1. Introduction and Who Guideline applies to

This guideline is for staff working in the haemophilia centre or involved in the emergency management of people with bleeding disorders in UHL

2. Guideline Standards and Procedures

People with bleeding disorders have an equal risk of traumatic injuries as the general population, but there may be an increased risk of severe bleeding from lesser trauma, and the consequences of trauma may be more dramatic with increased bleeding into injured tissue. In addition, there will be an increase risk of pathological bleeding. As a result it is crucial that this group are provided with early access to expert advice and management of the bleeding disorder, in addition to the usual measures to treat an injury or bleed episode.

People with bleeding disorders should be registered at a haemophilia centre and provided with a card stating their diagnosis, as well as emergency contact details.

The pathway for seeking medical advice in the event of the bleed/trauma is as follows:

Recognition of bleed /trauma

Severe – Emergency department

Not severe – During working hours, patient/carer to call haemophilia centre – telephone advice / review in haemophilia centre. During out of hours, patient or carer to ring on call haematology registrar - telephone advice or review in appropriate place eg. ED.

On attending to hospital, clinical assessment and provision of haemostatic support as required. If assessed as severe by haemophilia team, treat and send onwards to emergency dept.

If presenting to emergency department, hospital electronic system alert (Nerve centre) prompts the treating ED clinician to assess the patient and to contact haemostasis team or haematology registrar on call.

General measures: RICE

Rest

Ice/ immobilise

Compress

Elevate

Analgesia: Avoid NSAIDs

Contact physiotherapist as appropriate

Consider tranexamic acid

Specific measures:

These are highly specific to the bleeding disorder but options for haemostatic support include: DDAVP – for e.g. mild haemophilia A, Von Willebrand disease, platelet function disorders

Factor concentrate: for e.g. haemophilia A &B, deficiency of factors 1, VII, XI, X

SD- FFP(Octaplas) – for deficiency of factor V, XI

PCC – For deficiency of Factors II, X

Bypassing agents (FEIBA, NOVOSEVEN) – for patients with factor inhibitors, typically haemophilia A

N. B. AVOID CONCOMITANT ADMINISTRATION OF EMICIZUMAB AND FEIBA DUE TO RISKS OF THROMBOTIC MICROANGIOPATHY

For haemophilia A & B, please see the following table. This is a guide for intensity and duration of treatment for particular bleed episodes.

	Minimal therapeutic	**Dose (U kg^{-1} BW)**		
Site of haemorrhage	**Factor level**	**Factor VIII**	**Factor IX**	**Duration in days**
Joint	30-50%	20-30	30-50	1-2 (or until resolution)
Muscle	30-50%	20-30	30-40	1-2 (or until resolution)
Gastrointestinal tract	40-60%	30-40	40-60	7-10
Oral mucosa	30-50%	20-30	30-40	Until healing
Epistaxis	30-50%	20-30	40-60	Until healing
Haematuria	30-100%	25-50	70-100	Until healing
Central nervous system	60-100%	50	80-100	7-10
Retroperitoneal	50-100%	30-50	60-100	7-10
Trauma or surgery	50-100%	30-50	60-100	Until healing

Similar advice for the rarer bleeding disorders is beyond the scope of this guideline due to the rarity and heterogeneity of the conditions. Specific management plans should be made by those with experience in managing bleeding disorders at the haemophilia centre.

Pathway for patients with inherited bleeding disorders and attending ED

This group of patients are at increased risk of bleeding including fatal bleeding events
INVOLVE HAEMOPHILIA TEAM IN THE CARE OF THESE PATIENTS AT ALL TIMES

Patient with a known bleeding disorder presents to ED

- Self identifies
- Carries a medical card
- Nerve centre alert

INITIAL TRIAGE

Triaged by ED Nurse

- Nature of Complaint/Injury
- Type of bleeding disorder and usual treatment
- Have they self-treated; **when** was the last dose of clotting factor given; **how much** and **what** product was given?
- ED triage nurse calls haemophilia team immediately

Contact details:

- Monday to Friday 08:30am – 4:30pm, Haemophilia Centre Nurses Office – Ext.:6500 or Haematology Registrar on call through switchboard
- Out of hours – Haematology Registrar on call through switchboard

Patient to be reviewed by ED TEAM while waiting for haemophilia input

INITIAL BLEEDING DISORDER TREATMENT

Haematology registrar

- Identifies patients who require immediate treatment with clotting factor concentrate and informs ED triage Nurse of the management plan
- Accesses treatment records in Haemophilia Centre or liaise with Haematology SHO to do this (out of hours)
- Identifies the correct product and dose for patient as per individual patient prescription. Liaise with Haemophilia Consultant if needed.
- Attends ED and documents assessment and plan, prescribes clotting factor.

