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The Integrated Care model, also known as the Comprehensive Care model or the Hemophilia Treatment Centre (HTC) model

“This term refers to the continuous supervision of all medical and psychosocial aspects affecting the haemophilia patient and family, including supervision of home-based treatment (either prophylaxis or on-demand). It generally demands that all modalities of care – access to care providers, as well as diagnostic and therapeutic facilities – are available and delivered in a single specialized centre, by a team of health care providers. The composition of the integrated care team varies, but generally adheres to key components recommended by the World Federation of Haemophilia, including a medical director, a nurse coordinator, a physical therapist, a psychosocial expert (e.g. social worker) and a specialized coagulation laboratory.”

– NHF-McMaster Guideline on Care Models for Haemophilia Management

The comprehensive management of hemophilia began as “the Hemophilia Demonstration Project” at the Los Angeles Orthopedic Hospital in 1964. “The project represented a ‘first’ in hemophilic rehabilitation since it combined in one coordinated setting all the professional specialists required to diagnose and to treat the multiple problems manifested by this disease.”

It has since become the accepted model of hemophilia care throughout the world and is recommended by the World Federation of Hemophilia in its Guidelines for the Management of Hemophilia, 2nd Edition.

In Canada, the comprehensive care model was first promulgated in the conference entitled “Comprehensive Care for the Canadian Hemophiliac”, held in Winnipeg in May 1978, organized by the Canadian Hemophilia Society (CHS) in close collaboration with its own multidisciplinary Medical and Scientific Advisory Committee (MSAC).

[N.B. The balance of this document will be inclusive and refer to the entire family of inherited bleeding disorders rather than exclusively to hemophilia, to people with bleeding disorders [PwBD] and to treatment centres rather than Hemophilia Treatment Centres.]

The First Edition of the Canadian Comprehensive Care Standards for Hemophilia and Other Inherited Bleeding Disorders was published in June 2007. It has proven invaluable in guiding health care providers in treatment centres, hospital administrators, ministries of health and the patient association in the coordination and delivery of “integrated” or “comprehensive” care for PwBD.

Much, however, has changed since then, notably:

- a doubling in the number of diagnosed patients over 15 years to more than 10,000, largely through increased recognition of abnormal bleeding symptoms and improved diagnosis;
- an increase in co-morbidities due to an ageing patient population, necessitating enhanced collaboration with specialty services such as cardiology, oncology, nephrology and vascular surgery;
- the number and complexity of treatment regimens (e.g. immune tolerance induction to eradicate inhibitors, extended half-life factor concentrates, non-factor replacement therapies and gene therapy);
increasing recognition of the need to individualize treatment regimens;

the introduction of new clinical assessment tools such as point-of-care ultrasound and standardized hemophilia-specific outcome measures;

the development and adoption of improved tools to monitor and manage care, such as comprehensive registries, expanded pharmacokinetic capabilities, electronic patient files and tele-medicine;

an emphasis on increased participation in physical activities, physical therapy and an increase in orthopedic interventions;

increased health system requirements for accountability for utilization of health resources (e.g., treatments, human resources, laboratory testing);

an increasing obligation to collect real-world evidence on health outcomes and an expectation that treatment centres participate in registries and research;

an increased recognition of the impact of bleeding disorders in women, resulting in increased diagnosis, and necessitating improved collaboration with specialties such as obstetrics and anesthesiology to coordinate care plans;

an increased recognition of the complexity and impact of mild bleeding disorders;

an increased recognition of the level of pain experienced by many PwBD and the need to manage it;

an increased recognition of the psychosocial issues for both patients and caregivers related to being diagnosed and living with a chronic condition;

the introduction in Canada of frequent product tendering and wholesale treatment product switching for patients, imposing on treatment centres the obligation to review, consent and implement new treatments in hundreds of patients every two to three years;

an increasing population of new Canadians with bleeding disorders who arrive with joint damage and whose care requires cultural and linguistic sensitivity.

This updated version is needed to reflect current best practices.

