Policy Directive

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Management of Haemophilia and Related Bleeding Disorders

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Functional Sub group	Clinical/ Patient Services - Medical Treatment Population Health - Health Promotion
Summary	The document is the NSW Ministry of Health's policy in relation to the treatment of patients with haemophilia and related bleeding disorders.
Replaces Doc. No.	Factor VIII Treatment/Supply to Patients with Haemophilia and von Willebrands Disorder [PD2005_168] Factor VIII Usage - January 1997 - Guidelines [PD2005_106] Bleeding Disorders - Charging for Consumables Used in Home Based Treatment [PD2005_253]
Author Branch	Office of the Chief Health Officer
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Applies to	Local Health Districts, Board Governed Statutory Health Corporations, Chief Executive Governed Statutory Health Corporations, Specialty Network Governed Statutory Health Corporations, Affiliated Health Organisations, Public Health System Support Division, Community Health Centres, Dental Schools and Clinics, Government Medical Officers, NSW Ambulance Service, Ministry of Health, Private Hospitals and Day Procedure Centres, Public Health Units, Public Hospitals
Audience	Clinicians and surgeons who treat patients who have haemophilia and related bleeding disorders.
Distributed to	Public Health System, Divisions of General Practice, Government Medical Officers, Health Associations Unions, NSW Ambulance Service, Ministry of Health, Private Hospitals and Day Procedure Centres, Tertiary Education Institutes
Review date	21-Aug-2018
Policy Manual	Patient Matters
File No.	05/7125-2
Status	Active

Director-General

This Policy Directive may be varied, withdrawn or replaced at any time. Compliance with this directive is **mandatory** for NSW Health and is a condition of subsidy for public health organisations.

POLICY STATEMENT

MANAGEMENT OF HAEMOPHILIA AND RELATED BLEEDING DISORDERS

PURPOSE

This document articulates the NSW Health policy in relation to the treatment of patients with Haemophilia and related bleeding disorders.

MANDATORY REQUIREMENTS

Compliance with NSW Ministry of Health's policy in relation to the treatment of patients with Haemophilia and related bleeding disorders is mandatory in public facilities.

It is recommended that licensed private facilities also comply with requirements of the Policy Directive.

IMPLEMENTATION

Chief Executives of Local Health Districts must ensure:

- the principles and requirements of this policy are applied, achieved and sustained.
- local protocols are in place in the relevant facilities to support implementation.
- all relevant staff are made aware of their obligations regarding this Policy Directive.

REVISION HISTORY

Version	Approved by	Amendment notes
August 2013 (PD2013_027)	Deputy Director General, Population and Public Health	Replaces PD2005_168, PD2005_106 and PD2005_253.

ATTACHMENTS

1. Management of Inheritable Bleeding Disorders: Procedures.

Management Of Haemophilia And Related Bleeding Disorders

NSW GOVERNMENT Health

PROCEDURES

Issue date: August-2013

PD2013_027

CONTENTS

BACKGROUND

This Policy Directive embodies the recommendations of the NSW/ACT Haemophilia Advisory Council Clinical Committee, members of which provide expert clinical advice to the NSW Ministry of Health in relation to the treatment of patients with haemophilia and related bleeding disorders. The recommendations of this Committee reflect best practice in the field; they encompass recommendations of the World Federation of Haemophilia and they align clinical practice in NSW with that in the rest of Australia the USA and Europe.

INTRODUCTION

The information provided in this Policy Directive is intended for clinicians (medical practitioners, nurses and midwives) and surgeons who treat patients with haemophilia and related bleeding disorders.

All patients in NSW or the ACT with haemophilia or a related bleeding disorder that may or may not require treatment must be registered with a Haemophilia Treatment Centre (HTC; see Appendix 1 for details) and their details should be entered on the Australian Bleeding Disorders Registry.

1 TREATMENT

1.1 On demand treatment

Factor concentrates may be given on demand for bleeding episodes.

1.2 Prophylaxis

The following information is based on Guidelines developed by the Australian Health Ministers Advisory Council[1].

