WHAT PRECAUTIONS DO I NEED TO TAKE?

It is important to treat bleeds as they occur and prevent bleeds as much as possible.

- Learn how to identify a "bleed" – a bleeding episode related to mild haemophilia
- All bleeds could potentially be serious. Treat a bleed early. Learn when to treat yourself and when to seek help
- Contact your Haemophilia Centre immediately after significant injury or before having medical or dental procedures
- Learn how to prevent bleeds.

Your Haemophilia Centre team will help you with this.

The aim of learning how to manage your bleeds is to enjoy the best quality of life and have as few complications as possible.

HAEMOPHILIA CENTRES

Registering and staying in contact with the local Haemophilia Centre is an important part of managing mild haemophilia.

There is at least one specialist Haemophilia Centre or Service in every state or territory in Australia, located in a major public hospital. Haemophilia Centres have a team of health professionals with expertise in providing treatment and care to people with bleeding disorders including haemophilia.

How can the Haemophilia Centre team help you or your child?

They can:

- Work with you to make a treatment plan
- Help you or your child to learn how to recognise, treat and prevent bleeds
- Liaise with other doctors and health services to make sure care is appropriate for your or your child's bleeding disorder
- Review your or your child's bleeding disorder regularly and adjust the treatment plan
- Update you on the latest information about haemophilia and treatment
- Advise on ways to live well with mild haemophilia, including managing sport, travel, childcare, school, working, relationships and having families.

The team includes:

- Haematologists: doctors who specialise in blood disorders
- Haemophilia nurses
- Social workers or counsellors
- Physiotherapists
- Access to specialised laboratory services
- Other specialist health professionals

Stay in touch with your Haemophilia Centre and check with them regularly about new information and advances in treatment and care. Update the Centre if you change your contact details or move.

WHAT CAN I EXPECT WITH MILD HAEMOPHILIA?

Many people who have mild haemophilia do not experience problems with bleeding, or only have bleeding episodes occasionally. People with severe haemophilia can have spontaneous bleeding - the bleeding that takes place in joints and muscles without any obvious cause. However, in mild haemophilia this is extremely rare.

Usually persistent bleeding will only follow more serious injury or medical and dental procedures or surgery. Some girls and women may have bleeding problems with menstruation and after childbirth, but this can often be prevented or managed with appropriate treatment.

HOW TO RECOGNISE A BLEED

Because people with mild haemophilia have bleeding problems so rarely, they sometimes do not recognise the symptoms and delay seeking treatment.

It is important not to ignore bleeds or think they will be OK if they are left untreated.

Bleeds that are not treated quickly can take longer to stop and to heal. Serious bleeds can be dangerous and need prompt medical attention.

The Haemophilia Centre team will help you with learning how to recognise a bleed and how to deal with it.

When you need to seek advice from your Haemophilia Centre

Mild haemophilia can complicate even small injuries or medical and dental procedures and surgery. If you have any of the following problems, call your Haemophilia Centre who can help you decide whether they can be managed by your local doctor or you need specialist care at the Haemophilia Centre:

- **Bruising:** seems to be growing larger or swelling, is painful and limits movement

- **Mouth, tongue or nose bleeding:** continues to ooze or bleed at times for several days

- **Muscles and joints:** bleeds might occur if a muscle or joint (knees, ankles, elbows, etc) is over-extended, twisted, overworked or receives a hard hit:
  - Feeling of tightness, heat or swelling
  - Stiffness or tingling, difficult to move or extend the limb
  - Pain or movement problems that keep you awake overnight after the RICE procedure (see page 21).
  Contact the Haemophilia Centre if there is still pain in the calf or forearm after several hours

- **Stomach, bowels, urinary tract:**
  - Pink, red or brown urine
  - Urinating more often or difficulty/pain with urinating
  - Abdominal or back pain
  - Bright red bleeding from the bowel
  - Blood in bowel motions or black bowel motions

