

Caring for people with inherited bleeding disorders

Information for staff working in residential care facilities



Australian Haemophilia Nurses' Group (AHNG)
Australia/New Zealand Haemophilia Social Workers'
and Counsellors' Group (ANZHSWCG)

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Table of Contents

Background	3
Objectives	3
Bleeding disorder basics	4
Living with an inherited bleeding disorder	5
Managing an inherited bleeding disorder day-to-day	7
Management after an injury	10
Injury care plan – site specific	11
Facts about “factor”	14
Treatment plan	16
HTC contact information	17
Haemophilia Foundation contact information	18

Background

Due to the advances in the management of inherited bleeding disorders such as haemophilia, people with bleeding disorders are living longer than ever before. We are now beginning to see an ageing population and will increasingly see people entering residential care facilities with these conditions.

The aim of this booklet is to provide staff at residential care facilities with an understanding of the specific needs of these residents in relation to their inherited bleeding disorder.

Objectives

The information contained in this booklet aims to:

- Provide staff and carers with information about the care of people with inherited bleeding disorders entering care facilities
- Assist the transition of clients into aged care facilities
- Provide education about the management of bleeding events in people with inherited bleeding disorders

Bleeding disorder basics

Definition of a bleeding disorder – a disorder where there is a deficiency or impaired functioning of one of the proteins necessary for blood clots to form and healing to occur. They are characterised by extended bleeding after injury, surgery, trauma or menstruation.

Bleeding disorders can be inherited or acquired. Inherited disorders are passed down from parent to child through genetics while acquired disorders can develop or occur spontaneously later in life. They are not contagious.

Disorders include:

- haemophilia A
- haemophilia B
- von Willebrand disease
- and other rare bleeding disorders.

These disorders affect both men and women, although men are more likely to have haemophilia.

There are varying severities (mild, moderate and severe) of each specific inherited bleeding disorder which reflect the impact of the bleeding disorder on symptoms in day to day life. For example, a person with severe haemophilia may have spontaneous bleeding into a joint or muscle which occurs at times for no obvious reason, where as a person with mild haemophilia may only have bleeding with surgery, procedures or trauma.

Please note that when it comes to any surgery, procedure, dentistry or injury ALL residents with an inherited bleeding disorder require review by the Haemophilia Treatment Centre (HTC) and possible intervention. As such, staff should inform the Centre as soon as they are aware of any upcoming invasive procedures.

For further information regarding the specific bleeding disorder of your resident refer to disorder specific booklets. These booklets may be available through Haemophilia Treatment Centres or the Haemophilia Foundation Australia website - www.haemophilia.org.au.

Living with an inherited bleeding disorder

For the majority of people, living with an inherited bleeding disorder is only an issue when there is planned surgery/intervention (including dental work) or at times of accident and injury. Women in earlier years may have suffered from menorrhagia (heavy periods) and perhaps have been troubled with iron deficiency.

However, for those with severe bleeding disorders, repeated bleeding into joints (typically the elbows, knees and ankles) results in joint deformity and pain and often requires surgical intervention. You may notice that some people with inherited bleeding disorders, particularly those with a severe bleeding disorder, require residential care at an earlier age due to early onset arthritis and the resulting physical disability.

The majority of people with bleeding disorders manage their condition well and are very independent. Many have faced years of living with a debilitating chronic illness, but they remain very resilient and are a resourceful group of people. Being respectful of their knowledge and sensitive to their past experiences will most certainly ensure a smooth transition to residential care.

As with many chronic diseases, people with inherited bleeding disorders are encouraged to be involved in their medical care and treatments, and are used to working with health professionals to achieve good health care outcomes. It would be valuable to continue to involve the resident and/or their partner/carer in decisions related to managing their disorder. Some residents may find it very difficult to change or put in place new ways in managing their disorder and some may feel they have lost their sense of control. Involving them and their partner/carer in discussions and developing a plan together which takes into account their experience and knowledge may help with this.