The goal of prophylaxis is to improve the quality of life for patients with severe bleeding disorders (Haemophilia A or B and Von Willebrand Disorders) by maintaining sufficient coagulation factor levels to prevent spontaneous joint bleeding and the morbidity associated with complications of joint bleeds.

Factor prophylaxis is recommended for all patients with severe Haemophilia and Von Willebrand Disorders who are at risk of, or who have experienced, joint and other serious bleeding.

For the purposes of this policy children have a severe bleeding disorder when their **factor level is <5% and they have significant bleeding**, for example, intracranial haemorrhage.

The age at which prophylaxis therapy is introduced will vary depending on the patient's bleeding phenotype and whether the patient and their family are willing to comply with the prophylactic treatment regimen. Most children with severe haemophilia (A or B) start prophylaxis between the ages of 1 and 5 years.

The recommended dose range for prophylaxis in children depends on the underlying factor deficiency and is influenced by the recovery and the expected half-life of the factor. The usual dose range recommended for:

- Factor VIII deficiency is 25 – 40 International Units (IU) /Kg three times a week; and
- Factor IX is 40 – 75 IU/Kg two times a week.

These doses may need to be increased or given more frequently in some patients in order to prevent spontaneous bleeding.

Factor VIII/IX usage and selected clinical and laboratory outcome indicators should be routinely monitored and evaluated for all patients on prophylaxis.

[1] The Australian Health Minister's Advisory Council Evidence-based Clinical Practice Guidelines for the Use of Recombinant and Plasma-derived Factor VIII and Factor IX Products (2006)

It is mandatory that all patients receiving prophylaxis have their treatment co-ordinated and monitored by a designated NSW/ACT HTC.

It is expected that people with severe bleeding disorders will exercise reasonable precautions in managing their voluntary exposure to high risk of injury. Prophylactic treatment should be accompanied by patient/parent education about such risks.

1.3 Patients with inhibitors

Patients with inhibitors should be treated at an HTC.

2. SURGERY

Surgical intervention maybe elective or occasionally emergency in nature.

2.1 Emergency surgery

Where emergency surgery is conducted at a facility that is not an HTC the hospital should contact the Director of the HTC at the Royal Prince Alfred Hospital if the patient is an adult or the Director of the Sydney Children's Hospital Network- Westmead Campus if the patient is a child (see Attachment 1 for contact details of the relevant HTCs). It is likely that urgent transfer of the patient would be required.

2.2 Elective surgery

General matters

Elective surgery on patients with bleeding disorders must take place in consultation with a NSW/ACT HTC.

Applications for supply of coagulation factor for surgery must be approved by the Haemophilia Advisory Council Clinical Committee. Once approval has been given, factor concentrates will be made available by the Australian Red Cross Blood Service.

Application for coagulation factors for surgery should be made on the form at Attachment 2 of this Policy Directive.

Principles relating to elective surgery

The following Guidelines govern the management of elective surgery in patients with haemophilia and other bleeding disorders:

- the Australian Health Minister's Advisory Council Evidence-based Clinical Practice Guidelines for the Use of Recombinant and Plasma-derived Factor VIII and Factor IX Products (2006);
- the Australian Haemophilia Centre Directors' Organisation Guideline for the Management of Patients with Haemophilia undergoing surgical procedures (2005); and
- the World Federation of Haemophilia Guidelines for the Management of Haemophilia (2012).

In this Policy Directive the principles outlined in the Guidelines have been adapted to balance patient safety with patient preference and equity of access.

In the interests of patient safety, patients with:

- Factor VIII < 30%
- Factor IX < 30%
- Factor XI < 30%
- Von Willebrand Factor activity < 30%; and
- other rare bleeding disorders

should have elective surgery performed in a designated NSW/ACT Haemophilia Treatment Centre since they are "high risk patients" with an increased tendency to bleed, both early and late, and they require management by clinicians experienced in managing haemophilia and related bleeding disorders.

If a patient has a factor level that is over 30% and there is a specific reason for a procedure to be conducted at a hospital that is not a designated NSW/ACT HTC, the patient/parent(s) or carer **MUST** be made aware of the potential risks attached to having surgery in a hospital that is not a designated or affiliated NSW/ACT HTC and the NSW/ACT Haemophilia Advisory Council Clinical Committee **MUST** be advised.