The intended audience for these standards is:

- treatment centre team members
- health care administrators including:
  - department and program managers
  - institutional executives
  - regional and provincial health authorities
  - ministries of health
- PwBD and caregivers
- researchers
- Canadian Blood Services and Héma-Québec
- the Provincial/Territorial Blood Liaison Committee.
Multidisciplinary input
This version of the Standards was written by the Canadian Inherited Bleeding Disorder Standards Working Group (CIBDSWG). It includes:

- Association of Hemophilia Clinic Directors of Canada
  - Lawrence Jardine, (co-chair), Ontario
  - Natasha Pardy, Newfoundland and Labrador
- Canadian Association of Nurses in Hemophilia Care
  - Karen Sims, British Columbia
  - Marie-Hélène Thompson, Québec
- Canadian Hemophilia Society
  - Kathy Mulder (co-chair), Manitoba
  - David Page, Québec
  - Bojan Pirnat, Ontario
  - Wendy Quinn, Saskatchewan
  - Pamela Wilton, Ontario
- Canadian Physiotherapists in Hemophilia Care
  - JoAnn Nilson, Saskatchewan
- Canadian Social Workers in Hemophilia Care
  - Jennifer King, Saskatchewan
- Ontario Hemophilia Coordinator
  - Sarah Crymble, Ontario

These representatives consulted widely with the members of their respective organizations.

International standards
The CIBDSWG reviewed standards and guidelines produced by the World Federation of Hemophilia, the UK Haemophilia Alliance, the European Haemophilia Network and the U.S. National Hemophilia Foundation, (see Appendix 1, International standards for the care and treatment of inherited bleeding disorders) as well as Canadian standards from other conditions, for example, cystic fibrosis.

Literature review
Literature searches were conducted to identify research support for the standards proposed.

External review
External reviews of these standards were conducted by:

- Dr. Paul Giangrande, former medical vice-president of the World Federation of Hemophilia (2000-2004) and former centre director of the Oxford (U.K.) Haemophilia Centre;
- Dr. Carol Kasper, former medical vice-president of the World Federation of Hemophilia (1996-2000) and former medical director of the Orthopaedic Hemophilia Treatment Center in Los Angeles, California;
GOALS

These standards are designed to:

- ensure all Canadians with an inherited bleeding disorder, regardless of geographical location or sex at birth, are registered with treatment centres providing equitable, evidence-based and patient-centric care.

- ensure delivery of team-based integrated care.

- promote value-based health care using measurement tools to:
  - allow benchmarking of quality assurance indicators across Canada;
  - promote consistent measurement of patient-relevant health outcomes;
  - transparently share measured indicators and outcomes in order to generate improvements in care delivery;
  - reduce health-related risks such as joint disease, inhibitors and treatment-related complications that may result from non-integrated or outdated practices, through the comprehensive care model and open collaboration among treatment centres, hospitals, health authorities and governments.

- provide guidance to those responsible for coordinating the delivery of care to PwBD.

- ensure that local point-of-care health care providers can access and consult with experienced health care providers in those treatment centres.

- empower PwBD and caregivers to be engaged in their care and assist treatment centre personnel to equitably and consistently meet their expectations.

- ensure that health resources are used efficiently and effectively.

POPULATIONS SERVED

This document applies to bleeding disorder treatment centres who serve people, caregivers and families living with ...

- hemophilia A and B, including carriers;
- von Willebrand disease;
- inherited platelet function disorders;
- rare factor deficiencies;
- inhibitors to clotting factors;
- acquired coagulopathies.

If the services of a treatment centre are expanded to treat other congenital disorders associated with a bleeding tendency, additional resources will be required; these standards do not apply to these conditions.
These standards are based on the following principles:

- Treatment centres shall strive to register and oversee care to all PwBD living in the treatment centre's geographical region.

- Treatment centres shall respond to the complexity and rarity of inherited bleeding disorders, and the associated lifelong needs by delivering care through an integrated, multi- and inter-disciplinary chronic disease management model.
  - Treatment centres shall monitor for complications, including joint disease, psychosocial issues, inhibitors, adverse drug reactions and transfusion reactions.
  - Health promotion and disease prevention in the context of bleeding and its consequences shall be a priority.
  - Care shall be delivered in a way that aims to minimize the pain, incapacity, and physical and emotional disability that may result from inherited bleeding disorders and their complications.
  - A holistic approach to patient care shall be adopted, ensuring that all PwBD have access to a wide range of specialist services.
  - Care shall be provided equitably, regardless of sex at birth.

- Treatment centres shall provide care that is based on the best available evidence.

