organization, be created to advise the Ministry of Health.

Coagulation therapies

The cost of coagulation therapies (factors I, II, VII, VIII, IX, XI, XIII, VIII/VWF and factor VIII bypassing therapies) in 2013-14 was $204,000,000 for Canadian Blood Services and $46,000,000 for Héma-Québec for a total of $250,000,000, despite a significant decrease in the per unit cost of recombinant factor VIII—factor VIII represents close to 50% of the total

coagulaton product budget —compared to previous years. Prescription of these biological products requires the expertise of hematologists experienced in the care of people with inherited bleeding disorders, expertise and experience that resides only in physicians working in IBDCCCs. Approximately 90% of these products are infused by patients or their caregivers via home infusion programs to prevent bleeding, a Standard of Care for many patients in Canada and around the world for the last several decades. Many Programs, however, describe a lack of capacity to adequately monitor the optimal utilization of these clotting factor concentrates used in home infusion protocols and their key clinical outcomes. Systems to report home infusions in accurate and timely ways are inadequate. Nurses are unable to respond quickly to critical care issues as identified by accurate and timely reporting of factor utilization. Under- and over-utilization occur. Sub-optimal health outcomes, including unnecessary bleeding and joint damage, are the results. Product wastage occurs in such an environment.

The cost of coagulation therapies, therefore, represents 90 to 95 percent of the total costs for the care of people with bleeding disorders. A silo approach to funding the care of people with inherited bleeding disorders is a barrier to rational resource allocation; hospitals are unaware of and perhaps unconcerned with the cost of factor concentrates reimbursed via the provincial blood budgets, and Ministries of Health have little control over hospitals' human resources. The CHS views the current under-funding of Programs, whose role is to facilitate the best health outcomes with such valuable coagulation therapies, as "pound-foolish."

Why is this a critical time to correct the problems?

A key indicator of the level of hemophilia care in a country is the number of International Units (IUs) of factor VIII consumed per capita. Canada currently uses 5.7 IUs per capita, 25-50% less factor VIII than developed countries such as United States, United Kingdom, Australia, Ireland and Sweden, in part due to historically conservative prophylactic protocols. In Canada over the last five years, utilization of factor VIII has been increasing at a rate of 6% per year. The recent 40-50% decrease in the per unit cost of factor VIII in Canada is very likely to be accompanied by an increase in utilization, as patients and physicians adopt prophylactic regimens that more effectively protect against bleeding and improve quality of life. At the same time, extended half-life factor VIII and IX concentrates, the first advance in the efficacy of factor therapies in four decades, are being introduced. The abilities to document utilization and evaluate health outcomes as clinical practices evolve are more critical than ever. This can only be done with adequate human resources in the Centres.

It is current practice in some provinces that clotting factor concentrates are made available by prescription only to patients who are registered in a Hemophilia Program (key indicator 1-4). In this way, physicians prescribing concentrates and treating patients outside the Centre can be put in touch with the Program specialists and benefit from the Centre's high level of knowledge and experience. Product utilization is optimized.

Recommendation 13: That IBDCCCs be given a provincial mandate for the prescription of clotting factor concentrates such that, except in emergency, they can only be prescribed to patients registered in a Program; and that outlying hospitals that distribute clotting factor concentrates be required to report distribution to the Program (where this is not already the case).

More education is needed regarding accurate and timely home infusion reporting, which represents approximately 90% of clotting factor concentrate usage. With the launch of the Canadian Bleeding Disorder Registry in 2015, the timing is ideal for such an initiative.

Recommendation 14: That the Canadian Hemophilia Society and the Canadian Association of Nurses in Hemophilia Care develop education and awareness tools to improve home infusion reporting.

Tranexamic acid and desmopressin are key drug therapies used in the treatment of inherited bleeding disorders and should be included in all provincial drug formularies. Where not available because of cost barriers, patients may turn to expensive clotting factor concentrates, which are provided at no cost through the blood budget. Increased access would improve patient care and conserve valuable clotting factor concentrates.

Recommendation 15: Add tranexamic acid and desmopressin to provincial drug formularies (where this is not already the case) and remove any funding barriers to patients who require these drugs.

Continuing education

It is critical that all core team members be able to benefit from attendance at the annual national meetings of their associations. Hemophilia and bleeding disorders are rare conditions; continuing education is feasible only through quality interactions with peers across Canada (key indicators 1-15a, 1-15b). This training should be supported and facilitated by hospitals. Thirteen of the 25 Programs indicated there are barriers to core team members accessing continuing education opportunities.

N.B. The cost of attending annual meetings of health care provider associations is borne by the Canadian Hemophilia Society.

Recommendation 16: That IBDCCCs and the hospitals that house them support professional training and continuing education for core team members for these rare conditions through their national associations, namely: Canadian Association of Nurses in Hemophilia Care, Canadian Physiotherapists in Hemophilia Care and Canadian Social Workers in Hemophilia Care.

Research

Hemophilia care is changing quickly with the advent of a new generation of factor concentrates that will require long-term research to measure their impact on health outcomes. Provincial and territorial governments are requiring increased monitoring of the utilization of blood products and their recombinant alternatives. A new integrated information system will be introduced into Canadian IBDCCCs in 2015, which will result in increased data collection, both for clinical research purposes and utilization monitoring. Research and involvement in national databases are identified as two of the 11 responsibilities of an IBDCCC in the Standards document.

Recommendation 17: That hospitals and Ministries of Health recognize that research is a key responsibility of an IBDCCC and, where expertise exists, provide support for clinical research.

