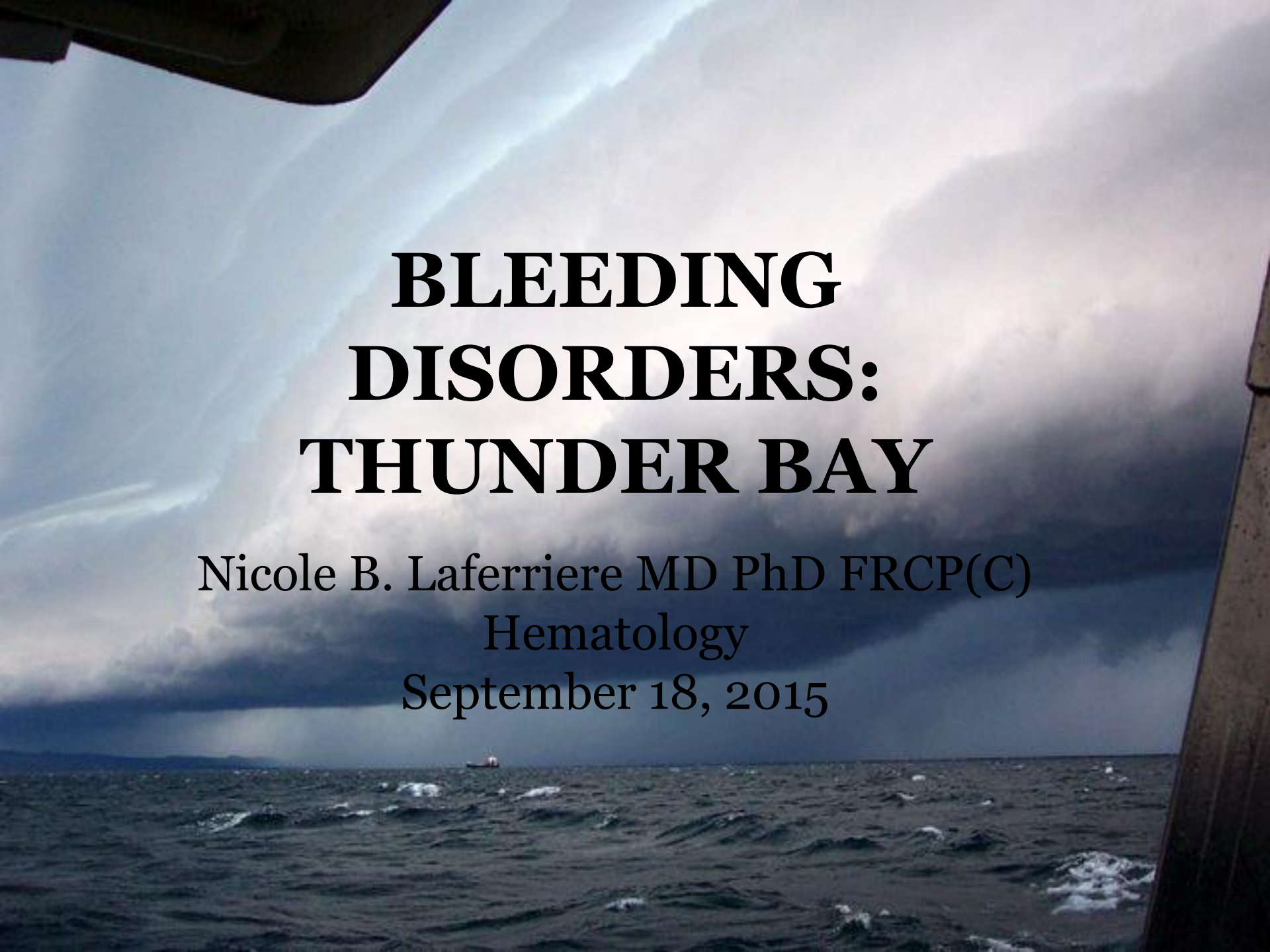
Disclosures

- Honoraria: Roche, Celgene, Amgen, Lundbeck, Novartis



BLEEDING DISORDERS: THUNDER BAY

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Hematology
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Diagnosis vWF Disease

- vWF disease reported to be the most common inherited bleeding disorder
- vWF deficiency or defects of vWF and increased mucocutaneous bleeding.
- Diagnosis remains a challenge for Hematologists and laboratories

Bleeding History

- Bleeding Assessment Tests (BATs)
- Heavy/prolonged bleeding following surgical procedures (tonsillectomy, wisdom teeth)
- Easy bruising/palpable purpura
- Spontaneous epistaxis requiring more than 10 min to stop or ER visit
- Prolonged bleeding following dental extractions
- Packed red blood cell transfusion requirement
- Heavy menses from menarche with clots > 1 inch in diameter/changing pad or tampon hourly.
- Medications: ASA, NSAIDS, Anticoagulants. Naturopathic/homeopathic medications

Laboratory Testing for vWD

- vWF Antigen
- vWF Activity
- Factor VIII
- vWF Multimers
- PFA 100 testing
- ABO type