Some specific topics:

Joint health

For people with severe bleeding disorders joint health is a primary concern. Joints may have become damaged in childhood if appropriate treatment was inaccessible or in short supply, as was the case when most residents with bleeding disorders were growing up. When repeated bleeding occurs into a specific joint, it becomes a target joint as the blood within the joint sparks a vicious cycle of re-bleeding. As mentioned, the elbows, knees and ankles are the main joints affected. For some older people all 6 joints may have some degree of damage.

Prior to entry to the residential facility patients may have undergone surgery to reduce pain and improve function. This may include knee replacements, ankle fusions and elbow resections. The use of walking aids may be encouraged. Transferring patients must be done with their assistance (and perhaps with their helpful instructions) as they will know best how not to aggravate the pain, and to avoid knocks and bruises if equipment is to be used (e.g., hoists). Please remember to protect the resident's skin and joints when using equipment to reduce the risk of skin tears, bruising or joint injuries.

Pain

Pain management in people with inherited bleeding disorders poses some challenges over and above the usual ageing person. Due to the risk of adverse effects including the risk of gastric ulcers and effects on platelets leading to increased risk of bleeding, non-steroidal anti-inflammatory drugs (NSAIDs) are usually avoided. However, COX-2 selective NSAIDs may be considered and can play a vital role in pain management related to arthritic joint pain. Paracetamol with/without narcotics may also be considered in the management of chronic pain in the older person with an inherited bleeding disorder. Any change to analgesia should be done in conjunction with the Haemophilia Treatment Centre.

Managing inherited bleeding disorders day-to-day

There are two ways to manage inherited bleeding disorders on a day-to-day basis (excluding management for planned procedures): **on-demand** and **prophylaxis**. The treatment approach for the individual will be determined by the treating haematologist.

1. On-demand (episodic): treatment only as required for the management of a bleeding episode, either related to trauma/injury or a spontaneous event. This type of treatment regimen is used in all types of bleeding disorders and severities.

2. Prophylaxis: treatment given at regular intervals to prevent spontaneous bleeding episodes. Prophylaxis is usually only used in people with severe bleeding disorders such as severe haemophilia A, severe haemophilia B and severe von Willebrand disease.



Butterfly needle used for infusions



Infusing factor replacement therapy

Factor replacement therapy: treatment may consist of factor concentrate, commonly referred to as “factor” by patients. The type of factor concentrate is specific to the bleeding disorder. Factor is given intravenously and some patients will be, or would have been, proficient in self administration in the home. Their partners/carers may also be proficient in factor administration.

Desmopressin (DDAVP) is also used to treat people with mild haemophilia (including some women) and mild type 1 von Willebrand disease. It is often given as an intravenous infusion over 30 minutes, but is also available in a concentrated vial strength which allows for subcutaneous administration. DDAVP is known to cause fluid retention for 24 hours following administration and a 1.0 L fluid restriction should be in place over this period. The use of DDAVP is not recommended in elderly patients over the age of 65.

Some patients may require the insertion of a PICC line (peripherally inserted central catheter) in order to receive the factor concentrate. If this is considered an appropriate option, the Haemophilia Treatment Centre will assist the Residential Care Facility in managing such a device through services such as Hospital in the Home.

Your residential facility will need to consider how the resident will be managed and should involve the resident (as appropriate), family, residential care facility staff, general practitioner and HTC staff, as required.

Partners/carers

Please be aware that due to the lifelong nature of the bleeding disorder, a resident’s significant other will have participated in caring for the resident. The partner or carer will often be able to provide an insight on how the bleeding disorder has impacted on the resident’s life. They may know how to prepare the treatment and may even be able to administer the treatment if required. It will be important to discuss the role of the partner or carer within your residential facility and include them as part of the team.