- Services shall be individualized and delivered such that PwBD and families are partners in care.
  - The risks and benefits of all relevant treatment options shall be presented and discussed.
  - Shared decision-making shall be promoted.
  - Home-based care shall be facilitated, where appropriate, and overseen on an ongoing basis.
  - Care shall be delivered/provided within the person’s own community, wherever achievable in a safe and suitable manner.

- Health care providers shall behave in a manner that is culturally and socially respectful.
  - Privacy and confidentiality shall be respected.

- Treatment centres shall participate regularly in quality assurance activities (e.g. contributing data to a national report with provincial/treatment centre breakdown that is then sent to program managers, health authorities, governments and patient associations).
A. ROLE OF TREATMENT CENTRES

The treatment centre shall ...

1. be supported by the host institution and health authority to achieve the obligations set out in this document, and together with health administration maintain/pursue support and recognition by the provincial Ministry of Health.

2. be affiliated with a health care institution and all core team members are encouraged to engage in relevant continuing education opportunities.

3. raise awareness in the local medical community to facilitate referrals and collaborate in care.

4. strive to register and provide care to all PwBD and their families in its respective region.

5. provide and/or facilitate access to the full range of expertise required to respond to the complex and diverse needs of PwBD, and to meet these standards. (See Appendix 2, Team members and roles: Core and extended.)

B. DIAGNOSIS

The treatment centre shall ...

1. have a triage process for new patient referrals.

2. have access to an accredited hemostasis laboratory with appropriate arrangements in place for 24/7 coagulation testing and establish formal links for specialized hemostasis testing when required. (See Appendix 6, Coagulation testing.)

3. have access to a diagnostic imaging department and establish formal links where specialized radiology services are required.

4. have a formal relationship with a genetic laboratory so that all patients and families have access to carrier testing and genetic services.

C. CARE

The treatment centre shall ...

1. provide care and treatment based on the most current clinical treatment guidelines. Where guidelines are not available, care shall be based on the most current scientific literature and best practices.

2. have access to staff with sufficient clinical skills to care for all PwBD in a manner that is appropriate to their age and condition.

3. provide and/or facilitate access to appropriate hematologic, obstetric and gynaecologic services to women with inherited bleeding disorders, including carriers of hemophilia.
develop, review and utilize policies and procedures for the treatment of non-urgent, urgent and emergency bleeding episodes.

5. have access to appropriate pharmacy, transfusion medicine and blood bank services.

6. make prophylaxis and home-based care available to PwBD as appropriate.

7. ensure that PwBD on home-based therapy have access to the required supply of factor concentrates and non-factor replacement therapies.

8. ensure that PwBD have access to infusion/injection equipment and supplies, either by supplying or facilitating procurement.

9. ensure 24/7 access to bleeding disorder expertise to PwBD through the centre, through partnerships, on call or consultative services.

10. coordinate treatment and follow-up for acute bleeds, including guidance regarding safe return to activities.

11. provide regular clinics for all PwBD, including women with bleeding disorders, to permit regular assessments on an appropriate schedule. (See Appendix 5, Recommended schedule for assessments.)

12. collect detailed health outcomes over time. (See Appendix 4, Health outcomes to be measured.)

13. maintain a record of patient/caregiver education.

14. coordinate care through transition (pediatric to adult, program to program, or province to province) to ensure continuity of care.

15. respect all professional, institutional and provincial guidelines regarding patient privacy and confidentiality.

D. PATIENT/CAREGIVER EDUCATION AND SUPPORT

The treatment centre shall ...

1. provide information to PwBD and their families about clinical phenotype and therapeutic options relevant to their diagnosis.

2. provide to PwBD/caregivers comprehensive information on treatment benefits and risks to make informed decisions.

3. provide information about potential risks and benefits of physical activities and sports.

4. provide access to genetic counselling delivered by an individual knowledgeable about inherited bleeding disorders.

5. provide, for PwBD/families on home-based care, comprehensive training about safe and responsible handling, storage, preparation and administration of treatment products, and safe disposal of used equipment and supplies.
6. facilitate education and learning about current issues related to bleeding disorders to PwBD and their families.

7. provide up-to-date documentation (e.g. FactorFirst wallet card) that identifies each person’s bleeding disorder, recommended treatment plan and treatment centre contact information.

8. inform PwBD and their caregivers whom to contact in the event of a bleeding episode.