The NSW/ACT Haemophilia Advisory Council Clinical Committee will review such requests on a case by case basis to ensure that the following requirements are met:

(1) The hospital where it is proposed the operation is to be conducted MUST have:

- Blood Bank/Pathology capable of providing appropriate support, even in emergency circumstances;
- Intensive Care Unit (ICU) (where relevant) – staffed 24 hours by qualified ICU specialists to supervise treatment; and
- Pathology – with ability to provide same day factor levels.

(2) The hospital must have access to a:

Clinical Haematologist who is available to supervise and review treatment daily and is on-call at all times.

(3) The treating Clinical Haematologist must:

- liaise with the Director of the relevant NSW/ACT HTC and must provide a written account of a:
  - treatment plan – including dosages and target levels; and
  - monitoring plan – the protocol must be approved by an HTC Director, and it must be followed and carried out in close liaison with the Director of the relevant NSW/ACT HTC.
- liaise daily (or as required clinically) during the treatment period with the Director of the NSW/ACT HTC; and
- provide the Director of the relevant NSW/ACT HTC with a summary of factor usage and outcomes of the patient under their management.

(4) The HTC Director must be:

- prepared to review and, if appropriate, approve the treatment and monitoring plan;
- liaise with the treating Clinical Haematologist if the patient's treatment and monitoring plan are approved; and
- be available to the treating Clinical Haematologist during the treatment period.

3. LIST OF ATTACHMENTS

1. List of Haemophilia Treatment Centres.
2. Application Form for the Supply of Coagulation Factors for Elective Surgery.

Attachment 1

HAEMOPHILIA TREATMENT CENTRES

The locations and contact details of the NSW/ACT Haemophilia Treatment Centres are as follows:

NSW

Royal Prince Alfred Hospital
The Director
Haemophilia Centre
Building 77, Level 5
Royal Prince Alfred Hospital
Missenden Road Camperdown NSW 2050
Telephone 02 9515 7013
Emergency 02 9515 6111
Fax 02 9515 8946

Westmead Hospital
The Director
Department of Haematology
Westmead Hospital
Telephone: (02) 9845 6274
After hours & weekends: Haematology Registrar 0409392151

The Sydney Children's Hospital Network- Westmead Campus
The Director
The Kids' Factor Zone
The Children's Hospital at Westmead
Cnr Hawkesbury Rd & Hainsworth St
Westmead NSW 2145
Telephone 02 9845 0839
After hours & weekends: (02) 9845 0000 and ask for the Haematologist on call Fax 02 9845 3332

Sydney Children's Hospital Network – Randwick Campus
Head of Paediatric Haematology
Centre for Children's Cancer and Blood Disorders
Sydney Children's Hospital
High Street
Randwick NSW 2031
(02) 9382 1690
After hours & weekends: (02) 9382 1111 and ask for the Haematologist on call
Fax (02) 9382 1789

Prince of Wales Hospital
Senior Staff Haematologist
Department of Haematology
SEALS Level 4
Prince of Wales Hospital
Barker Street
Randwick NSW 2031
(02) 9382 9013
After hours & weekends: (02) 9382 2222 & ask for the Haematologist on call
Fax: (02) 9382 9116

Calvary Mater Newcastle
The Director
Haemophilia Centre
Edith Street Waratah NSW 2298
Telephone 02 4921 1240
After hours & weekends: (02) 4921 1211 and ask for Haematologist on call
Fax 02 4960 2136

ACT

The Canberra Hospital
The Director
Haemophilia Clinic
The Canberra Hospital
Yamba Drive
Garran ACT 2605
Telephone (02) 6244 4048
After hours & weekends: (02) 6244 2222 and ask for Haematologist on call
Fax: (02) 6244 2271

NSW GOVERNMENT Health

Facility:

FAMILY NAME	MRN
GIVEN NAME	☐ MALE ☐ FEMALE
D.O.B. _____/_____/_____	M.O.
ADDRESS	
LOCATION / WARD	
COMPLETE ALL DETAILS OR AFFIX PATIENT LABEL HERE	