Blood borne viruses

Be aware that some residents with inherited bleeding disorders may have received contaminated blood products in the past and as a result, were infected with HIV, hepatitis B and/or hepatitis C. As those affected age, the risk of complications associated with these viral infections increases. Even if their hepatitis C has been cured, they may have developed liver cirrhosis and require ongoing monitoring. These residents may see specialists such as hepatologists or immunologists for surveillance and follow-up; however, some may have this role performed by their GP.

Please be aware that not all residents will have disclosed their diagnosis to friends and family and therefore, confidentiality is paramount. Only staff involved in clinical care need to be aware of a resident’s viral status. It may be valuable to ask the resident to clarify who knows about their HIV and/or hepatitis B or C. This could be recorded in the resident’s notes and mentioned at handover so that clinical staff, particularly new or agency staff, are aware each shift to avoid discussing it in front of anyone by mistake.

Facts about blood borne viruses:

- Infection is not acquired through everyday social contact, through crockery, cutlery, showering or touching
- Items such as nail clippers, scissors, razors, toothbrushes or other personal items that have the potential risk to be contaminated by blood should only be used by individual residents
- Wounds should be covered
- Spills of bodily fluids should be managed immediately as per standard universal blood and body fluid precautions. The intention of standard precautions is that they are used with every blood or body fluid spill, no matter who is involved. It is important to practice this consistently so that:
 - Staff and residents are confident that they are all being protected in all circumstances
 - Residents with blood borne viruses do not feel discriminated against.
- Staff should be familiar with their residential care facility's procedures.

Psychosocial impact of living with a blood borne virus

- Living with a blood borne virus can have a huge impact on the quality of life of an individual, and his/her physical and mental health
- Stigma associated with blood borne viruses can leave residents feeling rejected and isolated, and can lead to feelings of anxiety and depression
- Residents may feel discriminated against by staff who do not understand the mode of transmission and over-emphasise the risk of transmission
- Staff have a legal responsibility to maintain the residents' confidentiality. Some residents may not have disclosed their status to family and friends, therefore staff should assume this is the case.
- Be aware that while some residents will feel comfortable in discussing their viral status, some may not. Please be sensitive towards residents' feelings as some may have had a lifetime of hardship

Haemophilia Treatment Centre (HTC)

Each state and territory in Australia has at least one Haemophilia Treatment Centre (HTC). At these Centres are consultant haematologist(s) with an interest in inherited bleeding disorders, haemophilia nurse(s) and most HTCs have access to a social worker/psychologist/counsellor and physiotherapist.

The HTC provides routine follow-up appointments for review, surgical plans and management of acute bleeding episodes. The staff at the HTC can also provide education and information as required.

The majority of people with inherited bleeding disorders are known to an HTC, but some will be seen by a private haematologist.

Management after an injury

If a resident is injured always assess immediately. **Never keep them waiting.** A resident can feel a bleed before there are any obvious signs or symptoms. Treatment should be a priority if there is suspicion of injury. The resident's treatment plan should be followed after administering essential first aid measures.

Residents will require urgent medical assessment and management of their bleeding disorder if they suffer:

- Any fall or injury in which there is trauma to the head, eye, throat, chest, back, abdomen, hip or groin
- Broken bone (or are suspected of a broken bone)
- Cut requiring sutures
- Common bleeds from the mouth, nose, cuts and scrapes that do not stop bleeding after 20 minutes of general first aid

Signs and symptoms of a bleed:

- Complaints of pain, tingling, bubbling, filling sensation within a muscle or joint, stiffness or decreased range of movement (ROM) in limb
- Swollen body part or an area, joint or muscle that is warm to touch
- Favours an arm or leg more than usual
- Resident limps or refuses to use limb