9. provide psychosocial support of the PwBD and family regarding schooling, employment and relationships, and the provision of social supports and counselling services.

10. provide advice and support to ageing PwBD who require treatment for co-morbidities and to the health care providers who care for them.

E. PRODUCT MONITORING

The treatment centre shall …

1. maintain a list of all PwBD registered in the home therapy program.

2. establish formal links to Canadian Blood Services or Héma-Québec for product ordering.

3. be able to …
   a. track home inventory in real time
   b. enter home inventory in electronic reporting systems
   c. account for and report the use of all factor concentrates and non-factor replacement therapies distributed to their registered patients
   d. monitor expiry dates of all factor concentrates and non-factor replacement therapies distributed to their registered patients
   e. be able to recall products within 24 hours.

4. establish a mechanism whereby PwBD and their families record all bleeding episodes and treatments with clotting factor concentrates and non-factor replacement therapies so as to provide the treatment centre essential clinical data. As a condition of receiving home therapy, PwBD and their families shall be required to provide this information.

5. report adverse events that occur in association with the administration of coagulation factor concentrates or non-factor replacement therapies, as mandated by the regulator, manufacturer or other appropriate body.

F. HUMAN RESOURCES FOR INTEGRATED CARE

The treatment centre shall …

1. maintain a complete complement of core team members which is adequate to meet these standards. (See Appendix 2, Team members and roles: Core and extended.)
2. be staffed with core team members who have the appropriate training and qualifications to provide care to the patient population. (See Appendix 7, Standards of care for core disciplines in bleeding disorder care.)

3. have a process to monitor its ability to deliver care to the patient population and to request adjustments in human resources, when necessary.

4. provide a mechanism for team members to share knowledge with each other to promote best patient outcomes.

5. develop and maintain a referral list for extended team members and utilize their services as needed.

6. invite extended team members to team educational workshops and activities as appropriate.

7. demonstrate collaboration among all team members.

8. facilitate core team members being members of relevant organizations and/or working groups within the bleeding disorder community and taking part in relevant education and training activities.

9. facilitate access to reference materials for team members and students (e.g. AHCDC Clinical Practice Guidelines, journal articles and texts).

G. PHYSICAL RESOURCES

The treatment centre shall ...

1. have a clinical area sufficient for diagnosis and treatment that is age-appropriate, comfortable, quiet and adequately equipped, that respects privacy and confidentiality, and that is designed for people with disabilities or mobility aids.

2. be located in a facility that is linked with a day medicine department and an emergency department so that PwBD can obtain treatment 24/7.

H. INFORMATION SYSTEMS, HEALTH RECORDS, DATA COLLECTION

The treatment centre shall ...

1. follow provincial and institutional policies for maintaining records and assign a confidential identifier to each patient.

2. document each patient’s treatment plan, as appropriate, and review it annually.

3. be registered with a bleeding disorder registry database (e.g. CBDR, iCHIP) and ensure that all core team members have access to the information systems.

4. keep data current and routinely export data, as required, to the provincial and national databases.

5. collect detailed information concerning the outcomes of treatment. (See Appendix 4, Health outcomes to be measured.)
I. LINKAGES

The treatment centre shall ...

1. provide information on inherited bleeding disorders, contact information in case of emergency and treatment recommendations to emergency departments and primary care providers.

2. provide contact information and emergency treatment recommendations to the emergency department nearest to the PwBD's home.

3. provide education and treatment recommendations to other community professionals who provide services to PwBD (e.g. surgery, dentistry).

4. establish a process for referring PwBD to services not provided within the program.

5. have a process in place to meet and discuss issues of mutual concern with the Canadian Hemophilia Society and its provincial chapters.

6. maintain current contact information for the treatment centre in listings with the Canadian Hemophilia Society, the World Federation of Hemophilia and parent hospital.

J. ACCREDITATION, AUDIT, QUALITY ASSURANCE, RESEARCH

The treatment centre shall ...

1. participate in hospital or peer evaluation and respond to critical appraisal.

2. participate in a formal accreditation and evaluation process, when established.

3. participate in research activities relevant to patients with bleeding disorders, where patient numbers and clinic resources permit such participation. Where this is not possible, treatment centres should endeavour to inform interested patients about opportunities to participate in research studies conducted in other treatment centres. (See Appendix 3, point 5.)

4. make known the institution's process to allow PwBD and families to communicate concerns, complaints and appreciation.