- **Menorrhagia:** heavy bleeding with menstrual periods

See CARRYING THE HAEMOPHILIA GENE, page 40

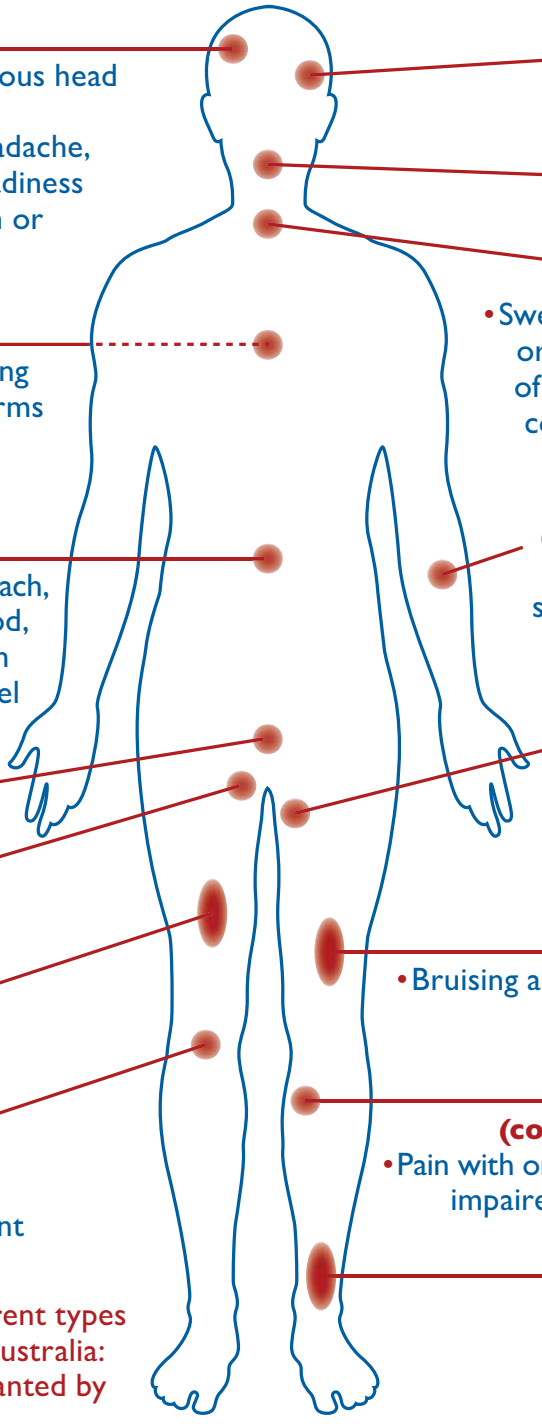
- **Babies and small children:** if you notice these signs, talk to your Haemophilia Centre:
  - Irritability, crying, pain (unrelated to hunger/needing a feed, teething or needing a nappy change)
  - Heat or swelling at a joint
  - Favouring one leg or arm
  - Avoiding some normal actions with one hand, leg or arm, or not wanting to walk.

"A couple of months ago I hurt my leg and did not go for treatment, despite my wife's pleading. A couple of days later I flew four hours on a plane, then spent the rest of the week having to do daily trips to hospital. I had to spend five weeks watching daytime television. It serves me right; it would have been so much easier to seek treatment!"

"It's a really good idea to visit your haematologist or Haemophilia Centre regularly and check treatment procedures before you need them. It's not the best time to negotiate the health system when you're in pain and need prompt attention."

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.



Head
- Blow to head or spontaneous head bleed
- Loss of consciousness, headache, vomiting, dizziness, unsteadiness
- Treat all head injuries with or without swelling

Spinal
- Back pain, weakness, tingling and numbness of legs or arms

Abdominal
- Swelling or pain in the stomach, vomiting, coughing up blood, unusual weakness, blood in bowel motions, black bowel motions

Urinary
- Blood in urine

Testes
- Blow to the groin

Muscles
- Swelling, lumps, pain, warmth

Joints
- Decreased or no movement

Eye
- Trauma to eye, pain, swelling

Tongue
- Bitten, bleeding

Throat
- Swelling, difficulty swallowing or breathing, discolouration of the skin around the neck, coughing or vomiting blood

Forearm (compartment syndrome)
- Pain with or without swelling, bruising, impaired mobility, tingling in hand

Groin (psoas or iliacus muscle buttock or groin)
- Pain, spontaneous flex, inability to extend the leg without pain

Bruising
- Bruising and significant swelling may require treatment

Calf (compartment syndrome)
- Pain with or without swelling, bruising, impaired mobility, tingling in foot

Skin
- Stitches

Source: adapted from Different types of serious bleeds. Wyeth, Australia: Sydney 2009. Permission granted by Pfizer Australia, 2011.

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

WHAT TO DO ABOUT AN INJURY OR BLEEDING

If bleeding symptoms, pain or swelling get worse in the first 24 hours or keep you awake overnight, contact your Haemophilia Centre for assessment and advice.

