Adults with Haemophilia and Related Bleeding Disorders
Acute Treatment Guidelines

Adult Comprehensive Care Centres (CCC) Ireland

- The National Centre for Hereditary Coagulation Disorders (NCHCD),
  St. James’s Hospital, Dublin 8.          Ph: 01 4162141
  http://www.stjames.ie/Departments/DepartmentsAZ/N/NationalCentreforHereditaryCoagulationDisorders

- Cork Coagulation Centre, Cork University Hospital.   Ph: 021 4922278
  www.cuh.hse.ie
Table of Contents

1.0 Introduction

2.0 Factor VIII Deficiency Management
   2.1 General Information
   2.2 Disease Severity
   2.3 Bleeding Episode Management
      Step 1 - Patient Assessment
      Step 2 - Communication to Comprehensive Care Centres (CCC)
      Step 3 - Treatment Selection & Administration
      Step 4 - Documentation / Records
      Step 5 - PRICE for Joint Bleeds
   2.4 Surgery Management
   2.5 Pregnancy Management

3.0 Factor IX Deficiency Management
   3.1 General Information
   3.2 Disease Severity
   3.3 Bleeding Episode Management
      Step 1 - Patient Assessment
      Step 2 - Communication to Comprehensive Care Centres (CCC)
      Step 3 - Treatment Selection & Administration
      Step 4 - Documentation / Records
      Step 5 - PRICE for Joint Bleeds
   3.4 Surgery Management
   3.5 Pregnancy Management

4.0 Von Willebrand Disease Management
   4.1 General Information
   4.2 Disease Severity
   4.3 Bleeding Episode Management
      Step 1 - Patient Assessment
      Step 2 - Communication to Comprehensive Care Centres (CCC)
      Step 3 - Treatment Selection & Administration
      Step 4 - Documentation / Records
      Step 5 - PRICE for Joint Bleeds
   4.4 Surgery Management
   4.5 Pregnancy Management

5.0 Platelet Function Disorders (PFDs) Management
   5.1 General Information
   5.2 Disease Severity
   5.3 Bleeding Episode Management
      Step 1 - Patient Assessment
      Step 2 - Communication to Comprehensive Care Centres (CCC)
Step 3 - Treatment Selection & Administration
Step 4 - Documentation / Records
Step 5 - PRICE for Joint Bleeds

5.4 Surgery Management
5.5 Pregnancy Management

6.0 Rare Bleeding Disorders (RBDs) Management
6.1 General Information
6.2 Disease Severity
6.3 Bleeding Episode Management
   Step 1 - Patient Assessment
   Step 2 - Communication to Comprehensive Care Centres (CCC)
   Step 3 - Treatment Selection & Administration
   Step 4 - Documentation / Records
   Step 5 - PRICE for Joint Bleeds
6.4 Surgery Management
6.5 Pregnancy Management

7.0 Appendices

Appendix 1: SOP Haemophilia Acute Pain Management

Appendix 2: FVIII Deficiency - Clotting Factor Concentrate Dose Calculation Guide

Appendix 3: FVIX Deficiency - Clotting Factor Concentrate Dose Calculation Guide
1.0 Introduction

The National Haemophilia Council (NHC) was set up in response to the findings of the Lindsay Tribunal in 2001 and established as a statutory body in 2004 (S.I. No. 451 of 2004.) The principal function of the NHC is to provide advice, information, support and education on all aspects of haemophilia to the Health Minister, Health Service Agencies and Persons with or affected by haemophilia. Under this remit the Council works continuously to provide Clinicians with current and comprehensive evidence-based guidelines for the safe and effective management of persons with haemophilia and related bleeding disorders.

Haemophilia refers to inherited bleeding disorders caused by the absence or low level of specific proteins called clotting factors (specifically factor VIII or factor IX in the blood). Related bleeding disorders are caused by deficiencies in other clotting factors such as VWF or by abnormalities in blood platelets. The most common bleeding disorders are:

- Factor VIII Deficiency (Haemophilia A)
- Factor IX Deficiency (Haemophilia B)
- Von Willebrand Disease (VWD)
- Platelet Function Disorders (PFDs)
- Rare Bleeding Disorders (RBDS0 i.e. Inherited deficiencies of Factors I, II, V, VII, X, XI, XIII)

Due to the complexity of haemophilia and its treatment, care of persons with these bleeding disorders should be co-ordinated by a specialist centre known as a Comprehensive Care Centre (CCC). The specialist multidisciplinary services and care that these centres provide have been shown to contribute significantly to improved outcomes and better quality of life for persons with bleeding disorders. The NHC recommends that all persons diagnosed with a bleeding disorder should be registered with and monitored by one of the designated CCCs in Ireland, which are:

- The National Centre for Hereditary Coagulation Disorders (NCHCD), St. James’s Hospital, Dublin 8
- Cork Coagulation Centre, Cork University Hospital.
- Paediatric CCC - Our Lady’s Children’s Hospital Crumlin (OLCHC), Dublin 12 – All persons <16 years

However, the NHC recognises that on occasion persons with haemophilia may present to a non-specialist service requiring treatment and/or intervention e.g. with a bleed. In these circumstances non-specialist clinicians are required to assess the patients and initiate management in collaboration with the patient’s CCC. Accordingly, the NHC has commissioned these guidelines to assist healthcare professionals in the immediate management of adult persons with haemophilia. The information is presented in condition-specific chapters in which the following information is included:

- General Information
- Disease Severity
- Bleed / Suspected Bleed Management
- Surgery / Interventional Procedure Management (Elective and/or Emergency)
- Pregnancy Management
Key Statements

- Acute treatment of all persons with an inherited bleeding disorder should be co-ordinated by the CCC with which the patient is registered.

- In the event a person with a diagnosis of haemophilia or related bleeding disorder presents to a hospital requiring assessment and/or treatment and/or intervention the treating Clinician should:
  - Contact the CCC (the patient should have a registration card detailing their diagnosis and CCC)
  - Confirm the bleeding disorder diagnosis, factor level and treatment of choice with the CCC
  - Agree a management and follow up plan with the CCC.

- Prescribers must ensure that they prescribe the correct clotting factor concentrate e.g. Advate for FVIII deficiency and Benefix for FIX deficiency.

  The Prescriber must note that not all patients with mild FVIII or FIX deficiency require clotting factor concentrate as the use of alternative treatments may be indicated e.g. DDAVP (some types of FVIII deficiency only) or Tranexamic Acid. The patient’s treatment of choice must be confirmed with the relevant CCC.

- These guidelines should be used in the management of persons with bleeding disorders as an adjunct to advice received from the CCC.

- Persons under the age of 16 years (Children) should be treated in accordance with paediatric guidelines.

Scope

These Guidelines apply to:

- Adults with Inherited Bleeding Disorders including the following:
  - Factor VIII Deficiency (Haemophilia A)
  - Factor IX Deficiency (Haemophilia B)
  - Von Willebrand Disease
  - Rare bleeding disorders (inherited deficiencies of factors I (Fibrinogen), II, V, VII, X, XI, XIII)
  - Inherited disorders of Platelet function

- Immediate treatment / intervention in non-specialist centres.

Definitions / Glossary
Throughout this document the following abbreviations / acronyms are used:

- VWD = Von Willebrand Disease
- VWF = Von Willebrand Factor
- CCC = Comprehensive Care Centre
- CFC = Coagulation Factor Concentrates