5. have adequate physical and human resources to meet these standards.
APPENDIX 1
INTERNATIONAL STANDARDS FOR THE CARE AND TREATMENT OF INHERITED BLEEDING DISORDERS

- U.K. Haemophilia Alliance: A National Service Specification for Haemophilia and Other Inherited Bleeding Disorders, 2006
- U.K. NHS Standard Contract for Haemophilia, 2013/14
- U.K. Standards of Care Service Provision of Physiotherapy for Children with Haemophilia & other Inherited Bleeding Disorders, 2017
- U.K. Standards of Care Service Provision of Physiotherapy for Adults with Haemophilia & other Inherited Bleeding Disorders, 2017
- U.S. National Hemophilia Foundation Standards and Criteria for the Care of Persons with Congenital Bleeding Disorders, 2002
Core team
In order to provide integrated care, the following disciplines are required in each treatment centre:

**Medical director** (an adult and/or pediatric hematologist or internist)
Provides subspecialty bleeding disorder medical expertise, supervises medical care and takes leadership in the treatment centre.

**Nurse** (RN or NP)
Provides expert clinical nursing care for PwBD, practices/acts within the domains of education, administration, research and policy to improve care delivery, may be the primary point of entry to facilitate access to care.

**Physiotherapist**
Monitors growth and development, offers a range of assessments and management for PwBD with acute and chronic conditions resulting from musculoskeletal bleeds, is responsible for monitoring joint function on a long-term basis, addresses prevention of bleeds and joint damage, promotes physical activity and coordinates rehabilitation.

**Social worker**
Provides support through psychosocial assessments, counselling/psychotherapy, advocacy, patient, family and community education, resources, supports and discharge planning.

**Coagulation laboratory staff**
Perform diagnostic testing, and accurate and precise coagulation assays and inhibitor testing.

**Blood bank staff**
Prepare, store and distribute coagulation products.

**Administrative assistant/data manager**
Supports the proper functioning of the treatment centre and collates clinical and financial data.

Extended team
In order to provide integrated care, the following disciplines should be available when they are needed for individual patients:

**Primary care provider**
Provides primary health care.

**Orthopedic surgeon**
Monitors joint and muscle problems and identifies the need for surgical intervention.

**Anaesthetist**
Provides anaesthesia and infuses coagulation products during surgery, and provides pain management post-surgery.

**Rheumatologist**
Provides diagnosis (detection) and treatment of musculoskeletal disease.
**Physical medicine specialist**
Provides care to enhance and restore functional ability and quality of life to people with physical impairments or disabilities.

**Hepatologist or gastroenterologist**
Monitors and treats the complications from HCV infection.

**Infectious disease specialist**
Monitors and treats the complications of transmissible and blood-borne infections such as HIV.

**Gynaecologist/obstetrician**
Manages the care for women with bleeding disorders: antenatal care for carriers, the delivery of a child who has a bleeding disorder, and other gynaecological issues.

**Geneticist**
Coordinates genetic testing and mutational analysis to enable confirmation of diagnosis, determination of carrier status and antenatal fetal testing.

**Genetic counsellor**
Supports PwBD and families regarding transmission, conception and family planning.

**Dentist**
Provides specialized dental services and advises community dentists.

**Expert in pain management**
Supports PwBD who need specialized pain management programs.

**Psychiatrist/psychologist**
Provides specialized psychosocial support and treatment.

**Child-life specialist**
Supports families in dealing with the challenges of a chronic disease in a young child.

**Athletic therapist/kinesiologist/sports medicine specialist**
Guide the PwBD/caregiver in activities that safely promote fitness.

**Orthotist**
Evaluates, designs, fabricates, fits and delivers supportive braces or orthotic devices to help patients improve function and achieve independence, safety and comfort.

**Geriatrician**
Provides expertise and care in health challenges due to ageing.

**Occupational therapist**
Promotes independence, meaningful occupations, and the PwBD’s functional ability to achieve daily routines/roles of choice.
Insufficient staffing levels may be apparent at an operational level if there is:
- inability to meet the recommended schedule for assessments for all persons with bleeding disorders
- above national average rate of factor wastage
- below national average rate of patients entering infusion records in their local database (MyCBDR/iCHIP)
- incomplete CBDR/iCHIP data sets
- no program designated to women with bleeding disorders
- poor attendance rates at clinics because patients do not perceive value in attending.