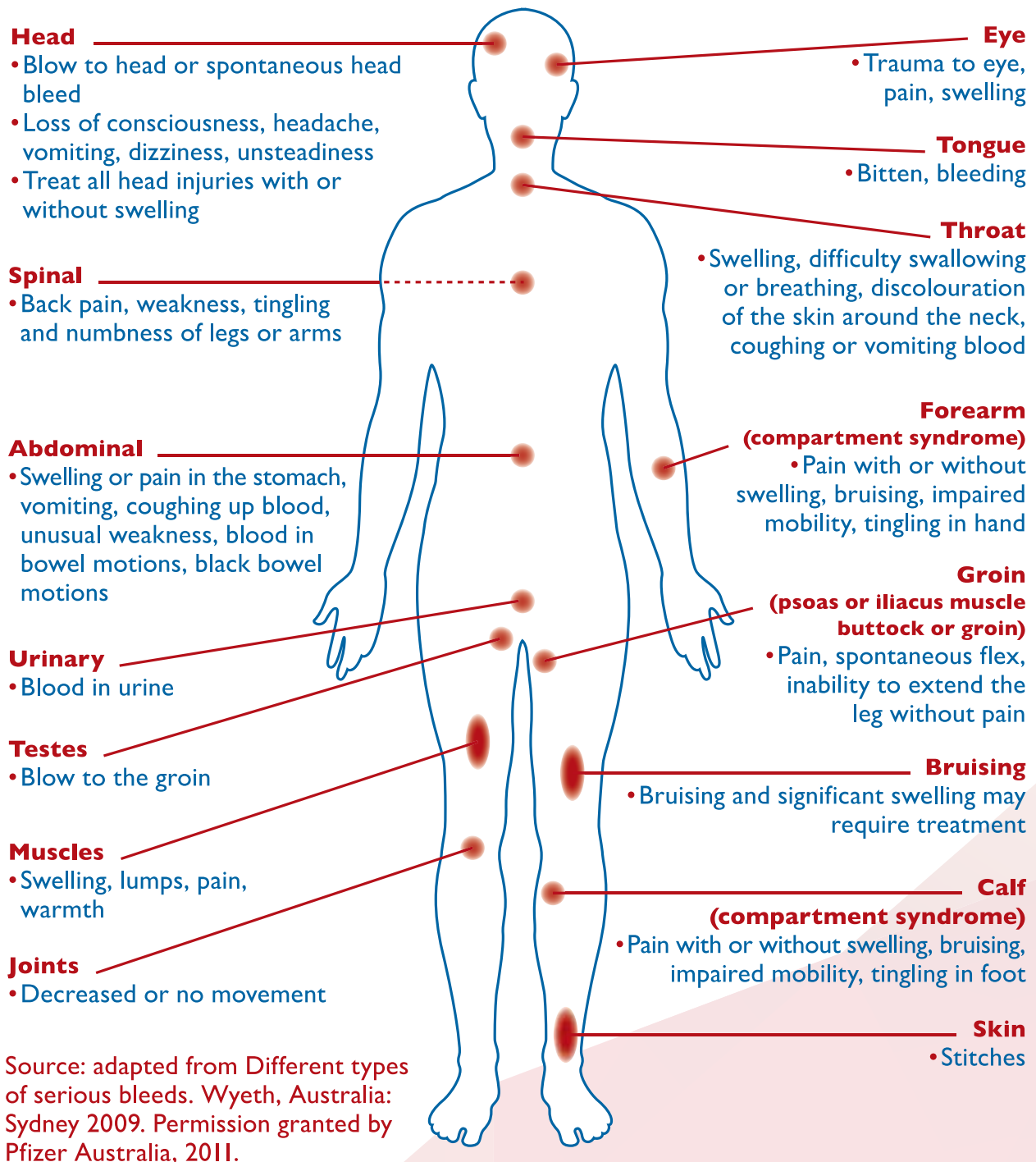
Use first aid guidelines and standard precautions:

- Wash hands, wear gloves
- Keep wound clean
- Clean blood spills
- If factor replacement therapy is used, dispose of needle as per unit policy.

