# Acquired Haemophilia

## Patient Information Leaflet

This leaflet has been designed to provide information and advice to people and their families who have been diagnosed with Acquired Haemophilia.
The information is a guide only and specific questions can be answered by your doctor or nurse specialist.

### What is Acquired Haemophilia?

Haemophilia describes a group of blood disorders in which there is a problem with clotting of the blood. Blood contains proteins called clotting factors, and these work to stop bleeding. The lack of clotting factor causes people with haemophilia to bleed for longer periods of time than people who have normal blood clotting levels.

The most common form of haemophilia is inherited and a person who is born with haemophilia will have it for life.

However, a much rarer form of haemophilia exists called **Acquired Haemophilia** and people with this type of haemophilia are not born with it but develop the condition later in life. This type of haemophilia usually affects the clotting factor VIII (eight).

In acquired haemophilia, the gene for factor VIII (eight) is normal. Instead, the person's own immune system starts to destroy their own Factor VIII as fast as it is made. It affects men and women equally and can present at any age, however acquired haemophilia is more likely to develop in older people.

### How is it diagnosed?

Usually, the first sign is significant bruising on the body; often the bleeding is spontaneous and not associated with knocks or injuries. The bleeding can occur in the muscles; this can be very painful and cause loss of movement. Other times, it can result in excessive bleeding following a surgical procedure, blood in the urine or from the bowels. A simple blood test that looks at the body's clotting system will identify that it is not working; extensive tests are then carried out by a specialist lab like the one here in St James's hospital.

## What causes it?

In many cases the cause is unknown; part of the immune system just stops working correctly. The body's immune system sees its own Factor VIII as foreign and produces antibodies which destroy the factor VIII.

In some cases, it can be associated with another illness, e.g., rheumatoid arthritis, ulcerative colitis, asthma, psoriasis, cancer and it can develop in women who have just had a baby.

Acquired haemophilia is a very rare condition and is usually managed in a specialist haemophilia centres.

## Is it Serious?

Acquired haemophilia can cause serious life-threatening bleeding. However, once it is diagnosed a number of treatments are available.

## What treatments are available?

There are two main goals in the management of acquired haemophilia: 1) Manage and treat bleeding. 2) Suppress the immune system to stop the destruction of factor VIII. You will be advised to stay in hospital until your factor levels return to safe levels. This normally takes many weeks; however individual patient response times vary so we cannot provide a specific time frame. Clotting factor concentrates (CFC) are given to treat bleeding and steroids and/or Rituximab are given to suppress the immune system.

- **Clotting Factor Concentrates (CFC)**

CFCs are given to treat bleeding. This involves injecting clotting factors directly in to your blood stream through your vein. These treatments may have to be given several times a day initially by the doctors or nurses. There are several types of clotting factors used; some come from blood plasma donated from other people; others are developed in labs and these are called recombinant products. While the CFCs are useful to control bleeding, they do not treat the antibodies being made by the immune system and therefore, other treatments which control the immune system are also needed (Steroids and Rituximab – see below).

- **Steroids**

Steroids are used in high doses initially to suppress the immune system and stop the body producing the antibodies that are destroying the factor VIII. In Acquired Haemophilia, steroid treatment is often needed for several weeks at high dose and once the factor levels return to normal, steroids are slowly reduced over a number of weeks.

Steroids are very effective drugs but sometimes have side effects, these include:

- Trouble sleeping if taken too late in the day,
- Change in mood,
- Increased appetite and weight gain
- Diabetes
- High blood pressure
- Increased risk of infection

The nurse caring for you can explain this in more detail. You will be also given some medication to try offset some of these complications

- Osteoporosis
- Stomach Ulcers

- **Rituximab**

Rituximab is a drug that works specifically by attaching itself to the cells that are producing the antibodies which are destroying the Factor VIII.

Usually, 4 treatments are given into a vein over a 4-week period.

In some incidences, Rituximab may cause an allergic reaction while it's being given. This is more common with the first treatment, so it will be given slowly over a few hours. The nurse will give you drugs to help prevent or reduce this.

In the event that you do have a reaction the nurse will administer medication promptly to reverse this. Signs of a reaction can include:

- Feeling unwell or different to before infusion started
- Shortness of breath
- Pain in your back, tummy or chest
- Tickle in your throat
- Flu-like symptoms (headaches, a high temperature or chills)
- Feeling sick
- Rash
- Feeling itchy

- **Emicizumab (Hemlibra)**

This medication can be given in conjunction with other medications for example steroids.

In acquired FVIII deficiency Emicizumab works by acting like a bridge between other clotting proteins in your body helping the blood to clot even when your own FVIII is not working. This reduces the risk of bleeding. This medication is given once weekly as an injection under the skin (a subcutaneous

injection) It can be injected into the tummy, thigh or upper arm. In most cases the patient or their caregiver can learn how to administer this medication. Depending on their treatment plan most patients or caregivers can administer their injections at home.

This medication is very effective but can have side effects such as headache, redness around the injection site and in rare cases there can be an increased risk of blood clots.

Your health care team will discuss these risks with you and you will be closely monitored by our team. It is very important that you tell your nurse straight away if you have any of these symptoms.

You may be on one or all of the above treatments above when diagnosed with acquired haemophilia.

## What to do when you are discharged?

You will be under regular follow up from the Haemophilia Centre initially. Contact the treatment centre if you have any signs of bleeding. Signs of bleeding include bruising, blood in the urine, blood from the back passage, pain, swelling or loss of movement of limbs.

If you need any invasive procedures or dental work, please contact the National Coagulation Centre for advice beforehand. It is important to note that injections in to your muscles (IM) are not advised, unless advice is sought from your haemophilia doctor or nurse. You should also avoid NSAIDS (Aspirin/Ibuprofen/similar drugs)

**General Contact Information for Haemophilia Treatment Centres**

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| **NCC Treatment Centre**<br>Working hours, Mon-Fri, 0830-1700 hrs<br>01 416 2141<br>Email – NCC@STJAMES.IE<br><br>Out of hours and Bank holidays – 01 410 3132<br><br>Alternatively, phone (01) 410 3000 and ask for the Haematology SHO on call. | **CUH Treatment Centre**<br>Working hours, Mon-Fri, 0830-1700 hrs<br>021 4922278 or 0879683246<br><br>After 5pm & bank holidays - 021 4546400 and ask for Haematology Registrar On Call |
| **CHI Treatment Centre (<16years only)**<br>Working hours, Mon-Fri, 0830-1700 hrs<br>Haemophilia Nurse Direct Line - 01 4096939/40<br>Pager 01 4096100 Bleep 8732/8733<br><br>After 5pm or at weekend/bank holidays, phone the hospital and ask to speak to the Haematology Registrar on call | **GUH Treatment Centre**<br>Mon – Fri, 0800 - 1600 hrs. Direct line 091 542348 or<br><br>Ring the work mobile 087 035 7499<br><br>After 4 pm and weekend /bank holidays ring the hospital on 091 524222 and ask for doctor 'on call' for Haematology |