Insufficient staffing WILL result in the following adverse health outcomes:
- above national average annual bleed rate
- above national average loss of time from work/school due to bleeds
- above national average number of severe or life/limb threatening bleeds
- above national average of bleeds requiring hospitalization
- higher than normal percentage of patients regularly using prescription narcotics
- above average number of patients using or self medicating with non-prescription agents, including alcohol and/or cannabis
- above national average rate of bleeding complications post planned/elective procedures
- higher mortality rate than average.
It is recommended that the following factors be taken into account when establishing optimal staffing levels:

1. Geography – In provinces with one designated treatment centre, or where PwBD may live a considerable distance from the centre, there will be a need for outreach clinics and the availability of Telehealth.

2. Number of PwBD

3. Case mix of PwBD:
   a. Number of rare bleeding disorders
   b. The proportion of pediatric vs. adults vs. seniors: availability of appropriate facilities/expertise available for all
      i. Need for transition care
   c. The number of newly diagnosed
   d. The number on home infusion programs
   e. The number with inhibitors
   f. The number with co-morbidities including arthropathy, HIV, HCV infection, ageing
   g. The number with chronic pain issues
   h. The total number of procedures per year which will require additional coordination of external consultations, infusions, coagulation testing, pain management, rehabilitation, home care …
      i. The proportion of new Canadians: language and cultural issues, pre-existing multiple arthropathies
   j. The number with venous access issues, central venous access devices
   k. Education levels, literacy levels and active participation in their own care

4. Other:
   a. Changes in treatment products
   b. Social determinants of health

5. Research
   a. It is expected that research will be undertaken by all programs, but cannot be accommodated at the expense of clinical care. Specific external research funding is required.
The focus must shift from the volume and profitability of services provided—physician visits, hospitalizations, procedures, and tests—to the patient outcomes achieved. In order to do so, patient specific outcomes that have value to patients must be measured. Value is defined as the health outcomes achieved that matter to patients relative to the cost of achieving those outcomes.

**Patient health outcomes**
- Total number of annual bleeds
- Total number of severe bleeds (defined as life- or limb-threatening, requiring transfusion or requiring hospitalization)
- Inhibitor status
- Immune status (Hep C/HIV status)
- Bleeding outcomes following planned procedures
- MSK assessment (measure of impairment, activities, participation)
- Mental health
- Pain assessment
- Socio-economic assessment (employment vs. unemployment vs. disability)
- Time missed from school or work

**Service delivery outcomes**
- Date of clinic review
- Date of updated treatment plan
- Total units of clotting factor concentrate and non factor therapy used by each patient per year
- Total number of treatments received by each patient per year
- Amount of coagulation factor concentrate given for prophylaxis, on demand, surgery, immune tolerance induction
- Pharmacokinetic testing completion
- Number of inpatient days due to bleeds
- Time to diagnosis, first visit or initial treatment
Biologic outcome indicators

Recovery
Factor therapy half-life
Factor trough levels
Inhibitor titre
DDAVP response (resting)
Number of joints with impairments of structure or function
For patients with hemophilia, the World Federation of Hemophilia recommends a multidisciplinary check-up including hematologic, musculoskeletal, and quality-of-life assessments by the core comprehensive care team members at least yearly and every six months for children. These standards call for the same schedule of assessments for those people with other inherited bleeding disorders. Individuals with no bleeds, no recent treatment changes and no musculoskeletal or psychosocial concerns may be reviewed in person less frequently as deemed to be appropriate by the comprehensive care team.
List of recommended coagulation tests to be provided by Canadian bleeding disorder treatment centres and their associated laboratories:

1. Lab tests requiring timely turnaround time for patient monitoring
   a. Routine testing may not require results within 24 hours.
   b. BDTC-associated labs should have staff trained in special coagulation tests available 24/7, either in-house or on call, for urgent circumstances necessitating rapid turnaround for treatment decisions.
   c. Where patients of a BDTC are using extended half-life products and/or non-factor therapies, the BDTC-associated lab should possess capacity to appropriately monitor patients locally.