All bleeding and bruising should be dealt with promptly to make sure no permanent injuries result.

If you have recurring bleeds into any parts of the body discuss it with your Haemophilia Centre team to investigate the cause, how to manage it and how you might prevent further bleeds.

Minor cuts, scratches and bruises

These can usually be managed using standard first aid techniques.

- To stop bleeding from minor cuts, apply pressure and put a band aid® on it. When the cut has been cleaned, keep it dry until healed
- Small bruises may look unattractive but are not serious and usually fade over a few weeks without treatment.

Larger bruises, strains, sprains, muscle/joint pain and swelling

To reduce bruising, pain or swelling and to recover more quickly, start the **RICE** procedure promptly:

Rest – stop the activity and rest the injured body part

Ice – apply a cold pack (e.g., an ice pack or bag of frozen vegetables or a cool relief gel wrapped in a towel) for about 10-15 minutes, then remove and reapply about every two hours

Compression – after icing, wrap the injury with an elastic compression bandage or firm bandages. Remove the bandage only for icing

Elevate – where practical, raise the injured body part above the level of the heart.

Stopping bleeding into a joint or muscle quickly can help to prevent complications, including permanent injury. Over time repeated bleeds into joints and muscles can lead to problems such as arthritis in joints. This is unusual in someone with mild haemophilia - it occurs more often in people with moderate or severe haemophilia, but can occur if bleeds are unrecognised and untreated in a person with mild haemophilia.

Do not take aspirin or non steroidal anti-inflammatory drugs (NSAIDs – ie, ibuprofen, indomethacin and naproxen) to relieve the pain unless your Haemophilia Centre advises you to do so. These medications can interfere with blood clotting and delay healing.

Nosebleeds

Sit upright and pinch your nostrils together below the bridge of the nose in the soft tissue for 10 -15 minutes. If the bleeding doesn't stop, repeat this procedure a second time. Try to remain calm. A cold cloth on the back of the neck or on the bridge of the nose may also help.

Drinking hot liquids and strenuous exercise can cause the nose bleed to restart. Avoid hot soup, tea or coffee and lifting and straining for 24 hours after a nose bleed.

Bleeding from the bowel or in urine

Contact the Haemophilia Centre for assessment and treatment – untreated bleeds can lead to anaemia or low haemoglobin levels.

Heavy periods (menorrhagia)

Women with heavy periods should consult both their haematologist and gynaecologist as this may also have other causes.

See CARRYING THE HAEMOPHILIA GENE, page 40

Serious bleeding or accidents and injuries/emergencies

Serious bleeding can cause lasting damage and sometimes can be life-threatening.

- Seek emergency treatment
- Where possible, contact the Haemophilia Centre
- The Haemophilia Centre may be able to call ahead to the Emergency Department and prepare for your arrival
- Remember that haemophilia is a rare condition and not many doctors or hospital staff are familiar with it. Ask the health service you are attending to liaise with your Haemophilia Centre about your or your child's treatment
- Not all hospitals will have product for treatment available for emergencies and the Haemophilia Centre may advise on hospitals that hold treatment product or make arrangements to transport it to the hospital
- Talk to your Haemophilia Centre about whether a treatment product will be held in your local hospital in case of emergency.

Consider wearing a medical alert bracelet, carrying a Haemophilia Centre treatment wallet card and having an ICE (In Case of Emergency) number in your mobile phone:
Information about these is available from your Haemophilia Centre or state/territory Haemophilia Foundation. Show the haemophilia treatment card to medical centre or emergency department staff or ambulance drivers.

If you are at all uncertain about when or how to treat injuries or bleeding, contact your Haemophilia Centre. Learning to self-manage treatment for your bleeding disorder happens over time.

TREATMENT OPTIONS FOR MILD HAEMOPHILIA

Your Haemophilia Centre will advise on the best treatment for you.

If you think you need treatment or are not sure, contact your Haemophilia Centre as soon as possible.

There are several types of treatment. Different treatments may be useful in different situations and at different times. The haematologist will look at all of this when they work with you or your child to decide the best treatment option. They will need to reassess the situation each time treatment is needed.

Treatment may be given to prevent bleeding complications:

- In preparation for surgery, medical procedures, dental treatment or childbirth
- Or after an injury or accident
- Or if bleeding does not stop.