APPLICATION FOR THE SUPPLY OF COAGULATION FACTORS FOR ELECTIVE SURGERY

SMR030045

Elective Surgery Regn. No_____

NSW/ACT HAEMOPHILIA ADVISORY COUNCIL CLINICAL COMMITTEE

This application for the supply of the clotting factor required for elective surgery should be submitted in the first instance to the address below. Consideration will be then given to the request by the NSW/ACT Haemophilia Advisory Council Clinical Committee at its next available meeting.
If there is a shortage of the relevant coagulation factor, elective surgery may have to be deferred and be subject to the haemophilia elective surgery priority list.
Please tick the relevant box where indicated.

Patient details

Family name:_____ Given Name _____ **Date of birth:**____/____/____ (dd / mm / yyyy)

Sex: ☐ Male ☐ Female **Weight:**_____kgs

Haemophilia Treatment Centre_____

Haemophilia Treatment Physician _____ **ABDR ID Number** _____

Type of Bleeding Disorder

☐ **Haemophilia A** ☐ Mild ☐ Moderate ☐ Severe **Factor VIII** level:_____%
☐ **Haemophilia B** ☐ Mild ☐ Moderate ☐ Severe **Factor IX** level: _____%
☐ **von Willebrands disease Type** ☐ 1 ☐ 2A ☐ 2B ☐ 2M ☐ 2N ☐ 3
Factor VIII:C _____% **vWF Ag**:_____% **VWF:RiCoF:**_____%
☐ **Factor XI deficiency** ☐ **Factor XIII deficiency** ☐ **Factor VII deficiency**
☐ **Other Bleeding Disorder**_____

Inhibitor status:_____ ☐ Negative ☐ Positive_____ **Titre**_____ BU_____

Proposed Surgery

Indications for surgery:_____
Surgery proposed:_____
Proposed Hospital / location of surgery:_____
Proposed date of surgery:_____/_____/___20____ .**Degree of urgency:**_____
Coagulation Factor Requirements (Tick relevant box and provide product name)
☐ **Plasma-derived Factor VIII** _____
☐ **Recombinant Factor VIII** _____
☐ **Plasma-derived Factor IX** _____
☐ **Recombinant Factor IX** _____
☐ **Factor IX** ☐ **Factor XI** ☐ **Factor VIIa** ☐ **Factor VII** ☐ **FEIBA**
☐ **Other** _____

Estimated requirement: _____bottles or_____IUs
For ☐ **Bolus or** ☐ **Continuous infusion**
Nature of any complicating conditions:_____

Signed: _____ Print Name _____ (Applicant)

Designation: _____ Date _____

03/2011

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APPLICATION FOR THE SUPPLY OF COAGULATION FACTORS FOR ELECTIVE SURGERY SMR030.045

NSW GOVERNMENT Health

Facility:

FAMILY NAME	MRN
GIVEN NAME	☐ MALE ☐ FEMALE
D.O.B. _____/_____/_____	M.O.
ADDRESS	
LOCATION / WARD	
COMPLETE ALL DETAILS OR AFFIX PATIENT LABEL HERE	

APPLICATION FOR THE SUPPLY OF COAGULATION FACTORS FOR ELECTIVE SURGERY

Please place this application in an envelope marked Confidential and address it to:

NSW/ACT Transfusion Medicine Specialist
Medical Services
Australian Red Cross Blood Service
17, O'Riordan Street
Alexandria NSW 2015

Office use only

NSW/ACT HAEMOPHILIA ADVISORY COUNCIL CLINICAL COMMITTEE
Approval for the supply of coagulation factors for elective surgery

1. Further information to be sought: _____

2. Details of further information obtained: _____

3. Decision of the Clinical Committee: _____

4. Approval

☐ **Approved - ARCBS to supply product**

Or

☐ **Deferred - to Haemophilia Elective Surgery / Coagulation Factor Waiting list**

Or

☐ **Not approved - for following reason(s)**

_____

Signed: _____ Print Name _____

Designation: _____ Date _____

Holes punched as per AS2828-1999

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