Nurses will administer concentrate in line with trust IV guidelines on MEDUSA or the SPC at www.medicines.org.uk

OUTCOME

Immediate treatment required within 30 mins	**Assess within 30 mins Treatment commenced within 2 hours**

DOES THIS PATIENT NEEDS TO BE REVIEWED BY ANOTHER SPECIALITY?

YES	**NO**
ED team to refer to appropriate team	**Admit under Haematology as required**

If patient discharged, ensure they have a plan and follow up is arranged with the Haemophilia Centre

General Clinical Information

- Bleeding disorders include: Haemophilia, Von Willebrand's Disease, other clotting factor deficiencies e.g. Factor XI, Platelet function disorders.
- Early bleeding in the head, spine, abdomen and pelvis may not be clinically obvious.
- Prompt treatment of joint and muscle bleeding prevents compartment syndrome.

Clotting factors (CFC)

- CFCs are available in the Haemophilia Centre and can be accessed by the Haemophilia CNS/Haematology Registrar/Haematology SHO
- CFCs may be administered by any nurse assessed as competent in giving IV drugs

Treatment principle: first treat then investigate

- Patients with severe inherited bleeding disorders, generally self-treat: ask when they last had treatment and what dose and establish current need
- Avoid aspirin, NSAID's or IM injections.

ANY HEAD INJURY OR DETERIORATION IN CONSCIOUS LEVEL REQUIRES AN URGENT SCAN ALONG WITH IMMEDIATE ESCALATION TO THE ED CONSULTANT.

Pathway for patients with inherited bleeding disorders and Attending Out of Hours

This group of patients are at increased risk of bleeding including fatal bleeding events
INVOLVE HAEMOPHILIA TEAM IN THE CARE OF THESE PATIENTS AT ALL TIMES

Patient with a known bleeding disorder rings Haematology Registrar

- Self identifies
- Carries a medical card
- Nerve centre alert

INITIAL TRIAGE

Triaged by Haematology Registrar over the phone:

- Nature of Complaint/Injury
- Type of bleeding disorder and usual treatment
- Have they self-treated; **when** was the last dose of clotting factor given; **how much** and **what** product was given?

Contact details:

- Out of hours – Haematology Registrar on call through switchboard

IF DECIDED TO ADMIT PATIENT TO ED

All Paediatrics to go To Children's Emergency Department	Adults Adult Emergency Department	Adults Osborne Assessment Unit

Patient that go to ED to be reviewed by ED TEAM while waiting for haemophilia input (see separate ED Pathway)

INITIAL BLEEDING DISORDER TREATMENT

Haematology registrar

- Identifies patients who require immediate treatment with clotting factor concentrate and informs ED triage Nurse of the management plan
- Accesses treatment records in Haemophilia Centre or liaise with Haematology SHO to do this (out of hours)
- Identifies the correct product and dose for patient as per individual patient prescription. Liaise with Haemophilia Consultant if needed.
- Attends ED/OAU and documents assessment and plan, prescribes clotting factor. Clotting factors are all kept in the Haemophilia Centre and key is kept on Ward 41.

Nurses will administer concentrate in line with trust IV guidelines on MEDUSA or the SPC at www.medicines.org.uk

OUTCOME

Immediate treatment required within 30 mins	Assess within 30 mins Treatment commenced within 2 hours

DOES THIS PATIENT NEEDS TO BE REVIEWED BY ANOTHER SPECIALITY?

YES	NO
ED team to refer to appropriate team	Admit under Haematology as required

If patient discharged, ensure they have a plan and follow up is arranged with the Haemophilia Centre

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Education and Training

Nil

3. Monitoring Compliance

What will be measured to monitor compliance	How will compliance be monitored	Monitoring Lead	Frequency	Reporting arrangements
Annual review	MDT	MDT lead	annual	MDT minutes

4. Supporting References (maximum of 3)

Guideline for the diagnosis and management of the rare coagulation disorders A United Kingdom Haemophilia Centre Doctors' Organization guideline on behalf of the British Committee for Standards in Haematology Andrew D. Mumford et al Haemophilia 2013

Emergency and out of hours care for patients with bleeding disorders – Standards of care for assessment and treatment. John Hanley et al for UKHCDO. UKHCDO website. 2009

The diagnosis and management of von Willebrand disease: a United Kingdom Haemophilia Centre Doctors Organization guideline approved by the British Committee for Standards in Haematology Mike A. Laffan et al. Br Journal of Haematology 2014

Guidelines for the management of hemophilia Srivastava, A. et al , (2013),. Haemophilia, 19: e1-e47. doi:10.1111/j.1365-2516.2012.02909

5. Key Words

Haemophilia, Von Willebrand disease

CONTACT AND REVIEW DETAILS	
Guideline Lead (Name and Title) Sandhya Munireddy	**Executive Lead**
Details of Changes made during review: Nerve centre alert added	