Desmopressin (DDAVP) is a synthetic hormone used to treat haemophilia A. It works by releasing the body's stored factor VIII into the bloodstream to help blood clot. These stores are limited, and the body may need time to rebuild stores of factor VIII before another dose is given. If you or your child need more treatment and the body's stores of factor VIII are depleted, clotting factor concentrate may need to be used instead.

Desmopressin can be given as a slow injection into a vein, but may also be given as an injection subcutaneously (into the fatty tissue under the skin), or in special circumstances as a nasal spray.

Desmopressin can help to prevent or treat bleeding in many people. It is not suitable for everyone. The haematologist at the Haemophilia Centre may decide to give a test dose of desmopressin and evaluate whether it will work for you or your child. This test may need to be repeated at times as people's responses to desmopressin change at different times in their life.

Tranexamic acid and aminocaproic acid are medicines that act by strengthening blood clots that have formed. This prevents the blood clot from being dislodged and bleeding restarting. They can be used to stop bleeding in the mouth or nosebleeds, gut bleeding, bleeding after dental work, minor surgery or an injury.

Most commonly they are taken as tablets, syrup or as a mouthwash. They may be used by themselves or together with desmopressin or a clotting factor concentrate.

Clotting factor concentrates: there are factor VIII concentrates for people with factor VIII deficiency and factor IX concentrates for people with factor IX deficiency. Both types of concentrates come in two different forms:

Recombinant factor is the most widely used type of concentrate. This is made by genetic engineering and contains little or no material from human blood or animals. There are several brands available manufactured by different pharmaceutical companies.

Plasma factor concentrates are also used. These are manufactured from the plasma (pale yellow fluid part) in human blood.

Factor concentrates are infused (injected) into a vein in the arm.

Fibrin glue is a medical gel made from fibrinogen and thrombin, which are proteins in the body that help blood to clot. It can be applied directly onto a wound to stop bleeding.

Hormone treatment, such as **oral contraceptives (birth control pills)**, can help women who have heavy menstrual bleeding. The hormones can increase factor VIII levels.

Should you learn to treat at home?

Treatment that needs to be injected, such as DDAVP or factor concentrate, can be given at the Haemophilia Centre or it may be possible to learn to treat at home. This decision will be made by you and your Haemophilia Centre, depending on your situation and how appropriate it is.

Treatment complications

After treatment with a clotting factor concentrate product, a small percentage of people with mild haemophilia may develop antibodies – known as 'inhibitors' - which make treatment less effective. There are a number of ways to treat inhibitors and many people are successful in overcoming them while others have ongoing problems.

Like all medicines, treatments for bleeding disorders can have side-effects in some people. These will be discussed with you when they are prescribed. All prescription medicines also have a plain language information leaflet called Consumer Medicine Information (CMI), which includes use, side effects and precautions. Ask your pharmacist for a copy of the CMI for your medicine.

TREATMENT PRODUCT SAFETY

Haemophilia treatment product safety is a high priority for Australian regulatory authorities, blood bank services, manufacturers and the bleeding disorders community.

If they have treatment with clotting factor concentrates, most people with mild haemophilia in Australia use recombinant products, which are genetically engineered and contain little or no human material. There have been no reports that viruses or infectious agents such as vCJD (variant Creutzfeldt-Jakob disease, the human form of "mad cow disease") have been transmitted by recombinant products.

Some people with mild haemophilia may use human plasma factor concentrates.

In Australia manufacture of human plasma factor concentrates is carefully regulated and monitored to make sure the concentrates are now as safe as possible from infections that can be transmitted by blood. This includes steps such as:

- Screening blood donors and testing blood donations
- Strict controls when selecting blood donors
- Treating human plasma factor concentrates with several processes to remove or inactivate HIV and viral hepatitis and, as far as possible, exclude other known infectious agents passed on by blood, such as vCJD.

In Australia before the early 1990s some people with haemophilia acquired hepatitis C through contaminated plasma clotting factor concentrates they used for their treatment. Some also acquired HIV. As a result new safety measures were developed and put in place. Work to prevent the transmission of infections through blood products is ongoing.

The risk of new infections from using human blood products is now thought to be extremely low. In spite of this, the risk cannot be entirely excluded, particularly if the risk came from a new or unknown type of blood-borne virus or other micro-organisms causing disease. Because of this, people using these products and patient advocacy organisations such as Haemophilia Foundation Australia (HFA) continue to take a strong and watchful interest in product safety.

Hepatitis B vaccination is recommended for people with bleeding disorders who use plasma derived concentrates.