Injury Care Plans – site specific

Serious bleeding: injuries requiring prompt medical advice

There may be no visible signs of bleeding in a person with an inherited bleeding disorder, however bleeding episodes can be life and limb threatening. The following bleeds and injuries should be treated as serious and require prompt medical attention.



Contact the Haemophilia Treatment Centre for assessment if pain or swelling from an injury keeps the resident awake overnight following the Rest Ice Compress and Elevate procedure.

Remember: people with bleeding disorders do not bleed faster, only longer

The following information is generic – please refer to the resident’s HTC Treatment Plan for information regarding the administration of haemostatic management

Mouth bleeds

- Apply continuous pressure to site of bleed
- If possible encourage the resident to place a piece of ice in the mouth
- If mouth bleeds are frequent, contact the HTC for advice
- Soft, tepid diet and fluids
- HTC may consider tranexamic acid paste and/or mouthwash

Nose bleeds

- With the resident sitting upright, pinch the soft part of nose
- If possible encourage the resident to place a piece of ice in the mouth
- Tepid food and drinks
- If ongoing/frequent HTC may consider tranexamic acid tablets

Superficial cuts and scrapes

- Wash with antiseptic soap
- Apply firm pressure with gauze for 20 mins
- Apply non-adherent dressing

Joint and muscle bleeds

- Can be serious and possibly limb-threatening
- If clotting factor concentrate can be administered at the facility, administer the first dose and contact the HTC (or Emergency Department on weekends/public holidays)
- Use R.I.C.E. principles

R.I.C.E.

R – rest – keep the resident as still as possible

I – ice – apply ice to area for 20 mins per hour (ice pack wrapped in towel, or cool relief gel directly on the skin)

C – compression – wrap area with elastic bandage or tubigrip

E – elevation

Issue walking aids (Zimmer frame or crutches) or resting braces (Richards splint for knees).

Advise non-weight bearing if in doubt.

Call for a review by the Physiotherapist.

Facts about “factor”

Definition of “factor” (clotting factor concentrate used for treatment): A product made from human blood or non-blood sources that helps the blood clot by replacing the missing clotting factor

Factor facts

- There are different factors, different brands and strengths
- A doctor or haemophilia nurse practitioner must prescribe the factor
- Factor is stored as a concentrated powder which requires reconstitution
- Factor is measured in international units (IU)
- Batch number, expiration date and number units per vial are printed on the factor box
- Batch number allows tracking – the HTC may provide you with a treatment diary in which vial stickers can be placed. Check with the HTC on how they would like you to record usage
- Use factor with the shortest expiration date first
- If factor is expiring at the end of the month, contact the HTC for replacement stock
- Factor concentrate is only to be mixed with the diluent provided
- Factor is injected as an intravenous bolus using the butterfly needle provided with the concentrate
- The line must be primed prior to delivery of factor concentrate
- If using the provided butterfly, the line does not require flushing
- Contact your HTC for information regarding re-ordering of stock

Factor dosage

- Is based on the weight of the person
- Dosing may change over time as activity changes or body mass changes
- Dosage and frequency of factor administration varies between clients
- Based on the required dosage, you may need to use more than one vial of factor concentrate, these can be pooled into one syringe and administered as a bolus over 1-2 minutes
- Factor needs to be administered within 3 hours of reconstitution, therefore only reconstitute immediately prior to administration
- Call the HTC if any questions.

Storage

- Ideally factor concentrate should be stored in the fridge
- Storing it at room temperature reduces the expiration to as little as 3 months – check with the HTC if refrigeration not available
- Once reconstituted do not return to the fridge
- Never freeze factor
- If there is a fridge malfunction please contact the HTC for advice.