May require repeated testing to confirm a diagnosis

Many Factors Affect Baseline vWF levels

- Pregnancy/HRT/Oral contraceptives
- Breastfeeding
- Menstrual cycle variation
- Infection/inflammation/physiologic stress
- Exercise
- Blood Group O has a 30% lower normal range
- Levels tend to increase with age, approximately 1% per year

Diagnosis vWF Disease

- Significant impact on quality of life and medical care
- Important to understand challenges of diagnosis
- Once diagnosis is made: how to treat
- Imperfect tools to quantify bleeding history
- Fluctuating lab values/testing peril makes it challenging to accurately assess baseline vWF levels

Low vWF Levels

- >50% patient with mild decrease vWF levels (30-50 IU/dL) are asymptomatic or have minimal bleeding symptoms
- Presence of vWF levels Bw 30-50 IU/dL does not automatically confer a diagnosis of vWF disease.
- Individuals with blood group O have a 25-30% lower levels vWF

- Easy bruising reported in 65% of the population
- Possible diagnosis Type 1 vWD may not have vWD but mildly low vWF levels and independent mild increased bruising
- Without a significant bleeding history, mildly decreased vWF does not predict significant bleeding.


EDMUND FITZGERALD

Laboratory Testing Bleeding Disorders NWO

- Thunder Bay experience
- Large number of patients with vWD and mild Hemophilia A followed in Hematology Clinic
- On review many had no history abnormal bleeding
- Reflex ordering based on local practice

- Contact with the TBRHSC Clinical Laboratory Hematology Coordinator
- ?What is going on

Investigation:

- Clinical Indication for testing
- Collections protocol
- Transportation of samples
- Procedures

Discoveries

- TBRHSC was shipping properly frozen specimens and samples on dry ice
- Specimens sat thawing on accession bench at private lab's distribution center
- Community labs did not freeze samples, which sat at room temp all day

Actions

- Troubleshooting specimen condition/transportation issues resulted in systematic recall of >100 patients with a prior diagnosis of vWD.
- Systematic re-testing, with samples handled appropriately and bleeding histories documented.
- Fewer than 10 per cent of patients actually found to have a significant vWD.

Diagnosis Bleeding Disorders

- Significant impact on lifestyle and medical treatment
- Need for accurate diagnosis and education
- Limitations due to inability to inaccurately assess bleeding

Diagnosis Bleeding Disorders

- Validated Bleeding disorders screening questions.
- Test only those with a high pre-test probability
- Accurate testing
- Refer to a Hemophilia Clinic Treatment Center

FactorFirst

EMERGENCY MANAGEMENT OF

HEMOPHILIA

and

VON WILLEBRAND DISEASE

Hemophilia Treatment Centres (HTCs)

The first **HTC** opened in Canada in Montreal in 1969. Today there are 24 **HTC**s across Canada staffed by a comprehensive care team comprised of a hematologist, nurse coordinator, physiotherapist and social worker responsible for:

- Medical management and care of people with bleeding disorders
- Advice re pre-op and dental care
- Teaching families and individuals about home infusion
- Providing education to hospital staff, physicians, schools/daycares
- Distribution and monitoring of the usage of clotting factor concentrates

Home Infusion

- Historically, treatment for bleeding episodes was only available from a medical professional/hospital.

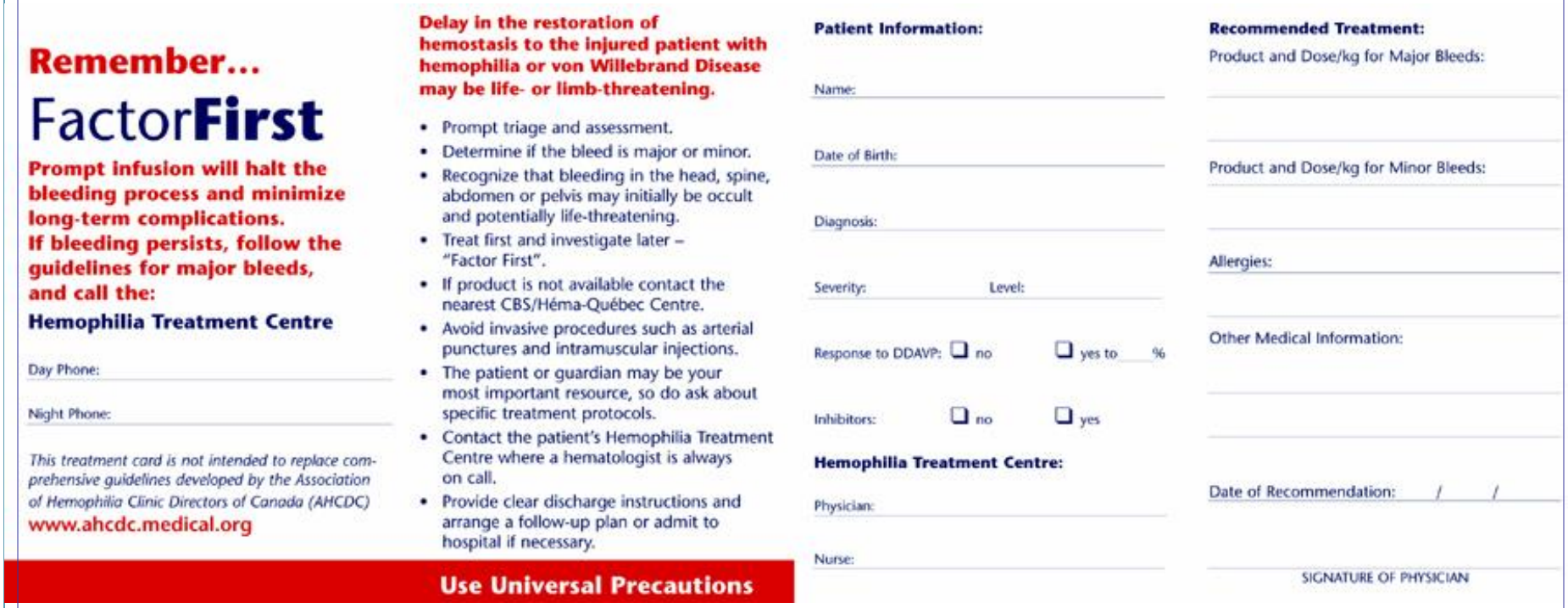
- The concept of home care began in California in 1962 and in Canada in the late 1960s.