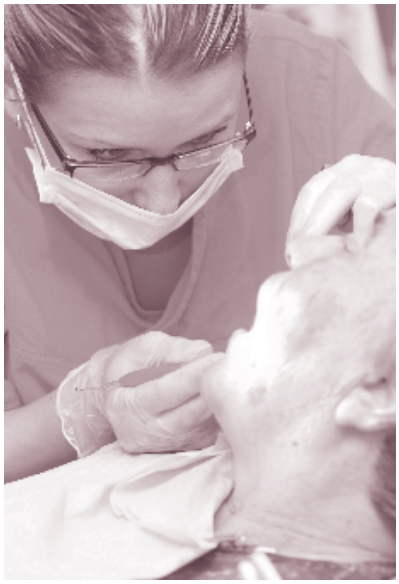
More information: Australian Red Cross Blood Service – http://www.donateblood.com.au

Australian Government. Department of Health & Ageing; National Health and Medical Research Council. Australian immunisation handbook. – http://www.health.gov.au

WHAT DO I TELL MY OTHER DOCTORS OR DENTIST?

Haemophilia is a relatively rare condition. Most doctors and dentists are not familiar with its treatment and will not be aware of your individualised treatment plan.

Ask doctors, dentists and other health care providers to liaise with your Haemophilia Centre about treatment or before having any surgery, dental work or medical procedures.



Make sure you know what type of haemophilia you have and ask the Haemophilia Centre to give you a treatment wallet card: haemophilia A and B require different treatment. The treatment card explains your diagnosis, what treatment should be given and who should be contacted for further advice. Keep the wallet card on you for quick reference.

Show your other doctors, dentist, and other health care providers the treatment card and ask them to liaise with your Haemophilia Centre. This will help with getting appropriate treatment. It will also make it easier to obtain treatment if you need it when you are away from your usual hospital or Haemophilia Centre, for example, if you are travelling or have moved interstate or overseas.

Always inform your doctor, dentist or surgeon that you have mild haemophilia before having any medical, dental or surgical procedures. This includes minor procedures, such as having sun spots or skin growths removed, and screening procedures such as colonoscopy or prostate biopsy.

Before you have any procedures, contact your Haemophilia Centre and discuss the medical support you may need to prevent bleeding complications. Where possible, plan this well ahead of time. The Haemophilia Centre team may also need to liaise with your surgical or dental team or other health professionals involved in your care to discuss the best approach for you individually and any pre- or post-treatment care you may need.

"Consult your Haemophilia Centre prior to any procedure - often your GP may not realise the true implications of a procedure from a haemophilia perspective."

"Ensure your health professionals (physiotherapists, GPs, podiatrists, specialists, optometrists, dentists, etc) are comfortable with the fact that you have haemophilia - if not, find one who is!"

"My local treatment team has issued me with a treatment card. It was really handy when I went interstate. Many of the staff at Emergency had never come across someone with haemophilia before, so it was a novelty for them."

INJECTIONS AND IMMUNISATIONS

Both children and adults with mild haemophilia can have all the normal immunisations. Informing the nurse or doctor giving the immunisation that you or your child has haemophilia is important. Injections may be given subcutaneously, into the fatty tissue under the skin, rather than into the muscle, and pressure put on the skin where you or your child was injected. This reduces the risk of bruising and bleeding.

However, it isn't always necessary to change the way of giving immunisations for children with mild haemophilia. If you or your child have mild haemophilia, contact the Haemophilia Centre for advice on immunisation methods.

Intramuscular injections, such as penicillin, can cause muscle bleeds and are not recommended for children or adults with mild haemophilia.

MEDICATION TO BE AVOIDED

Some medicines, vitamins and herbs interfere with the way platelets promote clotting and may delay healing. If you have mild haemophilia, consult with your haematologist before taking:

- Medicines containing aspirin
- Non-steroidal anti-inflammatory drugs, unless prescribed by a doctor with expertise in haemophilia (i.e., ibuprofen, indomethacin and naproxen – these have many brand names; ask your local pharmacist to check for you)
- Other blood thinners such as warfarin and heparin
- Capsules of fish oil containing omega-3 fatty acids (however, normal serves of fish should not cause a problem)
- Herbal or homeopathic medicines that affect platelet function or clotting, such as ginkgo biloba, ginger, ginseng and chondroitin
- Other medicines that claim to treat bleeding, bruising or improve clotting.

Also check with your doctor when starting new medications that could irritate your mucous membranes such as your nasal passages or stomach lining – any bleeding could be complicated by mild haemophilia.