Treatment Plan

Resident: _____

Type of bleeding disorder: Haemophilia A Haemophilia B Von Willebrand disease

Other: _____

Severity: mild moderate severe Inhibitor: Yes No

Do not give any aspirin, NSAIDs or aspirin-containing products
(possible exception: COX2-selective drugs; only in consultation with HTC)

Check all vitamins, minerals and supplements with the HTC staff prior to administration
as some can increase the bleeding risk

This resident may require management of bleeding disorder prior to any medical/surgery intervention

Common bleeding sites:

Target joints: _____

Oral mucosal: _____

Other sites: _____

Will factor be infused in the facility? YES NO N/A

If so, who will infuse the factor?

Where will the factor be stored?

Name of treatment to be infused:

Recommended treatment dose

Prophylaxis: _____ IU Frequency: _____

On-demand: Minor bleed _____ IU Major bleed _____ IU

Will Desmopressin be administered in the facility? YES NO N/A

If so, who will administer it? _____

On-demand: Minor bleed _____ mcg Route: IV Subcutaneous

Major bleed _____ mcg Route: IV Subcutaneous

HTC Contact Information

Contact	Address	Contact number
<p>Haemophilia Treatment Centre:</p> <p>HTC contact:</p> <p>After hours contact the hospital in which the HTC is located and ask to speak to:</p>		<p>Hospital number:</p> <p>Pager number:</p> <p>HTC Office:</p> <p>HTC Nurse:</p> <p>Mobile:</p>
<p>Haemophilia Social Worker/ Psychologist:</p>		<p>Office:</p> <p>Mobile:</p>

Haemophilia Foundations

Haemophilia Foundation Australia (HFA) represents the bleeding disorders community in Australia. HFA provides information and educational material relevant to people with bleeding disorders and has an email newsletter, a regular magazine, a web site, social media platforms including Facebook, Twitter, Instagram and YouTube, and print booklets.

T: 1800 807 173 (toll free)
E: hfaust@haemophilia.org.au
W: www.haemophilia.org.au

State and Territory Haemophilia Foundations provide information, newsletters, social activities and other support to people with haemophilia and other bleeding disorders, their families and the general public in their state/territory.

Haemophilia Foundation ACT (HFACT)

T: 0412 839 135
E: president@hfact.org.au
W: www.hfact.org.au

Haemophilia Foundation New South Wales (HFNSW)

T: 02 9280 2607
E: coordinator@hfnsw.org.au
W: www.hfnsw.org.au

Haemophilia Foundation Queensland (HFQ)

T: 07 3017 1778 or 0419 706 056 (office mobile)
E: info@hfq.org.au
W: www.hfq.org.au

Haemophilia Foundation South Australia (HFSA)

C/o HFA
T: 1800 807 173
E: hfaust@haemophilia.org.au

Haemophilia Foundation Tasmania (HFT)

T: 0431 344 575
E: haem_tas@yahoo.com.au
W: www.hft.org.au

Haemophilia Foundation Victoria (HFV)

T: 03 9555 7595
E: info@hfv.org.au
W: www.hfv.org.au

Haemophilia Foundation Western Australia (HFWA)

T: 08 9420 7294
E: office@hfwa.org
W: www.hfwa.org

Further reading

Australian Bleeding Disorders Registry (ABDR). **Annual Report**. www.blood.gov.au/data-analysis-reporting

Australian Haemophilia Centre Directors' Organisation (AHCDO). **Guidelines for the management of haemophilia in Australia**. 2016. www.ahcdo.org.au/guidelines/publications

Australian Haemophilia Nurses' Group. **Nursing management of haemophilia and von Willebrand disorder**. July 2013. www.haemophilia.org.au/publications

Haemophilia Foundation Australia. www.haemophilia.org.au/publications

A guide for people living with von Willebrand disorder. June 2010.

Haemophilia. July 2013.

Living with mild haemophilia: a guide. October 2011.

Acknowledgements

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More information

For more information and to contact Australian Haemophilia Nurses' Group (AHNG) or Australia/
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www.haemophilia.org.au/support-services/health-professional-groups

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