- Home infusion is the IV administration of clotting factor in the home setting, by a trained parent or the person with hemophilia himself. This program allows for more rapid treatment of bleeds thus decreasing the amount of permanent joint damage.

- Many individuals with severe hemophilia follow a regime of prophylaxis whereby they receive factor replacement 2-3 times per week to prevent bleeding episodes and the crippling effects of permanent joint damage.

- Not all patients are on home infusion. Those not needing frequent treatment are less able to maintain the required skills for safe and effective home management of bleeds.

Patient Treatment Card



Remember...

FactorFirst

Prompt infusion will halt the bleeding process and minimize long-term complications.
If bleeding persists, follow the guidelines for major bleeds, and call the:

Hemophilia Treatment Centre

Day Phone:

Night Phone:

This treatment card is not intended to replace comprehensive guidelines developed by the Association of Hemophilia Clinic Directors of Canada (AHCDC)

www.ahcdc.medical.org

Delay in the restoration of hemostasis to the injured patient with hemophilia or von Willebrand Disease may be life- or limb-threatening.

- Prompt triage and assessment.
- Determine if the bleed is major or minor.
- Recognize that bleeding in the head, spine, abdomen or pelvis may initially be occult and potentially life-threatening.
- Treat first and investigate later – "Factor First".
- If product is not available contact the nearest CBS/Héma-Québec Centre.
- Avoid invasive procedures such as arterial punctures and intramuscular injections.
- The patient or guardian may be your most important resource, so do ask about specific treatment protocols.
- Contact the patient's Hemophilia Treatment Centre where a hematologist is always on call.
- Provide clear discharge instructions and arrange a follow-up plan or admit to hospital if necessary.

Use Universal Precautions

Patient Information:

Name:

Date of Birth:

Diagnosis:

Severity: Level:

Response to DDAVP: ❑ no ❑ yes to %

Inhibitors: ❑ no ❑ yes

Hemophilia Treatment Centre:

Physician:

Nurse:

Recommended Treatment:

Product and Dose/kg for Major Bleeds:

Product and Dose/kg for Minor Bleeds:

Allergies:

Other Medical Information:

Date of Recommendation: / /

SIGNATURE OF PHYSICIAN

Care Team Roles

Emergency Hemophilia & VWD Care Providers

- Triage all bleeds as urgent.
- Refer to available resources (Factor**First** Treatment Card and poster, Emergency Care Manuals, www.hemophiliaemergencycare.com).
- Contact HTC hematologist on call for clarification of treatment.
- Coordinate follow-up with HTC.

HTC Providers

- Interdisciplinary resource for comprehensive hemophilia & VWD care
- Information sharing on individual hemophilia & VWD patient care plans

Hemophilia and VWD Families

- Maintain regular contact with HTC and be informed about their condition.
- Carry bleeding disorder related information (MedicAlert & Factor**First** card).
- Report to ER for assessment of bleeding episodes when required.
- Follow-up with HTC after ER visit.

Hematology Northwestern Ontario

- 3 Royal College Hematologists
- Association for Hematology Clinic Directors of Canada AHCDC
- Daily coverage Hematology Consultation TBRHSC and Regional partners

Hematology Services Northwestern Ontario Regional Cancer Centre

Benign Hematology Subgroup Committee

Advocacy for the complex knowledge required to diagnose and manage Hematologic disease

Investigation of

- Anemia
- ITP/AIHA
- Leukocytosis/thrombocytosis/polycythemia
- Leukopenia/thrombocytopenia
- Neutropenia
- Eosinophilia
- Felty's syndrome
- Cancer Associated Thrombosis
- Mastocytosis
- Lymphadenopathy