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<th>Locally accessible</th>
<th>24 hour service</th>
<th>Locally accessible or accessible through link with specialized lab</th>
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<td>ABO blood type</td>
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<td>PT, aPTT, thrombin time, fibrinogen</td>
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<td>Mixing studies</td>
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<td>Factor VIII and IX one-stage assays</td>
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<td>FVIII and FIX inhibitor assay</td>
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<td>Anti-porcine rFVIII inhibitor assay</td>
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<td>vWF antigen and activity assay</td>
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<td>Factor II, V, VII, X, XI and XIII activity assays</td>
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<td>Inhibitor assays for rare coagulation factor deficiencies</td>
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2. Lab tests used for diagnostic purposes (rapid results not required)

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<td>vWF Multimers</td>
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<td>Yes (If not available locally, process in place allowing patient to travel to an appropriate lab for required diagnostic testing)</td>
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<td>Platelet electron microscopy and flow cytometry</td>
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Canadian Association of Nurses in Hemophilia Care: Comprehensive Care Standards

Canadian Social Workers in Hemophilia Care: Standards of Practice

Standards of Physiotherapy Care for Persons with Bleeding Disorders, 2018 - Canadian Physiotherapists in Hemophilia Care

N.B. These standards may be password-protected and accessible only to members of the respective associations.
**Carrier** – The term “carrier” is used to describe a female of any age who is heterozygous for the gene for hemophilia A or B. It is generally used in the context of genetic testing and counselling. It is not intended to infer the presence or absence of an abnormal bleeding tendency. In a clinical context, females should be identified based on their factor VIII and IX levels, standardized bleeding scores and their bleeding phenotype, as is done for males.

**CBDR, iCHIP** – The Canadian Bleeding Disorders Registry and the Inherited Coagulopathy and Hemoglobinopathy Information Portal (BC) are information systems used by treatment centres to track product inventory and utilization, record patient outcomes, manage care and collect data for research.

**Clotting factor concentrates** – Plasma-derived and recombinant factors I, II, VII, VIIa, VIII, IX, X, XI, XIII and von Willebrand factor, used to treat many but not all bleeding disorders.

**Comprehensive care** – The continuous supervision of all medical and biopsychosocial aspects affecting the bleeding disorder patient and family, including supervision of home-based treatment (either prophylaxis to prevent bleeding or on-demand to control bleeding). It generally demands that all modalities of care – access to care providers, as well as diagnostic and therapeutic facilities – are delivered by a team of health care providers working in an interprofessional manner.

**Home-based care** – The regular prescribed administration of clotting factor concentrates, non-factor therapies and other coagulation products by the patient or caregiver outside of the hospital setting. This includes monitoring and reporting of outcomes.

**Immune tolerance induction** – The repeated and frequent administration of clotting factor concentrate to downregulate an already established antibody response to clotting proteins in order to eradicate the inhibitor.

**Non-factor therapies** – Therapies such as monoclonal antibodies (e.g. emicizumab and anti-tissue factor pathway inhibitors) and RNA interference technology (e.g. fitusiran) to treat bleeding disorders without factor replacement.

**Pharmacokinetic testing** – The process of collecting serial timed laboratory testing that allows personalization of dose and frequency of clotting factor infusions based on individual recovery, clearance and half-life.

**Prophylaxis** – The regular administration of clotting factor concentrates or non-factor therapies to prevent bleeding.

**PwBD** – People with bleeding disorders, both male and female, who are the focus of these standards.
Association of Hemophilia Clinic Directors of Canada, www.ahcdc.ca

Canadian Bleeding Disorders Registry, www.ahcdc.ca/cbdr

Canadian Hemophilia Registry, Data summaries, www.fhs.mcmaster.ca/chr

Canadian Hemophilia Society, www.hemophilia.ca


NHF-McMaster Guideline on Care Models for Haemophilia Management. Pai M et l. Haemophilia (2016), 22 (Suppl. 3), 6–16

MyCBDR, www.mycbdr.ca/MyCBDR/Account/Login?ReturnUrl=%2fMyCBDR%2fAccount%2fLogin%3freturnUrl%3d%252FMyCBDR%252F


National benchmarking of paediatric haemophilia treatment centres, Research and Practice in Thrombosis and Haemostasis. Conference: 27th Congress of the International Society on Thrombosis and Haemostasis. Australia. 3 (Supplement 1) (pp 890), 2019. Date of Publication: July 2019


The Inherited Coagulopathy and Hemoglobinopathy Information Portal (BC Provincial Blood Coordinating Office), www.ichip.ca