Hemostasis lab

While the assessors did not have the capacity to evaluate the quality of services in the hemostasis laboratory, staff in several Centres report that key tests cannot be done in a timely manner. A well functioning hemostasis lab able to perform all basic tests 24/7 and correct diagnoses is critical to a well-functioning Centre (key indicator 1-13a).

Recommendation 18: That Centre directors, laboratory directors and hospital officials review the capacity of the hemostasis laboratory with the goal of being able to perform key tests in a timely manner.

Services

24/7 on-call systems for hematologists and nurse coordinators have proven their worth in providing expert advice on treatment to patients and families, avoiding Emergency Department visits where possible and facilitating timely and appropriate care in Emergency Departments (key indicator 1-6a, b, c).

Recommendation 19: That hospitals support the availability of a 24/7 on-call service using qualified staff.

Patients' principal criticism of the care they receive is related to the Emergency Department. As care has become more and more home-based over the last three decades, the number of visits to Emergency Departments has decreased and Emergency Department staff have become less accustomed to treating people with inherited bleeding disorders. Patients and caregivers report that care is sometimes sub-optimal and wait times are long, especially in Emergency Departments outside the hospitals that house the Programs (key indicators 1-6c, 3-4a, 3-4b).

Recommendation 20: That hospitals allocate a portion of the nurses' time to training Emergency Department personnel, including in outlying Emergency Departments, and that this training be mandated.

Telemedicine has the potential to help with remote assessment of acute bleeds and as an alternative to in-person clinic visits for patients at a great distance from the Centre or outreach clinics (key indicators 3-4a, 3-4b).

Recommendation 21: That Centre personnel explore greater use of Telemedicine to overcome barriers to care for people living remotely from Centres.

Summer camps, organized and funded by all ten chapters of the Canadian Hemophilia Society are where most children learn to do self-infusions. They are the ideal places for children to become engaged in and educated about their own care. It is critical Program nurses be present to supervise this learning; unfortunately, in some provinces, nurses must use vacation time to attend camp. Time devoted to learning self-infusion at camp will reduce the time needed for infusion training during busy clinic visits.

Recommendation 22: That hospitals recognize summer camps are fundamental to children with inherited bleeding disorders gaining control over their own health care, and allocate time for IBDCCC nurses to attend and educate children.

Transitioning from a pediatric to an adult care model is a critical time for patients with chronic disease. It is a time when patients with bleeding disorders can abandon preventative care, ignore symptoms of bleeding and experience avoidable complications. While transition is facilitated in those 15 Programs that have common staff and facilities for both pediatric and adult populations, it can be especially challenging when patients "graduate" to new teams in different hospitals.

Recommendation 23: That pediatric and adult comprehensive care teams continue to work together to improve the formal transition programs (key indicator 3-9a). Preparations for transition should begin at the age of 10 or 12.

The Canadian Hemophilia Society

Some families have financial barriers to accessing optimal care. The provincial chapters of the Canadian Hemophilia Society should work with Program personnel to facilitate providing financial assistance to families in need to attend clinic and summer camp, and for specialized equipment such as helmets and mobility aids.

Recommendation 24: That a formal procedure for financial support for families (e.g. mobility aids, funds for travel to clinic and parking) be jointly developed between chapters and comprehensive care teams.

Closer ties and more frequent exchanges between the provincial chapters of the Canadian Hemophilia Society and the IBDCCC care team would be beneficial to discuss and improve services.

Recommendation 25: That provincial chapters of the Canadian Hemophilia Society and the IBDCCC care teams should set up formal procedures for regular meetings to discuss and improve services (where this does not already exist).

References

(1) Analysis of the cost of care for inherited bleeding disorders at St. Paul's Hospital, Vancouver. This analysis included non-labour costs, diagnostic and clinical support services, salaries, office space and the cost of Emergency Department services compared to factor VIII and IX usage over a five-year period. Factor VIII and IX costs alone were found to represent 93% of the total cost of care. The actual percentage of costs related to factor concentrate use is likely to be higher than 93% as bypassing agents (rFVIIa and FEIBA), VWF concentrates and concentrates to treat Factor I, II, VII, XI and XIII were not included in the St. Paul's costing. Extrapolation of the costs observed at St. Paul's (an annual average of $805,000 for 432 patients) to the rest of Canada yields an estimated cost of $19,953,000 for the more than 10,000 patients registered.

(2) Canadian Hemophilia Registry; Data summaries; **www.fhs.mcmaster.ca/chr/data.html**

(3) Canadian Blood Services 2013-14 Annual Report, p. 41

(4) Personal communication, Jean Lapierre, Directeur, produits stables et services logistiques, Héma-Québec

(5) Communications from CBS report savings of $160,000,000 for factor VIII alone in the years 2013-14, 2014-15 and 2015-16, compared to 2012-13.

(6) First analysis of 10-year trend in national factor concentrates usage, Canadian Hemophilia Assessment and Resource Management System, Dr Aïcha Traoré

(7) WFH Global Survey 2012; Map of Factor VIII Use per Capita; **www1.wfh.org/GlobalSurvey/Public_AGS/AGS_Factors_UIPerCapita_EN_Map.aspx**

(8) Canadian Blood Services, Coagulation Products, 2014, p. 3

(9) Personal communication, Dr. Alfonso Iorio, Associate Professor, Department of Clinical Epidemiology and Biostatistics, McMaster University; Chair, CHARMS Committee, Association of Hemophilia Clinic Directors of Canada

(10) Canadian Comprehensive Care Standards: results of a self-assessment survey of the Standards by 24 Canadian hemophilia treatment centres; Canadian Hemophilia Standards Group Survey – May, 2010

(11) World Federation of Hemophilia Global Survey 2013; Canadian Hemophilia Society report for Canada, June 2014